**2026 PacificSource Health Plans Prior Authorization Criteria**

Last Modified: 06/22/2026
(All criteria reviewed at least once per year. Updates occur every 1 to 2 months.)

PacificSource HEALTH PLANS

## Contents

POLICY NAME:
**ACTIMMUNE**
Affected Medications: ACTIMMUNE (interferon gamma 1b)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Chronic Granulomatous Disease (CGD)<br>  ○ Severe, malignant osteopetrosis (SMO)<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | • Patient's body surface area (BSA) must be documented along with the prescribed dose.<br>• Pediatrics with BSA less than 0.5 $m^2$: weight must be documented along with prescribed dose<br><br>**Chronic granulomatous disease**<br>• Diagnosis established by a molecular genetic test identifying a gene-related mutation associated with CGD<br><br>**Severe, malignant osteopetrosis**<br>• Diagnosis of severe infantile osteopetrosis established by ONE of the following:<br>  ○ Radiographic imaging consistent with osteopetrosis<br>  **OR**<br>  ○ Molecular genetic test identifying a gene-related mutation associated with SMO<br><br>**Oncology indications**<br>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | **Chronic Granulomatous Disease**<br>• Patient is on a prophylactic regimen with an antibacterial agent and an antifungal agent<br><br>**All indications**<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization:** documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • **CGD:** prescribed by, or in consultation with, an immunologist<br>• **SMO:** prescribed by, or in consultation with, an endocrinologist<br>• **Oncology indications:** prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **CGD and SMO:**<br>• Authorization: 12 months, unless otherwise specified |

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| | **<u>Oncology indications</u>:**<br>• Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ADDYI & VYLEESI**
Affected Medications: ADDYI (flibanserin), VYLEESI (bremelanotide injection)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br><br>**Vyleesi:** Premenopausal women with acquired, generalized hypoactive sexual desire disorder (HSDD)* as characterized by low sexual desire that causes marked distress or interpersonal difficulty that is NOT due to any of the following:<br>o A coexisting medical or psychiatric condition<br>o Problems within a relationship<br>o The effects of medication or other drug substance<br><br>**Addyi:** women less than 65 years of age with acquired, generalized hypoactive sexual desire disorder (HSDD)* as characterized by low sexual desire that causes marked distress or interpersonal difficulty and is NOT due to any of the following:<br>o A coexisting medical or psychiatric condition<br>o Problems within a relationship<br>o The effects of a medication or other drug substance<br><br>*Also known as female sexual interest/arousal disorder |
| **Required Medical Information:** | • Documented mental health diagnosis of acquired, generalized HSDD meeting the *Diagnostic and Statistical Manual of Mental Disorders*, Fifth Edition (DSM-5) criteria for female sexual interest/arousal disorder:<br>o Lack of, or significant reduction in, at least **3** of the following:<br>▪ interest in sexual activity<br>▪ sexual thoughts or fantasies<br>▪ initiation of sexual activity and responsiveness to a partner's initiation<br>▪ excitement or pleasure during all or almost all sexual activity<br>▪ interest or arousal in response to any sexual cues (e.g., written, verbal, visual)<br>▪ genital or non-genital sensations during sexual activity in all or almost all sexual encounters<br>o Symptoms have persisted for a minimum duration of 6 months<br>o Symptoms cause clinically significant distress<br>o Sexual dysfunction is not attributable to any of the following:<br>▪ A nonsexual medical or psychiatric condition<br>▪ Severe relationship distress (e.g., partner violence)<br>▪ The effects of medication or other substance use<br>▪ Other clinically significant and relevant stressors |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** will require documentation of treatment success and confirmation that patient is still premenopausal |
| **Exclusion Criteria:** | • Treatment of males<br>• Intended use is to enhance sexual performance |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a mental health provider |

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| | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 2 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ADENOSINE DEAMINASE (ADA) REPLACEMENT**
Affected Medications: REVCOVI (elapegademase-lvlr)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of adenosine deaminase severe combined immune deficiency (ADA-SCID) in pediatric and adult patients |
| **Required Medical Information:** | • Diagnosis of ADA-SCID confirmed by genetic testing showing biallelic pathogenic variants in the *ADA* gene<br>• Laboratory findings show at least **ONE** of the following:<br>  ○ Absent ADA levels in lysed erythrocytes<br>  ○ A marked increase in deoxyadenosine triphosphate (dATP) levels in erythrocyte lysates<br>  ○ A significant decrease in ATP concentration in red blood cells<br>  ○ Absent or extremely low levels of N adenosylhomocysteine hydrolase in red blood cells<br>  ○ Increase in 2'-deoxyadenosine in urine and plasma |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation showing that neither gene therapy nor a matched sibling or family donor for HCT (hematopoietic cell transplantation) is available, or that gene therapy or HCT was unsuccessful<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization** requires documentation of treatment success defined as disease stability and/or improvement as indicated by one or more of the following:<br>• Increase in plasma ADA activity<br>• Decrease in red blood cell dATP/dAXP level<br>• Improvement in immune function with diminished frequency/complications of infections |
| **Exclusion Criteria:** | • Other forms of autosomal recessive SCIDs<br>• All uses not listed under covered uses are considered experimental |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an immunologist or specialist experienced in the treatment of severe combined immune deficiency (SCID)<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**ADZYNMA**
Affected Medications: ADZYNMA (apadamtase alfa)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Congenital thrombotic thrombocytopenic purpura (cTTP) |
| **Required Medical Information:** | • Diagnosis of severe cTTP confirmed by BOTH of the following:<br>  ○ Molecular genetic testing confirming presence of homozygous or compound heterozygous variants in the ADAMTS13 gene<br>  ○ ADAMTS13 activity testing showing less than 10% of normal activity<br>• **For on-demand treatment:** documentation of current or past acute event with the following:<br>  ○ Reduction in platelet count by 50% or greater **OR** platelet count less than 100,000/microliter<br>  ○ Elevation in lactate dehydrogenase (LDH) level to more than 2x baseline or the upper limit of normal (ULN)<br>• **For prophylactic use:**<br>  ○ Must have history of at least one documented thrombotic thrombocytopenic purpura (TTP) event (past acute event or subacute event such as thrombocytopenia event or a microangiopathic hemolytic anemia event) |
| **Appropriate Treatment Regimen & Other Criteria:** | • **Dosing:**<br>  ○ Prophylactic: 40 IU/kg once every other week<br>  ▪ May be dosed weekly with documentation of appropriate prior dosing regimen or clinical response<br>  ○ On-demand therapy: 40 IU/kg on day 1, 20 IU/kg on day 2, and 15 IU/kg on day 3 and beyond until 2 days after the acute event is resolved<br><br>**<u>Reauthorization</u>**:<br>• For prophylactic use: documentation of treatment success defined as an improvement in the number or severity of TTP events, platelet counts, or clinical symptoms<br>• For on-demand use: documentation of treatment success, defined as an increase in platelet counts to at least 150,000/microliter, or counts returned to within 25% of baseline |
| **Exclusion Criteria:** | • Diagnosis of other TTP-like disorder, such as acquired or immune-mediated TTP |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist, oncologist, intensive care specialist, or specialist in rare genetic hematologic diseases<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**AFAMELANOTIDE**
Affected Medications: SCENESSE (afamelanotide injection)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of patients with erythropoietic protoporphyria (EPP) with phototoxic reactions |
| **Required Medical Information:** | **<u>Erythropoietic Protoporphyria (EPP)</u>**<br>• Documented diagnosis of EPP confirmed by biallelic loss-of-function mutation in the ferrochelatase (FECH) gene<br>• Documented increase in total erythrocyte protoporphyrin, with at least 85% metal-free protoporphyrin<br>• Documented symptoms of phototoxic reactions, resulting in dysfunction and significant impact on activities of daily living |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Reauthorization</u>:**<br>• Documentation of treatment success and clinically significant response to therapy (e.g., decreased severity and number of phototoxic reactions, increased duration of sun exposure, increased quality of life, etc.)<br>AND<br>• Continued implementation of sun and light protection measures during treatment to prevent phototoxic reactions |
| **Exclusion Criteria:** | • Cosmetic indications, such as vitiligo |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist at a recognized Porphyria Center<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**AFINITOR**
Affected Medications: AFINITOR, AFINITOR DISPERZ (everolimus), EVEROLIMUS SOLUBLE TABLET

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| **Covered Uses:** | • Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | **Oncology Indications**<br>• Documentation of performance status, all prior therapies used, and prescribed treatment regimen<br><br>**Tuberous Sclerosis Complex (TSC)**<br>• Documentation of treatment resistant epilepsy, defined as lack of seizure control with 2 different antiepileptic regimens and meeting following criteria:<br>  ○ Documentation of treatment failure with Epidiolex (cannabidiol solution) adjunct therapy<br>  ○ Documentation that Afinitor Disperz (only form approved for TSC-seizures) is being used as adjunct therapy for seizures<br>**OR**<br>• Documentation of symptomatic subependymal giant cell tumors (SGCTs) or TSC-associated subependymal giant cell astrocytoma (SEGA) in a patient who is not a good candidate for surgical resection |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | **Oncology Indications**<br>• Karnofsky Performance Status less than or equal to 50% or ECOG performance score greater than or equal to 3 |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • **Oncology Indication**: Prescribed by, or in consultation with, an oncologist<br>• **TSC Indication**: Prescribed by, or in consultation with, a neurologist or specialist in the treatment of TSC<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ALEMTUZUMAB**
Affected Medications: LEMTRADA (alemtuzumab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:<br>  ▪ Relapsing-remitting multiple sclerosis (RRMS)<br>  ▪ Active secondary progressive multiple sclerosis (SPMS) |
| **Required Medical Information:** | • Diagnosis confirmed with magnetic resonance imaging (MRI) per revised McDonald diagnostic criteria for MS<br>  ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of treatment failure with (or intolerance to) **ONE** of the following:<br>  ○ Rituximab (preferred biosimilar products: Riabni, Ruxience)<br>  ○ Ocrevus (Ocrelizumab), if previously established on treatment (excluding via samples or manufacturer's patient assistance programs)<br><br>**Reauthorization** requires provider attestation of treatment success<br>• Eligible for renewal 12 months after administration of last dose |
| **Exclusion Criteria:** | • Human immunodeficiency virus (HIV) infection<br>• Active infection<br>• Concurrent use of other disease-modifying medications indicated for the treatment of multiple sclerosis |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or a multiple sclerosis specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 5 doses for 5 days, unless otherwise specified<br>• Reauthorization: 3 doses for 3 days, unless otherwise specified |

POLICY NAME:
**ALGLUCOSIDASE ALFA**
Affected Medications: LUMIZYME (alglucosidase alfa)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Pompe Disease |
| **Required Medical Information:** | • Diagnosis of Pompe disease confirmed by an enzyme assay demonstrating a deficiency of acid α-glucosidase (GAA) enzyme activity or by DNA testing that identifies mutations in the GAA gene.<br>• Patient weight and planned treatment regimen |
| **Appropriate Treatment Regimen & Other Criteria:** | • One or more clinical signs or symptoms of Pompe disease, including but not limited to:<br>  ○ Readily observed evidence of glycogen storage (macroglossia, hepatomegaly, normal or increased muscle bulk)<br>  ○ Involvement of respiratory muscles manifesting as respiratory distress (such as tachypnea)<br>  ○ Profound diffuse hypotonia<br>  ○ Proximal muscle weakness<br>  ○ Reduced forced vital capacity (FVC) in upright or supine position<br>• Appropriate medical support is readily available when medication is administered in the event of anaphylaxis, severe allergic reaction, or acute cardiorespiratory failure<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Concurrent use of other enzyme replacement therapies such as Nexviazyme or Pombiliti and Opfolda |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a metabolic specialist, endocrinologist, biochemical geneticist, or physician experienced in the management of Pompe disease<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**ALOSETRON**
Affected Medications: ALOSETRON

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Women with severe diarrhea-predominant irritable bowel syndrome (IBS) |
| **Required Medical Information:** | • Female gender<br>• Chronic IBS symptoms lasting at least 6 months<br>• Diarrhea AND one or more of the following are present:<br>  ○ Frequent and severe abdominal pain/discomfort<br>  ○ Frequent bowel urgency or fecal incontinence<br>  ○ Disability or restriction of daily activities due to IBS<br>• Other anatomical or biochemical abnormalities of the gastrointestinal tract have been excluded as a cause of symptoms |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented inadequate response to all the following:<br>  ○ Loperamide<br>  ○ Dicyclomine<br>  ○ Hyoscyamine<br>  ○ Amitriptyline or nortriptyline<br><br>**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • History of chronic or severe constipation or sequelae from constipation, intestinal obstruction, stricture, toxic megacolon, gastrointestinal perforation, and/or adhesions, ischemic colitis, impaired intestinal circulation, thrombophlebitis, or hypercoagulable state, Crohn's disease or ulcerative colitis, diverticulitis, or severe hepatic impairment<br>• Concomitant use of fluvoxamine |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a gastroenterologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 2 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ALPHA-1 PROTEINASE INHIBITORS**
Affected Medications: ARALAST NP, GLASSIA, PROLASTIN-C, ZEMAIRA

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ Chronic augmentation and maintenance therapy in adults with clinically evident emphysema due to severe congenital alpha-1 antitrypsin (AAT) deficiency |
| **Required Medical Information:** | • Documented diagnosis of severe congenital AAT deficiency, confirmed by **BOTH** of the following (a and b):<br>  a. Baseline AAT serum concentration of less than or equal to 11 mmol/L (equivalent to 57 mg/dL or less via nephelometry, 80 mg/dL or less via radial immunodiffusion)<br>  b. One of the following high-risk phenotypic variants: PiZZ, PiSZ, Pi(null)(null), or other rare allelic mutation<br>2. Documentation of clinically evident emphysema or chronic pulmonary obstructive disease (COPD), confirmed by **ONE** of the following (a or b):<br>  a. Evidence of severe airflow obstruction, defined as forced expiratory volume in one second (FEV1) of 30-65% predicted<br>  b. Evidence of mild-moderate airflow obstruction, defined as an FEV1 between 66-80% of predicted, but has demonstrated a rapid decline by at least 100 mL/year |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of non-smoker status<br>  ○ Has not smoked for a minimum of 6 consecutive months leading up to therapy initiation and will continue to abstain from smoking during therapy<br>• Dosing: 60 mg/kg intravenously once weekly<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Use in the management of lung disease in which severe AAT deficiency has not been established<br>• Patients with IgA deficiency or with the presence of IgA antibodies<br>• Prior liver transplant |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**AMIFAMPRIDINE**
Affected Medications: FIRDAPSE (amifampridine phosphate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Lambert-Eaton myasthenic syndrome (LEMS) |
| **Required Medical Information:** | • Documented diagnosis of LEMS confirmed by **ONE** of the following:<br>  ○ Positive anti-P/Q-type voltage-gated calcium channel (VGCC) antibody test<br>  ○ Repetitive nerve stimulation (RNS) abnormalities, such as an increase in compound muscle action potential (CMAP) amplitude at least 60 percent after maximum voluntary contraction (i.e., post-exercise stimulation) or at high frequency (50 Hz)<br>• Documentation of clinical signs and symptoms consistent with LEMS, as follows: proximal muscle weakness (without atrophy), with or without autonomic features and areflexia |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of inadequate clinical response or intolerance to **ONE** of the following (except in active small cell lung carcinoma [SCLC]-LEMS):<br>  ○ Combination oral prednisone and azathioprine therapy<br>  ○ Combination intravenous immunoglobulin therapy with one of the following: oral prednisone or azathioprine<br><br>**Reauthorization:** documentation of treatment success, confirmed by improved or sustained muscle strength on clinical assessments |
| **Exclusion Criteria:** | • Seizure disorder<br>• Active brain metastases<br>• Clinically significant long QTc interval on ECG in previous year OR history of additional risk factors for torsade de pointes |
| **Age Restriction:** | • 6 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ANIFROLUMAB**
Affected Medications: SAPHNELO (anifrolumab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Systemic Lupus Erythematosus (SLE) |
| **Required Medical Information:** | • Documentation of SLE with moderate to severe disease (significant but non-organ threatening disease including constitutional, cutaneous, musculoskeletal, or hematologic involvement)<br>• Autoantibody-positive SLE, defined as positive for antinuclear antibodies (ANA) and/or anti-double-stranded DNA (anti-dsDNA) antibody |
| **Appropriate Treatment Regimen & Other Criteria:** | • Failure with at least 12 weeks of combination therapy including hydroxychloroquine OR chloroquine with one of the following:<br>  ○ Cyclosporine, azathioprine, methotrexate, or mycophenolate mofetil<br>• Documented failure with at least 12 weeks of subcutaneous Benlysta<br><br>**Reauthorization** requires documentation of treatment success or a clinically significant improvement such as a decrease in flares or corticosteroid use |
| **Exclusion Criteria:** | • Use in combination with other biologic therapies<br>• Use in severe active central nervous system lupus |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a rheumatologist or a specialist with experience in the treatment of systemic lupus erythematosus<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**ANTIEMETICS**
Affected Medications: VARUBI (rolapitant)

<table>
<tr><td>Covered Uses:</td><td>
<ul>
<li>All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
<ul>
<li>Prevention of delayed nausea and vomiting associated with initial and repeat courses of emetogenic cancer chemotherapy, including, but not limited to, highly emetogenic chemotherapy</li>
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<tr><td>Required Medical Information:</td><td>
<u>Chemotherapy Induced Nausea and Vomiting Prophylaxis</u>
<ul>
<li>Documentation of planned chemotherapy regimen</li>
<li>Documentation of a highly OR moderately emetogenic chemotherapy regimen</li>
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<table>
<tr><th colspan="4">Highly Emetogenic Chemotherapy</th></tr>
<tr><td>Any regimen that contains an anthracycline and cyclophosphamide</td><td>Cyclophosphamide</td><td>Fam-trastuzumab deruxtecan-nxki</td><td>Sacituzumab govitecan-hziy</td></tr>
<tr><td>Carboplatin</td><td>Dacarbazine</td><td>Ifosfamide</td><td>Streptozocin</td></tr>
<tr><td>Carmustine</td><td>Doxorubicin</td><td>Mechlorethamine</td><td>FOLFOX</td></tr>
<tr><td>Cisplatin</td><td>Epirubicin</td><td>Melphalan</td><td></td></tr>
<tr><th colspan="4">May be considered highly emetogenic in certain patients</th></tr>
<tr><td>Dactinomycin</td><td>Idarubicin</td><td>Methotrexate (250 mg/m$^2$ or greater)</td><td>Trabectedin</td></tr>
<tr><td>Daunorubicin</td><td>Irinotecan</td><td>Oxaliplatin</td><td></td></tr>
<tr><th colspan="4">Moderately Emetogenic Chemotherapy</th></tr>
<tr><td>Aldesleukin</td><td>Cytarabine</td><td>Idarubicin</td><td>Mirvetuximab soravtansine-gynx</td></tr>
<tr><td>Amifostine</td><td>Dactinomycin</td><td>Irinotecan</td><td>Naxitamab-gqgk</td></tr>
<tr><td>Bendamustine</td><td>Daunorubicin</td><td>Irinotecan (liposomal)</td><td>Oxaliplatin</td></tr>
<tr><td>Busulfan</td><td>Dinutuximab</td><td>Lurbinectedin</td><td>Romidepsin</td></tr>
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| | Clofarabine | Dual-drug liposomal encapsulation of cytarabine and daunorubicin | Methotrexate (250 mg/m$^2$ or greater) | Temozolomide |
| | Trabectedin | | | |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Chemotherapy induced Nausea and Vomiting Prophylaxis</u>**<br>• Documented treatment failure with a 5-HT3 receptor antagonist (e.g., ondansetron, granisetron) in combination with dexamethasone while receiving the current chemotherapy regimen<br><br>**<u>Quantity Limit:</u>**<br>• 1 dose per 14 days<br><br>**<u>Reauthorization</u>** requires documentation of treatment success and initial criteria to be met | | | |
| **Exclusion Criteria:** | • Treatment of acute or breakthrough nausea and vomiting | | | |
| **Age Restriction:** | • 18 years of age and older | | | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care | | | |
| **Coverage Duration:** | • Authorization: 6 months, unless otherwise specified | | | |

POLICY NAME:
**ANTIHEMOPHILIC FACTORS**
Affected Medications: Advate, Adynovate, Afstyla, Alphanate, Alphanate/VWF Complex/Human, Alphanine SD, Alprolix, Altuviiio, Benefix, Corifact, Eloctate, Esperoct, Feiba NF, Helixate FS, Hemofil M, Humate P, Idelvion, Ixinity, Jivi, Koate DVI, Kogenate FS, Kovaltry, Novoseven RT, NovoEight, Nuwiq, Obizur, Rebinyn, Recombinate, Riastap, Rixubis, Sevenfact, Tretten, Vonvendi, Wilate, Xyntha

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Documentation of dose based on reasonable projections, current dose utilization, product labeling, diagnosis, baseline factor level, circulating factor activity (% of normal or units/dL), and rationale for use<br>• Current weight<br>• Documentation of Bethesda Titer level and number of bleeds in the past 3 months with severity and cause of bleed<br><br>**<u>Documentation of one of the following diagnostic categories:</u>**<br>• Hemophilia A or Hemophilia B<br>  ○ Mild: factor levels greater than 5% and less than 30%<br>  ○ Moderate: factor levels of 1% to 5%<br>  ○ Severe: factor levels of less than 1%<br>• Von Willebrand disease (VWD), which must be confirmed with plasma von Willebrand factor (VWF) antigen, plasma VWF activity, and factor VIII activity<br><br>**<u>Documentation of one of the following indications:</u>**<br>• Acute treatment of moderate to severe bleeding in patients with:<br>  ○ Mild, moderate, or severe hemophilia A or B<br>  ○ Severe VWD<br>  ○ Mild to moderate VWD in clinical situations with increased risk of bleeding<br>• Perioperative prophylaxis and/or treatment of acute, moderate to severe bleeding in patients with hemophilia A, hemophilia B, or VWD<br>• Routine prophylaxis in patients with severe hemophilia A, severe hemophilia B, or severe VWD<br>  ○ For Wilate and Vonvendi for routine prophylaxis: documentation of severe Type 3 VWD |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Hemophilia A (factor VIII deficiency)</u>**<br>• Documentation indicates requested medication is to achieve or maintain but not to exceed maximum functional capacity in performing daily activities<br>• For mild disease: treatment failure or contraindication to Stimate (desmopressin)<br>• **Eloctate** and **Nuwiq** require documented inadequate response, or documented intolerable adverse event, with all preferred products (Kogenate FS, Kovaltry, Novoeight, Jivi, Adynovate)<br>• **Helixate FS** requires documented treatment failure with Kogenate FS due to an intolerable adverse event and the prescriber has a compelling medical rationale for not expecting the same event to occur with Helixate FS |

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| | • **Altuviiio** requires documentation of severe hemophilia or moderate hemophilia with a severe bleeding phenotype defined by frequent non-traumatic bleeds requiring prophylaxis<br><br>**<u>Hemophilia B (factor IX deficiency)</u>**<br>• For **Benefix**, **Idelvion,** and **Rebinyn**: documentation treatment failure or contraindication to Rixubis<br>• For **Alprolix**: documentation of contraindication to Rixubis for perioperative management<br><br>**<u>von Willebrand disease (VWD)</u>**<br>• For **Vonvendi**:<br>  ○ Documentation of treatment failure or contraindication to Humate P AND Alphanate for perioperative prophylaxis and/or treatment of acute, moderate to severe bleeding<br>  ○ Documentation of treatment failure or contraindication to Wilate for routine prophylaxis<br><br>**<u>All Indications</u>**<br>• Approval based on necessity and laboratory titer levels<br>• Coverage for a non-preferred product requires documentation of one of the following:<br>  ○ Documented intolerable adverse event to all preferred products, and the adverse event was not an expected adverse event attributed to the active ingredient<br>  ○ Currently receiving treatment with a non-preferred product, excluding via samples or manufacturer's patient assistance programs<br><br>**<u>Reauthorization</u>**: requires documentation of planned treatment dose, number of acute bleeds since last approval (with severity and cause of bleed), past treatment history, and titer inhibitor level to factor VIII and IX as appropriate |
| **Exclusion Criteria:** | • Acute thrombosis, embolism, or symptoms of disseminated intravascular coagulation<br>• Obizur for congenital hemophilia A or VWD<br>• Tretten for congenital factor XIII B-subunit deficiency<br>• Jivi and Adynovate for VWD<br>• Idelvion for immune tolerance induction in patients with Hemophilia B<br>• Vonvendi for congenital hemophilia A or hemophilia B<br>• Afstyla and Nuwiq for VWD |
| **Age Restriction:** | • Subject to review of FDA label for each product<br>• Jivi: 7 years of age and older<br>• Adynovate: 12 years of age and older<br>• Vonvendi: 18 years of age and older<br>• Wilate for routine prophylaxis with von Willebrand disease: 6 years and older |
| **Prescriber Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• Members who are on a State Based Drug List are required to utilize pharmacy benefits only |

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| | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified<br>• Perioperative management: 1 month, unless otherwise specified |

POLICY NAME:
**ANTITHYMOCYTE GLOBULINS**
Affected Medications: ATGAM (antithymocyte globulin – equine), THYMOGLOBULIN (antithymocyte globulin – rabbit)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of allograft rejection in renal transplant recipients **(Atgam, Thymoglobulin)**<br>  ○ Treatment of moderate to severe aplastic anemia in patients unsuitable for bone marrow transplantation **(Atgam)**<br>  ○ Prophylaxis of acute rejection in renal transplant recipients **(Thymoglobulin)**<br>• National Comprehensive Cancer Network (NCCN) indications with evidence level of 2A or better<br>• Compendia-supported uses that will be covered **(Thymoglobulin)**<br>  ○ Prophylaxis and treatment of acute rejection in:<br>  ▪ Heart transplant recipients<br>  ▪ Liver transplant recipients<br>  ▪ Lung transplant recipients<br>  ▪ Pancreas transplant recipients<br>  ▪ Intestinal transplant recipients<br>  ○ Prophylaxis of acute rejection in multivisceral transplant recipients<br>  ○ Prophylaxis of graft-versus-host disease in unrelated donor hematopoietic stem cell transplant recipients |
| **Required Medical Information:** | **<u>Oncology uses</u>**<br>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br><br>**<u>All Indications</u>**<br>• Documentation of a complete treatment plan with planned dose, frequency and duration of therapy<br>• Current patient weight<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**<u>Prophylaxis of acute transplant rejection</u>**<br>• Patient must be considered high risk for acute rejection or delayed graft function based on one or more of either the following donor/recipient risk factors:<br><u>Donor risk factors:</u><br>  ○ Donor cold ischemia for more than 24 hours<br>  ○ Donor age older than 50 years old<br>  ○ Donor without a heartbeat<br>  ○ Donor with ATN<br>  ○ Donor requiring high-dose inotropic support<br><u>Recipient risk factors:</u><br>  ○ Repeated transplantation<br>  ○ Panel-reactive antibody value exceeding 20% before transplant |

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| | ○ Black race<br>○ One or more HLA antigen mismatches with the donor |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Prophylaxis of acute transplant rejection</u>**<br>• Documented treatment failure, intolerable adverse event, or contraindication to the use of basiliximab<br><br>**<u>Treatment of allograft rejection in renal transplant recipients</u>**<br>• Requests for Atgam require documented treatment failure or rationale for avoidance of Thymoglobulin |
| **Exclusion Criteria:** | • Oncology uses: Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• Active acute or chronic infections which contraindicate additional immunosuppression<br>• Use in patients with aplastic anemia who are suitable candidates for bone marrow transplantation or in patients with aplastic anemia secondary to neoplastic disease, storage disease, myelofibrosis, Fanconi's syndrome, or in patients known to have been exposed to myelotoxic agents or radiation **(Atgam)** |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist in oncology, hematology, nephrology or transplant medicine as appropriate for diagnosis<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 1 month, unless otherwise specified |

POLICY NAME:
**ANTITHROMBIN III**
Affected Medications: ANTITHROMBIN III (THROMBATE III)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ Indicated in patients with hereditary antithrombin deficiency (hATd) for:<br>  ▪ Prevention of perioperative and peripartum thromboembolism<br>  ▪ Prevention and treatment of thromboembolism |
| **Required Medical Information:** | **<u>All Indications</u>**<br>• Documented diagnosis of hATd, confirmed by antithrombin (AT) activity levels below 70% on functional assay (not taken during acute illness, surgery, or thromboembolic event that could give falsely low antithrombin levels) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Prevention of Perioperative Thromboembolism</u>**<br>• Approved first-line for perioperative thromboprophylaxis in combination with heparin, with or without intent to use as bridge to warfarin therapy<br><br>**<u>Prevention of Peripartum Thromboembolism</u>**<br>• Documentation of **ONE** of the following:<br>  ○ Personal or family history of thrombosis<br>  ○ Insufficient response to heparin **AND** intolerance to direct oral anticoagulants (DOACs)<br><br>**<u>Prevention of Thromboembolism</u>**<br>• Documentation of inadequate clinical response, intolerance, or contraindication to **BOTH** of the following:<br>  ○ Warfarin<br>  ○ At least one DOAC<br><br>**<u>Treatment of Thromboembolism</u>**<br>• Approved first-line for treatment of thromboembolism as adjunct to anticoagulant therapy, unless coagulation is temporarily contraindicated |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist, geneticist, or obstetrician<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • **Perioperative/peripartum prevention; thromboembolism treatment:** 1 month, unless otherwise specified<br>• **Thromboembolism prevention:** 6 months, unless otherwise specified |

POLICY NAME:
**ANTI-AMYLOID MONOCLONAL ANTIBODY**
Affected Medications: LEQEMBI (lecanemab), LEQEMBI-IQLIK (lecanemab-irmb), KISUNLA (donanemab-azbt)

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| **Covered Uses:** | • Leqembi (lecanemab), Leqembi-Iqlik (lecanemab-irmb), and Kisunla (donanemab-azbt) are not considered medically necessary due to insufficient evidence of therapeutic value |
| **Required Medical Information:** | |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | |
| **Coverage Duration:** | |

# APOC-III TARGETED AGENTS

## Scope & Exclusions

### Included Indications:
All Food and Drug Administration (FDA)–approved indications not otherwise excluded by plan design.

### Prescriber Limits:
Prescribed by, or in consultation with, a cardiologist, endocrinologist, gastroenterologist, or specialist in lipid disorders.

## Initial Authorization Criteria

### Required Medical Information:

1. All indications must be FDA-supported for the requested product
2. Requested dosing must be according to the FDA label based on diagnosis, age, and weight.

### Familial Chylomicronemia Syndrome (FCS)
**Group 1 Drugs:** Redemplo
**Group 2 Drugs:** Tryngolza

1. Fasting triglyceride level of at least 880 mg/dL refractory to standard triglyceride therapies
2. Documentation of following a low-fat diet with less than 20 grams of fat per day
3. Group 1 Drugs require ONE of the following:
    a. History of acute pancreatitis
    b. History of recurrent abdominal pain without other known cause
    c. Genetic testing showing a pathogenic gene mutation in LPL, APOC2, APOA5, GPIHBP1, or LMF1 genes
    d. Group 2 drugs require genetic testing showing a pathogenic gene mutation in LPL, APOC2, APOA5, GPIHBP1, or LMF1 genes AND treatment failure to Group 1 Drugs.

## Renewal (Continuation) Criteria

1. Documentation of reduction in triglyceride levels from baseline

## Duration of Approval

Initial: 6 months
Renewal: 24 months

POLICY NAME:
**APOMORPHINE**
Affected Medications: APOKYN, APOMORPHINE SOLUTION

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Acute, intermittent treatment of hypomobility, "off" episodes in patients with advanced Parkinson's disease (PD) |
| **Required Medical Information:** | • Diagnosis of advanced PD<br>• Documentation of acute, intermittent hypomobility, "off" episodes occurring for at least 2 hours per day while awake despite an optimized treatment regimen |
| **Appropriate Treatment Regimen & Other Criteria:** | • Established on a stable dose of carbidopa-levodopa with intent to continue<br>• Documented treatment failure with concurrent use of levodopa-carbidopa and a second agent from one of the following classes:<br>  ○ Catechol-O-methyltransferase (COMT) inhibitors (e.g., entacapone)<br>  ○ Dopamine agonists (e.g., pramipexole, ropinirole)<br>  ○ Monoamine oxidase-B (MAO-B) inhibitors (e.g., selegiline, rasagiline)<br><br>**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Use as monotherapy or first line agent |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**APROCITENTAN**
Affected Medications: TRYVIO (aprocitentan)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of hypertension in combination with other antihypertensive drugs |
| **Required Medical Information:** | • Diagnosis of resistant hypertension<br>• Blood pressure remains above target goal (as determined by treating provider) despite adherence to antihypertensive therapies<br>• Documentation of intent to use as an adjunct to current antihypertensive therapies |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with concurrent use of at least four antihypertensive drugs (from different drug classes) at maximum tolerated doses, for a minimum of 12 weeks:<br>  ○ Angiotensin-converting enzyme (ACE) inhibitor OR angiotensin II receptor blocker (ARB)<br>  ○ Calcium channel blocker (e.g. amlodipine, nifedipine, diltiazem, verapamil)<br>  ○ Diuretic (e.g. hydrochlorothiazide, chlorthalidone)<br>  ○ Beta-blocker (e.g. atenolol, carvedilol)<br>  ○ Mineralocorticoid receptor antagonist (e.g. spironolactone, eplerenone)<br><br>**Reauthorization** requires documentation of treatment success and continued use of at least three background blood pressure therapies |
| **Exclusion Criteria:** | • Pregnancy<br>• Concurrent use with an endothelin receptor antagonist (e.g. ambrisentan, bosentan, Opsumit, Filspari) |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist, nephrologist, or endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ARIKAYCE**
Affected Medications: ARIKAYCE (Amikacin inhalation suspension)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of *Mycobacterium avium* complex (MAC) lung disease as part of a combination antibacterial drug regimen in adults who have limited or no alternative treatment options, and who do not achieve negative sputum cultures after a minimum of 6 consecutive months of a multidrug background regimen therapy |
| **Required Medical Information:** | • Diagnosis of MAC lung disease confirmed by BOTH of the following:<br>  ○ A MAC-positive sputum culture obtained within the last 3 months<br>  ○ Evidence of underlying nodular bronchiectasis and/or fibrocavity disease on a chest radiograph or chest computed tomography<br>• The MAC isolate is susceptible to amikacin with a minimum inhibitory concentration (MIC) of less than or equal to 64 µg/mL<br>• Documentation of failure to obtain a negative sputum culture after a minimum of 6 consecutive months of a multidrug background regimen therapy for MAC lung disease such as clarithromycin (or azithromycin), rifampin and ethambutol |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of BOTH of the following:<br>  ○ This drug has been prescribed as part of a combination antibacterial drug regimen<br>  ○ This drug will be used with the Lamira® Nebulizer System<br><br>**<u>Reauthorization</u>** requires documentation of negative sputum culture obtained within the last 30 days.<br><br>• The American Thoracic Society/Infectious Diseases Society of America (ATS/IDSA) guidelines state that patients should continue to be treated until they have negative cultures for 1 year. Treatment beyond the first reauthorization (after 18 months) will require documentation of a positive sputum culture to demonstrate the need for continued treatment. Patients that have had negative cultures for 1 year will not be approved for continued treatment. |
| **Exclusion Criteria:** | • Diagnosis of non-refractory MAC lung disease |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an infectious disease specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ASCIMINIB**
Affected Medications: SCEMBLIX (asciminib)

<table>
<tr><td>Covered Uses:</td><td>• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better</td></tr>
<tr><td>Required Medical Information:</td><td>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• Documentation of Philadelphia chromosome positive (Ph+) or BCR::ABL1- positive chronic myeloid leukemia (CML) in chronic phase (May be appropriate in some cases of advanced phase CML- Check NCCN guidelines)</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td><u>Philadelphia chromosome or BCR::ABL1- positive chronic myeloid leukemia (CML) in chronic phase (CP) meeting one of the following:</u><br><br><u>Low Risk Score</u><br>• Documented treatment failure with imatinib (if used as initial tyrosine kinase inhibitor [TKI]) AND one or more additional tyrosine kinase inhibitor (TKI) bosutinib, dasatinib, or nilotinib. (Note BCR::ABL1 kinase domain mutation status for drug specific contraindications)<br><br><u>Intermediate or high-risk score</u><br>• Documented treatment failure with a second-generation tyrosine kinase inhibitor (TKI), bosutinib, dasatinib, or nilotinib. (Note BCR:ABL1 kinase domain mutation status for drug specific contraindications)<br><br><table><tr><th>Drug</th><th>Contraindicated Mutations</th></tr><tr><td>Asciminib</td><td>A337T, P465S, M244V, or F359V/I/C</td></tr><tr><td>Bosutinib</td><td>T315I, V299L, G250E, or F317L</td></tr><tr><td>Dasatinib</td><td>T315I/A, F317L/V/I/C, or V299L</td></tr><tr><td>Nilotinib</td><td>T315I, Y253H, E255K/V, or F359V/C/I</td></tr><tr><td>Ponatinib</td><td>None</td></tr></table><br>OR<br>• Documented T315I positive mutation<br>AND<br>• Documented treatment failure with ponatinib<br><br><u>Reauthorization</u> requires documentation of disease responsiveness to therapy</td></tr>
<tr><td>Exclusion Criteria:</td><td>• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• Presence of either A337T, P465S, M244V, or F359V/I/C BCR::ABL1 kinase domain mutation</td></tr>
<tr><td>Age Restriction:</td><td></td></tr>
<tr><td>Prescriber/Site of Care Restrictions:</td><td>• Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care</td></tr>
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| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ATIDARSAGENE AUTOTEMCEL**
Affected Medications: LENMELDY (atidarsagene autotemcel)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Treatment of children with pre-symptomatic late-infantile (PSLI), pre-symptomatic early-juvenile (PSEJ), or early symptomatic early-juvenile (ESEJ) metachromatic leukodystrophy (MLD) |
| **Required Medical Information:** | • Diagnosis of metachromatic leukodystrophy (MLD) confirmed by the following:<br>  ○ Arylsulfatase (ARSA) activity below the normal range in peripheral blood mononuclear cells or fibroblasts<br>  ○ Presence of two disease-causing mutations of either known or novel alleles<br>  ○ Presence of sulfatides in a 24-hour urine collection (to exclude MLD carriers and patients with ARSA pseudodeficiency)<br>AND<br>• Diagnosis of the late-infantile subtype of MLD confirmed by two out of three of the following:<br>  ○ Age at onset of symptoms in the older sibling(s) less than or equal to 30 months<br>  ○ Two null (0) mutant ARSA alleles<br>  ○ Peripheral neuropathy as determined by electroneurographic study<br>OR<br>• Diagnosis of the early-juvenile subtype of MLD confirmed by two out of three of the following:<br>  ○ Age at onset of symptoms (in the patient or in the older sibling) between 30 months and 6 years (has not celebrated their seventh birthday)<br>  ○ One null (0) and one residual (R) mutant ARSA allele(s)<br>  ○ Peripheral neuropathy as determined by electroneurographic study |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | • Allogeneic hematopoietic stem cell transplantation in the previous six months<br>• Previous gene therapy<br>• Documented HIV infection<br>• Documented history of a hereditary cancer |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with a neurologist or hematologist/oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 2 months (for one time infusion), no reauthorization, unless otherwise specified |

POLICY NAME:
**AVACOPAN**
Affected Medications: TAVNEOS 10mg capsule

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ As an adjunctive treatment of adult patients with severe, active anti-neutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis (AAV), including granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA), in combination with standard therapy including glucocorticoids |
| **Required Medical Information:** | • Diagnosis supported by at least one of the following:<br>  ○ Tissue biopsy of kidney or other affected organs<br>  ○ Positive ANCA, clinical presentation compatible with AAV, and low suspicion for secondary vasculitis<br>  ○ Clinical presentation compatible with AAV, low suspicion for secondary vasculitis, and concern for rapidly progressive disease<br>• Documented severe, active disease (including major relapse), defined as: vasculitis with life- or organ-threatening manifestations (e.g., alveolar hemorrhage, glomerulonephritis, central nervous system vasculitis, subglottic stenosis, mononeuritis multiplex, cardiac involvement, mesenteric ischemia, limb/digit ischemia)<br>• Documentation of all prior therapies used and anticipated treatment course<br>• Baseline liver test panel: serum alanine aminotransferase, aspartate aminotransferase, alkaline phosphatase, and total bilirubin<br>• Current hepatitis B virus (HBV) status |
| **Appropriate Treatment Regimen & Other Criteria:** | • Will be used with a standard immunosuppressive regimen including glucocorticoids<br>• Will be used during induction therapy only<br>• Will be used in any of the following populations/scenarios:<br>  ○ In patients unable to use glucocorticoids at appropriate doses<br>  ○ In patients with an estimated glomerular filtration rate less than 30 mL/min/1.73 m2<br>  ○ In patients who have experienced relapse following treatment with two or more different induction regimens, including both rituximab- and cyclophosphamide-containing regimens (unless contraindicated)<br>  ○ During subsequent induction therapy in patients with refractory disease (failure to achieve remission with initial induction therapy regimen)<br>• Dosing: 30 mg (three 10 mg capsules) twice daily (once daily when used concomitantly with strong CYP3A4 inhibitors)<br>• Reauthorization: must meet criteria above (will not be used for maintenance treatment) |
| **Exclusion Criteria:** | • Treatment of eosinophilic-GPA (EGPA)<br>• Active, untreated and/or uncontrolled chronic liver disease (e.g., chronic active hepatitis B, untreated hepatitis C virus infection, uncontrolled autoimmune hepatitis) and cirrhosis<br>• Active, serious infections, including localized infections<br>• History of angioedema while receiving Tavneos, unless another cause has been established |

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| | • History of HBV reactivation while receiving Tavneos, unless medically necessary |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a rheumatologist, nephrologist, or pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 6 months with no reauthorization, unless otherwise specified |

POLICY NAME:
**AVALGLUCOSIDASE ALFA-NGPT**
Affected Medications: NEXVIAZYME (avalglucosidase alfa-ngpt)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Late-Onset Pompe Disease |
| **Required Medical Information:** | • Diagnosis of Pompe Disease confirmed by an enzyme assay demonstrating a deficiency of acid α-glucosidase (GAA) enzyme activity or by DNA testing that identifies mutations in the GAA gene.<br>• Patient weight and planned treatment regimen. |
| **Appropriate Treatment Regimen & Other Criteria:** | • One or more clinical signs or symptoms of Late-Onset Pompe Disease:<br>  ○ Progressive proximal weakness in a limb-girdle distribution<br>  ○ Delayed gross-motor development in childhood<br>  ○ Involvement of respiratory muscles causing respiratory difficulty (such as reduced forced vital capacity [FVC] or sleep disordered breathing)<br>  ○ Skeletal abnormalities (such as scoliosis or scapula alata)<br>  ○ Low/absent reflexes<br>• Appropriate medical support is readily available when medication is administered in the event of anaphylaxis, severe allergic reaction, or acute cardiorespiratory failure.<br>• Patients weighing less than 30 kilograms will require documented treatment failure or intolerable adverse event to Lumizyme.<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced.<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy. |
| **Exclusion Criteria:** | • Diagnosis of infantile-onset Pompe Disease<br>• Concurrent use of other enzyme replacement therapies such as Lumizyme or Pombiliti and Opfolda |
| **Age Restriction:** | • 1 year of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a metabolic specialist, endocrinologist, biochemical geneticist, or physician experienced in the management of Pompe disease<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**AVATROMBOPAG**
Affected Medications: DOPTELET (avatrombopag), DOPTELET Sprinkle (avatrombopag)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Thrombocytopenia in adult patients with chronic liver disease (CLD) who are scheduled to undergo a procedure<br>  ○ Thrombocytopenia in patients at least 1 year of age with chronic immune thrombocytopenia (ITP) who have had an insufficient response to a previous treatment |
| **Required Medical Information:** | **<u>Thrombocytopenia in patients with CLD undergoing a procedure</u>**<br>• Documentation of planned procedure including date<br>• Documentation of baseline platelet count of less than 50,000/microliter<br><br>**<u>Thrombocytopenia in patients with chronic ITP</u>**<br>• Documentation of **ONE** of the following:<br>  ○ Platelet count less than 20,000/microliter<br>  ○ Platelet count less than 30,000/microliter AND symptomatic bleeding<br>  ○ Platelet count less than 50,000/microliter AND increased risk for bleeding (such as peptic ulcer disease, use of antiplatelets or anticoagulants, history of bleeding at higher platelet count, need for surgery or invasive procedure) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Thrombocytopenia in patients with chronic (ITP):</u>**<br>• Documentation of inadequate response, defined as platelets did not increase to at least 50,000/microliter, to the following therapies:<br>  ○ **ONE** of the following:<br>  ▪ Inadequate response with at least 2 therapies for immune thrombocytopenia, including corticosteroids, rituximab, or immunoglobulin<br>  ▪ Splenectomy<br>  ○ Eltrombopag olamine<br><br>**<u>Reauthorization (chronic ITP only)</u>**<br>• Response to treatment with platelet count of at least 50,000/microliter (not to exceed 400,000/microliter)<br>**OR**<br>• The platelet counts have not increased to at least 50,000/microliter and the patient has NOT been on the maximum dose for at least 4 weeks |
| **Exclusion Criteria:** | • Use in combination with another thrombopoietin receptor agonist, spleen tyrosine kinase inhibitor, or similar treatments (Eltrombopag olamine, Nplate, Tavalisse) |
| **Age Restriction:** | • Doptelet tablets: 18 years of age and older<br>• Doptelet sprinkle: 1 year of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist or gastroenterology/liver specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **Thrombocytopenia in patients with CLD undergoing a procedure**:<br>• Authorization: 1 month (for a one time 5-day regimen), unless otherwise specified<br>**Thrombocytopenia in patients with chronic ITP**:<br>• Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**AXATILIMAB-CSFR**
Affected Medications: NIKTIMVO (axatilimab-csfr)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Chronic graft-versus-host disease (cGVHD)<br>  ○ NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | • Diagnosis of cGVHD following hematopoietic stem cell transplantation (HSCT)<br>• Documentation of refractory or recurrent active cGVHD<br>• Patient weight and planned treatment regimen |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with one from each category at maximally indicated doses:<br>  ○ Prednisone or methylprednisolone<br>  ○ Jakafi (ruxolitinib)<br>  ○ Imbruvica (ibrutinib) or Rezurock (belumosudil)<br><br>**Dosing** is in accordance with FDA labeling and does not exceed 0.3 mg/kg (maximum of 35 mg) every 2 weeks |
| **Exclusion Criteria:** | • Concurrent use with Jakafi, Imbruvica, or Rezurock<br>• Patient weight of less than 40 kg<br>• Platelet count of less than 50 x $10^9$/L<br>• Absolute neutrophil count of less than 1 × $10^9$/L<br>• ALT and AST greater than 2.5 times the upper limit of normal<br>• Total bilirubin greater than 1.5 times the upper limit of normal<br>• Creatinine clearance less than 30 mL/minute |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist or oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**AZTREONAM**
Affected Medications: CAYSTON (aztreonam)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Cystic fibrosis |
| **Required Medical Information:** | • Documentation of confirmed diagnosis of cystic fibrosis<br>• Culture and sensitivity report confirming presence of Pseudomonas aeruginosa in the lungs<br>• Baseline FEV1 greater than 25% but less than 75% predicted |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented failure, contraindication, or resistance to inhaled tobramycin.<br>• **Dosing:** 28 days on and 28 days off<br>**Reauthorization** requires documentation of improved respiratory symptoms and confirmed need for long-term use |
| **Exclusion Criteria:** | • Baseline FEV1 less than 25% or greater than 75% predicted |
| **Age Restriction:** | • Age 7 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**BCR-ABL TYROSINE KINASE INHIBITORS - SECOND GENERATION**
Affected Medications: TASIGNA (nilotinib capsules), DANZITEN (nilotinib tablets), DASATINIB, BOSULIF (bosutinib)

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | • Documentation of performance status, all prior therapies used, and prescribed treatment regimen<br>• Documentation Philadelphia chromosome or BCR::ABL1-positive mutation status |
| **Appropriate Treatment Regimen & Other Criteria:** | • For patients with chronic phase Chronic Myeloid Leukemia (CP-CML) and low-risk score:<br>o Documented clinical failure with imatinib<br><br>**Bosulif, Danziten**<br>• Coverage for Bosulif and Danziten requires documented treatment failure or intolerable adverse event with both dasatinib and nilotinib (Tasigna)<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy (as applicable, BCR-ABL1 transcript levels, cytogenetic response) |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist.<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**BELIMUMAB**
Affected Medications: BENLYSTA (belimumab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Systemic Lupus Erythematosus (SLE)<br>  ○ Lupus Nephritis (LN) |
| **Required Medical Information:** | • Documentation of current weight (intravenous requests only)<br><br>**Systemic Lupus Erythematosus:**<br>• Documentation of active SLE with moderate classification (significant but non-organ threatening disease including constitutional, cutaneous, musculoskeletal, or hematologic involvement)<br>• Autoantibody-positive SLE, defined as positive for antinuclear antibodies (ANA) and/or anti-double-stranded DNA (anti-dsDNA) antibody<br>• Baseline measurement of **ONE** or more of the following:<br>  ○ SLE Responder Index-4 (SRI-4), SLE Activity Index (SLEDAI) variant, or other validated scale<br>  ○ Frequency of flares requiring corticosteroid use<br><br>**Lupus Nephritis:**<br>• Documentation of biopsy-proven active Class III, IV, and/or V disease<br>• Baseline measurement of one or more of the following: urine protein-creatinine ratio (uPCR), urine protein, estimated glomerular filtration rate (eGFR), or frequency of flares or corticosteroid use |
| **Appropriate Treatment Regimen & Other Criteria:** | **All uses:**<br>• Use of intravenous formulation requires:<br>  ○ Documented inability to use subcutaneous formulation OR<br>  ○ Currently receiving treatment with the intravenous formulation, excluding via samples or manufacturer's patient assistance programs<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced (intravenous requests only)<br><br>**Systemic Lupus Erythematosus:**<br>• Failure with at least 12 weeks of combination therapy including hydroxychloroquine OR chloroquine with one of the following:<br>  ○ Cyclosporine, azathioprine, methotrexate, or mycophenolate mofetil<br><br>**Reauthorization** requires documentation of treatment success defined as **ONE** of the following:<br>• Clinically significant improvement in SRI-4, SLEDAI variant, or other validated scale for measurement of disease<br>• Decrease in frequency of flares or corticosteroid use<br><br>**Lupus Nephritis:** |

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| | • No dialysis in the past 12 months AND estimated glomerular filtration rate (eGFR) equal to or above 30 mL/min/1.73m$^2$<br>• Failure of at least 12 weeks of mycophenolate mofetil AND cyclophosphamide<br><br>**<u>Reauthorization</u>** requires documentation of treatment success defined as ONE of the following:<br>• Improvement in eGFR<br>• Reduction in urinary protein-creatinine ratio or urine protein<br>• Decrease in flares or corticosteroid use |
| **Exclusion Criteria:** | • Use in combination with other biologic therapies for LN or SLE<br>• Use in severe active central nervous system lupus |
| **Age Restriction:** | • 5 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a nephrologist, rheumatologist, or specialist with experience in the treatment of systemic lupus erythematosus or lupus nephritis<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**BELZUTIFAN**
Affected Medications: WELIREG (belzutifan)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | **<u>Von Hippel-Lindau (VHL) disease</u>**<br>• Diagnosis documented by the following:<br>  ○ Pathogenic VHL germline mutation diagnostic for VHL disease AND at least one of the following:<br>  ▪ Presence of solid, locoregional tumor in kidney showing accelerated tumor growth (growth of 5 mm or more per year)<br>  ▪ Presence of symptomatic and/or progressively enlarging central nervous system (CNS) hemangioblastomas not amenable to surgery<br>  ▪ Presence of pancreatic solid lesion or pancreatic neuroendocrine tumor (pNET) with rapid tumor growth<br><br>**<u>Treatment-refractory advanced or metastatic clear cell renal carcinoma</u>**<br>• Advanced disease after use of the following treatments (per NCCN guidelines):<br>  ○ A programmed death receptor-1 (PD-1) OR programmed death-ligand 1 (PD-L1) AND<br>  ○ A vascular endothelial growth factor tyrosine kinase inhibitor (VEGF-TKI)<br><br>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Reauthorization:</u>** documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• Metastatic pNET disease<br>• Not to be used in combination with other oncologic agents for the treatment of VHL disease |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**BEREMAGENE GEPERPAVEC-SVDT**
Affected Medications: VYJUVEK (beremagene geperpavec-svdt)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Dystrophic Epidermolysis Bullosa (DEB) |
| **Required Medical Information:** | • Diagnosis of recessive DEB confirmed by both of the following:<br>  ○ Skin biopsy of an induced blister with immunofluorescence mapping (IFM) and/or transmission electron microscopy (TEM)<br>  ○ Genetic test results documenting mutations in the COL7A1 gene<br>• Clinical signs and symptoms of DEB such as skin fragility, blistering, scarring, nail changes, and milia formation in the areas of healed blistering |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of receiving standard of care preventative or treatment therapies for wound care, control of infection, nutritional support<br>• Documented trial and failure of Filsuvez<br>• **Dosing** is in accordance with FDA labeling and does not exceed the following:<br>  ○ Maximum weekly volume of 2 mL<br>  ○ Maximum of 12-week course per wound<br>  ○ Maximum of 4 tubes per 28 days<br><br>**Reauthorization** will require documentation of treatment success defined as complete wound healing on a previous site and need for treatment on a new site |
| **Exclusion Criteria:** | • Evidence or history of squamous cell carcinoma in the area that will undergo treatment<br>• Concurrent use with Filsuvez (birch triterpenes topical gel)<br>• Dominant DEB (DDEB) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a dermatologist or a specialist experienced in the treatment of epidermolysis bullosa<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 3 months, unless otherwise specified |

POLICY NAME:
**BESREMI**
Affected Medications: BESREMI (ropeginterferon alfa-2b)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved or compendia supported indications not otherwise excluded by plan design<br>  ○ Polycythemia vera<br>  ○ Essential thrombocythemia<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | **<u>Polycythemia vera</u>**<br>• Diagnosis of polycythemia vera confirmed by all major criteria (1-3) OR the first 2 major criteria (1-2) plus the minor criterion:<br>  ○ **Major criteria:**<br>  (1). Elevated hemoglobin concentration (greater than 16 g/dL), elevated hematocrit (greater than 48 percent), or increased red blood cell mass (greater than 25% above mean normal predicted value)<br>  (2). Presence of *JAK2* V617F **or** *JAK2* exon 12 mutation<br>  (3). Bone marrow biopsy showing age-adjusted hypercellularity with trilineage proliferation (panmyelosis), including prominent erythroid, granulocytic, and increase in pleomorphic, mature megakaryocytes without atypia. May not be required in patients with sustained absolute erythrocytosis (hemoglobin over 18.5 g/dL and hematocrit over 55.5 percent in men; hemoglobin over 16.5 g/dL and hematocrit over 49.5 percent in women) with presence of a *JAK2* V617F or *JAK2* exon 12 mutation.<br>  ○ **Minor criterion:** Subnormal serum erythropoietin level.<br><br>**<u>Essential Thrombocythemia</u>**<br>• Diagnosis of essential thrombocythemia, confirmed by all major criteria (1-4) **OR** the first 3 major criteria (1-3) plus the minor criterion:<br>  ○ **Major criteria:**<br>  (1). Platelet count greater than or equal to 450,000 cells/mcL.<br>  (2). Bone marrow biopsy showing proliferation mainly of the megakaryocytic lineage, with hyperlobulated staghorn-like nuclei, infrequently dense clusters; no significant increase or left shift in neutrophil granulopoiesis or erythropoiesis; no relevant bone marrow fibrosis.<br>  (3). Diagnostic criteria for BCR::ABL1-positive chronic myeloid leukemia, polycythemia vera, primary myelofibrosis, or other neoplasms are not met.<br>  (4). Presence of *JAK2*, *CALR*, or *MPL* mutation.<br>  ○ **Minor criterion:** Presence of another clonal marker (e.g., *ASXL1*, *EZH2*, *TET2*, *IDH1/IDH2*, *SRSF2*, or *SRF3B1* mutation) **OR** no identifiable cause for thrombocytosis (such as iron deficiency, chronic infection, chronic inflammatory disease, prior splenectomy). |

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| | **<u>Oncology Indications</u>**<br>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Polycythemia Vera</u>**<br>• Documentation of treatment failure, intolerance, or contraindication to hydroxyurea<br><br>**<u>Essential Thrombocythemia</u>**<br>• Documented treatment failure, intolerance, or contraindication to both of the following: hydroxyurea and peginterferon alfa-2a (Pegasys)<br><br>**<u>Reauthorization</u>** requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist or hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**BETAINE**
Affected Medications: CYSTADANE (betaine), BETAINE

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Homocystinuria |
| **Required Medical Information:** | • Diagnosis of homocystinuria associated with one of the following:<br>  ○ Cystathionine beta-synthase (CBS) deficiency<br>  ○ 5,10-methylenetetrahydrofolate reductase (MTHFR) deficiency<br>  ○ Cobalamin cofactor metabolism (cbl) defect<br>• Baseline plasma homocysteine levels |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented trial and failure of **ONE** of the following forms of supplementation:<br>  ○ Vitamin B6 (pyridoxine)<br>  ○ Vitamin B9 (folate)<br>  ○ Vitamin B12 (cobalamin)<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy shown by lowering of plasma homocysteine levels |
| **Exclusion Criteria:** | • Uncorrected vitamin B12 or folic acid levels |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a metabolic or genetic disease specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**BETIBEGLOGENE AUTOTEMCEL**
Affected Medications: ZYNTEGLO (betibeglogene autotemcel)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of beta thalassemia in adult and pediatric patients who require regular red blood cell (RBC) transfusions |
| **Required Medical Information:** | • Documented diagnosis of transfusion dependent beta thalassemia (TDT), defined as:<br>  ○ Requiring at least 100 mL/kg per year of packed red blood cells (pRBCs) or at least 8 transfusions per year of pRBCs in the 2 years preceding therapy<br>  ○ Confirmed genetic testing based on the presence of biallelic mutations at the beta-globin gene (*HBB* gene)<br>• Clinically stable and eligible to undergo hematopoietic stem cell transplant (HSCT)<br>• Used as single agent therapy (not applicable to lymphodepleting or bridging therapy while awaiting manufacture)<br>• Females of reproductive potential must have negative pregnancy test prior to start of mobilization, reconfirmed prior to conditioning procedures, and again before administration of Zynteglo |
| **Appropriate Treatment Regimen & Other Criteria:** | • Patients must weigh a minimum of 6 kilograms and be able to provide a minimum number of cells (5,000,000 CD34+ cells/kilogram) |
| **Exclusion Criteria:** | • Prior HSCT or other gene therapy<br>• Severe iron overload warranting exclusion from therapy, as determined by the treating physician<br>• Uncorrected bleeding disorder<br>• Cardiac T2* less than 10 milliseconds by magnetic resonance imaging (MRI)<br>• White blood cell count less than $3x10^9$/L and/or platelet count less than $100x10^9$/L that is unrelated to hypersplenism<br>• Positive for human immunodeficiency virus 1 & 2 (HIV-1/HIV-2), hepatitis B virus, or hepatitis C virus, advanced liver disease, or current or prior malignancy |
| **Age Restriction:** | • 4 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months (one-time infusion), unless otherwise specified |

POLICY NAME:
**BEVACIZUMAB**
Affected Medications: AVASTIN, MVASI, ZIRABEV, ALYMSYS, VEGZELMA, JOBEVNE

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher<br>• For the Treatment of Ophthalmic disorders:<br>  ○ Neovascular (Wet) Age-Related Macular Degeneration (AMD)<br>  ○ Macular Edema Following Retinal Vein Occlusion (RVO)<br>  ○ Diabetic Macular Edema (DME)<br>  ○ Diabetic Retinopathy (DR) in patients with Diabetes Mellitus |
| **Required Medical Information:** | • Documentation of disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | **Stage III or IV Epithelial Ovarian, Fallopian Tube, or Primary Peritoneal Cancer following initial surgical resection**<br>• Approval will be limited for up to 22 cycles of therapy<br><br>**All Indications**<br>• Coverage for a non-preferred product (Avastin, Alymsys, Vegzelma, Jobevne) requires documentation of one of the following:<br>  ○ Use for an ophthalmic condition (Avastin only)<br>  ○ A documented intolerable adverse event to the preferred products, Mvasi and Zirabev, and the adverse event was not an expected adverse event attributed to the active ingredient<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Oncologic indication: prescribed by, on in consultation with, an oncologist<br>• Ophthalmic indication: prescribed by, on in consultation with, an ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**BIRCH TRITERPENES**
Affected Medications: FILSUVEZ (birch triterpenes topical gel)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Dystrophic Epidermolysis Bullosa (DEB)<br>  ○ Junctional Epidermolysis Bullosa (JEB) |
| **Required Medical Information:** | • Diagnosis of recessive DEB or JEB confirmed by skin biopsy of an induced blister with immunofluorescence mapping (IFM) and/or transmission electron microscopy (TEM)<br>• Genetic test results documenting mutations in one of the following genes: COL7A1, COL17A1, ITGB4, LAMA3, LAMB3, or LAMC2<br>• Clinical signs and symptoms of EB such as skin fragility, blistering, scarring, nail changes, and milia formation in the areas of healed blistering<br>• Presence of open partial-thickness wounds that have been present for at least 21 days |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of receiving standard of care preventative or treatment therapies for wound care, control of infection, nutritional support.<br>• **Dosing** does not exceed the following:<br>  ○ Maximum of 1 mm layer to affected area(s)<br>  ○ Maximum of 28 tubes per 28 days<br><br>**Reauthorization** requires documentation of treatment success defined as complete wound healing on a previous site and need for continued treatment on a new site |
| **Exclusion Criteria:** | • Concurrent use with Vyjuvek (beremagene geperpavec-svdt)<br>• Dominant DEB (DDEB) |
| **Age Restriction:** | • 6 months of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a dermatologist or a specialist experienced in the treatment of epidermolysis bullosa<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 3 months, unless otherwise specified |

# BOTULINUM TOXINS

Included Products: Botox, Dysport, Xeomin, Myobloc, Daxxify

## Scope & Exclusions

### Included Indications:

Coverage limited to FDA-approved and select medically necessary non-cosmetic indications. Drug Compendia supported indications may be covered.

**Botox** is approved first-line for: focal dystonia, hemifacial spasm, orofacial dyskinesia, upper/lower limb spasticity, or other conditions of focal spasticity wherein botulinum toxin is the preferred mode of therapy.
**Dysport** is approved first-line for: focal dystonia, hemifacial spasm, drug-induced orofacial dyskinesia, upper or lower limb spasticity.
**Xeomin** is approved first-line for: cervical dystonia, blepharospasm, upper limb spasticity, chronic sialorrhea.

### Exclusions:

Use in cosmetic procedures (e.g., facial rhytides, glabellar lines, platysmal bands)

Use for Temporomandibular joint disorder

Combined use with a calcitonin gene-related peptide (CGRP) inhibitor for the prevention of migraine.

### Prescriber limits:

Prescribed by, or in consult with, an appropriate specialist for the indication.

## Initial Authorization Criteria

### Required Medical Information:

1. All indications must be FDA-supported for the requested product or strongly supported in drug compendia (e.g. LexiComp or DRUGDEX).
2. Requested dosing must be according to the FDA label or medical literature/treatment guidelines based on diagnosis and age.
3. Requests for non-preferred products require inadequate response, intolerance, or an FDA-labeled *and* patient-specific contraindication to **all** preferred products as outlined within indication-specific criteria.
   FDA-labeled contraindications are defined as those listed within the package insert. Allergic reactions may be accepted as contraindicated when an ingredient found only in the preferred product is established as the causative agent.

### Achalasia (Cardiospasm)

**Preferred Drug:** Botox

1. Treatment failure to an interventional therapy (peroral endoscopic myotomy (POEM), laparoscopic Heller myotomy (LHM), or pneumatic dilation (PD)) or not a candidate for interventional therapy due to high risk of complications.

## Anal Fissure

**Preferred Drug:** Botox

1. Inadequate response or intolerance to both:
    a. Rectiv ointment
    b. Topical diltiazem OR topical nifedipine

## Cervical Dystonia

**Preferred Drugs:** Botox, Dysport, and Xeomin
**Nonpreferred Drug:** Myobloc, Daxxify

## Chronic Migraine

**Preferred Drug:** Botox

1. Patient experiences at least 15 headache days per month.
2. Inadequate response or intolerance to an adequate trial of at least 1 standard therapy listed below (or contraindication to all):
    a. Candesartan
    b. Antiepileptics (topiramate, valproic acid, divalproex sodium)
    c. Beta-Blockers
    d. Anti-depressants (amitriptyline, nortriptyline, venlafaxine, duloxetine)
3. Attestation that Botox will not be used in combination with an anti-calcitonin gene-related peptide (CGRP) monoclonal antibody or an oral CGRP antagonist for migraine prevention.

## Chronic Sialorrhea

**Preferred Drug:** Botox
**Nonpreferred Drug:** Myobloc

1. Documentation of treatment failure with glycopyrrolate oral tablets

## Overactive Bladder (OAB)/Neurogenic Detrusor Overactivity (NDO)

**Preferred Drug:** Botox

1. Inadequate response or intolerance to at least two urinary incontinence anticholinergic agents (e.g., oxybutynin, solifenacin, tolterodine)

## Primary Axillary Hyperhidrosis

**Preferred Drug:** Botox
**Nonpreferred Drug:** Myobloc

1. Inadequate response or intolerance to at least two alternative therapies:
    a. Topical aluminum chloride 20%
    b. Iontophoresis
    c. Oral glycopyrrolate
    d. Oral oxybutynin

## Duration of Approval

**Anal Fissure:** 3 months (one treatment), unless otherwise specified.
**Chronic Migraine:** 12-month initial approval, 24-month reauthorization unless otherwise specified.
**OAB/NDO:** 3-month initial approval, 12-month reauthorization unless otherwise specified.
**All Other Indications:** 12 months, unless otherwise specified.

POLICY NAME:
**BUROSUMAB**
Affected Medications**:** CRYSVITA (burosumab-twza)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ X-linked hypophosphatemia (XLH)<br>  ○ FGF23-related hypophosphatemia in tumor induced osteomalacia (TIO) associated with phosphaturic mesenchymal tumors |
| **Required Medical Information:** | **<u>All Indications</u>**<br>• Documentation of diagnosis by:<br>  ○ A blood test demonstrating ALL of the following (in relation to laboratory reference ranges):<br>  ▪ Low phosphate<br>  ▪ Elevated FGF23<br>  ▪ Low 1,25-(OH)2D<br>  ▪ Normal calcium or parathyroid hormone (PTH)<br>  ○ A urine test demonstrating decreased tubular reabsorption of phosphate corrected for glomerular filtration rate (TmP/GFR)<br>  ○ Evidence of skeletal abnormalities, confirmed by radiographic evaluation<br><br>**<u>Tumor-Induced Osteomalacia</u>**<br>• Documentation that tumor cannot be located or is unresectable<br>• Alternative renal phosphate-wasting disorders have been ruled out |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>All Indications</u>**<br>• Documentation of treatment failure with at least 12 months of oral phosphate and calcitriol supplementation in combination, unless contraindicated or not tolerated<br><br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**<u>Reauthorization</u>** requires:<br>• Documentation of normalization of serum phosphate levels<br>• If established on therapy for 12 months or more, improvement in radiographic imaging of skeletal abnormalities |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber Restrictions:** | • Prescribed by, or in consultation with, a nephrologist, endocrinologist, or a provider experienced in managing patients with metabolic bone disease<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**CALCIFEDIOL**
Affected Medications: RAYALDEE (calcifediol extended-release)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of secondary hyperparathyroidism in adult patients with stage 3 or 4 chronic kidney disease (CKD) and serum total 25-hydroxyvitamin D levels less than 30 ng/mL |
| **Required Medical Information:** | • A confirmed diagnosis of secondary hyperparathyroidism with persistently elevated or progressively rising serum intact parathyroid hormone (iPTH) that is at least 2.3 times above the upper limit of normal for the assay used<br>• Documentation of all of the following prior to treatment initiation:<br>  ○ Stage 3 or 4 CKD<br>  ○ Serum total 25-hydroxyvitamin D level is less than 30 ng/mL<br>  ○ Corrected serum calcium is below 9.8 mg/dL |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of persistent vitamin D deficiency (level below 30 ng/mL), despite at least 12 weeks of adherent treatment with each of the following at an appropriate dose, unless contraindicated or not tolerated:<br>  ○ Vitamin D2 (ergocalciferol) or vitamin D3 (cholecalciferol)<br>  ○ Calcitriol<br>  ○ Doxercalciferol<br>  ○ Paricalcitol<br><br>**Reauthorization** will require documentation of a clinically significant response to therapy, evidenced by increased serum total 25-hydroxyvitamin D level (to at least 30 ng/mL) and reduced plasma iPTH to goal therapeutic range (or an approximate 30% reduction compared to baseline) |
| **Exclusion Criteria:** | • A diagnosis of stage 1, 2, or 5 chronic kidney disease, or end-stage renal disease (ESRD) on dialysis |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a nephrologist or endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

# CALCITONIN GENE-RELATED PEPTIDE (CGRP) INHIBITORS

## Scope & Exclusions

### Included Indications:
All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.

### Excluded Uses:
Combined use with Botox for the prevention of migraine.
Combined use with another calcitonin gene-related peptide (CGRP) inhibitor (acute or preventative).

## Initial Authorization Criteria

1. All indications must be FDA-supported for the requested product.
2. Requested dosing must be according to the FDA label based on diagnosis.

### Preventative Treatment of Migraine
**Preferred Drugs:** Emgality
**Nonpreferred Drugs:** Vyepti

1. Diagnosis of migraine
2. Inadequate response or intolerance to an adequate trial of at least 1 standard therapy listed below (or contraindication to all) for migraine prophylaxis:
    a. Candesartan
    b. Antiepleptics (Topiramate, Valproic Acid, Divalproex Sodium)
    c. Beta-Blockers
    d. Anti-depressants (amitriptyline, nortriptyline, venlafaxine, duloxetine)

**Vyepti** requires inadequate response or intolerance to Emgality AND Botox

### Preventative Treatment of Episodic Cluster Headache
**Preferred Drug:** Emgality

1. History of episodic cluster headache with at least two cluster periods lasting from 7 days to 1 year (when untreated) separated by pain-free remission periods of at least one month
2. Documented treatment failure with verapamil (at least 480 mg daily for at least 3 weeks), or if unable to tolerate verapamil or contraindications apply, another oral preventative therapy (lithium, topiramate)

### Acute Treatment of Migraine
**Preferred Drug:** Ubrelvy

1. Diagnosis of migraine
2. Failure with BOTH of the following:
    a. One oral triptan, such as almotriptan, eletriptan, naratriptan, sumatriptan, rizatriptan, zolmitriptan, or frovatriptan tablets; rizatriptan or zolmitriptan oral-disintegrating tablet (ODT)
    b. One non-oral triptan, such as sumatriptan nasal spray or injection, zolmitriptan nasal spray

## Duration of Approval

Initial: 6 months, unless otherwise specified
Renewal: 24 months, unless otherwise specified

POLICY NAME:
**CANNABIDIOL**
Affected Medications: EPIDIOLEX (cannabidiol)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Lennox-Gastaut Syndrome (LGS)<br>  ○ Dravet Syndrome (DS)<br>  ○ Tuberous Sclerosis Complex (TSC) |
| **Required Medical Information:** | **All Indications**<br>• Patient weight<br>• Documentation that cannabidiol will be used as adjunctive therapy<br>• Baseline seizure type and seizure frequency |
| **Appropriate Treatment Regimen & Other Criteria:** | **LGS**<br>• Documented treatment failure with at least **two** antiepileptic drugs (e.g. valproate, lamotrigine, rufinamide, topiramate, felbamate, clobazam)<br>• Dosing not to exceed 20 mg/kg per day<br><br>**DS**<br>• Documented treatment failure with at least **two** antiepileptic drugs (e.g. valproate, clobazam, topiramate, levetiracetam)<br>• Dosing not to exceed 20 mg/kg per day<br><br>**TSC**<br>• Documented treatment failure with at least **two** antiepileptic drugs<br>• Dosing not to exceed 25 mg/kg per day<br><br>**Reauthorization** requires documentation of treatment success and a reduction in seizure severity, frequency, and/or duration. |
| **Exclusion Criteria:** | • Use as monotherapy for seizure control |
| **Age Restriction:** | • 1 year of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**CAPLACIZUMAB-YHDP**
Affected Medications: CABLIVI (caplacizumab-yhdp)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of adult and pediatric patients 12 years of age and older with acquired thrombotic thrombocytopenic purpura (aTTP), in combination with plasma exchange and immunosuppressive therapy |
| **Required Medical Information:** | • Diagnosis, or suspected diagnosis, of aTTP, meeting all of the following:<br>  ○ Severe thrombocytopenia (platelet count less than 100 x $10^9$/L)<br>  ○ Microangiopathic hemolytic anemia (MAHA) confirmed by red blood cell fragmentation (e.g., schistocytes) on peripheral blood smear<br>  ○ Baseline ADAMTS13 activity level of less than 10%<br>• Documentation of **ONE** of the following:<br>  ○ Failure of at least one initial treatment for aTTP, such as therapeutic plasma exchange (TPE), glucocorticoids, or rituximab<br>  ○ Documentation of high-risk disease meeting **ONE** of the following:<br>  ▪ Neurologic abnormalities (seizures, focal weakness, aphasia, dysarthria, confusion, coma)<br>  ▪ Altered mental status<br>  ▪ Elevated serum troponin levels<br>• Documentation that Cablivi will be used in combination with standard-of-care treatment for aTTP (TPE and glucocorticoid) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Total treatment duration will be limited to 58 days beyond the last TPE treatment<br><br>**Reauthorization** requires documented signs of ongoing disease (such as suppressed ADAMTS13 activity levels) and no more than 2 recurrences of aTTP while on Cablivi. Recurrence is defined as thrombocytopenia after initial recovery of platelet count (platelet count greater than or equal to 150,000) that requires re-initiation of daily plasma exchange. |
| **Exclusion Criteria:** | • Use for other causes of thrombocytopenia, such as other TTP-like disorders (congenital or hereditary TTP) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 3 months (for new episode), unless otherwise specified |

POLICY NAME:
**CAPSAICIN KIT**
Affected Medications: QUTENZA (capsaicin kit)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Neuropathic pain associated with postherpetic neuralgia (PHN)<br>  ○ Neuropathic pain associated with diabetic peripheral neuropathy (DPN) of the feet |
| **Required Medical Information:** | |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with at least 12 weeks of ALL of the following:<br>  ○ gabapentin<br>  ○ pregabalin<br>  ○ carbamazepine, oxcarbazepine, or valproic acid/divalproex sodium<br>  ○ amitriptyline or nortriptyline<br>  ○ topical lidocaine<br>• Dose limited to a single treatment (up to 4 patches) once every 90 days<br><br>**Reauthorization:** requires documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a pain management specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months (single treatment), unless otherwise specified<br>• Reauthorization: 12 months (up to 4 treatments), unless otherwise specified |

POLICY NAME:
**CARGLUMIC ACID**
Affected Medications: CARBAGLU, CARGLUMIC ACID

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Acute hyperammonemia due to one of the following:<br>  ▪ N-Acetylglutamate Synthase (NAGS) deficiency<br>  ▪ Propionic Acidemia (PA) or Methylmalonic Acidemia (MMA)<br>  ○ Chronic hyperammonemia due to N-Acetylglutamate Synthase (NAGS) deficiency |
| **Required Medical Information:** | • Diagnosis is confirmed by enzymatic, biochemical, or genetic testing<br>• Ammonia level above the upper limit of normal (ULN) reference range for the patient's age |
| **Appropriate Treatment Regimen & Other Criteria:** | • Current weight<br><br>**Acute hyperammonemia**<br>• Prescribed in combination with at least one other ammonia-lowering therapy (examples include: sodium phenylacetate and sodium benzoate, intravenous glucose, insulin, L-arginine, L-carnitine, protein restriction, dialysis)<br>• For disease due to PA or MMA: Prescribed treatment course does not exceed 7 days<br><br>**Reauthorization** for acute disease requires documentation of reoccurrence of acute hyperammonemia meeting initial criteria<br><br>**Chronic hyperammonemia due to N-Acetylglutamate Synthase (NAGS) deficiency**<br>• Prescribed in combination with a protein-restricted diet<br><br>**Reauthorization** for chronic disease requires:<br>• Documentation of treatment success and a clinically significant response to therapy as evidenced by reduction in ammonia levels<br>• Documentation of member's current weight and continuation of appropriate treatment course |
| **Exclusion Criteria:** | • Hyperammonemia caused by other enzyme deficiencies in the urea cycle:<br>  ○ Carbamyl phosphate synthetase I (CPSI) deficiency<br>  ○ Ornithine transcarbamylase (OTC) deficiency<br>  ○ Argininosuccinate synthetase (ASS) deficiency<br>  ○ Argininosuccinate lyase (ASL) deficiency<br>  ○ Arginase deficiency<br>• Chronic treatment (use beyond 7 days) of acute or chronic hyperammonemia due to MMA or PA |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a metabolic disease specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **Acute Hyperammonemia due to PA or MMA:**<br>• Authorization: 7 days, unless otherwise specified |

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| | **<u>Acute Hyperammonemia due to NAGs deficiency:</u>**<br>• Authorization: 1 month, unless otherwise specified<br><br>**<u>Chronic Hyperammonemia:</u>**<br>• Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**CERLIPONASE ALFA**
Affected Medications: BRINEURA (cerliponase alfa)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ To slow the loss of ambulation in pediatric patients with neuronal ceroid lipofuscinosis type 2 (CLN2), also known as tripeptidyl peptidase-1 (TPP1) deficiency |
| **Required Medical Information:** | • Diagnosis of CLN2 disease confirmed by BOTH of the following:<br>  ○ Enzyme assay demonstrating deficient TPP1 activity<br>  ○ Genetic testing that has detected two pathogenic variants/mutations in the TPP1/CLN2 gene (one on each parental allele of the TPP1/CLN2 gene)<br>• Documentation of mild to moderate functional impairment at baseline using the CLN2 Clinical Rating Scale, defined as ALL the following:<br>  ○ Combined score of 3 to 6 in the motor and language domains<br>  ○ Score of at least 1 in the motor domain<br>  ○ Score of at least 1 in the language domain |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing is in accordance with FDA labeling<br><br>**<u>Reauthorization</u>:**<br>• Documentation of clinical responsiveness to therapy defined as disease stabilization OR a score of at least 1 in the motor domain of the CLN2 Clinical Rating Scale |
| **Exclusion Criteria:** | • Any sign or symptom of acute or unresolved localized infection on or around the device insertion site (e.g., cellulitis or abscess); or suspected or confirmed CNS infection (e.g., cloudy CSF or positive CSF gram stain, or meningitis)<br>• Any acute intraventricular access device-related complication (e.g., leakage, extravasation of fluid, or device failure)<br>• Other forms of neuronal ceroid lipofuscinosis<br>• Patients with ventriculoperitoneal shunts |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist with expertise in the diagnosis of CLN2<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 6 months, unless otherwise specified |

POLICY NAME:
**CFTR MODULATORS**
Affected Medications: ALYFTREK (vanzacaftor/tezacaftor/deutivacaftor), KALYDECO (ivacaftor), ORKAMBI (lumacaftor/ivacaftor), SYMDEKO (tezacaftor/ivacaftor), TRIKAFTA (elexacaftor/tezacaftor/ivacaftor)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Cystic fibrosis (CF) in patients with mutation(s) in the F508del cystic fibrosis transmembrane conductance regulator (CFTR) gene or another responsive mutation in the CFTR gene<br>  ○ CF in patients who are homozygous for the F508del mutation in the CFTR gene (Orkambi) |
| **Required Medical Information:** | • Documentation of cystic fibrosis (CF) diagnosis confirmed by appropriate genetic or diagnostic testing (FDA-approved CF mutation test)<br>  ○ Please provide the diagnostic testing report and/or Cystic Fibrosis Foundation Patient Registry Report<br>• Documentation of mutation(s) in the CFTR gene for which the drug has been FDA-approved to treat |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** will require documentation of treatment success |
| **Exclusion Criteria:** | • Kalydeco: Homozygous F508del mutation<br>• Concurrent use with another CFTR modulator |
| **Age Restriction:** | • Alyftrek: 6 years of age and older<br>• Kalydeco: one month of age and older<br>• Orkambi: 1 year of age and older<br>• Symdeko: 6 years of age and older<br>Trikafta: 2 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a pulmonologist or provider who specializes in CF<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 12 months, unless otherwise specified<br>• Reauthorization: 24 months unless otherwise specified |

POLICY NAME:
**CHELATING AGENTS**
Preferred drugs: deferasirox soluble tablet, deferasirox tablet
Non-Preferred drugs: Ferriprox (deferiprone), deferiprone

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| 1. Is the request for continuation of therapy currently approved through insurance? | Yes – Go to renewal criteria | No – Go to #2 |
| 2. Is the request to treat a diagnosis according to one of the Food and Drug Administration (FDA)-approved indications? | Yes – Go to appropriate section below | No – Criteria not met |
| **Chronic Iron Overload Due to Blood Transfusions in Myelodysplastic Syndromes**<br>**Preferred Drugs –** deferasirox soluble tablet, deferasirox tablet<br>**Non -Preferred drugs:** Ferriprox (deferiprone), deferiprone | | |
| 1. Documentation of International Prognostic Scoring System (IPSS) low or intermediate-1 risk level? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Documentation of a history of more than 20 red blood cell (RBC) transfusions OR that it is anticipated that more than 20 would be required? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Documentation of serum ferritin levels greater than 2500 ng/ml? | Yes – Document and go to # 4 | No – Criteria not met |
| 4. Is the request for generic formulation of deferasirox (oral or soluble tablet)? | Yes – Go to #6 | No- Go to #5 |
| 5. Is there documented failure to deferasirox and deferoxamine (Desferal)? | Yes – Document and go to #6 | No – Criteria not met |
| 6. Is the drug prescribed by, or in consultation with, a hematologist specialist? | Yes – Go to #7 | No – Criteria not met |
| 7. Is the requested dose within the Food and Drug Administration (FDA) approved label? | Yes – Approve up to 12 months | No – Criteria not met |
| **Chronic Iron Overload Due to Blood Transfusions in Thalassemia syndromes, Sickle Cell Disease, or other anemias**<br>**Preferred Drugs –** deferasirox soluble tablet, deferasirox tablet<br>**Non -Preferred drugs:** Ferriprox (deferiprone), deferiprone | | |

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| 1. Documentation of pretreatment serum ferritin level within the last 60 days of at least 1000 mcg/L? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is the request for generic formulation of deferasirox (oral or soluble tablet)? | Yes – Document and go to #4 | No – Go to #3 |
| 3. Is there documented failure to deferasirox and deferoxamine (Desferal)? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Documentation of platelet counts greater than 50,000 per microliter? | Yes – Go to #5 | No – Criteria not met |
| 5. Is the drug prescribed by, or in consultation with, a hematologist specialist? | Yes – Document and go to #6 | No – Criteria not met |
| 6. Is the requested dose within the Food and Drug Administration (FDA) approved label? | Yes – Approve up to 12 months | No – Criteria not met |
| **Chronic Iron Overload in Non-Transfusion Dependent Thalassemia Syndromes**<br>**Preferred Drugs –**deferasirox soluble tablet, deferasirox tablet | | |
| 1. Documentation of liver iron (Fe) concentration (LIC) levels consistently greater than or equal to 5 mg Fe per gram of dry weight | Yes – Document and go to #2 | No – Criteria not met |
| 2. Documentation of serum ferritin levels consistently greater than 300 mcg/L prior to initiation of treatment | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is the requested dose within the Food and Drug Administration (FDA) approved label? | Yes – Approve up to 12 months | No – Criteria not met |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy defined as a reduction from baseline liver iron concentration (LIC) or serum ferritin level (LIC and serum ferritin must still be above 3 mg Fe per gram of dry weight and 500 mcg/L, respectively) | Yes – Go to #2 | No – Criteria not met |
| 2. Is the requested dose within the Food and Drug Administration (FDA)-approved label and PacificSource quantity limitations? | Yes – Approve up to 12 months | No – Criteria not met |

| Quantity Limitations |
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| • **Exjade (deferasirox soluble tablet) – available in 125mg, 250mg, 500mg tablets**<br>  ○ **20-40 mg/kg/day**<br>• **Jadenu (deferasirox tablet or granules) – available in 90mg, 180mg, 360mg tablets**<br>  ○ **14-28 mg/kg/day**<br>• **Ferriprox (deferiprone) – 100mg/ml oral solution, 500mg, 1000mg tablets**<br>  ○ **75-99 mg/kg/day**<br>  ○ **Can be used in adult and pediatric patients 8 years of age and older (tablets), or 3 years of age and older (solution)** |

POLICY NAME:
**CHENODIOL**
Affected Medications: CTEXLI (chenodiol)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Treatment of cerebrotendinous xanthomatosis (CTX) in adults. |
| **Required Medical Information:** | • Diagnosis of CTX (Bogaert-Scherer-Epstein syndrome) confirmed by genetic testing that detects pathogenic variants in the *CYP27A1* gene<br>• Documentation with all the following:<br>  ○ Plasma cholestanol 5 to 10 times greater than normal<br>  ○ Elevated urine bile alcohol levels (23s-pentol) |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires improvement or stabilization of cognitive function and decrease in cholestanol or urine bile alcohol levels compared to baseline |
| **Exclusion Criteria:** | • Combined use with Chenodal (chenodeoxycholic acid) or Cholbam (cholic acid) |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist, endocrinologist, or other metabolic specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**CHOLBAM**
Affected Medications: CHOLBAM (cholic acid)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of bile acid synthesis disorders due to single enzyme defects (SEDs)<br>  ○ Adjunctive treatment of peroxisomal disorders, including Zellweger spectrum disorders, in patients who exhibit manifestations of liver disease, steatorrhea, or complications from decreased fat-soluble vitamin absorption |
| **Required Medical Information:** | • Documentation of all prior therapies, patient weight and anticipated treatment course<br>• Baseline liver function tests (AST, ALT, GGT, ALP, total bilirubin, INR)<br><br>**Bile acid synthesis disorder**<br>• Diagnosis confirmed by assessment of serum or urinary bile acid levels using mass spectrometry (Fast Atom Bombardment ionization - Mass Spectrometry (FAB-MS) analysis)<br><br>**Peroxisomal disorders including Zellweger spectrum disorders**<br>• Diagnosis confirmed by clinical features, elevated very long-chain fatty acid (VLCFA) levels, peroxisomal biomarkers, genetic testing<br>• Prothrombin time (vitamin K), serum levels of vitamins A, D, and E<br>• Hepatic injury or at risk of liver injury (elevations in liver enzymes or atypical bile acids) OR<br>• If normal liver function tests, must show manifestations of liver disease, steatorrhea, or complications from decreased fat-soluble vitamin absorption |
| **Appropriate Treatment Regimen & Other Criteria:** | • Will not be used for treatment of extrahepatic manifestations (such as neurologic symptoms) of bile acid synthesis disorders<br><br>**Reauthorization** requires documentation of clinically significant improvement in liver function as determined by meeting TWO of the following criteria:<br>• Improvement in abnormal liver chemistries (AST, ALT, bilirubin)<br>• Reduction or stabilization of hepatic inflammation and fibrosis<br>• Reduced levels of the toxic C27-bile acid intermediates dihydroxycholestanoic acid (DHCA) and trihydroxycholestanoic acid (THCA) in plasma and urine<br>• Improvement in prothrombin time (as a result of improved vitamin K absorption) and serum levels of vitamins A, D, and E<br>• No evidence of cholestasis on liver biopsy<br>• Body weight increased or stabilized<br><br>• Treatment should be discontinued if liver function does not improve after 3 months of start of treatment |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |

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| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hepatologist, gastroenterologist, or metabolic specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**CHOLESTATIC LIVER DISEASE**
Affected Medications: BYLVAY (odevixibat), LIVMARLI (maralixibat)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Pruritus due to progressive familial intrahepatic cholestasis (PFIC)<br>  ○ Cholestatic pruritus in patients with Alagille syndrome (ALGS) |
| **Required Medical Information:** | • Documentation of experiencing moderate to severe pruritis associated with PFIC or ALGS<br>• Documentation of serum bile acid concentration above the upper limit of normal (ULN) reference range for the reporting laboratory<br><br>**PFIC**<br>• Documentation of confirmed molecular diagnosis of PFIC type 1 or type 2<br>  ○ Documentation of absence of ABCB11 gene variant if PFIC type 2<br><br>**ALGS**<br>• Documentation of ALGS confirmed by:<br>  ○ Genetic test detecting a JAG1 or NOTCH2 mutation **OR**<br>  ○ Liver biopsy and at least three clinical features:<br>  ▪ Chronic cholestasis<br>  ▪ Cardiac disease<br>  ▪ Ocular or skeletal abnormalities<br>  ▪ Characteristic facial features<br>  ▪ Renal and vascular disease |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of current weight and dosing in accordance with FDA labeling<br>• Documented treatment failure with **ALL** of the following for at least 30 days:<br>  ○ Rifampin<br>  ○ Ursodiol<br>  ○ Cholestyramine (or colesevelam if requesting for ALGS)<br><br>**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Prior hepatic decompensation events<br>• Decompensated cirrhosis (such as ALT or total bilirubin greater than 10-times the ULN)<br>• Concomitant liver disease (e.g., biliary atresia, liver cancer, non- PFIC related cholestasis)<br>• Prior liver transplant |
| **Age Restriction:** | • Age is in accordance with FDA labeling |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hepatologist or a specialist with experience in the treatment of PFIC or ALGS<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified |

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| | • Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**CIALIS**
Affected Medications: CIALIS (2.5 mg, 5 mg), TADALAFIL (2.5 mg, 5 mg)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of symptomatic benign prostatic hyperplasia (BPH)<br>  ○ Mental health diagnosis of erectile disorder (ED) meeting sexual dysfunction criteria |
| **Required Medical Information:** | **Benign Prostatic Hyperplasia**<br>• Documented diagnosis of benign prostatic hyperplasia (BPH)<br><br>**Mental Health Diagnosis of Erectile Dysfunction**<br>• Documentation of a mental health diagnosis of erectile dysfunction meeting the *Diagnostic and Statistical Manual of Mental Disorders*, Fifth Edition (DSM-5) criteria:<br>  ○ At least one of the three following symptoms must be experienced with 75% to 100% of occasions of sexual activity:<br>  ▪ Marked difficulty in obtaining an erection during sexual activity<br>  ▪ Marked difficulty in maintaining an erection until the completion of sexual activity<br>  ▪ Marked decrease in erectile rigidity<br>  ○ The above symptoms have persisted for a minimum duration of approximately 6 months<br>  ○ The above symptoms cause clinically significant distress in the individual<br>  ○ The sexual dysfunction is not attributable to any of the following:<br>  ▪ A nonsexual medical or psychiatric condition<br>  ▪ Severe relationship distress (e.g., partner violence)<br>  ▪ The effects of medication or other substance use<br>  ▪ Other clinically significant and relevant stressors |
| **Appropriate Treatment Regimen & Other Criteria:** | **Benign Prostatic Hyperplasia**<br>• Documented treatment failure with at least two of the following: alfuzosin, doxazosin, silodosin, finasteride, tamsulosin, terazosin<br><br>**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy<br><br>• Limited to 1 tablet per day |
| **Exclusion Criteria:** | • Erectile dysfunction unrelated to a mental health diagnosis of sexual dysfunction according to the DSM-5 diagnostic criteria |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Mental health diagnosis of erectile dysfunction: prescribed by, or in consultation with, a mental health provider<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**CLADRIBINE**
Affected Medications: MAVENCLAD (cladribine)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:<br>  ▪ Clinically isolated syndrome (CIS)<br>  ▪ Relapsing-remitting multiple sclerosis (RRMS)<br>  ▪ Active secondary progressive disease (SPMS) |
| **Required Medical Information:** | • Diagnosis confirmed with magnetic resonance imaging (MRI) per revised McDonald diagnostic criteria for MS<br>  ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with (or intolerance to) a minimum 12-week trial of at least two disease-modifying therapies for MS<br><br>**Reauthorization (one time only)** requires provider attestation of treatment success<br>• Eligible to initiate second treatment cycle 43 weeks after last dose was administered |
| **Exclusion Criteria:** | • Concurrent use of other disease-modifying medications indicated for the treatment of MS<br>• Current malignancy<br>• Human immunodeficiency virus (HIV) infection<br>• Active chronic infections (e.g., hepatitis, tuberculosis)<br>• Pregnancy<br>• Treatment beyond 2 years |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or MS specialist<br>• All approved are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 2 months, unless otherwise specified<br>• Reauthorization: 2 months, unless otherwise specified |

POLICY NAME:
**COAGADEX**
Affected Medications: COAGADEX (Factor X)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Indicated in children and adults with hereditary Factor X (FX) deficiency for:<br>  ▪ Routine prophylaxis to reduce frequency of bleeding episodes<br>  ▪ On-demand treatment and control of bleeding episodes<br>  ▪ Perioperative management of bleeding in mild, moderate, or severe disease |
| **Required Medical Information:** | **<u>All Indications</u>**<br>• Documented diagnosis of hereditary Factor X (FX) deficiency, confirmed by baseline plasma FX levels (FX:C) less than or equal to 10%<br>• Patient weight<br><br>**<u>Routine Prophylaxis</u>**<br>• Documented baseline frequency of bleeding episodes<br><br>**<u>Perioperative Management</u>**<br>• Documentation of scheduled procedure with intent to use Coagadex for perioperative management of bleeding episodes |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**<u>Reauthorization</u>**<br>• **Prophylaxis:** Reauthorization requires documentation of treatment plan and responsiveness to therapy, defined as a reduction in spontaneous bleeds requiring treatment<br>• **Treatment:** Reauthorization requires documentation of treatment plan, number of acute bleeds since last approval, and number of doses on-hand (not to exceed 6 total doses) |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • **Prophylaxis/On-demand:**<br>  ○ Initial Authorization: 3 months, unless otherwise specified<br>  ○ Reauthorization: 12 months, unless otherwise specified<br>• **Perioperative:**<br>  ○ Authorization: 1 month, unless otherwise specified |

POLICY NAME:
**COMPOUNDED MEDICATION**
Affected Medications: ALL COMPOUNDED MEDICATIONS

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | • All compounded ingredients must be submitted on the pharmacy claim |
| **Appropriate Treatment Regimen & Other Criteria:** | • Compounded medications will only be payable after **ALL** commercially available or formulary products have been exhausted<br>• In the case of payable claim, only compound ingredients that are covered on the applicable formulary will be reimbursed under this policy<br>  ○ Compounds above a certain dollar threshold will be stopped by the claim adjudication system |
| **Exclusion Criteria:** | • Compounds for experimental or investigational uses will not be covered<br>• Compounds containing non-Food and Drug Administration (FDA) approved ingredients will not be covered<br>• Compounded medications will not be covered when an Food and Drug Administration (FDA) approved, commercially available medication is on the market for treatment of requested condition |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 3 months, unless otherwise specified |

POLICY NAME:
**CONCIZUMAB**
Affected Medications: ALHEMO (concizumab-mtci)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Routine prophylaxis to prevent or reduce the frequency of bleeding episodes in adult and pediatric patients 12 years of age and older with:<br>  ▪ Hemophilia A (congenital factor VIII deficiency) with or without FVIII inhibitors<br>  ▪ Hemophilia B (congenital factor IX deficiency) with or without FIX inhibitors |
| **Required Medical Information:** | • Diagnosis of FVIII deficiency (hemophilia A) or FIX deficiency (hemophilia B)<br>• Documentation of baseline factor level less than 1% AND prophylaxis required<br>OR<br>• Baseline factor level 1% to 3% and a documented history of at least two episodes of spontaneous bleeding into joints<br>• Prescribed for routine prophylaxis to prevent or reduce the frequency of bleeding episodes<br>• Documentation if inhibitors present<br>• Number of bleeds in the past 3 months with severity and cause of bleed<br>• Documentation of current weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Prophylactic agents must be discontinued<br>• Documentation of planned treatment dose based on reasonable projections, current dose utilization, and disease severity<br><br>**Hemophilia A**:<br>• Documentation of treatment failure or contraindication to FVIII prophylaxis with 1 or more preferred therapies: Advate, Adynovate, Eloctate, Altuviiio, Kogenate FS, Kovaltry, Novoeight, Jivi (with bypassing agent if inhibitors present) OR Hemlibra<br><br>**Hemophilia B**:<br>• Documentation of treatment failure or contraindication to FIX prophylaxis with 1 or more preferred therapies: Rixubus, BeneFIX, Alprolix, Idelvion, Rebinyn (with bypassing agent if inhibitors present)<br><br>**<u>Reauthorization</u>:**<br>• Documentation of bleeding episodes (number and severity) showing reduction in spontaneous bleeds requiring treatment<br>• Documentation that Alhemo plasma concentration is above 200 ng/mL to decrease the risk of bleeding episodes<br>• Documentation of planned treatment dose, past treatment history, and titer inhibitor level to factor VIII and FIX as appropriate |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

# CONTINUOUS GLUCOSE MONITORS

## Scope & Exclusions

### Included Indications:

All Food and Drug Administration (FDA)–approved indications not otherwise excluded by plan design.

### Exclusions:

Type 2 diabetes not using insulin therapy

### Other Limits:

1. Continuous Glucose Monitoring System (CGM) coverage is limited to the prescription benefit unless otherwise specified by benefit design
2. Coverage for a CGM that is not Dexcom or Freestyle Libre requires current use of an insulin pump that is only compatible with a non-preferred continuous glucose monitor

## Initial Authorization Criteria

### Management of Insulin-dependent Diabetes

**Preferred Products:** Dexcom, Freestyle Libre

- The patient is currently using insulin therapy to manage their diabetes OR is experiencing gestational diabetes (with or without insulin use)

## Duration of Approval

2 years, unless otherwise specified

**POLICY NAME:**
**COPPER HISTIDINATE**
**Affected Medications:** ZYCUBO (copper histidinate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Treatment of Menkes disease (MD) in pediatric patients |
| **Required Medical Information:** | Diagnosis of Menkes Disease (MD) confirmed by ONE of the following:<br>• Biochemical (plasma catecholamine analysis, elevated DOPAC:DHPG and dopamine: norepinephrine ratios) or genetic confirmation of ATP7A mutation consistent with a severe MD phenotype<br>• Documentation of clinical signs and symptoms (including but not limited to characteristic Menkes kinky hair, skin abnormalities, progressive neurologic impairment, growth and developmental delays, and multisystem complications affecting the brain, bones, and connective tissues) that are strongly consistent with MD when biochemical or genetic results are pending, and delay would result in harm<br>• Documentation of age-appropriate dosing<br>  ○ Twice daily dosing may be approved for patients less than 1 year of age |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Reauthorization</u>**<br>• If not previously provided, genetic confirmation of ATP7A mutation consistent with a severe phenotype<br>• Positive therapeutic response through improved survival, attainment of developmental milestones, physical gains, and normalization of copper-related biomarkers<br>• For use beyond 3 years: documentation of continued benefit-to-risk |
| **Exclusion Criteria:** | • Use for other copper transport syndromes, such as occipital horn syndrome |
| **Age Restriction:** | |
| **Prescriber Restrictions:** | • Prescribed by or in consultation with neurologist or other specialty with experience treating MD<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization if genetic test results are pending: 3 months<br>• Authorization: 12 months, unless otherwise stated |

POLICY NAME:
**CORLANOR**
Affected Medications: CORLANOR (ivabradine), IVABRADINE

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Stable, symptomatic chronic heart failure with reduced ejection fraction in adult patients (adjunctive therapy)<br>  ○ Stable, symptomatic heart failure due to dilated cardiomyopathy (DCM) in pediatric patients 6 months and older<br>• Inappropriate sinus tachycardia |
| **Required Medical Information:** | **Chronic heart failure in adult patients**<br>• Documentation of chronic heart failure with left ventricular ejection fraction (LVEF) 35% or less AND<br>• Resting heart rate of at least 70 beats per minute (bpm)<br><br>**Heart failure in pediatric patients**<br>• Documentation of stable symptomatic disease due to DCM<br>• Currently in sinus rhythm with an elevated heart rate<br><br>**Inappropriate sinus tachycardia**<br>• Documented resting heart of at least 100 beats per minute, with a mean heart rate of at least 90 beats per minute over 24 hours, that is not due to appropriate physiologic response or primary abnormality (such as hyperthyroidism or anemia)<br>• Symptoms are present (such as palpitations, shortness of breath, dizziness, and/or decreased exercise capacity)<br>• Documented absence of identifiable causes of sinus tachycardia and exclusion of atrial tachycardia |
| **Appropriate Treatment Regimen & Other Criteria:** | **Chronic heart failure in adult patients**<br>• Documented treatment failure with a beta blocker (metoprolol succinate extended release, carvedilol, or carvedilol extended release) at the maximally tolerated dose for heart failure treatment OR<br>• Documentation of contraindication to beta-blocker use<br><br>**Heart failure in pediatric patients**<br>• Treatment failure with beta blocker or digoxin, or contraindication to beta blocker and digoxin use.<br><br>**All Indications**<br>• Requests for brand Corlanor tablets will require documentation of an adverse event with generic ivabradine tablets (and the adverse event was not an expected adverse event attributed to the active ingredient)<br>• Requests for Corlanor oral solution will require at least **ONE** of the following:<br>  ○ Request is for a pediatric patient<br>  ○ Request is for an adult patient who is unable to swallow tablets<br>  ○ Documentation of an adverse event with generic ivabradine tablets (and the adverse event was not an expected adverse event attributed to the active |

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| | ingredient)<br><br>**<u>Reauthorization</u>** requires documentation of treatment success and a clinically significant response to therapy<br>• Development of atrial fibrillation while on therapy will exclude patient from reauthorization |
| **Exclusion Criteria:** | • Acute, decompensated heart failure<br>• Blood pressure less than 90/50 mm Hg<br>• Sick sinus syndrome, sinoatrial block, third-degree atrioventricular block (unless stable with functioning demand pacemaker)<br>• Severe hepatic impairment (Child-Pugh class C)<br>• Heart rate maintained exclusively by pacemaker |
| **Age Restriction:** | • Heart failure due to DCM: 6 months to less than 18 years of age |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**COVERAGE OF SELECT HIGH INTENSITY STATINS AT TIER 0 COPAY**
Affected Medications: ATORVASTATIN (40 mg, 80 mg), ROSUVASTATIN (20 mg, 40 mg), SIMVASTATIN (80 mg)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Primary prevention of cardiovascular disease |
| **Required Medical Information:** | **<u>Primary prevention of cardiovascular disease (must meet all of the following):</u>**<br>• 40 to 75 years of age<br>• Presence of at least one cardiovascular risk factor such as:<br>  ○ Dyslipidemia<br>  ○ Diabetes<br>  ○ Hypertension<br>  ○ Smoking<br>• Estimated 10-year risk of cardiovascular event of at least 10% or higher |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**CRINECERFONT**
Affected Medications: CRENESSITY (crinecerfont)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>Congenital adrenal hyperplasia (CAH) |
| **Required Medical Information:** | • Confirmed diagnosis of classic CAH due to 21-hydroxylase deficiency (21-OHD) confirmed by one of the following<br>○ Elevated 17-hydroxyprogestone level<br>○ Confirmed cytochrome CYP21A2 genotype<br>○ Positive newborn screening with confirmatory second-tier testing (such as liquid chromatography tandem mass spectrometry)<br>○ Cosyntropin stimulation test<br>• Documentation of being used concurrently with a systemic glucocorticoid (such as hydrocortisone, prednisone, prednisolone, dexamethasone)<br>• Body surface area (BSA) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Requests for oral solution must have documented inability to swallow tablets<br>• Documentation of being on a supraphysiologic systemic glucocorticoid dose to control disease (total glucocorticoid dose of at least 10 mg/m$^2$/day in hydrocortisone dose equivalents)<br>• Dosing is in accordance with FDA labeling<br><br>**Reauthorization** requires documentation of treatment success defined by a reduction in serum androstenedione (A4) or reduction in glucocorticoid dose |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 4 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization:12 months, unless otherwise specified |

POLICY NAME:
**CRIZANLIZUMAB**
Affected Medications**:** ADAKVEO (crizanlizumab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ To reduce the frequency of vaso-occlusive crises (VOCs) in adults and pediatric patients aged 16 years and older with sickle cell disease |
| **Required Medical Information:** | • Diagnosis of sickle cell disease confirmed by genetic testing<br>• Two or more sickle cell-related crises in the past 12 months<br>• Therapeutic failure of 6-month trial on maximum tolerated dose of hydroxyurea or intolerable adverse event to hydroxyurea |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization** requires documentation of treatment success defined by a decrease in the number of vaso-occlusive crises |
| **Exclusion Criteria:** | • Long-term red blood cell transfusion therapy<br>• Hemoglobin is less than 4.0 g/dL<br>• Chronic anticoagulation therapy (such as warfarin, heparin) other than aspirin<br>• History of stroke within the past 2 years<br>• Combined use with Endari (L-glutamine) |
| **Age Restriction:** | • 16 years of age and older |
| **Prescriber Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**CROVALIMAB**
Affected Medications: PIASKY (crovalimab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Paroxysmal nocturnal hemoglobinuria (PNH) |
| **Required Medical Information:** | • Detection of PNH clones of at least 5% by flow cytometry diagnostic testing<br>  ○ Presence of at least 2 different glycosylphosphatidylinositol (GPI) protein deficiencies (e.g., CD55, CD59, etc.) within at least 2 different cell lines (e.g., granulocytes, monocytes, erythrocytes)<br>• Baseline lactate dehydrogenase (LDH) levels greater than or equal to 2 times the upper limit of normal range<br>• One of the following PNH-associated clinical findings:<br>  ○ Presence of a thrombotic event<br>  ○ Presence of organ damage secondary to chronic hemolysis<br>  ○ History of 4 or more blood transfusions required in the previous 12 months<br>• Body weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented inadequate response, contraindication, or intolerance to ravulizumab-cwvz (Ultomiris)<br>• Dosing is in accordance with FDA labeling and most recent body weight<br><br>**Reauthorization** requires documentation of treatment success defined as a decrease in serum LDH, stabilized/improved hemoglobin, decreased transfusion requirement, and reduction in thromboembolic events compared to baseline |
| **Exclusion Criteria:** | • Concurrent use with other biologics for PNH (Soliris, Ultomiris, Empaveli, Fabhalta)<br>• Current meningitis infection or other unresolved serious infection caused by encapsulated bacteria |
| **Age Restriction:** | • 13 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**CYSTARAN, CYSTADROPS**
Affected Medications: CYSTARAN SOLUTION 0.44 % OPHTHALMIC (cysteamine hydrochloride solution), CYSTADROPS SOLUTION 0.37% OPHTHALMIC (cysteamine hydrochloride solution)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ Ocular Cystinosis |
| **Required Medical Information:** | • Diagnosis of ocular cystinosis<br>• Documentation of slit-lamp examination showing corneal cystine crystal accumulation |
| **Appropriate Treatment Regimen & Other Criteria:** | • Reauthorization requires documentation of treatment success defined as reduction in cystine crystals compared to baseline |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**CYSTEAMINE**
Affected Medications: PROCYSBI (cysteamine bitartrate delayed release)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Nephropathic cystinosis |
| **Required Medical Information:** | • Diagnosis of nephropathic cystinosis confirmed by **ONE** of the following:<br>  ○ Molecular genetic testing showing mutations in the CTNS gene<br>  ○ Leukocyte cystine concentration above the laboratory reference range<br>  ○ Presence of cysteine corneal crystals by slit lamp examination |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure or intolerable adverse event with Cystagon |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**DANICOPAN**
Affected Medications: VOYDEYA (danicopan)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Treatment of extravascular hemolysis (EVH) in adults with paroxysmal nocturnal hemoglobinuria (PNH) |
| **Required Medical Information:** | • Patients must be administered a meningococcal vaccine at least two weeks prior to initiation of the requested therapy and revaccinated according to current Advisory Committee on Immunization Practices (ACIP) guidelines |
| **Appropriate Treatment Regimen & Other Criteria:** | • Must be used in combination with ravulizumab-cwvz (Ultomiris) or eculizumab (Soliris) [separate authorization required]<br>• Documentation of clinically significant extravascular hemolysis (EVH) defined as persistent anemia (Hgb less than or equal to 9.5 gram/deciliter) with absolute reticulocyte count greater than or equal to 120 x $10^9$/liter despite use of Ultomiris or Soliris for at least 6 months<br><br>**Reauthorization** requires documentation of treatment success defined as a decrease in serum LDH, stabilized/improved hemoglobin, decreased transfusion requirement, and reduction in thromboembolic events compared to baseline |
| **Exclusion Criteria:** | • Use without Ultomiris or Soliris<br>• Concurrent use with biologics for PNH other than Ultomiris and Soliris (such as pegcetacoplan or iptacopan)<br>• Current meningitis infection |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**DEFIBROTIDE**
Affected Medications: DEFITELIO (defibrotide sodium)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of adult and pediatric patients with hepatic veno-occlusive disease (VOD), also known as sinusoidal obstruction syndrome (SOS), with renal or pulmonary dysfunction following hematopoietic stem-cell transplantation (HSCT) |
| **Required Medical Information:** | • Diagnosis of, or high suspicion for, classical or late-onset hepatic VOD<br>• Weight prior to HSCT, dose, and frequency |
| **Appropriate Treatment Regimen & Other Criteria:** | • Requested dose within the FDA-approved label |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 2 months with no reauthorization, unless otherwise specified |

POLICY NAME:
**DEFLAZACORT**
Affected Medications: DEFLAZACORT

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Duchenne muscular dystrophy (DMD) in patients 2 years of age and older |
| **Required Medical Information:** | • Laboratory confirmation of Duchenne muscular dystrophy (DMD) diagnosis by genetic testing and serum creatinine kinase at least 10 times the upper limit of normal prior to starting treatment<br>• Baseline motor function assessment from one of the following:<br>  ○ 6-minute walk test<br>  ○ North Star Ambulatory Assessment (NSAA)<br>  ○ Motor Function Measure (MFM)<br>  ○ Hammersmith Functional Motor Scale (HFMS) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with a 6-month trial of prednisone, or intolerable adverse event causing one of the following:<br>  ○ Clinically significant weight gain defined as greater than or equal to 10% of body weight gain over a 6-month period<br>  ○ Psychiatric/behavioral issues (e.g., abnormal behavior, aggression, irritability) that persists beyond the first six weeks of prednisone treatment<br><br>**Reauthorization** requires a documented improvement from baseline or stabilization of motor function demonstrated by a motor function assessment tool |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 2 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 12 months, unless otherwise specified<br>• Reauthorization: 24 months, unless otherwise specified |

POLICY NAME:
**DELANDISTROGENE MOXEPARVOVEC-ROKL**
Affected Medications: ELEVIDYS (delandistrogene moxeparvovec-rokl)

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| **Covered Uses:** | • Some Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of ambulatory pediatric patients ages 4 and up with Duchenne muscular dystrophy (DMD) |
| **Required Medical Information:** | • Confirmed mutation of DMD gene between exons 18-58<br>• Documentation of being ambulatory without needing an assistive device such as a wheelchair, walker, or cane<br>• North Star Ambulatory Assessment (NSAA) scale total score of 17 or more<br>• Receiving physical and/or occupational therapy<br>• Baseline anti-AAVrh74 total binding antibody titer of less than 1:400 as measured by ELISA<br>• Current weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of being on a stable dose of an oral corticosteroid such as prednisone for at least 12-weeks, and will continue prior to and following Elevidys infusion, according to FDA approved labeling<br>• Does not exceed FDA approved dosing based on weight and maximum of 70 vials<br><br>Number of vials needed = patient body weight (kg) rounded to nearest number of vials |
| **Exclusion Criteria:** | • Exon 8 and/or exon 9 deletion in DMD gene<br>• Concomitant therapy or within the past 6 months with DMD-directed antisense oligonucleotides such as golodirsen, casimersen, viltolarsen, eteplirsen<br>• Current active infection<br>• Previous Elevidys treatment in their lifetime<br>• Acute liver disease or impaired liver function<br>• Treatment in non-ambulatory patients – at this time, this indication is not considered medically necessary due to insufficient available evidence of therapeutic value |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 1 month (one-time dose, no reauthorization), unless otherwise specified |

POLICY NAME:
**DENOSUMAB**
Affected Medications: PROLIA (denosumab), JUBBONTI (denosumab-bbdz), STOBOCLO (denosumab-bmwo), CONEXXENCE (denosumab-bnht), BILDYOS (denosumab-nxxp), OSPOMYV (denosumab-dssb), BOSAYA (denosumab-kyqq), ENOBY (denosumab-qbde)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ Osteoporosis/bone loss |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose: 60 mg once every 6 months |
| **Coverage Duration:** | • Initiation Authorization: 24 months, unless otherwise specified<br>• Reauthorization: 24 months, unless otherwise specified |

POLICY NAME:
**DIAZOXIDE CHOLINE**
Affected Medications: VYKAT XR (diazoxide choline)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded:<br>  ○ Hyperphagia in adults and pediatric patients 4 years of age and older with Prader-Willi syndrome (PWS) |
| **Required Medical Information:** | • Diagnosis of PWS confirmed by genetic testing<br>• Provider attests that patient experiences moderate to severe symptoms of hyperphagia related to PWS and provides documentation of associated symptoms (e.g. food-seeking behaviors)<br>• Caregiver has implemented and intends to continue strategies to establish a food-secure environment (e.g. locked food storage)<br>• Patient is able to swallow tablets whole<br>• Recent weight documentation (within 30 days) to ensure appropriate dose |
| **Appropriate Treatment Regimen & Other Criteria:** | **Dosing**: within FDA-approved label<br><br>**Reauthorization** will require documentation of the following:<br>• Patient has experienced an improvement in hyperphagic symptoms, such as a decrease in food-related aggression or manipulation, or lessened food preoccupation that interferes with normal daily activities, etc.<br>• Patient is adherent to therapy and able to successfully swallow the prescribed number of tablets daily<br>• Recent weight documentation (within 30 days) to ensure appropriate dose<br>• For adult patients, provider has determined that patient is likely to still benefit from therapy (i.e. patient has not entered the phase of symptom improvement sometimes observed in adulthood) |
| **Exclusion Criteria:** | • Use for eating disorders without PWS |
| **Age Restriction:** | • 4 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with an endocrinologist, psychiatrist, or specialist with experience in the treatment of PWS<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 6 months, unless otherwise specified |

POLICY NAME:
**DIFELIKEFALIN**
Affected Medications: KORSUVA (difelikefalin)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Chronic kidney disease-associated pruritus (CKD-aP) during hemodialysis (HD) |
| **Required Medical Information:** | • Documentation of chronic kidney disease confirmed by presence of kidney damage or decreased kidney function for three or more months<br>• Documentation of moderate to severe pruritus associated with HD<br>• Documentation of normal serum parathyroid hormone (PTH), phosphate, calcium, and magnesium levels<br>• Documentation of patient's current dry body weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of inadequate relief with trial of all of the following therapies (minimum 1 month trial each):<br>  ○ A topical agent (such as an emollient or analgesic)<br>  ○ An oral antihistamine (such as hydroxyzine or diphenhydramine)<br>  ○ Gabapentin or pregabalin<br><br>**Reauthorization** will require documentation of clinically significant improvement or stabilization in pruritus from baseline and continued hemodialysis use |
| **Exclusion Criteria:** | • Peritoneal dialysis<br>• Severe hepatic impairment |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a nephrologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**DOJOLVI**
Affected Medications: DOJOLVI (triheptanoin oral liquid)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ A source of calories and fatty acids for the treatment of pediatric and adult patients with molecularly confirmed long-chain fatty acid oxidation disorders |
| **Required Medical Information:** | • Diagnosis of long chain fatty acid oxidation disorder (LC-FAOD) confirmed by molecular genetic testing or enzyme assay<br>• Documentation of total prescribed daily caloric intake<br>• Documentation of severe disease as evidenced by one of the following:<br>  ○ Hypoglycemia after short periods of fasting<br>  ○ Evidence of functional cardiomyopathy with poor ejection fraction requiring ongoing management<br>  ○ Frequent severe major medical episodes requiring emergency room visits, acute care, or hospitalization (3 events within the past year, or 5 events within the past 2 years)<br>  ○ Elevated creatinine kinase (chronic or episodic) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of persistent symptoms despite dietary management and use of an over the counter (OTC) medium-chain triglyceride (MCT) product<br>• Dose not to exceed 35% of daily caloric intake<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Concurrent use of another medium chain triglyceride product |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist or provider experienced in the management of metabolic disorders<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**DONISLECEL**
Affected Medications: LANTIDRA (donislecel solution)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Diagnosis of type 1 diabetes for 5 or more years<br>• Documentation of inability to achieve target HbA1c despite adherence to intensive insulin management with all the following:<br>  ○ Multiple daily injections of prandial and basal insulin or on an insulin pump<br>  ○ Performing at least four blood glucose tests per day or using a continuous glucose monitor<br>• Documentation of 2 or more episodes of severe hypoglycemia (blood glucose level less than 50 mg/dL) in the past three years requiring assistance of another person with either an oral carbohydrate, intravenous glucose, or glucagon administration<br>• Documentation of hypoglycemia unawareness, defined by the absence of adequate autonomic symptoms during an episode of severe hypoglycemia |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires documentation of not achieving exogenous insulin independence within one year of infusion or within one year of losing independence from exogenous insulin (maximum of three infusions per lifetime) |
| **Exclusion Criteria:** | • Pregnancy<br>• Malignancy<br>• Active infection<br>• Previous kidney or pancreas transplant<br>• Prior portal vein thrombosis |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 3 months (single treatment), unless specified otherwise |

POLICY NAME:
**DORNASE ALFA**
Affected Medications: PULMOZYME (dornase alfa)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • The diagnosis of Cystic Fibrosis (CF) has been confirmed by appropriate diagnostic or genetic testing<br>• Additional testing should include evaluation of overall clinical lung status and respiratory function (e.g., pulmonary function tests, lung imaging, etc.) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Pulmozyme will be used in conjunction with standard therapies for cystic fibrosis<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 1 month of age or older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 24 months, unless otherwise specified |

POLICY NAME:
**DROXIDOPA**
Affected Medications: DROXIDOPA

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of orthostatic dizziness with symptomatic neurogenic orthostatic hypotension (nOH) caused by:<br>  ▪ Primary autonomic failure (Parkinson's disease [PD], multiple system atrophy [MSA], pure autonomic failure [PAF])<br>  ▪ Dopamine beta-hydroxylase deficiency<br>  ▪ Non-diabetic autonomic neuropathy |
| **Required Medical Information:** | • Diagnosis of nOH caused by one of the following:<br>  ○ Primary autonomic failure (such as PD, MSA, PAF)<br>  ○ Dopamine beta-hydroxylase deficiency<br>  ○ Non-diabetic autonomic neuropathy<br>• Documentation of severe symptomatic orthostatic hypotension, demonstrated by **both** of the following:<br>  ○ Minimum 20 mmHg decrease in systolic blood pressure **OR** minimum 10 mmHg decrease in diastolic blood pressure within 3 minutes of standing<br>  ○ Documentation of significant symptoms affecting activities of daily living |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure or intolerable adverse event with a minimum 30-day trial to both fludrocortisone and midodrine<br><br>**Reauthorization** requires documentation of treatment success as determined by treating provider |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or cardiologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 1 month, unless otherwise specified<br>• Reauthorization: 3 months, unless otherwise specified |

POLICY NAME:
**DUOPA**
Affected Medications: DUOPA (carbidopa-levodopa enteral suspension)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of motor fluctuations in patients with advanced Parkinson's disease (PD) |
| **Required Medical Information:** | • Documentation of all the following:<br>  ○ Diagnosis of advanced PD<br>  ○ Clear response to levodopa treatment with evidence of "On" periods<br>  ○ Persistent motor fluctuations with "Off" time occurring 3 hours or more per day while awake despite an optimized PD treatment regimen<br>  ○ Has undergone or has planned placement of a nasojejunal (NJ) tube for temporary administration of Duopa OR gastrostomy-jejunostomy (PEG-J) tube for long-term administration of Duopa |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with both of the following:<br>  ○ Oral levodopa/carbidopa<br>  ○ Two additional agents from different anti-PD drug classes:<br>  ▪ Monoamine oxidase-B (MAO-B) inhibitors (ex: selegiline, rasagiline)<br>  ▪ Dopamine agonists (ex: amantadine, pramipexole, ropinirole)<br>  ▪ Catechol-O-methyltransferase (COMT) inhibitors (ex: entacapone)<br><br>**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Atypical Parkinson's syndrome ("Parkinson's Plus" syndrome) or secondary Parkinson's<br>• Non-levodopa responsive PD<br>• Contraindication to percutaneous endoscopic gastro-jejunal (PEG-J) tube placement or long-term use of a PEG-J<br>• Concomitant use with nonselective MAO inhibitors or have recently (within 2 weeks) taken a nonselective MAO inhibitor |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**ECULIZUMAB**
Affected Medications: SOLIRIS (eculizumab), EPYSQLI (eculizumab-aagh), BKEMV (eculizumab-aeeb)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis<br>  ○ Atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy<br>  ○ Generalized myasthenia gravis (gMG) in adult and pediatric patients six years of age and older who are anti-acetylcholine receptor (AChR) antibody positive<br>  ○ Neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive |
| **Required Medical Information:** | **PNH**<br>• Detection of PNH clones of at least 5% by flow cytometry diagnostic testing<br>  ○ Presence of at least 2 different glycosylphosphatidylinositol (GPI) protein deficiencies (e.g., CD55, CD59, etc.) within at least 2 different cell lines (e.g., granulocytes, monocytes, erythrocytes)<br>• Baseline lactate dehydrogenase (LDH) levels greater than or equal to 1.5 times the upper limit of normal range<br>• One of the following PNH-associated clinical findings:<br>  ○ Presence of a thrombotic event<br>  ○ Presence of organ damage secondary to chronic hemolysis<br>  ○ History of 4 or more blood transfusions required in the previous 12 months<br><br>**aHUS**<br>• Clinical presentation of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury<br>• Patient shows signs of thrombotic microangiopathy (TMA) (e.g., changes in mental status, seizures, angina, dyspnea, thrombosis, increasing blood pressure, decreased platelet count, increased serum creatinine, increased LDH, etc.)<br>• ADAMTS13 activity level greater than or equal to 10%<br>• Shiga toxin E. coli related hemolytic uremic syndrome (ST-HUS) has been ruled out<br>• History of 4 or more blood transfusions required in the previous 12 months<br><br>**gMG**<br>• Diagnosis of gMG confirmed by **ONE** of the following:<br>  ○ A history of abnormal neuromuscular transmission test<br>  ○ A positive edrophonium chloride test<br>  ○ Improvement in gMG signs or symptoms with an acetylcholinesterase inhibitor<br>• Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV<br>• Positive serologic test for AChR antibodies<br>• Documentation of **ONE** of the following:<br>  ○ MG-Activities of Daily Living (MG-ADL) total score of 6 or greater<br>  ○ Quantitative Myasthenia Gravis (QMG) total score of 12 or greater<br><br>**NMOSD** |

<table>
<tr><td></td><td>
<ul>
<li>Diagnosis of seropositive aquaporin-4 immunoglobulin G (AQP4-IgG) NMOSD confirmed by all the following:
<ul>
<li>Documentation of AQP4-IgG-specific antibodies on cell-based assay</li>
<li>Exclusion of alternative diagnoses (such as multiple sclerosis)</li>
<li>At least ONE core clinical characteristic:
<ul>
<li>Acute optic neuritis</li>
<li>Acute myelitis</li>
<li>Acute area postrema syndrome (episode of otherwise unexplained hiccups or nausea/vomiting)</li>
<li>Acute brainstem syndrome</li>
<li>Symptomatic narcolepsy OR acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesion on magnetic resonance imaging (MRI) [see table below]</li>
<li>Acute cerebral syndrome with NMOSD-typical brain lesion on MRI [see table below]</li>
</ul></li>
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</ul>
<table>
<tr><th>Clinical presentation</th><th>Possible MRI findings</th></tr>
<tr><td>Diencephalic syndrome</td><td>• Periependymal lesion<br>• Hypothalamic/thalamic lesion</td></tr>
<tr><td>Acute cerebral syndrome</td><td>• Extensive periependymal lesion<br>• Long, diffuse, heterogenous, or edematous corpus callosum lesion<br>• Long corticospinal tract lesion<br>• Large, confluent subcortical or deep white matter lesion</td></tr>
</table>
</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td>
<u>PNH</u>
<ul>
<li>Documented inadequate response, contraindication, or intolerance to ravulizumab-cwvz (Ultomiris)</li>
</ul>
<u>aHUS</u>
<ul>
<li>Failure to respond to plasma therapy within 10 days
<ul>
<li>Trial of plasma therapy not required if one of the following is present:
<ul>
<li>Life-threatening complications of HUS such as seizures, coma, or heart failure</li>
<li>Confirmed presence of a high-risk complement genetic variant (e.g., CFH or CFI)</li>
</ul></li>
</ul></li>
<li>Documented inadequate response, contraindication, or intolerance to ravulizumab-cwvz (Ultomiris)</li>
</ul>
<u>gMG</u>
<ul>
<li>Documentation of one of the following:
<ul>
<li>Treatment failure with an adequate trial (one year or more) of at least 2 immunosuppressive therapies (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)</li>
<li>Has required three or more courses of rescue therapy (plasmapheresis/plasma exchange and/or intravenous immunoglobulin), while on at least one immunosuppressive therapy, over the last 12 months</li>
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|---|---|
| | • Documented inadequate response, contraindication, or intolerance to each of the following:<br>  ○ Efgartigimod-alfa (Vyvgart)<br>  ○ Ravulizumab-cwvz (Ultomiris)<br><br>**NMOSD**<br>• Documented inadequate response, contraindication, or intolerance to **ALL** of the following:<br>  ○ Rituximab (preferred products: Riabni, Ruxience)<br>  ○ Satralizumab-mwge (Enspryng)<br>  ○ Inebilizumab-cdon (Uplizna)<br>  ○ Ravulizumab-cwvz (Ultomiris)<br><br>**Reauthorization:**<br>• gMG: documentation of treatment success defined as an improvement in MG-ADL and QMG scores from baseline<br>• NMOSD: documentation of treatment success defined as the stabilization or improvement in neurological symptoms as evidenced by a decrease in acute relapses, Expanded Disability Status Scale (EDSS) score, hospitalizations, or plasma exchange treatments<br>• PNH: documentation of treatment success defined as a decrease in serum LDH, stabilized/improved hemoglobin, decreased transfusion requirement, and reduction in thromboembolic events compared to baseline<br>• aHUS: documentation of treatment success defined as a decrease in serum LDH, stabilized/improved serum creatinine, increased platelet count, and decreased plasma exchange/infusion requirement compared to baseline |
| **Exclusion Criteria:** | • Concurrent use with other disease-modifying biologics for requested indication, unless otherwise indicated by the FDA for combination use with Soliris<br>• Current meningitis infection |
| **Age Restriction:** | • PNH and NMOSD: 18 years of age and older<br>• gMG: 6 years of age and older<br>• aHUS: 2 months of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist<br>  ○ PNH: hematologist<br>  ○ aHUS: hematologist or nephrologist<br>  ○ gMG: neurologist<br>  ○ NMOSD: neurologist or neuro-ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**EDARAVONE**
Affected Medications: RADICAVA (edaravone), RADICAVA ORS

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Amyotrophic lateral sclerosis (ALS) |
| **Required Medical Information:** | • Documentation of "definite" or "probable" ALS diagnosis based on revised El Escorial (Airlie House) or Awaji criteria<br>• Disease duration of 2 years or less<br>• Normal respiratory function defined as percent-predicted forced vital capacity values (% FVC) of at least 80%<br>• Patient currently retains most activities of daily living (ADLs), defined as at least 2 points on all 12 items of the ALS functional rating scale-revised (ALSFRS-R) |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires both of the following:<br>• Documentation of treatment success, as determined by prescriber (e.g., retention of most ADLs)<br>• Patient is not dependent on invasive mechanical ventilation (e.g., intubation, tracheostomy) |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or provider with experience in treating ALS<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ELADOCAGENE EXUPARVOVEC-TNEQ**
Affected Medications: KEBILIDI (eladocagene exuparvovec-tneq)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Treatment of aromatic L-amino acid decarboxylase (AADC) deficiency |
| **Required Medical Information:** | • Diagnosis of AADC deficiency confirmed by genetic testing showing bilateral/biallelic mutations in the DDC gene<br>• Reduced AADC enzyme activity in plasma<br>• Cerebrospinal fluid (CSF) shows all of the following:<br>  ○ Reduced levels of 5-hydroxyindoleacetic acid (5-HIAA), homovanillic acid (HVA), and 3-methoxy-4-hydroxyphenylglycol (MHPG)<br>  ○ Elevated levels of 3-O-methyldopa (3-OMD), levodopa (L-Dopa), and 5-hydroxytryptophan (5-HTP)<br>  ○ Normal levels of pterins (neopterin and biopterin)<br>• Clinical symptoms of AADC deficiency such as movement disorders, hypotonia, autonomic dysfunction, and developmental delay<br>• Documented achieved skull maturity assessed by neuroimaging |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing is in accordance with FDA labeling |
| **Exclusion Criteria:** | • Prior gene therapy administration<br>• Anti-AAV2 neutralizing antibody titer over 1,200 folds |
| **Age Restriction:** | • 1 to 17 years of age |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or geneticist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 3 months (one-time infusion only), unless otherwise specified |

POLICY NAME:
**ELAGOLIX**
Affected Medications: ORILISSA (elagolix), ORIAHNN (elagolix/estradiol/norethindrone acetate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Moderate to severe endometriosis-associated pain (Orilissa)<br>  ○ Heavy menstrual bleeding associated with uterine leiomyomas (Oriahnn) |
| **Required Medical Information:** | **<u>Pain due to endometriosis</u>**<br>• Documentation of both of the following:<br>  ○ Diagnosis of moderate to severe pain associated with endometriosis<br>  ○ Attestation that patient is premenopausal<br><br>**<u>Heavy menstrual bleeding due to uterine leiomyomas</u>**<br>• Documentation of both of the following:<br>  ○ Diagnosis of heavy menstrual bleeding associated with uterine leiomyomas<br>  ○ Attestation that patient is premenopausal |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Pain due to endometriosis</u>**<br>• Documentation of a trial and inadequate relief (or contraindication) after at least 3 months of both of the following first-line therapies:<br>  ○ Nonsteroidal anti-inflammatory drugs (NSAIDs)<br>  ○ Continuous (no placebo pills) hormonal contraceptives<br><br>**<u>Reauthorization</u>** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • History of osteoporosis<br>• Pregnancy<br>• Severe (Child-Pugh Class C) hepatic impairment (Orilissa)<br>• Mild, moderate, and severe (Child-Pugh Class A, B, and C) hepatic impairment (Oriahnn) |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist in obstetrics/gynecology or reproductive endocrinology<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 18 months (Orilissa 150 mg once daily* and Oriahnn only), unless otherwise specified<br><br>• *Maximum treatment duration for Orilissa 150 mg once daily in patients with moderate hepatic impairment (Child-Pugh Class B) and Orilissa 200 mg twice daily is 6 months. Reauthorization not allowed |

POLICY NAME:
**ELAMIPRETIDE**
Affected Medications: FORZINITY (elamipretide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ To improve muscle strength in adult and pediatric patients with Barth syndrome weighing at least 30 kilograms |
| **Required Medical Information:** | • Diagnosis of Barth Syndrome confirmed by genetic testing that detects mutations in the TAZ gene or an elevated monolysocardiolipin:cardiolipin (MLCL:CL) ratio<br>• Documented weight of greater than or equal to 30 kilograms<br>• Documentation of an estimated glomerular filtration rate (eGFR) of at least 30mL/minute and that the patient is not on dialysis |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires documentation of improvement or continued benefit in muscle strength, fatigue, or other clinical symptoms of the disease |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist, endocrinologist, hematologist, geneticist, or neurologist.<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months<br>• Reauthorization: 6 months |

POLICY NAME:
**ELIVALDOGENE AUTOTEMCEL**
Affected Medications: SKYSONA (elivaldogene autotemcel)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Early, active cerebral adrenoleukodystrophy (CALD) in male patients |
| **Required Medical Information:** | • Confirmed diagnosis of CALD with all of the following:<br>  ○ Confirmed *ABCD1* gene mutation<br>  ○ Elevated very-long-chain fatty acid (VLCFA) values for ALL of the following:<br>  ▪ Concentration of C26:0<br>  ▪ Ratio of C24:0 to C22:0<br>  ▪ Ratio of C26:0 to C22:0<br>  ○ Neurologic function score (NFS) less than or equal to 1 (asymptomatic or mildly symptomatic disease)<br>  ○ Active central nervous system disease established by central radiographic review of brain magnetic resonance imaging (MRI) demonstrating both of the following:<br>  ▪ Gadolinium enhancement on MRI of demyelinating lesions<br>  ▪ Loes scores between 0.5 and 9 on the 34-point scale |
| **Appropriate Treatment Regimen & Other Criteria:** | • Coverage of Skysona is provided if the patient does not have access to a hematopoietic stem cell transplant with a matched sibling donor<br><br>**Approved for one-time single infusion only** |
| **Exclusion Criteria:** | • Female gender<br>• Previously received an allogeneic transplant or gene therapy |
| **Age Restriction:** | • 4 to 17 years of age |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist, endocrinologist, or hematologist/oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified (one infusion only) |

POLICY NAME:
**ELTROMBOPAG DERIVATIVES**
Affected Medications: ELTROMBOPAG OLAMINE, ELTROMBOPAG OLAMINE PACKET, ALVAIZ (eltrombopag choline)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of thrombocytopenia in patients with persistent or chronic immune thrombocytopenia (ITP)<br>  ○ Treatment of thrombocytopenia in patients with hepatitis C infection<br>  ○ Treatment of severe aplastic anemia |
| **Required Medical Information:** | **<u>Thrombocytopenia in patients with chronic ITP</u>**<br>• Documentation of **ONE** of the following:<br>  ○ Platelet count less than 20,000/microliter<br>  ○ Platelet count less than 30,000/microliter AND symptomatic bleeding<br>  ○ Platelet count less than 50,000/microliter AND increased risk for bleeding (such as peptic ulcer disease, use of antiplatelets or anticoagulants, history of bleeding at higher platelet count, need for surgery or invasive procedure)<br><br>**<u>Thrombocytopenia in patients with chronic hepatitis C</u>**<br>• Documentation of plan to initiate interferon-based therapy<br>• Documentation of platelet count less than 75,000/microliter<br><br>**<u>Severe aplastic anemia</u>**<br>• Diagnosis confirmed by bone marrow biopsy<br>• Documentation of at least two of the following:<br>  ○ Absolute reticulocyte count (ARC) less than 60,000/microliter<br>  ○ Platelet count less than 20,000/microliter<br>  ○ Absolute neutrophil count (ANC) less than 500/microliter |
| **Appropriate Treatment Regimen & Other Criteria:** | • Eltrombopag olamine packet formulation requires documented medical inability to use oral tablet formulation<br><br>**<u>Thrombocytopenia in patients with persistent or chronic ITP</u>**<br>• Documentation of inadequate response, defined as platelets did not increase to at least 50,000/microliter, to the following therapies:<br>  ○ **ONE** of the following:<br>  ▪ Inadequate response with at least 2 therapies for immune thrombocytopenia, including corticosteroids, rituximab, or immunoglobulin<br>  ▪ Splenectomy<br><br>**<u>Reauthorization</u>:**<br>• Response to treatment with platelet count of at least 50,000/microliter (not to exceed 400,000/microliter)<br>**OR**<br>• The platelet counts have not increased to a platelet count of at least 50,000/microliter and the patient has NOT been on the maximum dose for at least 4 weeks<br><br>**<u>Thrombocytopenia in patients with chronic hepatitis C</u>** |

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| | **Reauthorization:**<br>• Response to treatment with platelet count of at least 90,000/microliter (not to exceed 400,000/microliter) and eltrombopag is used in combination with antiviral therapy<br><br>**Severe aplastic anemia**<br>• Documentation of refractory severe aplastic anemia as indicated by insufficient response to at least one prior immunosuppressive therapy<br>**OR**<br>• For those less than 40 years of age without a rapidly available matched related donor (MRD) or 40 years of age and older: documentation that eltrombopag is being used as first line treatment in combination with standard immunosuppressive therapy (Atgam and cyclosporine)<br><br>**Reauthorization (refractory severe aplastic anemia only) r**equires hematologic response to treatment defined as meeting **ONE** or more of the following criteria:<br>• Platelet count increases to 20,000/microliter above baseline, or stable platelet counts with transfusion independence for a minimum of 8 weeks<br>• Hemoglobin increases by greater than 1.5 g/dL or a reduction in greater than or equal to 4 units red blood cell (RBC) transfusions for 8 consecutive weeks<br>• ANC increase of 100% or an ANC increase greater than 500/microliter |
| **Exclusion Criteria:** | • Use in combination with another thrombopoietin receptor agonist, spleen tyrosine kinase inhibitor, or similar treatments (Doptelet, Nplate, Tavalisse) |
| **Age Restriction:** | **Thrombocytopenia in patients with ITP**<br>• 1 year of age and older (eltrombopag olamine)<br>• 6 years of age and older (Alvaiz)<br><br>**Thrombocytopenia in patients with chronic hepatitis C and patients with severe aplastic anemia**<br>• 18 years of age and older (eltrombopag olamine and Alvaiz)<br><br>**Severe Aplastic Anemia (initial therapy)**<br>• 2 years of age and older<br>• 18 years of age and older (Alvaiz) |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist or gastroenterology/liver specialist<br>• All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **Thrombocytopenia in patients with ITP**<br>• Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br><br>**Thrombocytopenia in patients with chronic hepatitis C**<br>• Initial Authorization: 2 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br><br>**Severe aplastic anemia**<br>• Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br><br>**Severe aplastic anemia in combination with cyclosporine and Atgam** |

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| | • Authorization: 6 months, no reauthorization, unless otherwise specified |

POLICY NAME:
**EMAPALUMAB**
Affected Medications: GAMIFANT (emapalumab-lzsg)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of primary hemophagocytic lymphohistiocytosis (HLH) in patients (newborn and older) intolerant to conventional HLH therapy or with refractory, recurrent, or progressive disease<br>  ○ Treatment of HLH/macrophage activation syndrome (MAS) in known or suspected Still's disease, including systemic Juvenile Idiopathic Arthritis (sJIA) in patients (newborn and older) with an inadequate response or intolerance to glucocorticoids, or with recurrent MAS |
| **Required Medical Information:** | • Documentation confirming status as a hematopoietic stem cell transplant (HSCT) candidate<br><br>**HLH**<br>• Diagnosis confirmed by presence of a genetic mutation known to cause primary HLH (e.g., PRF1, UNC13D, STX11, STXBP2) **OR** documentation showing at least 5 of the following are present:<br>  ○ Prolonged fever (lasting over 7 days)<br>  ○ Splenomegaly<br>  ○ **Two** of the following cytopenias in the peripheral blood:<br>  ▪ Hemoglobin less than 9 g/dL<br>  ▪ Platelet count less than 100,000/mcL<br>  ▪ Neutrophils less than 100/mcL<br>  ○ **One** of the following:<br>  ▪ Hypertriglyceridemia defined as fasting triglycerides 3 mmol/L or higher (equivalent to 265 mg/dL or higher)<br>  ▪ Hypofibrinogenemia defined as fibrinogen 1.5 g/L or lower<br>  ○ Hemophagocytosis in bone marrow, spleen, or lymph nodes (with no evidence of malignancy)<br>  ○ Low or absent natural killer cell activity (according to local laboratory reference)<br>  ○ Ferritin 500 mcg/L or higher<br>  ○ Soluble CD25 (i.e., soluble IL-2 receptor) 2,400 U/mL or higher<br><br>**HLH with MAS**<br>• Diagnosis of HLH and documentation of active MAS in the setting of Adult Onset Still's disease or sJIA with ferritin levels greater than 684 ng/mL<br>• Documentation showing at least 2 of the following are present:<br>  ○ Platelet count is 181,000/mcL or lower<br>  ○ AST is greater than 48 U/L<br>  ○ Triglycerides is greater than 156 mg/dL<br>  ○ Fibrinogen is 360 mg/dL or lower |
| **Appropriate Treatment Regimen & Other Criteria:** | **HLH**<br>• Documentation of refractory, recurrent, or progressive disease (or intolerable adverse event) on conventional HLH therapy (e.g., dexamethasone, etoposide, methotrexate, hydrocortisone) |

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| | • Must be used in combination with dexamethasone, unless currently established on and planning to continue one of the following: cyclosporine, glucocorticoids, and/or intrathecal methotrexate<br><br>**HLH with MAS**<br>• Documentation of refractory, recurrent, or progressive disease (or intolerable adverse event) on high dose intravenous glucocorticoids and Kineret (anakinra)<br><br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy AND patient has not yet received HSCT |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist, oncologist, transplant specialist, or provider with experience in the management of HLH<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 2 months, unless otherwise specified<br>• Reauthorization: 4 months, unless otherwise specified |

POLICY NAME:
**EMICIZUMAB**
Affected Medications: HEMLIBRA (emicizumab-kxwh)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | • Documented diagnosis of hemophilia A with or without inhibitors<br>• Prescribed for routine prophylaxis to prevent or reduce the frequency of bleeding episodes |
| **Appropriate Treatment Regimen & Other Criteria:** | • Baseline factor level less than 1% AND prophylaxis required OR<br>• Baseline factor level 1% to 3% AND a documented history of at least two episodes of spontaneous bleeding into joints<br>• Prophylactic agents must be discontinued<br>▪ Factor VIII Inhibitors: after the first week of HEMLIBRA<br>▪ Bypassing Agents: one day before starting HEMLIBRA<br><br>**Loading Dose:**<br>• 3 mg/kg once every week for 4 weeks<br>• Maximum 1,380 mg per 28 day supply<br><br>**Maintenance dose:**<br>• 1.5 mg/kg once every week or<br>• 3 mg/kg once every 2 weeks or<br>• 6 mg/kg once every 4 weeks<br>• Any increases in dose must be supported by an acceptable clinical rationale (i.e. weight gain, increase in breakthrough bleeding when patient is fully adherent to therapy, etc.)<br><br>**Product Availability**<br>• Single-dose vials for injection: 30 mg/mL, 60 mg/0.4 mL, 105 mg/0.7 mL, 150 mg/mL<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>Reauthorization requires documentation of treatment success defined as a reduction in spontaneous bleeds requiring treatment, as well as documentation of bleed history since last approval |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 6 months, unless otherwise specified |

POLICY NAME:
**EMSAM**
Affected Medications: EMSAM (selegiline)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of major depressive disorder (MDD) |
| **Required Medical Information:** | • Diagnosis of major depressive disorder (MDD) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure to an adequate trial (clinically sufficient doses for a minimum 6-week duration) to each of the following:<br>  ○ A selective serotonin reuptake inhibitor (SSRI)<br>  ○ A serotonin/norepinephrine reuptake inhibitor (SNRI)<br>  ○ A tricyclic antidepressant or mirtazapine<br>  ○ Bupropion<br>  ○ Antidepressant augmentation therapy (e.g., second generation antipsychotic, thyroid hormone, lithium)<br>**OR**<br>• Documentation of inability to take any oral preparations (including commercially available liquid antidepressants)<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Pheochromocytoma |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a psychiatrist or behavioral health specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**ENZYME REPLACEMENT THERAPY (ERT) FOR GAUCHER DISEASE TYPE 1**
Affected Medications: CERDELGA (eliglustat), VPRIV (velaglucerase alfa), CEREZYME (imiglucerase), ELELYSO (taliglucerase alfa)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Vpriv: Gaucher disease type 1 (GD1)<br>  ○ Elelyso: GD1 for ages 4 years and older<br>  ○ Cerdelga: GD1 in adults who are CYP2D6 extensive metabolizers (EMs), intermediate metabolizers (IMs), or poor metabolizers (PMs) as detected by an FDA-cleared test<br>  ○ Cerezyme:<br>  ▪ Gaucher disease type 3<br>  ▪ GD1 that results in one or more of the following conditions:<br>  • Anemia<br>  • Thrombocytopenia<br>  • Bone disease<br>  • Hepatomegaly or splenomegaly |
| **Required Medical Information:** | • Diagnosis confirmed by enzyme assay showing deficiency of beta-glucocerebrosidase glucosidase enzyme activity **OR** genetic testing indicating mutation of two alleles of the glucocerebrosidase genome<br>  ○ For Cerdelga, must also have documentation of cytochrome P450 2D6 (CYP2D6) genotype by an FDA-approved test indicating CYP2D6 EM, IM, or PM status<br>• Documentation of baseline tests such as hemoglobin level, platelet count, liver function tests, renal function tests<br><br>**<u>For GD1:</u>**<br>• Documentation of at least one clinically significant disease complication such as:<br>  ○ Anemia (low hemoglobin and hematocrit levels)<br>  ○ Thrombocytopenia (platelet count less than 120,000 $mm^3$)<br>  ○ Bone disease (T-score less than -2.5 or bone pain)<br>  ○ Hepatomegaly or splenomegaly<br>  ○ For symptomatic children: symptoms of early presentation, such as malnutrition, growth retardation, impaired psychomotor development, and/or fatigue |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Cerdelga</u>**<br><br><u>Extensive or Intermediate Metabolizers of CYP2D6</u><br>• Quantity limit - 84 mg capsules #60 per 30 days<br><br><u>Poor Metabolizers of CYP2D6</u><br>• Quantity limit - 84 mg capsules #30 per 30 days |

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| | **<u>Elelyso, Vpriv, and Cerezyme</u>**<br>• Dosing is in accordance with FDA labeling and patient's most recent weight<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Concomitant use with another ERT for GD1 or with miglustat<br><br>**<u>Cerdelga</u>:**<br>• CYP2D6 ultrarapid metabolizers<br>• Moderate or severe hepatic impairment<br>• Pre-existing cardiac disease (congestive heart failure, myocardial infarction, bradycardia, heart block, arrhythmias, and long QT syndrome)<br>• Presence of moderate to severe renal impairment or end stage renal disease |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist in the management of Gaucher disease (hematologist, oncologist, hepatologist, geneticist or orthopedic specialist)<br>• All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**EPLONTERSEN, PATISIRAN, VUTRISIRAN**
Affected Medications: WAINUA (eplontersen), ONPATTRO (patisiran), AMVUTTRA (vutrisiran)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of hereditary transthyretin amyloidosis with polyneuropathy (hATTR-PN) in adults<br>  ○ Treatment of the cardiomyopathy of wild-type or hereditary transthyretin-mediated amyloidosis (ATTR-CM) in adults to reduce cardiovascular mortality, cardiovascular hospitalizations and urgent heart failure visits |
| **Required Medical Information:** | **ATTR-CM (Amvuttra)**<br>• Diagnosis of ATTR-CM supported by **ONE** of the following (a, b, or c):<br>  a. Cardiac tissue biopsy confirms presence of ATTR amyloid deposits by immunohistochemistry (IHC) or mass spectrometry<br>  b. Documentation of **BOTH** of the following (i and ii):<br>  i. Noncardiac tissue biopsy confirms presence of ATTR amyloid deposits by IHC or mass spectrometry<br>  ii. Imaging consistent with cardiac amyloidosis (echocardiogram [ECG], cardiac magnetic resonance [CMR], or positron emission tomography [PET])<br>  c. Documentation of **ALL** the following (i, ii, and iii):<br>  i. Grade 2 to 3 uptake on cardiac scintigraphy (utilizing Tc-PYP, Tc-DPD, or Tc-HMDP radiotracers)<br>  ii. Normal serum kappa/lambda free light chain (sFLC) ratio, serum protein immunofixation, **AND** urine protein immunofixation<br>  iii. Imaging consistent with cardiac amyloidosis (ECG, CMR, or PET)<br>• Documentation of New York Heart Association (NYHA) Functional Class I to III<br><br>**ATTR-PN**<br>• Documented diagnosis of hATTR confirmed by **BOTH** of the following:<br>  ○ Amyloid deposition on biopsy<br>  ○ Presence of pathogenic transthyretin (TTR) variant on genetic testing<br>• Presence of clinical manifestations of the disease, confirmed by presence of peripheral neuropathy on nerve conduction studies **OR** 2 of the following:<br>  ○ Autonomic dysfunction (bladder/urinary tract infections, gastrointestinal disturbances, erectile dysfunction, orthostatic hypotension)<br>  ○ Documented symptoms of sensorimotor polyneuropathy (e.g., paresthesia, balance issues, weakness/numbness in the hands/feet, or loss of sensation for pain, temperature, proprioception)<br>  ○ Cardiomyopathy, ocular involvement, or renal involvement<br>• Documentation of **ONE** of the following:<br>  ○ Baseline polyneuropathy disability (PND) score of less than or equal to IIIb<br>  ○ Baseline neuropathy impairment score (NIS) between 10 and 130<br>  ○ Baseline familial amyloid polyneuropathy (FAP) stage 1 or 2 |

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| **Appropriate Treatment Regimen & Other Criteria:** | • **Amvuttra** requests require one of the following:<br>  ○ ATTR-PN diagnosis<br>  ○ ATTR-CM diagnosis only: treatment failure with Attruby (acoramidis) evidenced by worsening of heart failure signs/symptoms, increase in NYHA class, and increase in cardiovascular related hospitalizations<br>• **Onpattro:** Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**<u>Reauthorization</u>:**<br><br>**<u>ATTR-CM (Amvuttra)</u>**<br>• Documentation of disease responsiveness (improvement in symptoms, quality of life, or 6-Minute Walk Test; slowing or stabilization of disease progression; reduced cardiovascular-related hospitalizations, etc.)<br><br>**<u>ATTR-PN</u>**<br>• Documentation of a positive clinical response (e.g., stabilized or improved neurologic impairment, motor function, cardiac function, quality of life assessment, serum TTR levels) |
| **Exclusion Criteria:** | • Prior or planned liver transplantation<br>• NYHA Functional Class III or IV (Wainua)<br>• NYHA Functional Class IV (Amvuttra)<br>• Combined use with TTR-lowering or stabilizing therapy |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or specialist experienced in the treatment of amyloidosis<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**EPOPROSTENOL**
Affected Medications: EPOPROSTENOL, VELETRI (epoprostenol), FLOLAN (epoprostenol)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1 |
| **Required Medical Information:** | **<u>Pulmonary Arterial Hypertension (PAH) WHO Group 1</u>**<br>• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:<br>  ○ Mean pulmonary artery pressure of at least 20 mm Hg<br>  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg<br>  ○ Pulmonary vascular resistance of at least 2.0 Wood units<br>• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class III or higher symptoms<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blockers) unless there are contraindications:<br>  ○ Low systemic blood pressure (systolic blood pressure less than 90)<br>  ○ Low cardiac index<br>  OR<br>  ○ Presence of severe symptoms (functional class IV)<br>• Documentation of current patient weight<br>• Documentation of a clear treatment plan |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of inadequate response or intolerance to the following therapy classes is required:<br>  ○ PDE5 inhibitors **AND**<br>  ○ Endothelin receptor antagonists (exception WHO Functional Class IV)<br><br>**<u>Reauthorization</u>** requires documentation of treatment success defined as one or more of the following:<br>• Improvement in walking distance<br>• Improvement in exercise ability<br>• Improvement in pulmonary function<br>• Improvement or stability in WHO functional class |
| **Exclusion Criteria:** | • Congestive heart failure due to severe left ventricular systolic dysfunction<br>• Long-term use in patients who develop pulmonary edema during dose initiation |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months unless otherwise specified |

POLICY NAME:
**ERECTILE DYSFUNCTION**
Affected Medications: VIAGRA, SILDENAFIL (25 mg, 50 mg, 100 mg), CIALIS (10 mg and 20 mg), EDEX KIT, LEVITRA, STAXYN, STENDRA, TADALAFIL (10 mg, 20 mg), VARDENAFIL, CAVERJECT

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment for a mental health diagnosis of erectile dysfunction (ED), also known as erectile disorder, meeting sexual dysfunction criteria |
| **Required Medical Information:** | • Documentation of a mental health diagnosis of erectile dysfunction meeting the *Diagnostic and Statistical Manual of Mental Disorders*, Fifth Edition (DSM-5) criteria:<br>  ○ At least one of the three following symptoms must be experienced with 75% to 100% of occasions of sexual activity:<br>  ▪ Marked difficulty in obtaining an erection during sexual activity<br>  ▪ Marked difficulty in maintaining an erection until the completion of sexual activity<br>  ▪ Marked decrease in erectile rigidity<br>  ○ The above symptoms have persisted for a minimum duration of approximately 6 months<br>  ○ The above symptoms cause clinically significant distress in the individual<br>  ○ The sexual dysfunction is not attributable to any of the following:<br>  ▪ A nonsexual medical or psychiatric condition<br>  ▪ Severe relationship distress (e.g., partner violence)<br>  ▪ The effects of medication or other substance use<br>  ▪ Other clinically significant and relevant stressors |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of treatment failure with tadalafil 2.5 mg or 5 mg tablets |
| **Exclusion Criteria:** | • Erectile dysfunction unrelated to a mental health diagnosis of sexual dysfunction according to the DSM-5 diagnostic criteria |
| **Prescriber/Site of Care Restrictions** | • Prescribed by, or in consultation with, a mental health provider<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Age Restriction:** | |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**ERGOT ALKALOIDS**
Affected Medications: DIHYDROERGOTAMINE MESYLATE INJECTION, DIHYDROERGOTAMINE MESYLATE NASAL SOLUTION

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Documentation of moderate to severe migraines |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of treatment failure, intolerance, or contraindication to all of the following:<br>  ○ At least **two** prescription strength non-steroidal anti-inflammatory drugs (NSAIDs) or combination analgesics (such as ibuprofen, naproxen, acetaminophen/aspirin/caffeine)<br>  ○ At least **one** oral 5-hydroxytryptamine-1 (5-$HT_1$) receptor agonist (such as sumatriptan, naratriptan, rizatriptan, zolmitriptan)<br>  ○ At least **one** non-oral 5-$HT_1$ receptor agonist (such as sumatriptan, zolmitriptan)<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Hemiplegic or basilar migraine<br>• Uncontrolled hypertension<br>• Ischemic heart disease (e.g., angina pectoris, history of myocardial infarction, history of silent ischemia)<br>• Peripheral artery disease<br>• Pregnancy or breastfeeding<br>• Documented severe chronic liver disease<br>• Severe renal impairment<br>• Use in combination with 5HT1 receptor agonist such as sumatriptan |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**ENDOTHELIN RECEPTOR ANTAGONISTS**
Affected Medications: BOSENTAN, AMBRISENTAN, OPSUMIT (macitentan), OPSYNVI (macitentan and tadalafil)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Pulmonary artery hypertension (PAH) World Health Organization (WHO) Group 1 |
| **Required Medical Information:** | • Documentation of Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1 confirmed by right heart catheterization meeting the following criteria:<br>  ○ Mean pulmonary artery pressure of at least 20 mm Hg<br>  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg<br>  ○ Pulmonary vascular resistance of at least 2.0 Wood units<br>• New York Heart Association (NYHA)/WHO Functional Class II or higher symptoms<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blocker), unless there are contraindications:<br>  ○ Low systemic blood pressure (systolic blood pressure less than 90)<br>  ○ Low cardiac index<br>  ○ Presence of severe symptoms (functional class IV) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation that the drug will be used in combination with a phosphodiesterase-5 (PDE-5) inhibitor<br>• Documentation of inadequate response or intolerance to oral calcium channel blocking agents if positive Acute Vasoreactivity Test<br>• For Opsumit (macitentan) and Opsynvi (macitentan and tadalafil) requests: documentation of inadequate response or intolerance to ambrisentan AND bosentan for 12 weeks is required<br><br>**Reauthorization** requires documentation of treatment success defined as one or more of the following:<br>• Improvement in walking distance<br>• Improvement in exercise ability<br>• Improvement in pulmonary function<br>• Improvement or stability in WHO functional class |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**ERYTHROPOIESIS STIMULATING AGENTS (ESAs)**
Affected Medications: ARANESP (darbepoetin alfa), EPOGEN (epoetin alfa), MIRCERA (methoxy polyethylene glycol-epoetin beta), PROCRIT (epoetin alfa)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>**Epogen & Aranesp & Procrit & Mircera**<br>• Treatment of anemia due to chronic kidney disease (CKD), including patients on dialysis and not on dialysis to decrease the need for red blood cell (RBC) transfusion<br>**Epogen & Procrit & Aranesp**<br>• Treatment of anemia in patients with non-myeloid malignancies where anemia is due to the effect of concomitant myelosuppressive chemotherapy, and upon initiation, there is a minimum of two additional months of planned chemotherapy<br>**Epogen & Procrit only**<br>• To reduce the need for allogeneic RBC transfusions among patients with perioperative hemoglobin greater than 10 to 13 or less g/dL who are at high risk for perioperative blood loss from elective, noncardiac, nonvascular surgery<br>• Treatment of anemia due to zidovudine administered at ≤ 4200 mg/week in patients with HIV-infection with endogenous serum erythropoietin levels of ≤ 500 mUnits/mL<br>**Compendia-supported uses**<br>• Symptomatic anemia in Myelodysplastic syndrome<br>• Allogenic bone marrow transplantation<br>• Anemia associated with Hepatitis C (HCV) treatment<br>• Anemia associated with rheumatoid arthritis (RA)/ rheumatic disease |
| **Required Medical Information:** | • One of the following in accordance with FDA (Food and Drug Administration)-approved label or compendia support:<br>○ Anemia associated with chronic renal failure<br>○ Anemia secondary to chemotherapy with a minimum of two additional months of planned chemotherapy<br>○ Anemia secondary to zidovudine-treated Human Immunodeficiency Virus (HIV) patients<br>○ Anemia in patients scheduled to undergo elective, non-cardiac, nonvascular surgery<br>○ Symptomatic anemia in Myelodysplastic syndrome<br>○ Allogenic bone marrow transplantation<br>○ Anemia associated with Hepatitis C (HCV) treatment<br>○ Anemia associated with rheumatoid arthritis (RA)/ rheumatic disease |
| **Appropriate Treatment Regimen & Other Criteria:** | • Coverage for the non-preferred drugs (Epogen and Mircera) is provided when the following criteria is met:<br>○ A documented intolerable adverse event to the preferred products Procrit and Retacrit, and the adverse event was not an expected adverse event attributed to the active ingredient |
| **Exclusion Criteria:** | • Use in combination with another erythropoiesis stimulating agent (ESA) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist, oncologist, or nephrologist |
| **Coverage Duration:** | • Authorization: 6 months, unless otherwise specified |

POLICY NAME:
**ETA RECEPTOR ANTAGONISTS**
Affected Medications: FILSPARI (sparsentan), VANRAFIA (atrasentan)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Reduce proteinuria in adults with primary immunoglobulin A nephropathy (IgAN) at risk of disease progression |
| **Required Medical Information:** | • Diagnosis of primary immunoglobulin A nephropathy (IgAN) confirmed with biopsy<br>• Documentation of proteinuria equal to or greater than 0.5 g/day (labs taken within 30 days of request)<br>• Documented estimated glomerular filtration rate (eGFR) equal to or greater than 30 mL/min/1.73 $m^2$ |
| **Appropriate Treatment Regimen & Other Criteria:** | • Persistent proteinuria (greater than or equal to 0.5 g/day) despite a minimum 12-week trial with all the following:<br>  ○ Maximally tolerated angiotensin-converting enzyme (ACE) inhibitor **OR** angiotensin receptor II blocker (ARB)<br>  ○ Glucocorticoid therapy, such as prednisone or methylprednisolone (or adverse effect with two or more glucocorticoid therapies, which is not associated with the corticosteroid class)<br>• For Vanrafia requests: documented trial and failure of Filspari (sparsentan)<br><br>**Reauthorization** requires documentation of treatment success, defined as reduction in proteinuria |
| **Exclusion Criteria:** | • Concurrent use of an endothelin A (ETA) receptor antagonist<br>• The requested medication should not be administered in combination with other medications indicated for immunoglobulin A nephropathy due to lack of clinical trial data supporting additive efficacy |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a nephrologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ETELCALCETIDE**
Affected Medications: PARSABIV (etelcalcetide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Secondary hyperparathyroidism in adults with chronic kidney disease (CKD) on dialysis |
| **Required Medical Information:** | • Documentation of both of the following:<br>  ○ Currently on dialysis<br>  ○ Intact parathyroid hormone (iPTH) level greater than 300 pg/mL<br>• Documentation of iPTH that is persistently elevated above target range despite at least 12 weeks of adherent treatment with each of the following at an appropriate dose, unless contraindicated or not tolerated:<br>  ○ Calcitriol<br>  ○ Doxercalciferol<br>  ○ Paricalcitol<br>  ○ Cinacalcet |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Diagnosis of parathyroid carcinoma, primary hyperparathyroidism or with chronic kidney disease who are not on hemodialysis |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist or nephrologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**ETRANACOGENE**
Affected Medications: HEMGENIX (etranacogene dezaparvovec-drlb)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Hemophilia B (congenital factor IX deficiency) |
| **Required Medical Information:** | • Documentation of diagnosis of Hemophilia B<br>• Documentation of baseline circulating level of factor IX less than or equal to 2% as attested by the managing physician AND requiring prophylactic Factor IX treatment<br>• Documentation of negative Factor IX inhibitor titers (if test result is positive, re-test within 2 weeks with negative result)<br>• Baseline lab values (less than 2 times upper limit of normal):<br>  ○ ALT<br>  ○ AST<br>  ○ Total bilirubin<br>  ○ Alkaline phosphatase (ALP) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of plan to discontinue Factor IX prophylaxis therapy upon achieving circulating factor IX levels of 5%<br><br>**Dosing:**<br>• 2 x $10^{13}$ genome copies (gc) per kilogram of body weight |
| **Exclusion Criteria:** | • Prior gene therapy administration |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care<br>• Prescribed by, or in consultation with, a hematologist or specialist with experience in the treatment of hemophilia |
| **Coverage Duration:** | • Authorization: 2 months (one-time infusion only), unless otherwise specified |

**POLICY NAME:**
**ETRIPAMIL**
**Affected Medications:** Cardamyst (etripamil nasal spray)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Conversion of acute symptomatic episodes of paroxysmal supraventricular tachycardia (PSVT) to sinus rhythm in adults |
| **Required Medical Information:** | • Diagnosis of atrioventricular nodal-dependent PSVT (e.g. AVNRT, AVRT)<br>• History of sustained episodes (20 minutes or greater) of PSVT<br>• History of at least one emergency department or provider visit for the treatment of an acute episode of PSVT |
| **Appropriate Treatment Regimen & Other Criteria:** | • If requesting a greater treatment frequency than the posted quantity limit: history of PSVT episodes requiring treatment over the past 12 months |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber Restrictions:** | • Prescribed by or in consultation with a specialist in the management of PSVT (e.g. cardiologist or electrophysiologist)<br>• All approvals are subject to utilization of the most-cost effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise stated<br>• Reauthorization: 12 months, unless otherwise stated |

POLICY NAME:
**EVKEEZA and JUXTAPID**
Affected Medications: EVKEEZA (evinacumab-dgnb), JUXTAPID (lomitapide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Homozygous familial hypercholesterolemia (HoFH) |
| **Required Medical Information:** | • Documentation of baseline untreated low-density lipoprotein cholesterol (LDL-C)<br>• Diagnosis confirmed by **ONE** of the following:<br>  ○ Baseline LDL-C greater than 560 mg/dL<br>  ○ Baseline LDL-C of 400 mg/dL and at least 1 parent with familial hypercholesterolemia<br>  ○ Baseline LDL-C of 400 mg/dL with aortic valve disease or xanthomata in ages less than 20 years<br>  ○ Presence of two abnormal LDL-C-raising gene defects (excluding double-null LDL receptor [LDLR] mutations) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented intent to take alongside maximally tolerated doses of statin and/or ezetimibe, unless otherwise contraindicated<br>OR<br>• History of statin intolerance requires documentation of **ONE** of the following:<br>  ○ Statin-associated rhabdomyolysis occurred with statin use and was confirmed by a creatinine kinase (CK) level at least 10 times the upper limit of normal<br>  ○ Statin-associated muscle symptoms (e.g., myopathy, myalgia) occurred with statin use and was confirmed by **BOTH** of the following:<br>  ▪ A minimum of two different statin trials, with at least one being a hydrophilic statin (rosuvastatin, pravastatin)<br>  ▪ A re-challenge of each statin (muscle symptoms stopped when each was discontinued and restarted upon re-initiation)<br>• Documented treatment failure, defined as an inability to achieve LDL-C reduction of 50% or greater **OR** LDL-C less than 100 mg/dL despite at least six months of adherent therapy with all of the following, unless contraindicated or not tolerated:<br>  ○ Maximally tolerated statin therapy<br>  ○ Ezetimibe<br>  ○ PCSK9 monoclonal antibody, unless double-null or LDLR activity 15% or less<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**<u>Reauthorization</u>** requires documentation of treatment success and a clinically significant response to therapy defined by an LDL-C level at goal or decreased by at least 30% from baseline |
| **Exclusion Criteria:** | • Combination therapy with Juxtapid and Evkeeza is considered experimental and is not a covered benefit |
| **Age Restriction:** | • Evkeeza: 1 year of age and older<br>• Juxtapid: 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist, cardiologist, or lipid specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**EVOLOCUMAB**
Affected Medications: REPATHA (evolocumab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Prevention of clinical atherosclerotic cardiovascular disease (ASCVD)<br>  ○ Primary hyperlipidemia (including heterozygous familial hypercholesterolemia [HeFH])<br>  ○ Homozygous familial hypercholesterolemia (HoFH) |
| **Required Medical Information:** | **<u>All Indications</u>**<br>• Documentation of current complete lipid panel within last 3 months<br>• Documentation of baseline (untreated) low-density lipoprotein cholesterol (LDL-C)<br><br>**<u>Primary Hyperlipidemia (non-familial)</u>**<br>• Documentation of baseline (untreated) LDL-C of at least 160 mg/dL<br><br>**<u>HeFH</u>**<br>• Diagnosis confirmed by **ONE** of the following:<br>  ○ Minimum baseline LDL-C of 160 mg/dL in adolescents or 190 mg/dL in adults **AND** 1 first-degree relative affected<br>  ○ Presence of one abnormal LDL-C-raising gene defect (e.g., LDL receptor [LDLR], apolipoprotein B [apo B], proprotein convertase subtilisin kexin type 9 [PCSK9] gain-of-function mutation, LDL receptor adaptor protein 1 [LDLRAP1])<br>  ○ World Health Organization (WHO)/Dutch Lipid Network criteria score of at least 8 points<br>  ○ Definite FH diagnosis per the Simon Broome criteria<br><br>**<u>HoFH</u>**<br>• Diagnosis confirmed by **ONE** of the following:<br>  ○ Baseline LDL-C greater than 560 mg/dL<br>  ○ Baseline LDL-C of 400 mg/dL and at least 1 parent with familial hypercholesterolemia<br>  ○ Baseline LDL-C of 400 mg/dL with aortic valve disease or xanthomata in ages less than 20 years<br>  ○ Presence of two abnormal LDL-C-raising gene defects (excluding double-null LDLR mutations) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>All Indications</u>**<br>• History of statin intolerance requires documentation of **ONE** of the following:<br>  ○ Statin-associated rhabdomyolysis occurred with statin use and was confirmed by a creatinine kinase (CK) level at least 10 times the upper limit of normal<br>  ○ Statin-associated muscle symptoms (e.g., myopathy, myalgia) occurred with statin use and was confirmed by **BOTH** of the following:<br>  ▪ A minimum of two different statin trials, with at least one being a hydrophilic statin (rosuvastatin, pravastatin)<br>  ▪ A re-challenge of each statin (muscle symptoms stopped when each |

<table>
<tr><td></td><td>was discontinued and restarted upon re-initiation)<br><br><u>Clinical ASCVD</u><br>• Documented treatment failure with minimum 12 weeks of statin/ezetimibe combination therapy at maximally tolerated doses with consistent use, as shown by ONE of the following:<br>  ○ Current LDL-C of at least 70 mg/dL<br>  ○ Current LDL-C of at least 55 mg/dL in patients at very high risk of future ASCVD events, based on history of multiple major ASCVD events OR 1 major ASCVD event + multiple high-risk conditions<br><br><table><tr><th>Major ASCVD Events</th><th>High-Risk Conditions</th></tr><tr><td>• ACS within the past 12 months<br>• History of MI (distinct from ACS event)<br>• Ischemic stroke<br>• Symptomatic PAD</td><td>• Age 65 years and older<br>• HeFH<br>• Prior coronary artery bypass or percutaneous intervention (outside of major ASCVD events)<br>• Diabetes<br>• Hypertension<br>• Chronic kidney disease<br>• Currently smoking<br>• History of congestive heart failure</td></tr></table><br><u>Primary Hyperlipidemia/HeFH/HoFH</u><br>• Documented treatment failure, defined as an inability to achieve LDL-C reduction of 50% or greater OR LDL-C less than 100 mg/dL, with minimum 12 weeks of statin/ezetimibe combination therapy at maximally tolerated doses with consistent use</td></tr>
<tr><td>Exclusion Criteria:</td><td>• Concurrent use with Leqvio</td></tr>
<tr><td>Age Restriction:</td><td></td></tr>
<tr><td>Prescriber/Site of Care Restrictions:</td><td>• Prescribed by, or in consultation with, a cardiologist, endocrinologist, or lipid specialist<br>• All approvals are subject to utilization of the most cost-effective site of care</td></tr>
<tr><td>Coverage Duration:</td><td>• Authorization: 12 months, unless otherwise specified</td></tr>
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POLICY NAME:
**EXAGAMGLOGENE AUTOTEMCEL**
Affected Medications: CASGEVY (exagamglogene autotemcel)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of sickle cell disease in adults and pediatric patients at least 12 years of age with recurrent vaso-occlusive crises.<br>  ○ Treatment of transfusion-dependent beta-thalassemia in adults and pediatric patients at least 12 years of age. |
| **Required Medical Information:** | **<u>SICKLE CELL DISEASE</u>**<br><br>• Documentation of sickle cell disease confirmed by genetic testing to show the presence of βS/βS, βS/β0 or βS/β+ genotype as follows:<br>  ○ Identification of significant quantities of HbS with or without an additional abnormal β-globin chain variant by hemoglobin assay<br>  **OR**<br>  ○ Identification of biallelic *HBB* pathogenic variants where at least one allele is the p.Glu6Val or p.Glu7Val pathogenic variant on molecular genetic testing<br>  **AND**<br>  ○ Patient does NOT have disease with more than two α-globin gene deletions<br>• Documentation of severe disease defined as 2 or more severe vaso-occlusive crises (VOCs) or vaso-occlusive events (VOEs) within the previous year (4 events over 2 years will also meet this requirement) VOC/VOEs defined as:<br>  ○ Acute pain event requiring a visit to a medical facility and administration of pain medications (opioids or IV NSAIDs) or RBC transfusions<br>  ○ Acute chest syndrome<br>  ○ Priapism lasting more than 2 hours and requiring visit to medical facility<br>  ○ Splenic sequestration<br>• Clinically stable and eligible to undergo hematopoietic stem cell transplant (HSCT) but unable to find a human leukocyte antigen (HLA) matched, related donor<br>• Adequate bone marrow, lung, heart, and liver function to undergo myeloablative conditioning regimen<br><br>**<u>TRANSFUSION DEPENDENT BETA THALASSEMIA</u>**<br><br>• Documented diagnosis of homozygous beta thalassemia or compound heterozygous beta thalassemia including β-thalassemia/hemoglobin E (HbE) (excludes alpha-thalassemia and hemoglobin S/ß-thalassemia variants) as outlined by the following:<br>  ○ Patient diagnosis is confirmed by HBB sequence gene analysis showing biallelic pathogenic variants<br>  **OR**<br>  ○ Patient has severe microcytic hypochromic anemia, anisopoikilocytosis with nucleated red blood cells on peripheral blood smear, and hemoglobin analysis that reveals decreased amounts or complete absence of hemoglobin A and increased amounts of hemoglobin F<br>• Documented transfusion-dependent disease defined as a history of transfusions of at least 100 mL/kg/year of packed red blood cells (pRBCs) or with 10 or more transfusions of pRBCs *per year* in the 2 years preceding therapy<br>• Clinically stable and eligible to undergo hematopoietic stem cell transplant (HSCT) but unable to find a human leukocyte antigen (HLA) matched, related donor |

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| **Appropriate Treatment Regimen & Other Criteria:** | • Must weigh a minimum of 6 kilograms and able to provide a minimum number of cells (3 × $10^6$ CD34+ cells/kg)<br>• Documentation that cardiac iron overload has been evaluated and there is no evidence of severe iron overload. (cardiac T2* less than 10 msec by magnetic resonance imaging [MRI] or left ventricular ejection fraction [LVEF] less than 45% by echocardiogram)<br>• No evidence of advanced liver disease [i.e., AST or ALT more than 3 times the upper limit of normal (ULN), or direct bilirubin value more than 2.5 times the ULN, or if a liver biopsy demonstrated bridging fibrosis or cirrhosis] |
| **Exclusion Criteria:** | • Prior HSCT or other gene therapy |
| **Age Restriction:** | • 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 6 months (one time infusion), unless otherwise specified |

POLICY NAME:
**FABRY DISEASE AGENTS**
Affected Medications: ELFABRIO (pegunigalsidase alfa), FABRAZYME (agalsidase beta), GALAFOLD (migalastat)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Fabry disease |
| **Required Medical Information:** | • Diagnosis of Fabry disease confirmed by one of the following:<br>  ○ Males: enzyme assay demonstrating undetectable (less than 3 percent) alpha-galactosidase A enzyme activity<br>  ○ Males: deficiency of alpha-galactosidase A enzyme activity (less than 35 percent) and genetic testing showing a mutation in the galactosidase alpha (GLA) gene<br>  ○ Females: genetic testing showing a mutation in the GLA gene<br>• For Galafold: Genetic testing confirming the presence of at least one amenable GLA variant<br><br>• Clinical signs and symptoms of Fabry disease, such as:<br>  ○ Severe neuropathic pain<br>  ○ Dermatologic manifestations (telangiectasias and angiokeratomas)<br>  ○ Corneal opacities<br>  ○ Kidney manifestations (proteinuria, polyuria, polydipsia)<br>  ○ Cardiac involvement (left ventricular hypertrophy, myocardial fibrosis, heart failure)<br>  ○ Cerebrovascular involvement (transient ischemic attacks, ischemic strokes)<br>  ○ Other manifestations common in Fabry disease (sweating abnormalities, hearing loss, or intolerance to heat, cold, or exercise) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Concurrent use with another agent on this policy (Galafold or enzyme replacement therapy for Fabry disease)<br>• For Galafold: Severe renal impairment (eGFR less than 30) or end-stage renal disease requiring dialysis |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a geneticist or a specialist experienced in the treatment of Fabry disease<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**FECAL MICROBIOTA**
Affected Medications: REBYOTA (fecal microbiota, live-jslm), VOWST (fecal microbiota spores, live-brpk)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Prophylaxis of *Clostridioides difficile* (C.diff) infection recurrence following antibiotic treatment |
| **Required Medical Information:** | • Documentation confirming a current diagnosis of recurrent C.diff infection (CDI) with a history of at least 2 recurrent episodes (initial episode + a minimum of 2 recurrences)<br>  ○ Recurrent CDI is defined as a resolution of CDI symptoms while on appropriate therapy, followed by a reappearance of symptoms within 8 weeks of discontinuing treatment<br>• Current episode of CDI must be controlled (less than 3 unformed or loose stools per day for 2 consecutive days)<br>• Administration will occur following completion of antibiotic course for CDI treatment<br>  ○ Within 24 to 72 hours for Rebyota<br>  ○ Within 2 to 4 days for Vowst<br>• Positive stool test for C.diff within the 30 days prior to request |
| **Appropriate Treatment Regimen & Other Criteria:** | **Rebyota**<br>• Previous treatment with at least **TWO** of the following in the setting of CDI recurrence: oral vancomycin, fidaxomicin, or fecal microbiota transplant (FMT)<br><br>**Vowst**<br>• Previous treatment with at least **TWO** of the following in the setting of CDI recurrence: oral vancomycin, fidaxomicin, or FMT<br>• Documented treatment failure with Rebyota |
| **Exclusion Criteria:** | • Retreatment with Rebyota or Vowst |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an infectious disease specialist or gastroenterologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 1 month with no reauthorization, unless otherwise specified |

POLICY NAME:
**FENFLURAMINE**
Affected Medications: FINTEPLA (fenfluramine)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of seizures associated with Dravet syndrome (DS)<br>  ○ Treatment of seizures associated with Lennox-Gastaut syndrome (LGS) |
| **Required Medical Information:** | • Documented diagnosis of Dravet syndrome (DS) or Lennox-Gastaut Syndrome (LGS)<br>• Current weight<br>• Documentation that therapy is being used as adjunct therapy for seizures<br><br>**Dravet Syndrome**<br>• Documentation of at least 6 convulsive seizures in the last 6 weeks while on stable antiepileptic drug therapy<br><br>**Lennox-Gastaut Syndrome (LGS)**<br>• Documentation of at least 8 drop seizures per month while on stable antiepileptic drug therapy |
| **Appropriate Treatment Regimen & Other Criteria:** | **Dravet Syndrome**<br>• Documented treatment and inadequate control of seizures with Epidiolex AND at least four of the following therapies:<br>  ○ Valproate, clobazam, clonazepam, levetiracetam, zonisamide, or topiramate<br><br>**Lennox-Gastaut Syndrome (LGS)**<br>• Documented treatment and inadequate control of seizures with Epidiolex AND at least three guideline directed therapies:<br>  ○ Valproate, lamotrigine, rufinamide, topiramate, felbamate, or clobazam<br><br>• **Dosing:** not to exceed 26 mg daily<br><br>**Reauthorization** requires documentation of treatment success and a reduction in seizure severity, frequency, or duration |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**FINERENONE**
Affected Medications: KERENDIA (finerenone)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Chronic kidney disease (CKD) associated with type 2 diabetes (T2DM) to reduce the risk of:<br>  ▪ Sustained estimated glomerular filtration rate (eGFR) decline<br>  ▪ End-stage kidney disease<br>  ▪ Cardiovascular death<br>  ▪ Non-fatal myocardial infarction<br>  ▪ Hospitalization for heart failure<br>  ○ Heart failure with left ventricular ejection fraction (LVEF) ≥ 40% to reduce the risk of:<br>  ▪ Cardiovascular death<br>  ▪ Hospitalization for heart failure<br>  ▪ Urgent heart failure visits |
| **Required Medical Information:** | **<u>CKD associated with T2DM</u>**<br>• Documentation of all the following:<br>  ○ eGFR greater than or equal to 25 mL/min/1.73 $m^2$<br>  ○ Urine albumin-to-creatinine ratio (UACR) greater than or equal to 30 mg/g<br>  ○ Serum potassium level less than or equal to 5.0 mEq/L<br><br>**<u>Heart Failure with LVEF ≥ 40%</u>**<br>• Documentation of all the following:<br>  ○ Heart failure with LVEF of 40% or more<br>  ○ eGFR greater than or equal to 25 mL/min/1.73 $m^2$<br>  ○ Serum potassium level less than or equal to 5.0 mEq/L |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>CKD associated with T2DM</u>**<br>• Currently receiving maximally tolerated dosage of an angiotensin converting enzyme (ACE) inhibitor or angiotensin receptor blocker (ARB), unless intolerant or contraindicated<br>• Documented treatment failure or intolerable adverse event to at least 12 weeks of sodium-glucose cotransporter 2 (SGLT2) inhibitor therapy<br><br>**<u>Heart Failure with LVEF ≥ 40%</u>**<br>• Documented treatment failure or intolerable adverse event to at least 12 weeks of each of the following, unless intolerant or contraindicated:<br>  ○ Sodium-glucose cotransporter 2 (SGLT2) inhibitor therapy<br>  ○ Mineralocorticoid receptor antagonist (MRA) therapy, such as spironolactone or eplerenone<br><br>**<u>Reauthorization</u>** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a nephrologist, endocrinologist, or cardiologist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**FLUCYTOSINE**
Affected Medications: FLUCYTOSINE

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved or compendia supported indications not otherwise excluded by plan design<br>  ○ Treatment of systemic Candida infections<br>  ▪ Cardiac infection, native or prosthetic valve endocarditis, or device infection<br>  ▪ Central nervous system (e.g., meningitis)<br>  ▪ Endophthalmitis<br>  ▪ Urinary tract infection (symptomatic cystitis, pyelonephritis)<br>  ○ Treatment of systemic Cryptococcus infections<br>  ▪ Meningitis<br>  ▪ Disseminated disease<br>  ▪ Severe pulmonary infection |
| **Required Medical Information:** | • Susceptibility cultures matching flucytosine activity<br>• Candida urinary tract infection: Documentation of fluconazole-resistant *C. glabrata*<br>• Endophthalmitis: Documentation of fluconazole- or voriconazole-resistant isolates |
| **Appropriate Treatment Regimen & Other Criteria:** | • FDA-approved or compendia supported dose, frequency and duration of therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an infectious disease specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 8 weeks, unless otherwise specified |

POLICY NAME:
**FLUOCINOLONE OCULAR IMPLANT**
Affected Medications: ILUVIEN, RETISERT, YUTIQ

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Diabetic macular edema (DME)<br>  ○ Chronic, non-infectious posterior uveitis |
| **Required Medical Information:** | **<u>DME (Iluvien)</u>**<br>• Diagnosis of clinically significant diabetic macular edema<br>• Documentation of past treatment with corticosteroids without a clinically significant rise in intraocular pressure<br><br>**<u>Uveitis (Retisert, Yutiq, and Iluvien)</u>**<br>• Diagnosis of chronic, non-infectious posterior uveitis confirmed by slit lamp and fundoscopic examination |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>DME (Iluvien)</u>**<br>• Documentation of inadequate response or intolerance to an intravitreal vascular endothelial growth factor (VEGF) inhibitor (preferred products: Avastin, Byooviz)<br>• Documentation of inadequate response to laser photocoagulation<br><br>**<u>Uveitis (Retisert, Yutiq, and Iluvien)</u>**<br>• Documentation of inadequate response or intolerance to all of the following:<br>  ○ Minimum 12-week trial with oral systemic corticosteroid<br>  ○ At least one corticosteroid-sparing immunosuppressive therapy (methotrexate, azathioprine, or mycophenolate mofetil)<br>  ○ At least one calcineurin inhibitor (cyclosporine, tacrolimus)<br>• For Retisert: Documentation of treatment failure with Yutiq |
| **Exclusion Criteria:** | • Active or suspected ocular or periocular infections<br>• Concurrent use of intravitreal implants or injections (corticosteroid, anti-VEGF)<br>• **Iluvien:** Glaucoma (with cup to disc ratios greater than 0.8) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **Iluvien**<br>• Authorization: 36 months, unless otherwise specified<br>**Retisert**<br>• Authorization: 30 months, unless otherwise specified<br>**Yutiq**<br>• Authorization: 36 months, unless otherwise specified |

POLICY NAME:
**FDA APPROVED DRUG – Drug or Indication Not Yet Reviewed By Plan for Formulary Placement**
Affected Medications: New Medications, Formulations, or Indications of Existing Drugs that are Under Review by Plan for Formulary Placement

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| **Covered Uses:** | • Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Documentation of disease state, level of control, and therapies failed<br>• Documentation of failure with all available formulary products for treatment of disease state<br>• Documentation that delay in treatment will cause loss of life, limb, function or other extreme pain |
| **Appropriate Treatment Regimen & Other Criteria:** | • Drug must be dosed according to package insert requirements |
| **Exclusion Criteria:** | • Exclusion based on package insert requirements |
| **Age Restriction:** | • Age based on package insert requirements |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a provider experienced in the management of the diagnosis<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Case by case based on member need |

POLICY NAME:
**FOSTAMATINIB**
Affected Medications: TAVALISSE (fostamatinib)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Thrombocytopenia in adults with chronic immune thrombocytopenia (ITP) who have had an insufficient response to a previous treatment |
| **Required Medical Information:** | **<u>Thrombocytopenia in patients with chronic ITP</u>**<br>• Documentation of **ONE** of the following:<br>  ○ Platelet count less than 20,000/microliter<br>  ○ Platelet count less than 30,000/microliter AND symptomatic bleeding<br>  ○ Platelet count less than 50,000/microliter AND increased risk for bleeding (such as peptic ulcer disease, use of antiplatelets or anticoagulants, history of bleeding at higher platelet count, need for surgery or invasive procedure) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Thrombocytopenia in patients with chronic ITP</u>**<br>• Documentation of inadequate response, defined as platelets did not increase to at least 50,000/microliter, to the following therapies:<br>  ○ **ONE** of the following:<br>  ▪ Inadequate response with at least 2 therapies for immune thrombocytopenia, including corticosteroids, rituximab, or immunoglobulin<br>  ▪ Splenectomy<br>  ○ Eltrombopag olamine<br><br>**<u>Reauthorization</u>:**<br>• Response to treatment with platelet count of at least 50,000/microliter or above (not to exceed 400,000/microliter) |
| **Exclusion Criteria:** | • Use in combination with a thrombopoietin receptor agonist, spleen tyrosine kinase inhibitor, or similar treatment for thrombocytopenia (such as eltrombopag olamine, Doptelet, or Nplate) |
| **Age Restriction:** | |
| **Prescriber Restrictions:** | • Prescribed by, or consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**FUMARATES FOR MULTIPLE SCLEROSIS**
Affected Medications: BAFIERTAM (monomethyl fumarate), VUMERITY (diroximel fumarate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:<br>  ▪ Clinically isolated syndrome (CIS)<br>  ▪ Relapsing-remitting multiple sclerosis (RRMS)<br>  ▪ Active secondary progressive disease (SPMS) |
| **Required Medical Information:** | • Diagnosis confirmed with magnetic resonance imaging (MRI) per revised McDonald diagnostic criteria for MS<br>  ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS |
| **Appropriate Treatment Regimen & Other Criteria:** | • Coverage of Bafiertam (monomethyl fumarate) or Vumerity (diroximel fumarate) requires documentation of **ONE** of the following:<br>  ○ Treatment failure with (or intolerance to) **TWO** of the following: dimethyl fumarate, fingolimod, teriflunomide<br>  ○ Currently receiving treatment with Bafiertam (monomethyl fumarate) or Vumerity (diroximel fumarate), excluding via samples or manufacturer's patient assistance program<br><br>**Reauthorization** requires provider attestation of treatment success |
| **Exclusion Criteria:** | • Concurrent use of other disease-modifying medications indicated for the treatment of MS |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or MS specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 24 months, unless otherwise specified |

POLICY NAME:
**FYARRO**
Affected Medications: FYARRO (nab-sirolimus)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | **Perivascular Epithelioid Cell Tumor (PEComa)**<br>• Presence of malignant locally advanced unresectable or metastatic disease confirmed by pathology.<br>• History of intolerable adverse event with trial of each of the following agents:<br>  ○ Sirolimus oral tablet<br>  ○ Everolimus or temsirolimus<br><br>Reauthorization: documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• History of disease progression with prior mechanistic target of rapamycin (mTOR) inhibitor treatment. |
| **Age Restriction:** | |
| **Prescriber Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**GABA-A RECEPTOR MODULATORS**
Affected Medications: ZULRESSO (brexanolone), ZURZUVAE (zuranolone)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of postpartum depression (PPD) |
| **Required Medical Information:** | • Documented major depressive episode with peripartum onset as defined by the *Diagnostic and Statistical Manual of Mental Health Disorders*, Fifth Edition (DSM-5) criteria:<br>  ○ At least **five** of the following symptoms have been present during the same 2-week period and represent a change from previous functioning (must include either (1) depressed mood or (2) lack of interest or pleasure):<br>  (1). Depressed mood most of the day, nearly every day, as indicated by either subjective report or observation made by others (in adolescents, may present as irritable mood)<br>  (2). Markedly diminished interest or pleasure in all (or almost all) activities most of the day, nearly every day, as indicated by either subjective account or observation<br>  (3). Significant weight loss when not dieting, weight gain, or decrease or increase in appetite nearly every day (in adolescents, consider failure to make expected weight gain)<br>  (4). Insomnia or hypersomnia nearly every day<br>  (5). Psychomotor agitation or retardation nearly every day (observable by others, not merely subjective feelings of restlessness or being slowed down)<br>  (6). Fatigue or loss of energy nearly every day<br>  (7). Feelings of worthlessness, or excessive or inappropriate guilt nearly everyday<br>  (8). Diminished ability to think or concentrate, or indecisiveness, nearly everyday (subjective account or observed by others)<br>  (9). Recurrent thoughts of death (not just fear of dying), recurrent suicidal ideation without a specific plan, or a suicide attempt or a specific plan for committing suicide<br>  ○ Symptoms cause clinically significant distress or impairment in social, occupational, or other important areas of functioning<br>  ○ Episode is not attributable to the direct physiological effects of a substance or to another condition<br>• Major depressive episode began no earlier than the third trimester and no later than the first 4 weeks following delivery<br>• Moderate to severe postpartum depression documented by one of the following rating scales:<br>  ○ Hamilton Rating Scale for Depression (HAM-D) score of greater than 17<br>  ○ Patient Health Questionnaire-9 (PHQ-9) score of greater than 10<br>  ○ Montgomery-Åsberg Depression Rating Scale (MADRS) greater than 20 points<br>  ○ Edinburgh Postnatal Depression Scale (EPDS) score of greater than 13 |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented trial with an oral antidepressant for at least 8 weeks unless contraindicated or documentation shows that the severity of the depression would place the health of the mother or infant at significant risk<br>• For Zulresso requests: documented treatment failure with Zurzuvae |

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| **Exclusion Criteria:** | • Greater than 6 months postpartum |
| **Age Restriction:** | • 15 years of age and older for Zulresso<br>• 18 years of age and older for Zurzuvae |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a psychiatrist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 1 month, one time approval per pregnancy, unless otherwise specified |

POLICY NAME:
**GANAXOLONE**
Affected Medications: ZTALMY (ganaxolone)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of seizures associated with cyclin-dependent kinase-like 5 (CDKL5) deficiency disorder (CDD) in patients 2 years of age and older |
| **Required Medical Information:** | • Documentation of CDKL5 mutation confirmed by genetic testing<br>• Documentation of inadequately controlled seizures despite current treatment |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with at least two therapies for seizure management<br><br>**Reauthorization** will require documentation of treatment success defined as a reduction in seizure frequency when compared to baseline |
| **Exclusion Criteria:** | • West syndrome<br>• Seizures of a predominantly infantile spasm type |
| **Age Restriction:** | • 2 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**GIVINOSTAT**
Affected Medications: DUVYZAT (givinostat)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Duchenne muscular dystrophy (DMD) in patients 6 years of age and older |
| **Required Medical Information:** | • Genetically confirmed diagnosis of DMD<br>• Documentation of being ambulatory without needing an assistive device such as a wheelchair, walker, or cane<br>• North Star Ambulatory Assessment (NSAA) scale total score of 17 or more<br>• Baseline motor function assessment from one of the following:<br>  ○ 4-stair climb (4SC) test<br>  ○ Time to Stand Test (TTSTAND)<br>  ○ 6-minute walk test (6MWT)<br>  ○ North Star Ambulatory Assessment (NSAA)<br>  ○ Motor Function Measure (MFM)<br>  ○ Hammersmith Functional Motor Scale (HFMS)<br>• Current weight and planned treatment regimen |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of being on a stable dose of an oral corticosteroid such as prednisone for at least 6 months, and will continue while on Duvyzat unless contraindicated<br><br>**Reauthorization** requires a documented improvement from baseline or stabilization of motor function demonstrated by a motor function assessment tool |
| **Exclusion Criteria:** | • Concomitant therapy or within the past 6 months with DMD-directed antisense oligonucleotides such as golodirsen, casimersen, viltolarsen, eteplirsen<br>• Prior to starting therapy, platelet count less than 150,000 cells/microliter<br>• During therapy, QTc interval exceeds 500 ms or increases by more than 60 ms from baseline |
| **Age Restriction:** | • 6 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**GIVOSIRAN**
Affected Medications: GIVLAARI (givosiran)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of adults with acute hepatic porphyria (AHP) |
| **Required Medical Information:** | • Documentation of elevated urine porphobilinogen (PBG) levels based on specific lab test utilized<br>• Diagnosis confirmed based on Porphyria Genomic testing<br>• Documentation of baseline acute attack frequency<br>• Evaluation and elimination of exacerbating factors of porphyria attacks including certain medications, smoking, drinking, and infections |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of active disease defined as at least 2 documented porphyria attacks within the last six months, which can include hospitalization, urgent healthcare visits, or requiring intravenous Hemin administration<br><br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization** will require documentation of positive clinical response and a reduction in acute attack frequency from baseline |
| **Exclusion Criteria:** | • Active HIV, hepatitis C, or hepatitis B infection(s)<br>• History of pancreatitis<br>• Concomitant use with prophylactic hemin<br>• History of liver transplant |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, specialist in the treatment of acute hepatic porphyria<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**GLUCAGON-LIKE PEPTIDE (GLP-1) RECEPTOR AGONIST**
Affected Medications: TRULICITY, OZEMPIC, RYBELSUS, MOUNJARO, LIRAGLUTIDE

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Diabetes Mellitus, Type 2 (T2DM)<br>  ○ T2DM and cardiovascular disease<br>  ○ T2DM and chronic kidney disease (CKD)<br>• Compendia-supported uses that will be covered<br>  ○ Metabolic dysfunction-associated steatohepatitis (MASH)<br>  ○ Moderate to severe obstructive sleep apnea (OSA) |
| **Required Medical Information:** | • Available information is reviewed including claims history, ICD10 codes, and previous fill history<br><br>**Type 2 Diabetes (T2DM)**<br>• Diagnosis of Type 2 diabetes confirmed with lab testing<br><br>**Metabolic dysfunction–associated steatohepatitis (MASH):**<br>• Diagnosis of noncirrhotic nonalcoholic steatohepatitis (NASH) or MASH with moderate to advanced (F2 to F3) liver fibrosis confirmed by ONE of the following:<br>  ○ Conclusive result from a well-validated non-invasive test such as:<br>  ▪ Fibroscan-AST (FAST) score<br>  ▪ MAST (score from MRI–proton density fat fraction, Magnetic resonance elastography [MRE], and serum AST)<br>  ▪ MEFIB (Fibrosis-4 Index greater than or equal to 1.6 and MRE greater than or equal to 3.3 kPa)<br>  ○ Liver biopsy (also required if non-invasive testing is inconclusive or other causes for liver disease have not been ruled out)<br>• Other causes for liver steatosis have been ruled out (such as alcohol-associated liver disease, chronic hepatitis C, Wilson disease, drug-induced liver disease)<br>• Baseline lab values for AST and ALT<br><br>**Obstructive sleep apnea (OSA)**<br>• Diagnosis of moderate to severe obstructive sleep apnea (OSA) with Apnea-Hypopnea Index (AHI) of at least 15 on polysomnography or home sleep study from the past 5 years<br>• Body mass index (BMI) of 30 $kg/m^2$ or greater<br>• Current symptoms of OSA such as excessive daytime sleepiness, loud snoring, choking, gasping, pauses in breathing, sleep arousals, and difficulty maintaining sleep throughout the night<br>• Documentation of being used in combination with a physician-directed weight loss program that involves a reduced calorie diet, increased physical activity, and behavioral modification |
| **Appropriate Treatment Regimen & Other Criteria:** | **MASH:**<br>• Documentation of abstinence from alcohol consumption<br>• Documentation of comprehensive comorbidity management being undertaken, including all the following:<br>  ○ Use of diet and exercise for weight management |

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| | ○ Medications to manage associated comorbid conditions, such as thyroid disease (must not have active disease), diabetes, dyslipidemia, hypertension, or cardiovascular conditions.<br><br>**Obstructive sleep apnea (OSA)**<br>• Documentation of one of the following:<br>○ Treatment failure with positive airway pressure (PAP) therapy, defined as having continued symptoms of OSA despite four or more hours of PAP use per night for 70 percent or more of nights<br>○ Contraindication to PAP therapy (such as upper airway anatomic abnormalities, pneumothorax or invasive mechanical ventilation)<br>• Documentation of being prescribed concurrently with positive airway pressure (PAP) therapy, unless contraindicated or clinically significant adverse effects are experienced<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy<br>• **MASH:**<br>○ Documentation of disease responsiveness to therapy based on improvements or stability in laboratory results, such as ALT and AST, or fibrosis as evaluated by a non-invasive test<br>• **OSA:**<br>○ Improvement in AHI score or OSA symptoms such as excessive daytime sleepiness, loud snoring, choking, gasping, pauses in breathing, sleep arousals, and difficulty maintaining sleep throughout the night<br>○ Achieved and maintained 10 percent or greater weight loss after starting Zepbound (tirzepatide)<br>○ Continued adherence to a reduced calorie diet, increased physical activity, and behavioral modification program |
| **Exclusion Criteria:** | • Use for weight loss or other excluded diagnosis<br>• Dosing above Food and Drug Administration (FDA) approved label for treatment of diabetes<br>• Use in patients who have achieved remission of diabetes (defined as a return of HbA1c to less than 6.5% that occurs spontaneously or following an intervention and that persists for at least three months in the absence of usual glucose-lowering pharmacotherapy)<br>• Polycystic kidney disease or glomerulonephritis<br>• Diagnosis of central or mixed sleep apnea<br>• Diagnosis of obesity hypoventilation syndrome or daytime hypercapnia<br>• History of ketoacidosis<br>• Concurrent use of other glucagon-like peptide-1 (GLP-1) receptor agonists |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 24 months, unless otherwise specified |

POLICY NAME:
**GONADOTROPIN**
Affected Medications: CHORIONIC GONADOTROPIN, PREGNYL, NOVAREL

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Hypogonadotropic hypogonadism secondary to a pituitary deficiency in males<br>  ○ Prepubertal cryptorchidism not caused by anatomic obstruction<br>• Perioperative use in male infants/toddlers with hypospadias and chordee OR total epispadias and bladder exstrophy |
| **Required Medical Information:** | **Hypogonadotropic hypogonadism secondary to a pituitary deficiency in males:**<br>• Documentation confirming the diagnosis |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Use for the diagnosis or treatment of infertility (if benefit exclusion)<br>• Obesity<br>• Prevention of recurrent or habitual miscarriage<br>• Treatment or prevention of breast cancer |
| **Age Restriction:** | • Prepubertal cryptorchidism: generally, between 4 and 9 years of age<br>• Hypospadias or epispadias: infant or toddler |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**GOSERELIN ACETATE IMPLANT**
Affected Medications: ZOLADEX (goserelin acetate implant)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Endometriosis<br>  ○ Endometrial thinning<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | **Endometriosis:**<br><br>• Documentation of moderate to severe pain due to endometriosis |
| **Appropriate Treatment Regimen & Other Criteria:** | **Endometriosis:**<br><br>• Documentation of a trial and inadequate relief (or contraindication) after at least 3 months of both of the following first-line therapies:<br>  ○ Nonsteroidal anti-inflammatory drugs (NSAIDs)<br>  ○ Continuous (no placebo pills) hormonal contraceptives<br><br>**Endometrial thinning:**<br><br>• Documentation of both of the following:<br>  ○ Diagnosis of dysfunctional uterine bleeding<br>  ○ Planning to use as an endometrial-thinning agent prior to endometrial ablation<br><br>**Reauthorization for oncologic uses** require documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• For endometriosis, prior use of Zoladex for a 6-month period |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • For oncologic uses: Prescribed by, or in consultation with, an oncologist<br>• For gynecologic uses: Prescribed by, or in consultation with, a gynecologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **Oncologic uses:**<br>• Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br><br>**Endometriosis:**<br>• Authorization: 6 months with no reauthorization, unless otherwise specified<br><br>**Endometrial thinning:**<br>• Authorization: 4 months (up to 2 doses only), unless otherwise specified |

POLICY NAME:
**GROWTH HORMONES**
Affected Medications: GENOTROPIN, GENOTROPIN MINIQUICK, HUMATROPE, NORDITROPIN FLEXPRO, NUTROPIN AQ NUSPIN, OMNITROPE, SAIZEN, SKYTROFA, ZOMACTON, SOGROYA, NGENLA

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | **<u>All indications:</u>**<br>• Documentation of baseline height, height velocity, bone age (pediatrics), and patient weight<br><br>**<u>Growth hormone deficiency or Pituitary dwarfism</u>**<br>• For initial approval, documentation of the following is required:<br>o Diagnosis of growth hormone deficiency or pituitary dwarfism AND<br>o Low serum values for GH stimulation test, IGF-1, and IGFBP-3 with delayed bone age AND<br>▪ Height standard deviation score (SDS) of -2.5 (0.6$^{th}$ percentile) OR<br>▪ Height velocity impaired AND<br>▪ Height SDS of -2 (2.3rd percentile) for bone age<br><br>**<u>Turner's syndrome</u>**<br>• For initial approval, documentation of the following is required:<br>o Diagnosis of Turner Syndrome done through genetic testing AND<br>▪ For patients less than 2 years of age:<br>• Documented 50% delay in growth from projected based on World Health Organization (WHO) growth curves at equivalent age, AND<br>• No secondary factor present that would explain observed growth delays<br>▪ For patients greater than or equal to 2 years of age:<br>• Height below the 5th percentile for bone age, AND<br>• No secondary factor present that would explain observed growth delays<br><br>**<u>Noonan's syndrome</u>**<br>• For initial approval, documentation of the following is required:<br>o Diagnosis of Noonan's syndrome done through genetic testing AND<br>▪ Height standard deviation score (SDS) of -2.5 (0.6$^{th}$ percentile) OR<br>▪ Height velocity impaired AND<br>▪ Height SDS of -2 (2.3rd percentile) for bone age<br><br>**<u>Short stature homeobox-containing gene (SHOX) deficiency</u>**<br>• For initial approval, documentation of the following is required:<br>o Diagnosis of SHOX deficiency done through genetic testing<br>▪ Height standard deviation score (SDS) of -2.5 (0.6$^{th}$ percentile) OR<br>▪ Height velocity impaired AND<br>▪ Height SDS of -2 (2.3rd percentile) for bone age |

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| | **<u>Chronic kidney disease stage 3 and greater OR kidney transplant</u>**<br>• For initial approval, documentation of the following is required:<br>  ○ Diagnosis of chronic kidney disease stage 3 or higher (CrCl less than 60mL/min)<br>  ○ Height velocity (SDS) less than -1.88 for bone age.<br><br>**<u>Prader-Willi syndrome</u>**<br>• For initial approval, documentation of the following is required:<br>  ○ Diagnosis of Prader-Willi syndrome through genetic testing **AND**<br>  ○ Height velocity impaired<br><br>**<u>Short Stature born small for gestational age (SGA) with no catch-up growth by 2 years to 4 years of age</u>**<br>• Birth weight and/or length of at least 2 standard deviations (-2 SD) from the mean for gestational age and sex<br>• Height standard deviation score (SDS) of -2.5 (0.6$^{th}$ percentile)<br>• Age at start of growth hormone therapy cannot be greater than 10 years<br>• Exclusion of other causes of short stature including growth-inhibiting medication, chronic disease, endocrine disorders<br><br>**<u>Adult Growth Hormone Deficiency:</u>**<br>• For initial approval, documentation of the following is required:<br>  ○ Dose and frequency are appropriate **AND**<br>  ○ Documented Growth Hormone Deficiency **AND**<br>  ○ Documented IGF-1 outside reference range for patient's sex and age, AND the patient has failed one growth hormone stimulation test (insulin tolerance test-ITT or Glucagon stimulation test when ITT is contraindicated)<br><br>**<u>Reauthorization:</u>**<br>• **Pediatric Indications**: requires a documented growth rate increase of at least 2.5 cm over baseline per year **AND** evaluation of epiphyses (growth plates) documenting they remain open<br>• **Adult Growth Hormone Deficiency:** requires documented clinical improvement and IGF-I within normal reference range for age and sex |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented trial and failure of at least 12 weeks of Norditropin or Genotropin prior to any other daily growth hormone<br>• For Skytrofa, Sogroya, and Ngenla:<br>  ○ Documented trial and failure of at least 12 weeks of both of the following:<br>  ▪ Norditropin or Genotropin<br>  ▪ One additional daily growth hormone |
| **Exclusion Criteria:** | • Pregnancy<br>• Elderly adults with age-adjusted low IGF-1 levels and no history of pituitary or hypothalamic disease.<br>• Growth Hormone (GH) replacement to enhance athletic performance<br>• Diagnosis of: Idiopathic Short Stature (ISS), height standard deviation score (SDS) less than -2.25, and associated with growth rates unlikely to permit attainment of adult height in the normal range |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist<br>• All approvals are subjects to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**HEPATITIS C DIRECT-ACTING ANTIVIRALS**
Affected Medications: MAVYRET (glecaprevir & pibrentasvir), Vosevi (Sofosbuvir/Velpatasvir/Voxilaprevir), Sofosbuvir/Velpatasvir

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>• AASLD (American Association for the Study of Liver Diseases)-supported use with class I or class IIa-Level A recommendation |
| **Required Medical Information:** | • Documentation of acute (Mavyret only) or chronic hepatitis C virus (HCV) by liver biopsy or by Food and Drug Administration (FDA)-approved serum blood test<br>• Current HIV status<br>• Current Hepatitis B status<br>• Baseline HCV RNA level within last 3 months<br>• Documentation that patient is one of the following:<br>  ○ Treatment-naïve<br>  ○ Treatment experienced, including documentation of previous treatment regimen and outcome<br>• Current documentation of hepatic impairment severity with Child-Pugh Classification **OR** bilirubin, albumin, INR, ascites status, and encephalopathy status to calculate Child-Pugh score, within 12 weeks prior to anticipated start of therapy<br>• Expected survival from non-Hepatitis C-associated morbidity is greater than 12 months |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose/duration or according to the most recently updated AASLD guideline recommendation (See table below) |
| **Exclusion Criteria:** | • Mavyret is contraindicated in patients with moderate and severe hepatic impairment (Child-Pugh B and C)<br>• Vosevi is not recommended in patients with moderate or severe hepatic impairment (Child-Pugh class B or C)<br>• Concurrent use of Vosevi with rifampin is contraindicated |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • See Appropriate Treatment Regimen & Other Criteria |

**Recommended Treatment Regimens for Adults and Adolescents 12 years of age and older with Acute (Mavyret only) or Chronic Hepatitis C virus**

| Treatment History | Cirrhosis Status | Recommended Regimen |
|---|---|---|
| **Treatment Naïve (Genotype 1-6)** | | |
| DAA-Treatment naïve, confirmed reinfection or prior treatment with | Non-cirrhotic | SOF/VEL x 12 weeks<br>Mavyret x 8 weeks |

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| PEG/RBV | Compensated Cirrhosis | SOF/VEL x 12 weeks<br>Mavyret x 8 weeks |
| | Decompensated Cirrhosis | SOF/VEL + RBV x 12 weeks<br>SOF/VEL x 24 weeks (if ribavirin ineligible*) |
| **Treatment Experienced (Genotype 1-6)** | | |
| Sofosbuvir based regimen treatment failures, including:<br>- Sofosbuvir + ribavirin<br>- Ledipasvir/sofosbuvir (Harvoni)<br>- SOF/VEL | Non-cirrhotic or compensated cirrhosis | Vosevi x 12 weeks<br>Mavyret x 16 weeks (except genotype 3) |
| Elbasvir/grazoprevir (Zepatier) treatment failures | Non-cirrhotic or compensated cirrhosis | Vosevi x 12 weeks |
| Mavyret treatment failures | Non-cirrhotic or compensated cirrhosis | Mavyret + SOF + RBV x 16 weeks<br>Vosevi x 12 weeks (plus RBV if compensated cirrhosis) |
| Multiple DAA treatment failures, including:<br>- Vosevi<br>- Mavyret + sofosbuvir | Non-cirrhotic or compensated cirrhosis | Mavyret + SOF + RBV x 16-24 weeks<br>Vosevi + RBV x 24 weeks |
| Abbreviations: DAA = direct-acting antiviral; PEG = pegylated interferon; RBV = ribavirin; SOF/VEL = sofosbuvir/velpatasvir<br><br>*Ribavirin ineligible/intolerance may include: 1) neutrophils less than 750 mm3, 2) hemoglobin less than 10 g/dL, 3) platelets less than 50,000 cells/mm3, autoimmune hepatitis or other autoimmune condition, hypersensitivity or allergy to ribavirin | | |

**Recommended Treatment Regimens for children ages 3 to 12 years of age with Acute (Mavyret only) or Chronic Hepatitis C virus**

| Treatment History | Cirrhosis Status | Recommended Regimen |
|---|---|---|
| **Treatment Naïve (Genotype 1-6)** | | |
| DAA-Treatment naïve, confirmed reinfection or prior treatment with PEG/RBV | Non-cirrhotic or compensated cirrhosis | SOF/VEL x 12 weeks<br>Mavyret x 8 weeks |
| | Decompensated Cirrhosis | SOF/VEL + RBV x 12 weeks |
| **Treatment Experienced** | | |
| Efficacy and safety is extremely limited in treatment experienced patients in this population. Can consider recommended treatment regimens in adults if FDA approved for pediatric use. Recommend consulting with hepatologist. | | |
| Abbreviations: DAA = direct-acting antiviral; PEG = pegylated interferon; RBV = ribavirin; SOF/VEL = sofosbuvir/velpatasvir | | |

**Recommended dosage of SOF/VEL in pediatric patients 3 years of age and older**

| Body Weight | Dosing of SOF/VEL |
|---|---|
| Less than 17kg | One 150mg/37.5mg pellet packet once daily |
| 17kg to less than 30kg | One 200mg/50mg pellet packet OR tablet once daily |
| At least 30kg | Two 200mg/50mg pellet packets once daily OR one 400mg/100mg tablet once daily |

**Recommended dosage of Mavyret in pediatric patients 3 years of age and older**

| Body Weight | Dosing of Mavyret |
|---|---|
| Less than 20kg | Three 50mg/20mg pellet packets once daily |
| 20kg to less than 30kg | Four 50mg/20mg pellet packets once daily |
| 30kg to less than 45kg | Five 50mg/20mg pellet packets once daily |
| 45kg and greater OR 12 years of age and older | Three 100mg/40mg tablets once daily |

POLICY NAME:
**HISTRELIN**
Affected Medications: SUPPRELIN LA (histrelin acetate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Central precocious puberty (CPP)<br>• Gender dysphoria |
| **Required Medical Information:** | **Central Precocious Puberty:**<br>• Documentation of CPP confirmed by basal luteinizing hormone (LH), follicle-stimulating hormone (FSH), and either estradiol or testosterone concentrations |
| **Appropriate Treatment Regimen & Other Criteria:** | **All Indications:**<br>• Approval requires documented treatment failure with leuprolide<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 2 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Central Precocious Puberty: Prescribed by, or in consultation with, an endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**HEREDITARY ANGIOEDEMA**
Affected Medications: BERINERT, ICATIBANT ACETATE, SAJAZIR, RUCONEST, KALBITOR, CINRYZE, HAEGARDA, TAKHZYRO, ORLADEYO, ANDEMBRY, DAWNZERA

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Hereditary angioedema attacks, prophylaxis (Cinryze, Haegarda, Takhzyro, Orladeyo, Andembry, Dawnzera)<br>  ○ Hereditary angioedema attacks, acute treatment (Berinert, icatibant acetate, Sajazir, Kalbitor, Ruconest, Ekterly) |
| **Required Medical Information:** | **Diagnosis of hereditary angioedema (HAE) classified as one of the following:**<br>• Type I or II HAE confirmed by low C4 levels AND one of the following:<br>  ○ Low C1 inhibitor functional or antigenic level less than 50% of the lower limit of normal as defined by the laboratory performing test<br>• "Type III" HAE confirmed by normal C4, C1 inhibitor (functional and antigenic) with one of the following:<br>  ○ Genetic testing confirming presence of HAE causing mutation such as mutation of coagulation factor XII gene (F12 mutation), mutation in the angiopoietin-1 gene, mutation in the plasminogen gene, mutation in the kininogen 1 gene, mutation in the myoferlin gene, mutation in the heparan sulfate 3-Osulfotransferase 6 gene<br>  ○ Family history of HAE AND documented recurring angioedema attacks that are refractory to high dose antihistamines (four times the usual dose)<br>• Documented full treatment plan and current body weight<br>• Documentation of number of attacks requiring treatment in the past year |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Acute Treatment</u>:**<br>• Documented history of one of the following:<br>  ○ Non-inflammatory subcutaneous angioedema (without hives) which is recurrent and lasts greater than 12 hours<br>  ○ Abdominal pain without a clear organic cause lasting greater than 6 hours<br><br>Coverage for non-preferred products (Berinert, Kalbitor, Ruconest, Ekterly) requires documentation of one of the following:<br>• Documented treatment failure to one of the preferred products: icatibant acetate or Sajazir<br>• Currently receiving treatment with a non-preferred product, excluding via samples or manufacturer's patient assistance programs<br><br>For requests to treat more than 3 attacks per month:<br>• Documentation of current treatment with, or failure, intolerance, or clinical rationale for avoidance of, prophylactic therapies<br>• Authorization for acute treatment will provide a sufficient quantity to treat the average number of acute attacks per month plus 1 additional dose<br><br>**<u>Prophylaxis Treatment</u>:** |

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| | • History of TWO or more severe attacks per month for the past 3 months (airway swelling, debilitating cutaneous or gastrointestinal episodes) despite short term treatment and at least one of the following:<br>  ○ Disabling symptoms for at least 5 days per month<br>  ○ History of at least one laryngeal attack caused by HAE<br>• Avoidance of possible triggers for HAE attacks such as<br>  ○ estrogen containing oral contraceptives/hormone replacement<br>  ○ angiotensin-converting-enzyme (ACE) inhibitors<br>  ○ dipeptidyl peptidase IV (DPP-4) inhibitors<br>  ○ Neprilysin inhibitor<br><br>Coverage for non-preferred products (Cinryze, Orladeyo, Andembry, Dawnzera) requires documentation of one of the following:<br>• Documented treatment failure to the preferred products Haegarda and Takhzyro<br>• Currently receiving treatment with a non-preferred product, excluding via samples or manufacturer's patient assistance programs<br><br>**Reauthorization** requires documentation of number of acute HAE attacks treated in the past year AND documentation of treatment success defined as reduction of frequency and severity of HAE attack episodes requiring acute therapy by greater than or equal to 50% from baseline.<br><br>• Requested dose within the Food and Drug Administration (FDA)-approved label<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced for all medical infusion drugs |
| **Exclusion Criteria:** | • Concurrent use of multiple HAE prophylactic treatments (Orladeyo, Haegarda, Takhzyro, Cinryze, Andembry, Dawnzera)<br>• Concurrent use of multiple HAE acute treatments (Berinert, Kalbitor, Runconest, icatibant acetate, Sajazir, Ekterly) |
| **Age Restriction:** | • Product specific per FDA labeled indication |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an allergist, immunologist, or pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**NITISINONE**
Affected Medications: NITISINONE

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Hereditary tyrosinemia type 1 (HT-1)<br>  ○ Alkaptonuria (AKU) |
| **Required Medical Information:** | • Diagnosis of hereditary tyrosinemia type 1 confirmed by:<br>  ○ Presence of succinylacetone (SA) in urine or blood<br>  ○ Genetic testing showing a mutation in the gene encoding fumarylacetoacetate hydrolase (FAH)<br>• Diagnosis of alkaptonuria confirmed by:<br>  ○ Quantitative measurement of homogentisic acid (HGA) in urine<br>  ○ Genetic testing showing a mutation in the homogentisic acid dioxygenase (HGD) gene<br>• Current patient weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Use as an adjunct to dietary restriction of tyrosine and phenylalanine<br><br>**Reauthorization** requires documentation of treatment success confirmed by:<br>• Reduction in urine or plasma succinylacetone (for HT-1) or homogentisic acid (for AKU) from baseline<br>• Documentation of dietary restriction of tyrosine and phenylalanine |
| **Exclusion Criteria:** | • Use without dietary restriction of tyrosine and phenylalanine |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist in the treatment of hereditary tyrosinemia or related disorders<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**HYDROCORTISONE ORAL GRANULES**
Affected Medications: ALKINDI SPRINKLE (hydrocortisone oral granules)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Glucocorticoid replacement therapy in pediatric patients with adrenocortical insufficiency |
| **Required Medical Information:** | • Diagnosis of adrenal insufficiency confirmed with an adrenal stimulation test<br>• Current body surface area (or height and weight to calculate)<br>• Current height and weight velocity<br>• For adolescents, evaluation of epiphyses (growth plates) documenting they remain open<br>• Complete treatment plan including dose in mg/m$^2$/day |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with a 6-month trial of two or more of the following:<br>  ○ Hydrocortisone tablets<br>  ○ Cortisone acetate tablets<br>  ○ Prednisolone or prednisone tablets<br>  ○ Compounded hydrocortisone oral capsules or solution<br>• **Dosing** is in accordance with FDA labeling and does not exceed the following:<br>  ○ Starting dose: 8-10 mg/m$^2$/day in 3 divided doses<br>  ○ When switching from other oral hydrocortisone formulations, use the same total hydrocortisone dosage<br>  ○ Infants with Congenital Adrenal Hyperplasia may start at a dose of 8-15 mg/m$^2$/day in 3 divided doses<br><br>**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Use in adolescents who have achieved their adult height<br>• Use for stress dosing<br>• Use in acute treatment of adrenal crisis or acute adrenal insufficiency<br>• Long term use with strong CYP3A4 inducers, unless medically necessary |
| **Age Restriction:** | • Less than 18 years of age |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a pediatric endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**HYFTOR**
Affected Medications: HYFTOR (sirolimus gel)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ For the treatment of facial angiofibroma (FA) associated with tuberous sclerosis complex (TSC) |
| **Required Medical Information:** | • Documented diagnosis of FA associated with TSC which are:<br>  ○ Rapidly changing in size and/or number<br>  ○ Causing functional interference, pain or bleeding<br>  ○ Inhibiting social interactions<br>• Current and baseline description of FA including lesion count, associated symptoms and complications, and overall severity |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with laser therapy and/or surgery (such as shave excision, cryotherapy, radiofrequency ablation, or dermabrasion), unless contraindicated<br><br>**Reauthorization** requires documentation of a positive clinical response to therapy (decrease in size and/or redness of facial angiofibromas) |
| **Exclusion Criteria:** | • Concurrent use of systemic mammalian target of rapamycin (mTOR) inhibitors<br>• Treatment of non-facial angiofibroma |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a dermatologist, oncologist, or neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**HYPOXIA-INDUCIBLE FACTOR PROLYL HYDROXYLASE (HIF PH) INHIBITORS**
Affected Medications: JESDUVROQ (daprodustat), VAFSEO (vadadustat)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Anemia due to chronic kidney disease (CKD) in adults who have been receiving dialysis |
| **Required Medical Information:** | • Diagnosis of anemia due to CKD<br>• Documentation of dialysis use for:<br>  ○ Jesduvroq: 4 or more months<br>  ○ Vafseo: 3 or more months<br>• Documentation of pretreatment hemoglobin level greater than 8 g/dL and less than 12 g/dL<br>• Adequate iron stores as indicated by current (within the last three months) serum ferritin level greater than or equal to 100 mcg/L or serum transferrin saturation greater than or equal to 20% |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of **ONE** of the following:<br>  ○ Documented hypo-responsiveness to an erythropoiesis stimulating agent (ESA), defined as the need for **ONE** of the following:<br>  ▪ Greater than 300 IU/kg per week of epoetin alfa<br>  ▪ Greater than 1.5 mcg/kg per week of darbepoetin<br>  ○ Intolerance to **BOTH** preferred ESA products epoetin alfa-epbx (Retacrit) and darbepoetin alfa (Aranesp)<br><br>**Reauthorization** requires documentation of treatment success and hemoglobin of greater than 8 g/dL and less than 12 g/dL |
| **Exclusion Criteria:** | • Use in combination with ESAs<br>• Current uncontrolled hypertension<br>• Active malignancy<br>• For Jesduvroq: Major adverse cardiac events (such as myocardial infarction, acute coronary syndrome, stroke, transient ischemic attack, venous thromboembolism) within 3 months prior to starting treatment |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist, such as a hematologist or nephrologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**IBREXAFUNGERP**
Affected Medications: BREXAFEMME (ibrexafungerp)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of vulvovaginal candidiasis (VVC)<br>  ○ Reduction in the incidence of recurrent vulvovaginal candidiasis (RVVC) |
| **Required Medical Information:** | **All Indications**<br>• Documented presence of signs/symptoms of current acute vulvovaginal candidiasis with a positive potassium hydroxide (KOH) test<br>• Documentation confirming that the patient is not pregnant and is on contraceptive for length of planned treatment<br><br>**RVVC**<br>• Documentation of three or more episodes of symptomatic vulvovaginal candidiasis infection within the past 12 months |
| **Appropriate Treatment Regimen & Other Criteria:** | **VVC**<br>• Documented treatment failure with both of the following for the current VVC episode:<br>  ○ Vaginally administered treatment (such as clotrimazole cream, miconazole cream, terconazole cream or suppository)<br>  ○ A 7-day course of fluconazole taken orally every third day for a total of 3 doses (days 1, 4, and 7)<br><br>**RVVC**<br>• Documented disease recurrence following 10 to 14 days of induction therapy with a topical antifungal agent or oral fluconazole, followed by fluconazole 150 mg once per week for 6 months<br><br>**Reauthorization** requires documentation of treatment success defined as a reduction in symptomatic vulvovaginal candidiasis episodes, and documentation supporting the need for additional treatment |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization (VVC): 3 months, unless otherwise specified<br>• Authorization (RVVC): 6 months, unless otherwise specified |

POLICY NAME:
**ILARIS**
Affected Medications: ILARIS (canakinumab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS), Hyperimmunoglobulin D syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD), Familial Mediterranean Fever (FMF), Adult-Onset Still's Disease (AOSD), Systemic Juvenile Idiopathic Arthritis (SJIA), Cryopyrin-Associated Periodic Syndromes (CAPS), Gout Flares |
| **Required Medical Information:** | **<u>Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS)</u>**<br>• Confirmed diagnosis of TRAPS with frequent and/or severe recurrent disease (such as recurrent fevers, prominent myalgias, migratory rash, periorbital edema) AND documented genetic defect of TNFRSF1A gene<br><br>**<u>Hyperimmunoglobulin D syndrome (HIDS)/ Mevalonate Kinase Deficiency (MKD)</u>**<br>• Confirmed diagnosis with one of the following:<br>  ○ Elevated serum IgD with or without elevated IgA<br>  ○ Genetic testing showing presence of heterozygous or homozygous mutation in the mevalonate kinase (MVK) gene<br>• Documentation of 3 or more febrile acute flares within a 6-month period<br><br>**<u>Still's Disease</u>**<br>• Confirmed diagnosis of Still's Disease, including Adult-Onset Still's Disease (AOSD) and Systemic Juvenile Idiopathic Arthritis (SJIA) in patients 2 years of age and older<br>• Documented clinical signs and symptoms including fever, rash, arthritis, arthralgia, myalgia, pharyngitis, pulmonary disease, elevated liver enzymes, C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), serum ferritin<br>**<u>Cryopyrin-Associated Periodic Syndromes (CAPS)</u>**<br>• Confirmed diagnosis of CAPS in patients 4 years and older including Familial Cold Autoinflammatory Syndrome (FCAS) or Muckle-Wells Syndrome (MWS) with one of the following:<br>  ○ Elevated inflammatory markers such as CRP and serum amyloid A with two of the following manifestations:<br>  ▪ Urticaria-like rash, cold-triggered episodes, sensorineural hearing loss, musculoskeletal symptoms, chronic aseptic meningitis, skeletal abnormalities<br>  ○ Genetic testing showing presence of NALP3 mutations<br><br>**<u>Gout Flares</u>**<br>• Confirmed diagnosis of gout that is refractory to standard therapies<br>• Documentation of having 3 or more gout flares in the past 12 months |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>TRAPS</u>**<br>• Documented clinical failure to episodic treatment with nonsteroidal anti-inflammatory drugs (NSAIDs), glucocorticoids (prednisone or prednisolone), and a minimum 12-week trial with Enbrel |

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| | **HIDS/MKD**<br>• Documented treatment failure to episodic treatment with nonsteroidal anti-inflammatory drugs (NSAIDs), glucocorticoids, and anakinra<br><br>**FMF**<br>• Documented treatment failure with maximal tolerable dose of colchicine (3 mg daily in adults and 2 mg daily in children)<br>• Documentation of frequent and/or severe recurrence disease despite adequate treatment with at least 12 weeks of anakinra<br><br>**Still's Disease**<br>• Documentation of frequent and/or severe recurrent disease despite adequate treatment with a minimum 12-week trial with each of the following:<br>  ○ NSAIDs or glucocorticoids<br>  ○ Methotrexate or leflunomide<br>  ○ Kineret (anakinra)<br>  ○ Actemra (tocilizumab)<br><br>**CAPS**<br>• Documentation of treatment failure with a minimum 12-week trial with anakinra<br><br>**Gout Flares**<br>• Documented treatment failure with all of the following for the symptomatic treatment of gout flares:<br>  ○ Prescription strength NSAIDs (naproxen, indomethacin, diclofenac, meloxicam, or celecoxib)<br>  ○ Colchicine<br>  ○ Glucocorticoids (oral or intraarticular)<br><br>**Reauthorization** requires documentation of treatment success |
| **Exclusion Criteria:** | • Treatment of neonatal onset multisystem inflammatory disorder (NOMID) or chronic infantile neurological cutaneous and articular syndrome (CINCA), rheumatoid arthritis, chronic obstructive pulmonary disease (COPD), type 2 diabetes mellitus<br>• Use in combination with tumor necrosis factor (TNF) blocking agents (e.g., Enbrel, Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Cimzia, Remicade, Simponi), Kineret, or Arcalyst |
| **Age Restriction:** | • FMF, HIDS/MKD, juvenile idiopathic arthritis, TRAPS: 2 years of age and older<br>• CAPS: 4 years of age and older<br>• Gout Flares: 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an allergist, immunologist, or rheumatologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 6 months, unless otherwise specified |

POLICY NAME:
**ILOPROST**
Drug Name: VENTAVIS (iloprost)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1 |
| **Required documentation:** | **<u>Pulmonary Arterial Hypertension (PAH) WHO Group 1</u>**<br>• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:<br>  ○ Mean pulmonary artery pressure of at least 20 mm Hg<br>  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg<br>  ○ Pulmonary vascular resistance of at least 2.0 Wood units<br>• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class III or higher symptoms<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blockers) unless there are contraindications:<br>  ○ Low systemic blood pressure (systolic blood pressure less than 90)<br>  ○ Low cardiac index<br>  OR<br>  ○ Presence of severe symptoms (functional class IV) |
| **Appropriate Treatment Regimen:** | • Documentation of inadequate response or intolerance to the following therapy classes is required:<br>  ○ PDE5 inhibitors **AND**<br>  ○ Endothelin receptor antagonists (exception WHO Functional Class IV)<br><br>**<u>Reauthorization</u>** requires documentation of treatment success defined as one or more of the following:<br>• Improvement in walking distance<br>• Improvement in exercise ability<br>• Improvement in pulmonary function<br>• Improvement or stability in WHO functional class |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or a pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**IMMUNE GLOBULIN**
Affected Medications: ASCENIV, BIVIGAM, FLEBOGAMMA, GAMMAGARD LIQUID/S-D, GAMMAPLEX, GAMUNEX-C, OCTAGAM, PANZYGA, PRIVIGEN, GAMMASTAN, ALYGLO, YIMMUGO, GAMMAKED

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved and compendia-supported uses not otherwise excluded by plan design as follows:<br>  ○ Primary immunodeficiency (PID)/Wiskott - Aldrich syndrome<br>  ○ Idiopathic thrombocytopenia purpura (ITP)<br>  ○ Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)<br>  ○ Guillain-Barre Syndrome (Acute inflammatory polyneuropathy)<br>  ○ Multifocal Motor Neuropathy<br>  ○ Pediatric HIV: Bacterial control or prevention<br>  ○ Myasthenia Gravis<br>  ○ Dermatomyositis/Polymyositis<br>  ○ Complications of transplanted solid organ (kidney, liver, lung, heart, pancreas) and bone marrow transplant<br>  ○ Stiff-Person Syndrome<br>  ○ Allogeneic Bone Marrow or Stem Cell Transplant<br>  ○ Kawasaki's disease (Pediatric)<br>  ○ Fetal alloimmune thrombocytopenia (FAIT)<br>  ○ Hemolytic disease of the newborn<br>  ○ Auto-immune Mucocutaneous Blistering Diseases<br>  ○ Chronic lymphocytic leukemia with associated hypogammaglobulinemia (CLL)<br>  ○ Toxic Shock Syndrome<br>  ○ Pediatric Acute-Onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS) |
| **Initial Approval Criteria:** | **<u>Primary immunodeficiency (PID)/Wiskott - Aldrich syndrome</u>:**<br><br>Includes but not limited to: X-linked agammaglobulinemia, common variable immunodeficiency (CVID), transient hypogammaglobulinemia of infancy, IgG subclass deficiency with or without IgA deficiency, antibody deficiency with near normal immunoglobulin levels) and combined deficiencies (severe combined immunodeficiencies, ataxia-telangiectasia, x-linked lymphoproliferative syndrome)<br>• Documentation of one of the following:<br>  ○ IgG level less than 200<br>  ○ Low IgG levels (below the laboratory reference range lower limit of normal) AND a history of multiple hard to treat infections as indicated by at least one of the following:<br>  ▪ Four or more ear infections within 1 year<br>  ▪ Two or more serious sinus infections within 1 year<br>  ▪ Two or more months of antibiotics with little effect<br>  ▪ Two or more pneumonias within 1 year<br>  ▪ Recurrent or deep skin abscesses<br>  ▪ Need for intravenous antibiotics to clear infections<br>  ▪ Two or more deep-seated infections including septicemia<br>**AND**<br>• Documentation showing a deficiency in producing antibodies in response to vaccination including all the following:<br>  ○ Titers that were drawn before challenging with vaccination<br>  ○ Titers that were drawn between 4 and 8 weeks after vaccination |

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| | **Idiopathic thrombocytopenia purpura (ITP):**<br><br>For Acute disease state:<br>• Documented use to manage acute bleeding due to severe thrombocytopenia (platelet counts less than 30,000/microliter)<br>**OR**<br>• To increase platelet counts prior to invasive surgical procedures, such as splenectomy (platelet count less than 100,000/microliter)<br>**OR**<br>• Documented severe thrombocytopenia (platelet count less than 20,000/microliter) and is considered to be at risk for intracerebral hemorrhage<br><br>Chronic Immune Thrombocytopenia (CIT):<br>• Documentation of increased risk for bleeding as indicated by a platelet count less than 30,000/microliter<br>• History of failure, contraindication, or intolerance with corticosteroids<br>• Duration of illness more than 6 months<br><br>**Chronic Inflammatory Demyelinating Polyneuropathy (CIDP):**<br><br>• Documented baseline in strength/weakness using objective clinical measuring tool (INCAT, Medical Research Council (MRC) muscle strength, 6 MWT, Rankin, Modified Rankin)<br>• Documented disease course is progressive or relapsing and remitting for 2 months or longer<br>• Abnormal or absent deep tendon reflexes in upper or lower limbs<br>• Electrodiagnostic testing indicating demyelination with one of the following:<br>o Motor distal latency prolongation in 2 nerves<br>o Reduction of motor conduction velocity in 2 nerves<br>o Prolongation of F-wave latency in 2 nerves<br>o Absence of F-waves in at least 1 nerve<br>o Partial motor conduction block of at least 1 motor nerve<br>o Abnormal temporal dispersion in at least 2 nerves<br>o Distal CMAP duration increase in at least 1 nerve<br>• Cerebrospinal fluid (CSF) analysis indicates all the following (if electrophysiologic findings are nondiagnostic):<br>o CSF white cell count of less than 10 cells/mm3<br>o CSF protein is elevated (greater than 45 mg/dL)<br>• Refractory to or intolerant of corticosteroids (prednisolone, prednisone) given in therapeutic doses over at least three months<br><br>**Guillain-Barre Syndrome (Acute inflammatory polyneuropathy):**<br><br>• Documentation that the disease is severe (aid required to walk)<br>• Onset of symptoms are recent (less than 1 month)<br><br>**Multifocal Motor Neuropathy (MMN):**<br><br>• Slowly progressive or stepwise progressive, focal, asymmetric limb weakness over at least one month |

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| | • Partial conduction block or abnormal temporal dispersion conduction must be present in at least 2 nerves<br>• Absence of upper motor neuron signs and bulbar involvement<br>• Baseline in strength/weakness has been documented using objective clinical measuring tool (e.g., Inflammatory Neuropathy Cause and Treatment (INCAT) Disability Score, Medical Research Council (MRC) muscle strength, 6 Minute walk test, Rankin, Modified Rankin<br><br>**Pediatric HIV: Bacterial control or prevention:**<br><br>• Approved for those 13 years of age and younger with HIV diagnosis<br>• Documented hypogammaglobulinemia (IgG less than 400 mg/dL)<br>OR<br>• Functional antibody deficiency as demonstrated by either poor specific antibody titers or recurrent bacterial infections<br><br>**Myasthenia Gravis:**<br><br>• Documented myasthenic crisis (impending respiratory or bulbar compromise)<br>• Documented use for an exacerbation (difficulty swallowing, acute respiratory failure, functional disability leading to discontinuation of physical activity)<br>• Documented failure with conventional therapy alone (azathioprine, cyclosporine and/or cyclophosphamide)<br><br>**Dermatomyositis/Polymyositis:**<br><br>• Documented severe active disease state on physical exam<br>• Documentation of at least two of the following:<br>  ○ Proximal muscle weakness in all upper and/or lower limbs<br>  ○ Elevated serum creatine kinase (CK) or aldolase level<br>  ○ Interstitial lung disease (ILD)<br>  ○ Skin findings such as Gottron papules, Gottron sign, heliotrope eruption, poikiloderma<br>  ○ Nailfold abnormalities<br>  ○ Hyperkeratosis and fissuring of palms and lateral fingers<br>• Documented failure with a trial of corticosteroids (such as prednisone)<br>• Documented failure with a trial of an immunosuppressant (methotrexate, azathioprine, cyclophosphamide)<br><br>**Complications of transplanted solid organ (kidney, liver, lung, heart, pancreas) and bone marrow transplant:**<br><br>Coverage is provided for one or more of the following:<br>• Suppression of panel reactive anti-HLA antibodies prior to transplantation<br>• Treatment of antibody mediated rejection of solid organ transplantation<br>• Prevention of cytomegalovirus (CMV) induced pneumonitis<br><br>**Stiff-Person Syndrome:**<br><br>• Documented anti-GAD antibodies |

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| | • Documented failure with at least 2 of the following treatments: benzodiazepines, baclofen, phenytoin, clonidine and/or tizanidine<br><br>**<u>Allogeneic Bone Marrow or Stem Cell Transplant</u>:**<br><br>• Approved in use for prevention of acute Graft- Versus- Host Disease (GVHD) or infection (such as cytomegalovirus)<br>• Documentation that the bone marrow transplant (BMT) was allogeneic<br>• Transplant was less than 100 days ago<br><br>**<u>Kawasaki's Disease (Pediatric)</u>:**<br><br>• Diagnosis or suspected diagnosis of Kawasaki's disease<br>• 13 years of age and under<br><br>**<u>Fetal alloimmune thrombocytopenia (FAIT)</u>:**<br><br>• Documentation of one or more of the following:<br>  ○ Previous FAIT pregnancy<br>  ○ Family history of the disease<br>  ○ Screening reveals platelet alloantibodies<br>• Authorization is valid until delivery date only<br><br>**<u>Hemolytic disease of the newborn</u>:**<br><br>• Diagnosis or suspected diagnosis of hemolytic disease in newborn patient<br><br>**<u>Auto-immune Mucocutaneous Blistering Diseases</u>:**<br><br>• Diagnosis confirmed by biopsy of one of the following:<br>  ○ Pemphigus vulgaris<br>  ○ Pemphigus foliaceus<br>  ○ Bullous Pemphigoid<br>  ○ Mucous Membrane Pemphigoid (Cicatricial Pemphigoid)<br>  ○ Epidermolysis bullosa aquisita<br>  ○ Pemphigus gestationis (Herpes gestationis)<br>  ○ Linear IgA dermatosis<br>• Documented severe disease that is extensive and debilitating<br>• Disease is progressive and refractory to a trial of conventional combination therapy with corticosteroids and immunosuppressive treatment (azathioprine, cyclophosphamide, mycophenolate mofetil)<br><br>**<u>Chronic lymphocytic leukemia (CLL) with associated hypogammaglobulinemia</u>:**<br><br>• Documentation of an IgG level less than 500 mg/dL<br>• Documented history of recurrent or chronic infections that have required intravenous antibiotics or hospitalization<br><br>**<u>Toxic Shock Syndrome</u>:**<br><br>• Diagnosis or suspected diagnosis of toxic shock syndrome |

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| | **<u>Pediatric Acute-Onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS)</u>:**<br><br>• A clinically appropriate trial of two or more less-intensive treatments was either not effective, not tolerated, or did not result in sustained improvement in symptoms, as measured by a lack of clinically meaningful improvement on a validated instrument directed at the patient's primary symptom complex. Treatments may be given concurrently or sequentially and may include:<br>  ○ Selective-serotonin reuptake inhibitor SSRI (e.g., fluoxetine, fluvoxamine, sertraline)<br>  ○ Behavioral therapy<br>  ○ Nonsteroidal anti-inflammatory (NSAID) (e.g., naproxen, diclofenac, ibuprofen)<br>  ○ Oral and IV corticosteroids (e.g., prednisone, methylprednisolone)<br>• Documentation of a consultation with a pediatric subspecialist (or adult subspecialist for adolescents) and the consulted subspecialist and the patient's primary care provider recommend the treatment |
| **Renewal Criteria:** | **Primary immunodeficiency (PID)**<br>• Renewal requires disease response as evidenced by a decrease in the frequency and/or severity of infections<br>**Chronic Immune Thrombocytopenia (Chronic ITP or CIT)**<br>• Renewal requires disease response as indicated by the achievement and maintenance of a platelet count of at least 50 as necessary to reduce the risk for bleeding<br>**Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)**<br>• Renewal requires documentation of a documented clinical response to therapy based on an objective clinical measuring tool (e.g., INCAT, Medical Research Council (MRC) muscle strength, 6 Minute walk test, Rankin, Modified Rankin)<br>**Multifocal Motor Neuropathy (MMN)**<br>• Renewal requires documentation that there has been a demonstrated clinical response to therapy based on an objective clinical measuring tool (INCAT, Medical Research Council (MRC) muscle strength, 6 Minute walk test, Rankin, Modified Rankin)<br>**Pediatric HIV: Bacterial control or prevention**<br>• 13 years of age or less<br>**Dermatomyositis/Polymyositis**<br>• Renewal requires documentation that CPK (Creatine phosphokinase) levels are lower and documentation of clinically significant improvement above baseline per physical exam<br>**Complications of transplanted solid organ (kidney, liver, lung, heart, pancreas) and bone marrow transplant**<br>• Renewal requires documentation of clinically significant disease response<br>**Stiff Person Disease**<br>• Renewal requires documentation of a clinically significant improvement over baseline per physical exam<br>**Allogeneic Bone Marrow or Stem Cell Transplant**<br>• Renewal requires documentation that the IgG is less than or equal to 400mg/dL; AND<br>• Therapy does not exceed one year past date of allogeneic bone marrow transplantation<br>**Auto-immune mucocutaneous blistering diseases:** |

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<tr><td></td><td>
<ul>
<li>Renewal requires a documented clinically significant improvement over baseline per physical exam</li>
</ul>
Chronic lymphocytic leukemia (CLL) with associated hypogammaglobulinemia
<ul>
<li>Renewal requires disease response as evidenced by a decrease in the frequency and/or severity of infections</li>
</ul>
Pediatric Acute-Onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS)
<ul>
<li>Renewal requires all of the following:
<ul>
<li>Documentation of a clinical reevaluation at three months after treatment initiation</li>
<li>Documentation of clinically meaningful improvement in the results of clinical testing with a validated instrument (which must be performed pretreatment and posttreatment)</li>
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</td></tr>
<tr><td>Dosing and Coverage Duration:</td><td>
<ul>
<li>Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced</li>
<li>Authorization durations are as stated below, unless otherwise specified</li>
</ul>
<table>
<tr><th>Indication</th><th>Dose</th><th>Approval Duration</th></tr>
<tr><td>PID</td><td>Up to 800 mg/kg every 3 to 4 weeks</td><td>Initial: up to 3 months<br>Reauthorization: up to 12 months</td></tr>
<tr><td>CIDP</td><td>2 g/kg divided over 2-5 days for one dose then maintenance dosing of 1 g/kg every 21 days</td><td>Initial: up to 3 months<br>Reauthorization: up to 12 months</td></tr>
<tr><td>ITP</td><td>1 g/kg once daily for 1-2 days<br><br>May be repeated monthly for chronic ITP</td><td>Acute ITP:<br>• Approval: 1 month only<br>Chronic ITP:<br>• Initial: up to 3 months<br>• Reauthorization: up to 12 months</td></tr>
<tr><td>FAIT</td><td>1 g/kg/week until delivery</td><td>Authorization is valid until delivery date only</td></tr>
<tr><td>Kawasaki’s Disease (pediatric patients)</td><td>Up to 2 g/kg x 1 single dose</td><td>Approval: 1 month only</td></tr>
<tr><td>MMN</td><td>2 g/kg divided over 2-5 days in a 28-day cycle<br>May be repeated monthly</td><td>Initial approval: 1 month<br>Reauthorization: up to 12 months</td></tr>
<tr><td>CLL</td><td>400 mg/kg every 3 to 4 weeks</td><td>Approval: up to 6 months</td></tr>
<tr><td>Pediatric HIV</td><td>400 mg/kg every 28 days</td><td>Initial: up to 3 months<br>Reauthorization: up to 12 months</td></tr>
<tr><td>Guillain-Barre</td><td>400 mg/kg once daily for 5 days</td><td>Approval: maximum of 2 rounds of therapy within 6 weeks of onset; 2 months maximum</td></tr>
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| | Myasthenia Gravis | Up to 2 g/kg x 1 dose (acute attacks) | Approval: 1 month (one course of treatment) |
| | Auto-immune blistering diseases | Up to 2 g/kg divided over 5 days in a 28-day cycle | Approval: up to 6 months |
| | Dermatomyositis /Polymyositis | Up to 2 g/kg given over 2-5 days in a 28-day cycle | Initial: up to 3 months<br>Reauthorization: up to 6 months |
| | Allogeneic Bone Marrow or Stem Cell Transplant | 500 mg/kg/week x 90 days, then 500 mg/kg/month up to one-year post-transplant | Initial: up to 3 months<br>Reauthorization: until up to one-year post-transplant |
| | Complications of transplanted solid organ: (kidney, liver, lung, heart, pancreas) transplant | 2 g/kg divided over 5 days in a 28-day cycle | Initial: up to 3 months<br>Reauthorization: up to 12 months |
| | Stiff Person Syndrome | 2 g/kg divided over 5 days in a 28-day cycle | Initial: up to 3 months<br>Reauthorization: up to 12 months |
| | Toxic shock syndrome | 1 g/kg on day 1, followed by 500 mg/kg once daily on days 2 and 3 | Approval: 1 month (one course of treatment) |
| | Hemolytic disease of the newborn | 1 g/kg x 1 dose, may be repeated once if needed | Approval: 1 month (one course of treatment) |
| | PANS/PANDAS | Each dose: Up to 2 g/kg divided over 2-5 days | Initial: up to 3 months (3 monthly doses)<br>Reauthorization: up to 3 months (3 monthly doses)<br><br>Total 6 monthly doses only |
| **Prescriber/Site of Care Restrictions:** | • Must be prescribed by a specialist for the condition being treated (such as neurologist, rheumatologist, immunologist, hematologist)<br>• All approvals are subject to utilization of the most cost-effective site of care | | |

POLICY NAME:
**INCLISIRAN**
Affected Medications: LEQVIO (inclisiran subcutaneous injection)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Primary hyperlipidemia (including heterozygous familial hypercholesterolemia [HeFH])<br>  ○ Secondary prevention in atherosclerotic cardiovascular disease (ASCVD) |
| **Required Medical Information:** | **All Indications**<br>• Documentation of baseline (untreated) low-density lipoprotein cholesterol (LDL-C)<br><br>**Primary Hyperlipidemia (non-familial)**<br>• Documentation of baseline (untreated) LDL-C of at least 190 mg/dL<br><br>**HeFH**<br>• Diagnosis confirmed by **ONE** of the following:<br>  ○ Minimum baseline LDL-C of 160 mg/dL in adolescents or 190 mg/dL in adults **AND** 1 first-degree relative affected<br>  ○ Presence of one abnormal LDL-C-raising gene defect (e.g., LDL receptor [LDLR], apolipoprotein B [apo B], proprotein convertase subtilisin kexin type 9 [PCSK9] loss-of-function mutation, or LDL receptor adaptor protein 1 [LDLRAP1])<br>  ○ World Health Organization (WHO)/Dutch Lipid Network criteria score of at least 8 points<br>  ○ Definite FH diagnosis per the Simon Broome criteria<br><br>**Clinical ASCVD**<br>• Documentation of established ASCVD, confirmed by at least **ONE** of the following:<br>  ○ Acute coronary syndromes (ACS)<br>  ○ History of myocardial infarction (MI)<br>  ○ Stable or unstable angina<br>  ○ Coronary or other arterial revascularization<br>  ○ Stroke or transient ischemic attack<br>  ○ Peripheral artery disease (PAD) presumed to be of atherosclerotic origin |
| **Appropriate Treatment Regimen & Other Criteria:** | **All Indications**<br>• History of statin intolerance requires documentation of **ONE** of the following:<br>  ○ Statin-associated rhabdomyolysis occurred with statin use and was confirmed by a creatinine kinase (CK) level at least 10 times the upper limit of normal<br>  ○ Statin-associated muscle symptoms (e.g., myopathy, myalgia) occurred with statin use and was confirmed by **BOTH** of the following:<br>  ▪ A minimum of two different statin trials, with at least one being a hydrophilic statin (rosuvastatin, pravastatin)<br>  ▪ A re-challenge of each statin (muscle symptoms stopped when each was discontinued and restarted upon re-initiation) |

<table>
<tr><td></td><td><u>Primary Hyperlipidemia/HeFH</u><br>• Documented treatment failure with minimum 12-week trial with ALL of the following, shown by an inability to achieve LDL-C reduction of 50% or greater OR LDL-C less than 100 mg/dL:<br>  ○ Maximally tolerated statin/ezetimibe therapy<br>  ○ Repatha<br><br><u>Clinical ASCVD</u><br>• Documented treatment failure with minimum 12 weeks of consistent maximally tolerated combination statin/ezetimibe therapy, as shown by ONE of the following:<br>  ○ Current LDL-C of at least 70 mg/dL<br>  ○ Current LDL-C of at least 55 mg/dL in patients at very high risk of future ASCVD events, based on history of multiple major ASCVD events OR 1 major ASCVD event + multiple high-risk conditions (see below)<br>• Documented treatment failure or intolerance to minimum 12-week trial of Repatha<br><br><table><tr><th>Major ASCVD Events</th><th>High-Risk Conditions</th></tr><tr><td>• ACS within the past 12 months<br>• History of MI (distinct from ACS event)<br>• Ischemic stroke<br>• Symptomatic PAD</td><td>• Age 65 years and older<br>• HeFH<br>• Prior coronary artery bypass or percutaneous intervention (outside of major ASCVD events)<br>• Diabetes<br>• Hypertension<br>• Chronic kidney disease<br>• Current smoking<br>• History of congestive heart failure</td></tr></table><br><u>Reauthorization</u> requires an updated lipid panel showing a clinically significant reduction in baseline LDL-C and continued adherence to therapy</td></tr>
<tr><td>Exclusion Criteria:</td><td>• Concurrent use with PCSK9 monoclonal antibodies (e.g., Repatha, Praluent)</td></tr>
<tr><td>Age Restriction:</td><td>• 18 years of age and older</td></tr>
<tr><td>Prescriber/Site of Care Restrictions:</td><td>• Prescribed by, or in consultation with, a cardiologist, endocrinologist, or lipid specialist<br>• All approvals are subject to utilization of the most cost-effective site of care</td></tr>
<tr><td>Coverage Duration:</td><td>• Authorization: 12 months, unless otherwise specified</td></tr>
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POLICY NAME:
**INEBILIZUMAB-CDON**
Affected Medications: UPLIZNA (inebilizumab-cdon)

<table>
<tr><td>Covered Uses:</td><td>
<ul>
<li>All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
<ul>
<li>Neuromyelitis optica spectrum disorder (NMOSD) in adults who are anti-aquaporin-4 (AQP4) antibody positive</li>
<li>Immunoglobulin G4-related disease (IgG4-RD) in adults</li>
<li>Generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) or anti-muscle specific tyrosine kinase (MuSK) antibody positive</li>
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<tr><td>Required Medical Information:</td><td>
<u>NMOSD</u>
<ul>
<li>Diagnosis of seropositive aquaporin-4 immunoglobulin G (AQP4-IgG) NMOSD confirmed by all the following:
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<li>Documentation of AQP4-IgG-specific antibodies on cell-based assay</li>
<li>Exclusion of alternative diagnoses (such as multiple sclerosis)</li>
<li>At least one core clinical characteristic:
<ul>
<li>Acute optic neuritis</li>
<li>Acute myelitis</li>
<li>Acute area postrema syndrome (episode of otherwise unexplained hiccups or nausea/vomiting)</li>
<li>Acute brainstem syndrome</li>
<li>Symptomatic narcolepsy OR acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesion on magnetic resonance imaging (MRI) [see table below]</li>
<li>Acute cerebral syndrome with NMOSD-typical brain lesion on MRI [see table below]</li>
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<table>
<tr><th>Clinical presentation</th><th>Possible MRI findings</th></tr>
<tr><td>Diencephalic syndrome</td><td>• Periependymal lesion<br>• Hypothalamic/thalamic lesion</td></tr>
<tr><td>Acute cerebral syndrome</td><td>• Extensive periependymal lesion<br>• Long, diffuse, heterogenous, or edematous corpus callosum lesion<br>• Long corticospinal tract lesion<br>• Large, confluent subcortical or deep white matter lesion</td></tr>
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<ul>
<li>History of at least 1 attack in the past year, or at least 2 attacks in the past 2 years, requiring rescue therapy</li>
</ul>
<u>IgG4-RD</u>
<ul>
<li>Diagnosis of IgG4-RD per American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) classification criteria that meets inclusion criteria, has no exclusion criteria, AND has equal to or greater than 20 classification criteria inclusion points</li>
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| | • The condition affects two or more organs or sites at any time, including at least one of the following: pancreas, bile ducts/biliary tree, orbits, lungs, kidneys, lacrimal glands, major salivary glands, retroperitoneum, aorta, pachymeninges, or thyroid gland<br>• Member is experiencing (or has recently experienced) an IgG4-RD flare that requires glucocorticoid treatment<br><br>**gMG**<br>• Diagnosis of gMG confirmed by ONE of the following:<br>  ○ A history of abnormal neuromuscular transmission test<br>  ○ A positive edrophonium chloride test<br>  ○ Improvement in gMG signs or symptoms with an acetylcholinesterase inhibitor<br>• Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV<br>• Positive serologic test for AChR or MuSK antibodies<br>• Documentation of ONE of the following:<br>  ○ MG-Activities of Daily Living (MG-ADL) total score of 6 or greater<br>  ○ Quantitative Myasthenia Gravis (QMG) total score of 12 or greater |
| **Appropriate Treatment Regimen & Other Criteria:** | **NMOSD**<br>• Documentation of inadequate response, contraindication, or intolerance to each of the following:<br>  ○ Rituximab (preferred products: Riabni, Ruxience)<br>  ○ Satralizumab-mwge (Enspryng)<br><br>**IgG4-RD**<br>• Documentation of inadequate response, contraindication, or intolerance to each of the following:<br>  ○ Glucocorticoids<br>  ○ Rituximab (preferred products: Riabni, Ruxience)<br><br>**gMG**<br>• Documentation of one of the following:<br>  ○ Treatment failure with an adequate trial (one year or more) of at least 2 immunosuppressive therapies (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)<br>  ○ Has required three or more courses of rescue therapy (plasmapheresis/plasma exchange and/or intravenous immunoglobulin), while on at least one immunosuppressive therapy, over the last 12 months<br><br>**Reauthorization** requires documentation of treatment success |
| **Exclusion Criteria:** | • Active Hepatitis B Virus (HBV) infection<br>• Active or untreated latent tuberculosis<br>• Concurrent use with other disease-modifying biologics for requested indication |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • **NMOSD:** Prescribed by, or in consultation with, a neurologist or neuro-ophthalmologist<br>• **IgGR-RD:** Prescribed by, or in consultation with, a rheumatologist, immunologist, |

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| | endocrinologist, nephrologist, hepatologist, or specialist with experience in the treatment of IgG4-RD<br>• **gMG:** Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**INFUSIONS FOR ADVANCED PARKINSON'S DISEASE**
Affected Medications: ONAPGO (apomorphine hydrochloride infusion), VYALEV (carbidopa-levodopa infusion)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Treatment of motor fluctuations in adults with advanced Parkinson's disease (PD) |
| **Required Medical Information:** | • Diagnosis of advanced PD<br>• Clear response to levodopa treatment with evidence of "On" periods<br><br>**Onapgo**<br>• Persistent motor fluctuations with "Off" time occurring 3 hours or more per day while awake despite an optimized PD treatment regimen<br><br>**Vyalev**<br>• Persistent motor fluctuations with "Off" time occurring 2.5 hours or more per day while awake despite an optimized PD treatment regimen |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with both of the following:<br>  ○ Oral carbidopa/levodopa extended release<br>  ○ Two additional agents from different anti-PD drug classes:<br>  ▪ Monoamine oxidase-B (MAO-B) inhibitors (ex: selegiline, rasagiline)<br>  ▪ Dopamine agonists (ex: amantadine, pramipexole, ropinirole)<br>  ▪ Catechol-O-methyltransferase (COMT) inhibitors (ex: entacapone)<br>**Onapgo**<br>• Dosing is in accordance with FDA labeling and does not exceed 98 mg/20 mL per day<br><br>**Vyalev**<br>• Dosing is in accordance with FDA labeling and does not exceed 3,525 mg of foslevodopa component per day<br><br>**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | **Onapgo**<br>• PD not responsive to levodopa<br>• Use for atypical Parkinson's syndrome (such as "Parkinson's Plus" syndrome) or secondary PD<br>• Previous neurosurgical treatment for PD<br><br>**Vyalev**<br>• PD not responsive to levodopa<br>• Concomitant or recent (within 2 weeks) use of nonselective MAO inhibitors<br>• Concomitant use with carbidopa/levodopa extended release products |
| **Age Restriction:** | **Onapgo**<br>• 30 years of age and older<br>**Vyalev** |

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| | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**INHALED MANNITOL**
Affected Medications: BRONCHITOL (mannitol)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Add-on maintenance therapy to improve pulmonary function in cystic fibrosis |
| **Required Medical Information:** | • Documentation of cystic fibrosis (CF) diagnosis confirmed by appropriate genetic or diagnostic testing<br>  ○ Additional testing should include evaluation of overall clinical lung status and respiratory function (e.g., pulmonary function tests, lung imaging, etc.) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with 6-month trial of twice daily inhaled hypertonic saline (at least 80% adherence), unless contraindicated or intolerable. Treatment failure defined as one or more of the following:<br>  ○ Increased pulmonary exacerbations from baseline<br>  ○ Decrease in FEV1<br>• Requests for Bronchitol 7-day and 4-week treatment packs for add-on maintenance therapy:<br>  ○ Documentation confirming successful completion of the Bronchitol Tolerance Test (BTT)<br>  ○ Prescribed in conjunction with a short-acting bronchodilator and standard therapies for CF<br><br>**Reauthorization** requires documentation of a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**INTERFERONS FOR MULTIPLE SCLEROSIS**
Affected Medications: AVONEX (interferon beta-1a), BETASERON (interferon beta-1b), PLEGRIDY (pegylated interferon beta-1a), REBIF (interferon beta-1a)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:<br>  ▪ Clinically isolated syndrome (CIS)<br>  ▪ Relapsing-remitting multiple sclerosis (RRMS)<br>  ▪ Active secondary progressive disease (SPMS) |
| **Required Medical Information:** | • Diagnosis confirmed with magnetic resonance imaging (MRI) per revised McDonald diagnostic criteria for MS<br>  ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS |
| **Appropriate Treatment Regimen & Other Criteria:** | • **Avonex and Plegridy:** Documentation of treatment failure with (or intolerance to) **BOTH** of the following:<br>  ○ Glatiramer OR Glatopa<br>  ○ Dimethyl fumarate, fingolimod OR teriflunomide<br>• **Rebif and Betaseron:** Documentation of treatment failure with (or intolerance to) **ALL** the following:<br>  ○ Glatiramer OR Glatopa<br>  ○ Dimethyl fumarate, fingolimod OR teriflunomide<br>  ○ Avonex OR Plegridy<br><br>**Reauthorization** requires provider attestation of treatment success |
| **Exclusion Criteria:** | • Concurrent use of other disease-modifying medications indicated for the treatment of MS |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or MS specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 24 months, unless otherwise specified |

POLICY NAME:
**INTRAVITREAL ANTI-VEGF THERAPY**
Affected Medications: LUCENTIS (ranibizumab injection), EYLEA (aflibercept), EYLEA HD (aflibercept), BEOVU (brolucizumab), SUSVIMO (ranibizumab implant), PAVBLU (aflibercept-ayyh)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved, or compendia supported, indications not otherwise excluded by plan design<br>  ○ Neovascular (Wet) Age-Related Macular Degeneration (AMD)<br>  ▪ Eylea, Eylea HD, Pavblu, Lucentis, Susvimo, Beovu<br>  ○ Macular Edema Following Retinal Vein Occlusion (RVO)<br>  ▪ Eylea, Eylea HD, Pavblu, Lucentis<br>  ○ Diabetic Macular Edema (DME)<br>  ▪ Eylea, Eylea HD, Pavblu, Lucentis, Beovu, Susvimo<br>  ○ Diabetic Retinopathy (DR) in patients with Diabetes Mellitus<br>  ▪ Eylea, Eylea HD, Pavblu, Lucentis, Susvimo<br>  ○ Myopic Choroidal Neovascularization (mCNV)<br>  ▪ Lucentis<br>  ○ Retinopathy of Prematurity (ROP)<br>  ▪ Eylea, Lucentis |
| **Required Medical Information:** | • Anticipated treatment course with dose and frequency clearly stated in chart notes |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Eylea/Pavblu Dosing</u>**<br>• **Coverage for the non-preferred products Eylea or Pavblu is provided when one of the following criteria is met:**<br>  ○ Currently receiving treatment with Eylea or Pavblu, excluding when the product is obtained as samples or via manufacturer's patient assistance programs<br>  ○ A documented inadequate response or intolerable adverse event with **<u>TWO</u>** of the following preferred products: Avastin, Vabysmo, or Byooviz,<br>  ○ Documentation of treatment-naïve retinopathy of prematurity (ROP) in a preterm infant 32 weeks or younger<br><br>• **AMD –** 2 mg (0.05 mL) every 4 weeks for the first 3 injections followed by 2 mg (0.05 mL) every 8 weeks<br>  ○ Continued every 4-week dosing requires documented clinical failure to every 8-week maintenance dosing<br>• **RVO -** 2 mg (0.05 mL) every 4 weeks<br>• **DME and DR –** 2 mg (0.05 mL) every 4 weeks for the first 5 injections followed by 2 mg (0.05 mL) every 8 weeks<br>• **ROP** – 0.4 mg (0.01 mL) as a single injection per affected eye(s); dose may be repeated up to 2 times with a minimum treatment interval between doses of at least 10 days (maximum of 3 doses total)<br><br>**<u>Eylea HD Dosing</u>**<br>• **Coverage for the non-preferred product Eylea HD is provided when one of the following criteria is met:**<br>  ○ Currently receiving treatment with Eylea HD, excluding when the product is obtained as samples or via manufacturer's patient assistance programs.<br>  ○ A documented inadequate response or intolerable adverse event with **<u>TWO</u>** of the following preferred products: Avastin, Vabysmo, or Byooviz |

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| | • **AMD and DME** – 8 mg (0.07 mL) every 4 weeks for the first 3 injections, followed by 8 mg (0.07 mL) every 8 to 16 weeks<br>  ○ Every 4-week dosing is limited to the first 3 injections only<br>• **RVO** - 8 mg (0.07 mL) every 4 weeks for the first 3-5 injections, followed by 8mg (0.07 mL) every 8 weeks<br>• **DR -** 8 mg (0.07 mL) every 4 weeks for the first 3 injections, followed by 8 mg (0.07 mL) every 8 weeks to 12 weeks<br>  ○ Every 4-week dosing is limited to the first 3 injections only<br><br>**<u>Lucentis Dosing</u>**<br>• **Coverage for the non-preferred product Lucentis is provided when the following criteria is met:**<br>  ○ A documented inadequate response or intolerable adverse event with **<u>TWO</u>** of the following preferred products: Avastin, Vabysmo, or Byooviz<br>• **AMD and RVO –** maximum 0.5 mg every 4 weeks<br>• **DME and DR –** 0.3 mg every 4 weeks<br>• **mCNV-** 0.5 mg every 4 weeks for up to 3 months<br>• **ROP** – 0.1 to 0.3 mg as a single injection in the affected eye(s); dose may be repeated up to 2 times with a minimum treatment interval between doses of 28 days (maximum of 3 doses total)<br><br>**<u>Beovu Dosing</u>**<br>• **Coverage for the non-preferred product Beovu is provided when either of the following criteria is met:**<br>  ○ Currently receiving treatment with Beovu, excluding when the product is obtained as samples or via manufacturer's patient assistance programs.<br>  ○ A documented inadequate response or intolerable adverse event with **<u>TWO</u>** of the following preferred products: Avastin, Vabysmo, or Byooviz<br>• **AMD** – 6 mg every month for the first three doses followed by 6 mg every 8 to 12 weeks<br>• **DME** – 6 mg every six weeks for the first five doses followed by 6 mg every 8 to 12 weeks<br><br>**<u>Susvimo Dosing</u>**<br>• **Coverage for the non-preferred product Susvimo is provided when the following criteria is met:**<br>  ○ A documented inadequate response or intolerable adverse event with **<u>TWO</u>** of the following preferred products: Avastin, Vabysmo, or Byooviz<br>• Must be established on ranibizumab (such as Byooviz) injections with response to treatment for a minimum of 6 months at standard dosing (0.5 mg every 4 weeks)<br>• **AMD and DME** – 2 mg administered continuously via ocular implant with refills every 24 weeks<br>• **DR** – 2 mg administered continuously via ocular implant with refills every 36 weeks<br><br>**<u>Reauthorization</u>** requires documentation of vision stability defined as losing fewer than 15 letters of visual acuity and/or improvements in visual acuity with evidence of decreased leakage and/or fibrosis (central retinal thickness) |
| **Exclusion Criteria:** | • Evidence of a current ocular or periocular infections<br>• Active intraocular inflammation |
| **Age** | |

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| **Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **Macular Edema Following Retinal Vein Occlusion (RVO) for Vabysmo**<br>• Authorization: 6 months with no reauthorization, unless otherwise specified<br><br>**Retinopathy of Prematurity (ROP)**<br>• Authorization: 3 months with no reauthorization, unless otherwise specified<br><br>**All other indications**<br>• Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**INTRAVITREAL COMPLEMENT INHIBITORS**
Affected Medications: SYFOVRE (pegcetacoplan), IZERVAY (avacincaptad pegol)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of geographic atrophy (GA) secondary to age-related macular degeneration (AMD) |
| **Required Medical Information:** | • Diagnosis of geographic atrophy (GA) secondary to age-related macular degeneration (AMD) confirmed by all the following:<br>  ○ Fundus Autofluorescence (FAF) imaging showing:<br>  ▪ Total GA area size between 2.5 and 17.5 $mm^2$<br>  ▪ If GA is multifocal, at least 1 focal lesion that is 1.25 $mm^2$ or greater<br>• Best-corrected visual acuity (BCVA) between 24 and 83 Early Treatment Diabetic Retinopathy Study (ETDRS) letters (20/25 and 20/320 Snellen equivalent) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing not to exceed:<br>  ○ Every 25-day dosing for Syfovre<br>  ○ Every 28-day dosing for Izervay<br><br>**Reauthorization** requires documentation of treatment success and for BCVA to remain at 24 letters or better (20/320 Snellen equivalent) |
| **Exclusion Criteria:** | • Presence of choroidal neovascularization in the eye(s) receiving treatment |
| **Age Restriction:** | • Syfovre: 60 years of age and older<br>• Izervay: 50 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**INTRON-A**
Affected Medications: INTRON-A, INTRON-A WITH DILUENT (interferon alfa-2b)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher<br>• Hypereosinophilic Syndrome (HES) in patients that are consistently symptomatic or with evidence of end-organ damage. |
| **Required Medical Information:** | • For Hepatitis B and C: Documentation of intolerance to or clinical rationale for avoidance of PEGylated interferon.<br>• HES: documentation of steroid resistant disease OR disease responding only to high-dose steroids and the addition of a steroid-sparing agent would be beneficial.<br>  ○ Non-lymphocytic variants of HES will also require documented failure with at least 12 weeks of hydroxyurea prior to interferon-alfa approval.<br>• Recent liver function tests, comprehensive metabolic panel, complete blood count with differential, TSH (within past 3 months)<br>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• Reauthorization: documentation of disease responsiveness to therapy |
| **Appropriate Treatment Regimen & Other Criteria:** | • Patients with preexisting cardiac abnormalities and/or advanced cancer: recent electrocardiogram<br>• Chest X ray for patients with pulmonary disorders<br>• Recent ophthalmologic exam at baseline for all patients<br>• Uncontrolled severe mental health illness should be addressed before use and monitored during treatment |
| **Exclusion Criteria:** | • Autoimmune hepatitis<br>• Decompensated liver disease |
| **Age Restriction:** | • Hepatitis B: greater than or equal to 1 year of age<br>• Hepatitis C: greater than or equal to 3 years of age<br>• All other indications greater than or equal to 18 years of age |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ISAVUCONAZONIUM SULFATE**
Affected Medications: CRESEMBA (isavuconazonium sulfate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of invasive aspergillosis<br>  ○ Treatment of invasive mucormycosis |
| **Required Medical Information:** | **Invasive Aspergillosis**<br>• Documented diagnosis supported by clinical manifestations of disease and at least one of the following:<br>  ○ Features of *Aspergillus* spp. on histopathology or cytology (tissue or fluid staining)<br>  ○ Positive culture or biopsy<br>  ○ Two or more positive polymerase chain reaction (PCR) tests from serum, plasma, bronchoalveolar lavage (BAL), or a combination<br>  ○ Elevated galactomannan (GM) index<br>  ▪ ≥ 1.0 from serum, plasma, BAL, or cerebrospinal fluid (CSF)<br>  OR<br>  ▪ ≥ 0.7 from serum or plasma **and** ≥ 0.8 from BAL<br><br>**Invasive Mucormycosis**<br>• Documented diagnosis of invasive mucormycosis supported by clinical manifestations of disease and at least one of the following:<br>  ○ Positive tissue culture or biopsy<br>  ○ Features of Mucorales on histopathology<br>  ○ Positive polymerase chain reaction (PCR) test (serum, plasma, or histological specimens with fungal elements) |
| **Appropriate Treatment Regimen & Other Criteria:** | **Invasive Aspergillosis**<br>• Documentated treatment failure with minimum 2-week trial of the following, unless intolerable or contraindicated:<br>  ○ Voriconazole<br>  ○ Posaconazole<br><br>**Invasive Mucormycosis**<br>• Documented treatment failure or intolerable adverse event with one of the following:<br>  ○ Amphotericin B (if request is for initial therapy)<br>  ○ Posaconazole (if request is for oral step-down therapy after initial therapy)<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Familial short QT syndrome |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an infectious disease specialist, transplant physician, or oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 3 months, unless otherwise specified |

POLICY NAME:
**LAROTRECTINIB**
Affected Medications: VITRAKVI (larotrectinib)

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• Documentation of positive neurotrophic tyrosine receptor kinase (NTRK) gene-fusion without a known acquired resistance mutation, as determined by an FDA approved test |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of an intolerance to, or clinical rationale for avoidance of Rozlytrek (entrectinib)<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**LAZERTINIB**
Affected Medications: LAZCLUZE (lazertinib)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• Documentation of confirmed non-small cell lung cancer (NSCLC) that is metastatic or unresectable with epidermal growth factor receptor (EGFR) exon 19 deletions or exon 21 L858R substitution mutations |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented intolerable adverse event to Tagrisso (osimertinib) with or without chemotherapy<br><br>**Reauthorization:** documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**LENACAPAVIR**
Affected Medications: SUNLENCA (lenacapavir)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of human immunodeficiency virus type 1 (HIV-1) infection, in combination with other antiretrovirals, in heavily treatment-experienced adults with multidrug resistant HIV-1 infection failing their current antiretroviral regimen due to resistance, intolerance, or safety considerations |
| **Required Medical Information:** | • Documentation of multidrug resistance within at least 3 of the 4 following antiretroviral classes (as defined by resistance to at least 2 agents within each of the 3 classes), unless contraindicated or clinically significant adverse effects are experienced:<br>  ○ Nucleoside reverse-transcriptase inhibitors (NRTIs)<br>  ○ Non-nucleoside reverse-transcriptase inhibitors (NNRTIs)<br>  ○ Protease inhibitors (PIs)<br>  ○ Integrase strand transfer inhibitors (INSTIs)<br>• Documentation of current (within the past 30 days) HIV-1 RNA viral load of at least 200 copies/mL |
| **Appropriate Treatment Regimen & Other Criteria:** | • Must be used in combination with an optimized background antiretroviral regimen that contains at least one agent demonstrating full viral susceptibility, as confirmed by resistance testing<br><br>**<u>Reauthorization</u>** requires all of the following:<br>• Treatment plan includes continued use of optimized background antiretroviral regimen<br>• Documentation of treatment success, as evidenced by one of the following:<br>  ○ Reduction in viral load from baseline or maintenance of undetectable viral load<br>  ○ Absence of postbaseline emergence of lenacapavir resistance-associated mutations confirmed by resistance testing |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an infectious disease or HIV specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Oral Tablet Initial Authorization: 1 month, unless otherwise specified<br>• Injection Initial Authorization: 6 months, unless otherwise specified<br>• Injection Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**LENIOLISIB**
Affected Medications: JOENJA (leniolisib)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Activated phosphoinositide 3-kinase delta syndrome (APDS) |
| **Required Medical Information:** | • Documentation of an APDS-associated *PIK3CD/PIK3R1* mutation without concurrent use of immunosuppressive medication<br>• Presence of at least one measurable nodal lesion on a CT or MRI scan<br>• Documentation of both of the following:<br>  ○ Nodal and/or extranodal lymphoproliferation<br>  ○ History of repeated oto-sino-pulmonary infections and/or organ dysfunction (e.g., lung, liver)<br>• Current weight (must be at least 45 kg) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Females of reproductive potential should have pregnancy ruled out and use effective contraception during therapy<br><br>**Reauthorization** will require documentation of treatment success as shown by both of the following:<br>• Improvement in lymphoproliferation as measured by a change from baseline in lymphadenopathy<br>• Normalization of immunophenotype as measured by the percentage of naïve B cells out of total B cells |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 12 to 75 years of age |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an immunologist, hematologist/oncologist, or specialist with experience in the treatment of APDS<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**LETERMOVIR**
Affected Medications: PREVYMIS (letermovir)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Prophylaxis of cytomegalovirus (CMV) infection and disease in CMV-seropositive recipients [R+] of an allogeneic hematopoietic cell transplant for adults and pediatric patients 6 months of age and older and weighing at least 6 kg<br>  ○ Prophylaxis of CMV disease in kidney transplant recipients at high risk for adult and pediatric patients 12 years of age and older and weighing at least 40 kg |
| **Required Medical Information:** | • Has received an allogeneic hematopoietic stem cell transplant (HSCT)<br>• Is cytomegalovirus CMV-seropositive<br><br>OR<br>• Has received a kidney transplant and is at high risk (Donor CMV-seropositive/Recipient CMV-seronegative [D+/R-] of CMV infection |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented trial and failure (or intolerable adverse event) with an adequate trial (at least 14 days) of at least one of the following: ganciclovir, valganciclovir, Foscarnet (HSCT only)<br><br>**HSCT Dosing**: Up to 480 mg (or 240 mg) once daily beginning between Day 0 and Day 28 post-transplantation and continued through Day 100 post-transplantation<br><br>**Kidney Transplant Dosing**: Up to 480mg once daily beginning between Day 0 and Day 7 post kidney transplant for high-risk recipients (donor CMV-seropositive/recipient CMV-seronegative) and continue through day 200 post transplantation |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an infectious disease provider or a specialist with experience in the prevention and treatment of CMV infection<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **HSCT**<br>• Authorization: 4 months, unless otherwise specified<br><br>**Kidney Transplant**<br>• Authorization: 7 months, unless otherwise specified |

POLICY NAME:
**LEUPROLIDE**
Affected Medications: leuprolide acetate, LUPRON DEPOT, LUPRON DEPOT-PED, ELIGARD, LUPANETA (leuprolide-norethindrone), FENSOLVI, CAMCEVI

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Endometriosis<br>  ○ Uterine leiomyomata (fibroids)<br>  ○ Central precocious puberty (CPP)<br>• NCCN (National Comprehensive Cancer Network) indications level 2A or higher<br>• Gender dysphoria |
| **Required Medical Information:** | **Endometriosis:**<br>• Documentation of moderate to severe pain due to endometriosis<br><br>**Uterine leiomyomata (fibroids):**<br>• Documentation of all the following:<br>  ○ Preoperative anemia due to uterine leiomyomata (fibroids)<br>  ○ Planning to undergo leiomyomata-related surgery in the next 6 months or less<br>  ○ Planning to use in combination with iron supplements<br><br>**Central precocious puberty:**<br>• Documentation of CPP confirmed by basal luteinizing hormone (LH), follicle-stimulating hormone (FSH), and either estradiol or testosterone concentrations |
| **Appropriate Treatment Regimen & Other Criteria:** | **Endometriosis:**<br>• Documentation of a trial and inadequate relief (or contraindication) after at least 3 months of both of the following first-line therapies:<br>  ○ Nonsteroidal anti-inflammatory drugs (NSAIDs)<br>  ○ Continuous (no placebo pills) hormonal contraceptives<br><br>**Central precocious puberty:**<br>• Approval of Fensolvi requires rationale for avoidance of Lupron and Supprelin LA |
| **Exclusion Criteria:** | • Undiagnosed abnormal vaginal bleeding<br>• Management of uterine leiomyomata without intention of undergoing surgery.<br>• Pregnancy or breastfeeding<br>• Use for infertility (if benefit exclusion) |
| **Age Restriction:** | • Endometriosis and preoperative uterine leiomyomata: 18 years of age and older<br>• Central precocious puberty (CPP): 11 years of age or younger (females), 12 years of age or younger (males) |
| **Prescriber/Site of Care Restrictions:** | • All other indications: prescribed by, or in consultation with, an oncologist, endocrinologist, or gynecologist as appropriate for diagnosis<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Uterine leiomyomata: maximum of 6 months, unless otherwise specified<br>• Endometriosis: 6 months, unless otherwise specified<br>• All other diagnoses: 12 months, unless otherwise specified |

POLICY NAME:
**LEVOKETOCONAZOLE**
Affected Medications: RECORLEV (levoketoconazole)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Cushing syndrome |
| **Required Medical Information:** | • Diagnosis of Cushing's syndrome due to one of the following:<br>  ○ Adrenocorticotropic hormone (ACTH)-secreting pituitary adenoma (Cushing's disease)<br>  ○ Ectopic ACTH secretion by a non-pituitary tumor<br>  ○ Cortisol secretion by an adrenal adenoma<br>• Mean 24-hour urine free cortisol (mUFC) greater than 1.5 times the upper limit of normal (ULN) for the assay (at least two measurements) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation confirming surgery is not an option **OR** previous surgery has not been curative<br>• Documentation of **ONE** of the following:<br>  ○ Clinical failure to maximally tolerated dose of oral ketoconazole for at least 8 weeks<br>  ○ Intolerable adverse event to oral ketoconazole, and the adverse event was not an expected adverse event attributed to the active ingredient<br><br>**Reauthorization** requires documentation of treatment success defined as mUFC normalization (i.e., less than or equal to the ULN) |
| **Exclusion Criteria:** | • Adrenal or pituitary carcinoma |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist, neurologist, or adrenal surgeon<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**LIFILEUCEL**
Affected Medications: AMTAGVI (lifileucel)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Diagnosis of unresectable or Stage IV metastatic melanoma<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• ECOG PS of 0 or 1<br>• Left ventricular ejection fraction (LVEF) greater than 45%<br>• Forced expiratory volume (FEV1) greater than 60%<br>• New York Heart Association (NYHA) classification not more than Class I |
| **Appropriate Treatment Regimen & Other Criteria:** | • At least one resectable lesion (or aggregate of lesions resected) of 1.5 cm or more in diameter post-resection to generate tumor-infiltrating lymphocytes (TILs)<br>• Disease progression after 1 or more prior systemic therapy including:<br>  ○ a PD-1–blocking antibody; and<br>  ○ if BRAF V600 mutation–positive, a BRAF inhibitor or BRAF inhibitor plus a MEK inhibitor |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• Melanoma of uveal or ocular origin<br>• Untreated or active brain metastasis |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist.<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 6 months (one dose per patient's lifetime), unless otherwise specified |

POLICY NAME:
**LONAFARNIB**
Affected Medications: ZOKINVY (lonafarnib)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ To reduce risk of mortality in Hutchinson-Gilford Progeria Syndrome<br>  ○ For treatment of processing-deficient Progeroid Laminopathies |
| **Required Medical Information:** | • A diagnosis of Hutchinson-Gilford Progeria Syndrome (HGPS) confirmed by mutational analysis (G608G mutation in the lamin A gene)<br>OR<br>• A diagnosis of processing-deficient Progeroid Laminopathies with one of the following:<br>  ○ Heterozygous LMNA mutation with progerin-like protein accumulation<br>  ○ Homozygous or compound heterozygous ZMPSTE24 mutations |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented height and weight, or body surface area (BSA)<br>• Documentation of medication review and avoidance of drugs that significantly affect the metabolism of lonafarnib (e.g. strong or moderate CYP3A4 inhibitors/inducers)<br>• Females of reproductive potential should have pregnancy ruled out and use effective contraception during treatment<br><br><u>Labs</u>:<br>• Absolute Phagocyte Count (sum of absolute neutrophil count, bands, and monocytes) greater than 1,000/microliters<br>• Platelets greater than 75,000/microliters (transfusion independent)<br>• Hemoglobin greater than 9g/dl.<br><br><u>Dosing</u>:<br>• Available as oral capsules: 50 mg, 75 mg<br>• Initial, 115 mg/m2/dose twice daily for 4 months, then increase to 150 mg/m2/dose twice daily<br>  ○ Do not exceed 115 mg/m2/dose twice daily when used in combination with a weak CYP3A4 inhibitor<br>  ○ Round all total daily doses to the nearest 25 mg increment<br><br>**<u>Reauthorization</u>** <u>requires documentation of treatment success and initial criteria to be met.</u> |
| **Exclusion Criteria:** | • Use for other progeroid syndromes or processing-proficient progeroid laminopathies<br>• Concomitant use with strong or moderate CYP3A4 inhibitors/inducers, midazolam, lovastatin, atorvastatin, or simvastatin<br>• Overt renal, hepatic, pulmonary disease or immune dysfunction<br>• BSA less than to 0.39 m2 |
| **Age Restriction:** | • Age 12 months or older with a BSA of greater than or equal to 0.39 m2 |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a provider with experience in treating progeria and/or progeroid laminopathies |
| **Coverage Duration:** | • Initial Authorization: 4 months<br>• Reauthorization: 12 months |

POLICY NAME:
**LOVOTIBEGLOGENE AUTOTEMCEL**
Affected Medications: LYFGENIA (lovotibeglogene autotemcel)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Treatment of sickle cell disease in adults and pediatric patients at least 12 years of age with a history of recurrent vaso-occlusive crises |
| **Required Medical Information:** | • Documentation of sickle cell disease confirmed by genetic testing to show the presence of βS/βS, βS/β0 or βS/β+ genotype as follows:<br>  ○ Identification of significant quantities of HbS with or without an additional abnormal β-globin chain variant by hemoglobin assay<br>  **OR**<br>  ○ Identification of biallelic *HBB* pathogenic variants where at least one allele is the p.Glu6Val or p.Glu7Val pathogenic variant on molecular genetic testing<br>  **AND**<br>  ○ Patient does NOT have disease with more than two α-globin gene deletions<br>• Documentation of severe disease defined as 2 or more severe vaso-occlusive crises (VOCs) or vaso-occlusive events (VOEs) within the previous year (4 events over 2 years will also meet this requirement)<br>  ○ VOC/VOEs defined as an event requiring a visit to a medical facility for evaluation AND necessitating subsequent interventions such as opioid pain management, non-steroidal anti-inflammatory drugs, red blood cell (RBC) transfusions, which results in a diagnosis of such being documented due to one (or more) of the following:<br>  ▪ Acute pain event<br>  ▪ Acute chest syndrome<br>  ▪ Priapism lasting more than 2 hours<br>  ▪ Acute splenic sequestration<br>  ▪ Acute hepatic sequestration<br>• For patients under 18 years of age, the patient does not have a known and suitable (10/10) human leukocyte antigen (HLA) matched related donor willing to participate in an allogeneic hematopoietic stem cell transplant (HSCT)<br>• Adequate bone marrow, lung, heart, and liver function to undergo myeloablative conditioning regimen<br>• Confirmed HIV negative as confirmed by a negative HIV test prior to mobilization |
| **Appropriate Treatment Regimen & Other Criteria:** | • Able to provide the minimum recommended dose of Lyfgenia- $3 \times 10^6$ CD34+ cells/kg. |
| **Exclusion Criteria:** | • Previous treatment with gene therapy for sickle cell disease<br>• Prior hematopoietic stem cell transplant (HSCT)<br>• History of hypersensitivity to dimethyl sulfoxide (DMSO) or dextran 40 |
| **Age Restriction:** | • 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months (one-time infusion), unless otherwise specified |

POLICY NAME:
**LUSPATERCEPT-AAMT**
Affected Medications: REBLOZYL (luspatercept-aamt)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of anemia in adults with beta thalassemia who require regular red blood cell (RBC) transfusions<br>  ○ Treatment of anemia in adults without previous erythropoiesis stimulating agent use (ESA-naïve) with very low- to intermediate-risk myelodysplastic syndromes (MDS) who may require regular RBC transfusions<br>  ○ Treatment of anemia failing an ESA and requiring 2 or more RBC units over 8 weeks in adult patients with very low- to intermediate-risk MDS with ring sideroblasts (MDS-RS) or with myelodysplastic/myeloproliferative neoplasm with ring sideroblasts and thrombocytosis (MDS/MPN-RS-T) |
| **Required Medical Information:** | **Beta Thalassemia**<br>• Documented diagnosis of beta thalassemia OR hemoglobin E/beta thalassemia<br>• Documentation of transfusion dependence as evidenced by BOTH of the following in the previous 24 weeks:<br>  ○ Has required regular transfusions of at least 6 RBC units<br>  ○ No transfusion-free period greater than 35 days<br>• Pre-treatment or pre-transfusion hemoglobin (Hgb) level is less than or equal to 11 g/dL<br><br>**Myelodysplastic Syndromes**<br>• Documented diagnosis of MDS, MDS-RS or MDS/MPN-RS-T with very low, low, or intermediate risk as classified by the International Prognostic Scoring System-Revised (IPSS-R)<br>• Documentation of requiring at least 2 RBC units over the previous 8 weeks<br>• Pre-treatment or pre-transfusion level is less than or equal to 11 g/dL |
| **Appropriate Treatment Regimen & Other Criteria:** | **Myelodysplastic Syndromes**<br>• For those with MDS-RS or MDS/MPN-RS-T, must have documentation of treatment failure with an ESA (e.g., Retacrit, Procrit, Epogen, Mircera), unless intolerant or current endogenous serum erythropoietin (sEPO) level is greater than 500 U/L<br><br>**Reauthorization**<br>• **Beta thalassemia:** requires documentation of treatment success, defined as a reduction in RBC transfusion burden from baseline by at least 20%<br>• **MDS:** requires documentation of treatment success, defined as achieving transfusion independence and/or an improvement in Hgb level from baseline |
| **Exclusion Criteria:** | • Diagnosis of non-transfusion-dependent beta thalassemia<br>• Use as immediate correction as a substitute for RBC transfusions<br>• Diagnosis of alpha thalassemia<br>• Known pregnancy |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • **Beta thalassemia**: Prescribed by, or in consultation with, a hematologist<br>• **MDS**: Prescribed by, or in consultation with, a hematologist or oncologist |

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| | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**LUSUTROMBOPAG**
Affected Medications: MULPLETA (lusutrombopag)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Thrombocytopenia in adult patients with chronic liver disease who are scheduled to undergo a procedure |
| **Required Medical Information:** | • Documentation of ALL the following:<br>  ○ Planned procedure including date<br>  ○ Baseline platelet count of less than 50,000/microliter |
| **Appropriate Treatment Regimen & Other Criteria:** | • Approved for one time 7-day dosing regimen |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist or gastroenterology/liver specialist |
| **Coverage Duration:** | • Authorization: 1 month (7 days of treatment), based on planned procedure date, unless otherwise specified |

POLICY NAME:
**MARIBAVIR**
Affected Medications: LIVTENCITY (maribavir)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of adults and pediatric patients (12 years of age and older and weighing at least 35 kg) with post-transplant cytomegalovirus (CMV) infection/disease that is refractory to treatment (with or without genotypic resistance) with ganciclovir, valganciclovir, cidofovir or foscarnet |
| **Required Medical Information:** | • Documentation of treatment refractory CMV infection or disease following hematopoietic stem cell transplant (HSCT) or solid organ transplant (SOT)<br>• Documentation of current weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented clinical failure (defined as detectable plasma CMV DNA) after minimum 3-week trial with at least one of the following: valganciclovir, ganciclovir, foscarnet, cidofovir<br><br>**<u>Reauthorization</u>:**<br>• Documented treatment success and a clinically significant response to therapy and continued need for treatment |
| **Exclusion Criteria:** | • CMV infection involving the central nervous system, including the retina<br>• Prophylaxis of CMV infection/disease |
| **Age Restriction:** | • 12 years and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by an infectious disease provider or a specialist with experience in the treatment of CMV infection |
| **Coverage Duration:** | • Authorization: 2 months, unless otherwise specified |

POLICY NAME:
**MARSTACIMAB**
Affected Medications: HYMPAVZI (marstacimab hncq)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Hemophilia A (congenital factor VIII deficiency)<br>  ○ Hemophilia B (congenital factory IX deficiency) |
| **Required Medical Information:** | • Diagnosis of congenital factor VIII deficiency (hemophilia A) or congenital factory IX deficiency (hemophilia B) without inhibitors<br>• Documentation of baseline factor level less than 1% AND prophylaxis required **OR**<br>• Baseline factor level 1% to 3% and a documented history of at least two episodes of spontaneous bleeding into joints<br>• Prescribed for routine prophylaxis to prevent or reduce the frequency of bleeding episodes |
| **Appropriate Treatment Regimen & Other Criteria:** | **Hemophilia A**<br>• Documented treatment failure with Hemlibra (emicizumab-kxwh)<br><br>**Hemophilia B**<br>• Documented treatment failure to factor IX prophylaxis for at least 6 months<br><br>Dose escalation to 300 mg once weekly:<br>• Documented of weighing at least 50 kg and experiencing at least 2 breakthrough bleeds while on 150 mg dose for at least 6 months<br><br>**Reauthorization** requires documentation of treatment success defined as a reduction in spontaneous bleeds requiring treatment, and documentation of bleed history since last approval |
| **Exclusion Criteria:** | • Concurrent use with bypassing agents<br>• Prior gene therapy administration<br>• Pregnancy |
| **Age Restriction:** | • 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**MAVACAMTEN**
Affected Medications: CAMZYOS (mavacamten)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ Hypertrophic cardiomyopathy with left ventricular outflow tract obstruction |
| **Required Medical Information:** | • Documented diagnosis of obstructive hypertrophic cardiomyopathy (OHCM)<br>• New York Heart Association (NYHA) class II or III symptoms<br>• Left ventricular ejection fraction (LVEF) of 55% or greater prior to starting therapy<br>• Valsalva left ventricular outflow tract (LVOT) peak gradient of 50 mmHg or greater at rest or with provocation, prior to starting therapy |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of negative pregnancy test **AND** use of effective contraception in females of reproductive potential<br>• Documented treatment failure, intolerance, or contraindication, to ALL of the following:<br>  ○ Non-vasodilating beta-blocker (e.g., atenolol, metoprolol, bisoprolol, propranolol)<br>  ○ Non-dihydropyridine calcium channel blocker (e.g., verapamil, diltiazem)<br><br>**Reauthorization** will require documentation of symptomatic improvement and that LVEF remains above 50% |
| **Exclusion Criteria:** | • History of two measurements of LVEF less than 50% while on mavacamten 2.5 mg tablets |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or a specialist with experience in the treatment of obstructive hypertrophic cardiomyopathy<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**MAVORIXAFOR**
Affected Medications: XOLREMDI (mavorixafor)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of WHIM syndrome (warts, hypogammaglobulinemia, infections and myelokathexis) in patients 12 years of age and older to increase the number of circulating mature neutrophils and lymphocytes. |
| **Required Medical Information:** | • Diagnosis of WHIM syndrome confirmed by genotype variant of CXCR4 and ANC (absolute neutrophil count) of 400 cells/μL or less<br>• Documentation of symptoms and complications associated with WHIM syndrome requiring medical treatment |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of weight to assess appropriate dosing<br>• Documentation of baseline ALC (absolute lymphocyte count) and ANC (absolute neutrophil count) to assess clinical response to treatment<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy with sustained improvement in ALC and ANC |
| **Exclusion Criteria:** | • Concomitant use with drugs that are highly dependent on CYP2D6 for clearance. |
| **Age Restriction:** | • 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an immunologist or hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**MECASERMIN**
Affected Medications: INCRELEX (mecasermin)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Severe primary insulin-like growth factor-1 (IGF-1) deficiency (Primary IGFD)<br>  ○ Patient with growth hormone (GH) gene deletion with neutralizing antibodies to GH |
| **Required Medical Information:** | • Prior to starting therapy, a height at least 3 standard deviations below the mean for chronological age and sex, and an IGF-1 level at least 3 standard deviations below the mean for chronological age and sex.<br>• One stimulation test showing patient has a normal or elevated GH level |
| **Appropriate Treatment Regimen & Other Criteria:** | • Initial: 0.04-0.08 mg/kg subcutaneously twice daily.<br>• Maintenance: Up to 0.12 mg/kg subcutaneously twice daily.<br>• **Reauthorization**: requires a documented growth rate increase of at least 2.5 cm over baseline per year AND evaluation of epiphyses (growth plates) documenting they remain open. |
| **Exclusion Criteria:** | • Epiphyseal closure, active or suspected neoplasia malignancy, or concurrent use with GH therapy.<br>• Patient has secondary causes of IGF1 deficiency (e.g., hypothyroidism, malignancy, chronic systemic disease, skeletal disorders, malnutrition, celiac disease) |
| **Age Restriction:** | • For patients 2 to 18 years of age. |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a pediatric endocrinologist<br>• All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**MEDICAL NECESSITY**
Affected Medications: Abilify MyCitea, Abirtega, Abrilada, Absorica, Absorica LD, Acanya, Aciphex, Actemra SQ, Acthar Gel, Acuvail, Acyclovix, Aczone, Adalimumab-aacf, Adalimumab-aaty, Adalimumab-adbm, Adalimumab-fkjp, Adalimumab-ryvk, Adapalene pads, Adcirca, Adlarity, Adlyxin, Admelog, Advicor, Adzenys ER, Adzenys XR, Aerospan, Afrezza, Aimovig, AirDuo, AirDuo Digihaler, Airsupra, Ajovy, Aklief, Allopurinol 200 mg tablet, Allzital, Alprazolam Dispersible, Alprazolam Intensol, Altoprev, Alvesco, Ameluz, Amitiza, Amjevita, Amphetamine ER suspension, Ampyra, Amrix, Amturnide, Amzeeq, Ancobon, Androgel, Androxy, Anzupgo, Apadaz, APAP-Caff-Dihydrocodeine, Apidra, Aplenzin, Aptiom, Arazlo, Arbli, Aripiprazole Dispersible, Armonair Digihaler, Armonair Respiclick, Arymo ER, Asacol HD, Asmanex, Asmanex HFA, Aspruzyo, Astepro solution, Atorvaliq, Aubagio, Auvelity, Aveed, Azathioprine tablet (75 mg, 100 mg), Azelex, Azesco, Azmiro, Azstarys, Baclofen Oral Suspension, Basaglar, Basaglar Tempo pen, Baxdela, Beconase, Belbuca, Beser, Bevespi Aerophere, Bexagliflozin, BiDil, Biifenac, Bildyos, Bimzelx, Bismuth Subcitrate-Metronidazole-Tetracycline, Blujepa, Brekiya, Brenzavvy, Breztri, Bridion, Brilinta, Brinsupri, Brisdelle, Briviact, Bryhali, Brynovin, Bucapsol, Budesonide 9 mg ER tablet, Bunavail, Bupap, Buphenyl, Bupropion XL 450 mg, Butisol, Butrans patch, Bydureon, Bydureon BCise, Byetta, Bynfezia, Byvalson, Cabtreo, Calcipotriene-Betamethasone Dipropionate suspension, Cambia, Capex shampoo, Capital-Codeine, Carac, Carbinoxamine 6 mg tablet, Carisoprodol-ASA, Carisoprodol-ASA-Codeine, CaroSpir, Carticel implant, Cataflam, Cephalexin 750 mg capsule, Cephalexin tablet, Cequa, Chlorpheniramine-Codeine, Chlorzoxazone 250 mg tablet, Cibinqo, Ciloxan, Cimzia, Ciprodex OTIC, Cipro HC Otic, Clemastine syrup, Clindamycin Phosphate-Benzoyl Peroxide gel 1.2-2.5 %, Clindavix, Clobetasol ophthalmic suspension, Clobetex, Clonidine ER 0.17 mg tablet, Codar AR, Colazal, Conjupri, Consensi, Conzip, Copaxone, Coreg CR, Cosopt PF, Cotempla XR ODT, Coxanto, Crexont, Crinone, Cuprimine, Cuvposa, Cyanocobalamin Nasal Spray, Cyclobenzaprine ER, Cyclosporine in Klarity, Cyltezo, Dapagliflozin, Dapagliflozin-Metformin ER, Dartisla ODT, Debacterol, Degludec, Delzicol, Demser, Depen, DermacinRx Lexitral cream pack, Dermalid, Desmoda, Desonate gel, Desonide gel, Desonide lotion, DesRx gel, Dexilant, Dhivy, Dichlorphenamide, Diclofenac 1.3 % patch, Diclofenac Potassium capsule, Diclofenac Potassium packet, Diclofenac Potassium 25 MG tablet, Diclofenac Sod soln 1.5 % & Capsaicin cream 0.025 % therapy pack, Diclofex DC cream, Diclopak, Diclosaicin cream, Diclotral pack, Diclotrex, Diclovix DM pak, Dificid, Diflorasone Diacetate, Dipentum, Doryx MPC, Doxepin 5 % cream, Doxycyline Hyclate 50 mg tablet, Doxycycline Hyclate DR tablet (50 mg, 80 mg, 200 mg), Doxycycline Monohydrate DR 40 mg capsule, Duaklir Pressair, Duetact, Duexis, Dulera, Duobrii, Durlaza, Dutoprol, Duzallo, Dxevo, Dyanavel XR, Dymista, Dynabec, Ebglyss, Econasil, Edarbi, Edarbyclor, Edurant PED, Egaten, Egrifta WR, Elepsia XR, Elidel, Elyxyb, Emflaza, Emflaza Suspension, Emrosi, Enalapril oral solution, Enbumyst, Enstilar foam, Entadfi, Entresto, Entyvio SQ, Eohilia, Epaned, Epanova, Epclusa, Eprontia, Equetro, Ergomar, Esbriet, Eskata, Evzio, Exdensur, Exjade, Exservan, Extina foam 2 %, Exxua, Fabior foam, Faslodex, Fenofibrate 120 mg, Fenortho, Firazyr, First-lansoprazole, Flector patch, Fleqsuvy, Flolipid, Flowtuss, Fluopar kit, Fluorouracil 0.5 % cream, Flurandrenolide, Fluoxetine (PMDD) tablet, Forfivo XL, Fortamet, Fortesta gel, Fosamax Plus D, Fulyzaq, Furoscix, Fycompa, Gabacaine pak, Gabapal, Giazo, Gilenya, Gimoti, Gleevec, Gloperba, Glumetza, Glycate, Glycopyrrolate 1.5 mg tablet, Gocovri, Gonitro, GPL pak, Halog, Halcinonide cream, Harliku, Harvoni, Harvoni pak, Helidac, Hemady, Hemangeol, Hemiclor, Hetlioz capsule, Hulio, Humalog, Humalog Junior KwikPen, Humatin, Humira, Humulin, Humulin 70/30 KwikPen, Humulin N, Humulin R-100, Hycofenix, Hyrimoz (Sandoz), Ibsrela, Ibuprofen-Famotidine, Idacio, Igalmi, Iheezo, Ilumya, Imbruvica 70 mg capsule, Imbruvica 140 mg & 280 mg tablet, Imiquimod 3.75 %, Imkeldi, Impeklo, Impoyz, Imuldosa, Imvexxy, Inbrija, Inderal LA, Indocin suppository, Indomethacin 20 mg capsule, Inflatherm kit, Inflatherm pak, Infugem, Ingrezza, Ingrezza Sprinkle, Innolet Insulin, Inpefa, Insulin Aspart, Insulin Aspart Protamine & Aspart 70/30, Insulin Degludec, Insulin Glargine, Insulin Glargine-yfgn, Insulin Lispro, Intrarosa, Invega ER, Invokamet, Invokamet XR, Invokana, Inzirqo, Isordil Titradose, Isosorbide Dinitrate-Hydralazine, Isotretinoin 25 mg and 35 mg capsule, Iyuzeh, Jadenu, Jadenu sprinkle packet, Jascayd, Javadin, Jentadueto, Jentadueto XR, Jublia, Jylamvo, Karbinal ER, Katerzia, Kazano, K-bicarb, Kenalog aerosol, Kenalog susp, Keragel, KeragelT, Kerydin, Kesimpta, Ketek, Ketorolac nasal spray, Keveyis, Kevzara, Khindivi, Kineret, Kirsty, Klisyri, Kombiglyze XR, Konvomep, Korlym, Kyzatrex, Lampit, Lasix Onyu, Latuda, Lescol XL, Letairis, Levamlodipine, Levorphanol Tartrate, Lexette, Lexuss, Lialda, Libervant, Licart, Lido GB 300 kit, Lidostream, Lidotin Pak, Lifems, Likmez, Lipritin Pak, Liptruzet, Lithostat, LMR Plus Lidocaine, Lodoco, Lofena, Lonhala Magnair, Lopressor solution, Loreev XR, Lotilaner, Lotronex, Lucemyra, Luzu, Lybalvi, Lynkuet, Lyrica, Lyrica CR tablet, Lyumjev, Lyumjev Kwikpen, Lyvispah, Meclofen, Meloxicam capsule, Mentax cream 1 %, Merilog, Mesalamine DR 800 mg tablet, Mesnex, Metaclopramide disintegrating tablet, Metaxall, Metaxall CP, Metformin ER (OSM), Metformin solution, Methadone Intensol,

MethylTESTOSTERone capsule, Metyrosine, Miebo, Mifepristone, Migraine pack, Minocycline ER, Minolira, Mitigare, mNexspike, Monocycline ER, MorphaBond, MorphaBond ER, Motegrity, Motofen, Motpoly XR, Mycapssa, Myfembree, Myhibbin, Myqorzo, Myrbetriq, Mytesi, Nalocet, Namenda XR, Namzaric, Naprelan, Naproxen-Esomeprazole, Nascobal, Natesto gel, Neo-Synalar cream, Nesina, Nexiclon XR, Nexletol, Nexlizet, Nilotinib-D, Nitisinone, Nityr, Nocdurna, Noctiva, Nolix, Nopioid TC kit, Norgesic Forte, Noritate, Norliqva, Noroxin, Northera, Nourianz, Novolin 70/30 Relion, Novolin N Relion, Novolin R Relion, Noxafil, NuDiclo Solupak, Nurtec, Nuvakaan kit, Nuvakaan II kit, Nuvigil, Nuzyra, Ofloxacin tablet, Ohtuvayre, Olpruva, Olumiant, Olysio, Omeprazole-Sodium Bicarb, Omlonti, Omnaris, Omvoh SQ, Ondansetron 24 mg tablet, Onexton, Onfi, Onglyza, Ontralfy, Onyda XR, Onzetra Xsail, Opipza, Oracea, Oralair, Orencia SQ, Orfadin, Orlynvah, Ormalvi, Orphenadrine-Aspirin-Caffeine tablet, Orphengesic Forte, Ortikos, Oseni, Otrexup, Otulfi, Oxaprozin capsule, Oxaydo, Oxycodone-Acetaminophen (2.5 mg-300 mg, 5 mg-300 mg, 7.5 mg-300 mg, 10 mg-300 mg), Ozobax, Palsonify, Pamelor, Panlor, Panretin gel, Paromomycin, Pazeo, Pedizolpak, Penicillamine tablet, Pennsaid solution, Pentican pak, Percocet, Pertzye, Pheburane, Pivya, Phyrago, Picato, Pioglitazone-Glimepiride, Pirfenidone 534 mg tablet,Pivya, Plaquenil, Pradaxa, Praluent, Prevacid SoluTab, Prevpac, Prialt, Prilo Patch, Prilopentin, Primlev, Primsol, Pristiq, ProAir Digihaler, Prolate, Promacta, Prudoxin, Purified Cortrophin gel, Purixan, Pyzchiva, Qbrelis, Qbrexza, Qdolo, Qelbree, Qfitlia, Qmiiz, QNASL, Qtern, Qudexy XR, QuilliChew ER, Quillivant XR, Quinixil, Quinosone, Qulipta, Qwo, Raldesy, Ranexa, Rasuvo, Rayos, Recarbrio, Reditrex, Relexxii, Relion Insulins, Relprevv, Reltone, Retin-A Micro pump gel (0.06 %, 0.08 %), Revatio, Rezvoglar, Rhapsido, Rhofade, Ribasphere, Ridaura, Riomet, Riomet ER, Rocklatan, Ryaltris, Ryvent, Ryzodeg 70/30, Sabril, Samsca, Saphris, Sarafem, Savaysa, Saxagliptin-Metformin ER, Sdamlo, Seconal, Seebri Neohaler, Seglentis, Segluromet, Semglee, Sensipar, Sephience, Sernivo, Seysara, Siklos, Silenor, Sila III pak, Siliq subcutaneous injection, Simlandi, Simponi, Simvastatin suspension, Skelaxin, Skelid, Soaanz, Sofdra, Soliqua, Solodyn, Solosec, Soolantra, Sorilux, Sotyktu, Sovaldi, Sovaldi pak, Spevigo Subcutaneous, Spironolactone suspension, Sporanox solution, Spritam, Sprix, Sprycel, Starjemza, Steglatro, Steglujan, Stelara, Steqeyma, Striant, Striant buccal, Suboxone, Subvenite, Sumatriptan-Naproxen, Sure Result DSS premium pack, Symbravo, Symbyax, Sympazan, Symproic, Synalar, Syndros, Syprine, Taclonex suspension, Talicia, Taltz, Tanzeum, Targadox, Tascenso ODT, Tasoprol, Tavaborole, Tazarotene foam, Tazarotene cream 0.05%, Tazorac Cream, Tazorac Gel, Tecfidera, Technivie, Tezruly, Thalitone, Thiola, Thiola EC, Thyquidity, Ticlopidine, Tiglutik, Tiopronin, Tivorbex, Tolak, Tolsura, Tonmya, Topiramate ER, Tosymra, Tovet kit, Tracleer, Tradjenta, Tramadol oral solution, Tretinoin Microsphere Gel 0.08 %, Treximet, Tri-Luma, Trixylitral kit, Trokendi XR, Trudhesa, Trulance, Tryptyr, Tudorza Pressair, Twyneo, Tyenne subcutaneous injection, Tyruko, Tyrvaya, Tyzeka, Tyzine, Ultravate, Ultresa, Umeclidinium bromide-Vilanterol, Uptravi, Ursodiol capsule (200 mg, 400 mg), Ustekinumab,Utibron Neohaler, Uzedy, Valsartan oral solution, Vanatol LQ, Vanos, Varophen, Vasotec, Vecamyl, Vectical, Veltassa, Venlafaxine Besylate ER, Veozah, Veramyst, Veregen, Verkazia, Versacloz, Vesicare LS, Vevye, Vexasyn, Vexasyn gel, Vfend oral suspension, V-Go, Viberzi, Vibramycin, Victoza, Victrelis, Viekira, Vigafyde, Viibryd, Viibryd Starter Pack, Vimovo, Viokace, Vivlodex, Vizz, Vogelxo, Voquezna dual pak, Voriconazole oral suspension, Vtol LQ solution, Vuity, Vybrique, Vyscoxa, Vyzulta, Wakix, Wegovy, Wezlana, Winlevi, Wynorza, Xaciato, Xadago, Xartemis XR, Xatmep, Xcopri, Xdemvy, Xelitral pack, Xeloda, Xelstrym, Xenazine, Xenleta, Xerese, Xermelo, Xhance, Ximino, Xromi, Xtampza ER, Xultophy, Xyosted, Yosprala, Yuflyma, Yupelri, Yusimry, Yutrepia, Yuvezzi, Yuviwel, Zanaflex capsule, Zavzpret, Zcort, Zebutal, Zecuity, Zelnorm, Zembrace, Zenevix, Zepatier, Zepbound, Zetonna, Zileuton ER, Zinbryta, Zipsor, Zituvimet, Zituvimet XR, Zituvio, Zolpak, Zolpidem capsule, Zolpimist, Zonalon, Zonisade, Zorvolex, ZTLido, Z-Tuss, Zunveyl, Zyclara, Zymfentra, Zypitamag, Zytiga

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Documented intolerance or treatment failure with the formulary alternatives for the submitted diagnosis |
| **Appropriate Treatment Regimen & Other Criteria:** | • Food and Drug Administration (FDA)-approved or compendia supported dosing |
| **Exclusion Criteria:** | • Uses that are considered experimental, such as concomitant use with other drugs in a manner considered experimental |

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| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Dependent on expected duration of therapy and necessity of documentation of response to therapy |

POLICY NAME:
**MEDICAL NECESSITY– ZEPBOUND**
Affected Medications: ZEPBOUND (tirzepatide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Treat moderate to severe obstructive sleep apnea (OSA) in combination with a reduced-calorie diet and increased physical activity in adults with obesity |
| **Required Medical Information:** | • Diagnosis of moderate to severe obstructive sleep apnea (OSA) with Apnea-Hypopnea Index (AHI) of at least 15 on polysomnography or home sleep study from the past 5 years<br>• Body mass index (BMI) of 30 $kg/m^2$ or greater<br>• Current symptoms of OSA such as excessive daytime sleepiness, loud snoring, choking, gasping, pauses in breathing, sleep arousals, and difficulty maintaining sleep throughout the night<br>• Documentation of being used in combination with a physician-directed weight loss program that involves a reduced calorie diet, increased physical activity, and behavioral modification |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented intolerance or treatment failure with the formulary alternatives for the submitted diagnosis<br>• Documentation of one of the following:<br>  ○ Treatment failure with positive airway pressure (PAP) therapy, defined as having continued symptoms of OSA despite four or more hours of PAP use per night for 70 percent or more of nights<br>  ○ Contraindication to PAP therapy (such as upper airway anatomic abnormalities, pneumothorax or invasive mechanical ventilation)<br>• Documentation of being prescribed concurrently with positive airway pressure (PAP) therapy, unless contraindicated or clinically significant adverse effects are experienced<br><br>**Reauthorization** requires documentation of treatment success defined by all the following:<br>• Improvement in AHI score or OSA symptoms such as excessive daytime sleepiness, loud snoring, choking, gasping, pauses in breathing, sleep arousals, and difficulty maintaining sleep throughout the night<br>• Achieved and maintained 10 percent or greater weight loss after starting Zepbound (tirzepatide)<br>• Continued adherence to a reduced calorie diet, increased physical activity, and behavioral modification program |
| **Exclusion Criteria:** | • Use for weight loss (no OSA diagnosis) or other excluded diagnosis<br>• Diagnosis of type 1 or type 2 diabetes with or without OSA<br>• Diagnosis of central or mixed sleep apnea<br>• Diagnosis of obesity hypoventilation syndrome or daytime hypercapnia<br>• History of ketoacidosis<br>• Personal or family history of medullary thyroid carcinoma (MTC) or Multiple Endocrine Neoplasia syndrome type 2 (MEN 2)<br>• Concurrent use of other glucagon-like peptide-1 (GLP-1) receptor agonists |
| **Age Restriction:** | • 18 years of age or older |

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| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months |

POLICY NAME:
**MEDICAL NECESSITY POLICY - WEGOVY**
Affected Medications: WEGOVY (semaglutide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Treatment of noncirrhotic metabolic dysfunction-associated steatohepatitis (MASH), formerly known as nonalcoholic steatohepatitis (NASH), with moderate to advanced liver fibrosis (consistent with stages F2 to F3 fibrosis) in adults in combination with a reduced calorie diet and increased physical activity<br>  ○ To reduce the risk of major adverse cardiovascular (CV) events (CV death, non-fatal myocardial infarction, or non-fatal stroke) in adults with established CV disease and either obesity or overweight |
| **Required Medical Information:** | **<u>Major Adverse Cardiovascular Event (MACE) Risk Reduction:</u>**<br>• Documented history of prior cardiovascular event defined as one of the following:<br>  ○ Myocardial infarction<br>  ○ Stroke (ischemic or hemorrhagic stroke)<br>  ○ Symptomatic peripheral artery disease (PAD) such as intermittent claudication with ankle-brachial index (ABI) less than 0.85 at rest, or history of peripheral arterial revascularization procedure<br>• Body mass index (BMI) of 27 kg/m2 or greater<br>• Used in combination with caloric restriction (diet), increased physical activity, and behavioral modification<br><br>**<u>Metabolic dysfunction-associated steatohepatitis (MASH):</u>**<br>• Diagnosis of noncirrhotic nonalcoholic steatohepatitis (NASH) or MASH with moderate to advanced (F2 to F3) liver fibrosis confirmed by ONE of the following:<br>  ○ Conclusive result from a well-validated non-invasive test such as:<br>  ▪ Fibroscan-AST (FAST) score<br>  ▪ MAST (score from MRI–proton density fat fraction, Magnetic resonance elastography [MRE], and serum AST)<br>  ▪ MEFIB (Fibrosis-4 Index greater than or equal to 1.6 and MRE greater than or equal to 3.3 kPa)<br>  ○ Liver biopsy (also required if non-invasive testing is inconclusive or other causes for liver disease have not been ruled out)<br>• Other causes for liver steatosis have been ruled out (such as alcohol-associated liver disease, chronic hepatitis C, Wilson disease, drug-induced liver disease)<br>• Baseline lab values for AST and ALT |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>MACE Risk Reduction:</u>**<br>• Documented intolerance or treatment failure with the formulary alternatives for cardiovascular disease, including:<br>  ○ Lipid lowering agents<br>  ○ Anti-platelets and anticoagulants<br>  ○ Antihypertensive agents |

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| | **Reauthorization:**<br>• Documentation of treatment success (MACE risk reduction)<br><br>**<u>MASH:</u>**<br>• Documented treatment failure (or intolerable adverse event) with at least 12 weeks of one of the following: Mounjaro, Ozempic, Trulicity, liraglutide, or Rybelsus<br>• Documentation of abstinence from alcohol consumption<br>• Documentation of comprehensive comorbidity management being undertaken, including all the following:<br>  ○ Use of diet and exercise for weight management<br>  ○ Medications to manage associated comorbid conditions, such as thyroid disease (must not have active disease), diabetes, dyslipidemia, hypertension, or cardiovascular conditions.<br><br>**Reauthorization:**<br>• Documentation of disease responsiveness to therapy based on improvements or stability in laboratory results, such as ALT and AST, or fibrosis as evaluated by a non-invasive test |
| **Exclusion Criteria:** | • Use for weight loss (no MACE or MASH diagnosis) or other excluded diagnosis<br>• Diagnosis of type 1 or type 2 diabetes without MACE or MASH |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist (MACE reduction)<br>• Prescribed by, or in consultation with, a gastroenterologist or hepatologist (MASH/NASH) |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**MEK INHIBITORS FOR NEUROFIBROMATOSIS TYPE 1 (NF1)**
Affected Medications: KOSELUGO (selumetinib), GOMEKLI (mirdametinib)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Neurofibromatosis type 1 with symptomatic, inoperable plexiform neurofibromas in pediatric patients 1 year of age and older<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | • Documented body surface area (BSA) and requested dose (all indications)<br><br>**<u>Neurofibromatosis type 1 (NF1) with inoperable plexiform neurofibromas</u>**<br>• Documentation of diagnosis of symptomatic and/or progressive, inoperable NF1, defined as one or more plexiform neurofibromas that cannot be completely removed without risk for substantial morbidity due to encasement of, or close proximity to, vital structures, invasiveness, or high vascularity<br>• Documentation of 2 or more of the following clinical diagnostic criteria as evaluated by a multidisciplinary specialist care team (a child of a parent with NF1 can be diagnosed if one or more of these criteria are met):<br>  ○ Six or more café-au-lait macules over 5 mm in greatest diameter in prepubertal individuals and over 15 mm in greatest diameter in post pubertal individuals<br>  ○ Freckling in the axillary or inguinal region<br>  ○ Two or more neurofibromas of any type or one plexiform neurofibroma<br>  ○ Optic pathway glioma<br>  ○ Two or more iris Lisch nodules identified by slit lamp examination or two or more choroidal abnormalities<br>  ○ A distinctive osseous lesion such as sphenoid dysplasia, anterolateral bowing of the tibia, or pseudarthrosis of a long bone<br>  ○ A heterozygous pathogenic NF1 variant with a variant allele fraction of 50% in apparently normal tissue such as white blood cells<br><br>**<u>NCCN Indications</u>**<br>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | • Coverage of Gomekli requires documentation of the following:<br>  ○ Documentation of intolerable adverse event to Koselugo OR 18 years of age and older<br>  ○ Dosing is limited to 2 $mg/m^2$<br><br>**<u>Reauthorization</u>** requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | **<u>Neurofibromatosis type 1 (NF1) with inoperable plexiform neurofibromas</u>**<br>• 1 to 18 years of age (Koselugo)<br>• 2 years of age and older (Gomekli) |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified |

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| | • Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**METHYLNALTREXONE**
Affected Medications: RELISTOR (methylnaltrexone bromide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Opioid-induced constipation in adult patients with advanced illness or pain caused by active cancer who require opioid dosage escalation for palliative care<br>  ○ Opioid-induced constipation in adult patients with chronic non-cancer pain, including patients with chronic pain related to prior cancer or its treatment who do not require frequent (e.g., weekly) opioid dosage escalation |
| **Required Medical Information:** | • Documentation of treatment of opioid-induced constipation (OIC) in an adult with:<br>  ○ Advanced illness who is receiving palliative care<br>  **OR**<br>  ○ Chronic non-cancer pain who has taken opioids for at least 4 weeks |
| **Appropriate Treatment Regimen & Other Criteria:** | **OIC in adults with chronic non-cancer pain**<br>• Documented treatment failure or contraindication to a trial of all of the following:<br>  ○ Lubiprostone<br>  ○ Linzess<br>  ○ Movantik<br><br>**Reauthorization** will require documentation of treatment success, a clinically significant response to therapy, and documentation of continued opioid use |
| **Exclusion Criteria:** | • Known or suspected mechanical gastrointestinal obstruction or increased risk for recurrent obstruction |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**METRELEPTIN**
Affected Medications: MYALEPT (metreleptin)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Congenital or acquired generalized lipodystrophy as a result of leptin deficiency |
| **Required Medical Information:** | • Current weight<br>• Baseline serum leptin levels, hemoglobin A1c (HbA1c), fasting glucose, fasting triglycerides, fasting serum insulin<br>• Prior Myalept use will require testing for anti-metrepeptin antibodies<br>• Documented leptin deficiency confirmed by laboratory testing (serum leptin of less than 12 ng/mL)<br>• Documentation of congenital or acquired generalized lipodystrophy with least **ONE** of the following:<br>  ○ Concurrent hypertriglyceridemia<br>  ○ Concurrent diabetes |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Generalized lipodystrophy with concurrent hypertriglyceridemia</u>**<br>• Triglycerides of 500 mg/dL or higher despite optimized therapy with at least two triglyceride-lowering agents from different classes (e.g., fibrates, statins) at maximum tolerated doses for at least 12 weeks each<br><br>**<u>Generalized lipodystrophy with concurrent diabetes</u>**<br>• Persistent hyperglycemia (HbA1c 7 percent or greater) despite dietary intervention and optimized insulin therapy at maximally tolerated doses for at least 12 weeks<br><br>**<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy documented by increased metabolic control defined by improvement in HbA1c, fasting glucose, and fasting triglyceride levels |
| **Exclusion Criteria:** | • Partial lipodystrophy<br>• General obesity not associated with leptin deficiency<br>• HIV-related lipodystrophy<br>• Metabolic disease, including diabetes mellitus and hypertriglyceridemia, without concurrent documentation of generalized lipodystrophy |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist<br>• All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**MIACALCIN**
Affected Medications: MIACALCIN injection (calcitonin-salmon)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Paget's disease of bone<br>  ○ Hypercalcemia |
| **Required Medical Information:** | **<u>Hypercalcemia</u>**<br>• Documented calcium level greater than or equal to 14 mg/dL (3.5 mmol/L)<br><br>**<u>Paget's disease of bone</u>**<br>• Documented baseline radiographic findings of osteolytic bone lesions<br>• Abnormal liver function test (LFT), including alkaline phosphatase<br>• Documented lack of malignancy within the past 3 months |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Hypercalcemia</u>**<br>• Documentation that additional methods for lowering calcium (such as intravenous fluids) did not result in adequate efficacy **OR**<br>• Clinical judgement necessitated immediate administration without waiting for other methods to show efficacy<br><br>**<u>Paget's disease of bone</u>**<br>• Documented trial and failure (or intolerable adverse event) with an adequate trial of both of the following:<br>  ○ Zoledronic acid (at least one dose)<br>  ○ Oral bisphosphonate (e.g., alendronate, risedronate) for at least 8 weeks<br>**OR**<br>• Documentation that the patient has severe renal impairment (e.g., creatinine clearance less than 35 mL/min)<br>**AND**<br>• Documentation of all of the following:<br>  ○ Normal vitamin D and calcium levels and/or supplementation<br>  ○ Symptoms that necessitate treatment with medication (e.g., bone pain, bone deformity)<br><br>**<u>Reauthorization – Paget's disease of bone:</u>**<br>• Documentation of treatment success and a clinically significant response to therapy (such as stable or lowered alkaline phosphatase level, resolution of bone pain or other symptoms) |
| **Exclusion Criteria:** | • Related to Paget's disease of bone<br>  ○ History of a skeletal malignancy or bone metastases<br>  ○ Concurrent use of zoledronic acid or oral bisphosphonates<br>  ○ Asymptomatic Paget's Disease of the bone<br>• Treatment or prevention of osteoporosis |
| **Age Restriction:** | • 18 years of age or older - for Paget's disease of bone only |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**MIGLUSTAT**
Affected Medications: MIGLUSTAT

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of adult patients with mild to moderate type 1 Gaucher disease<br>• Compendia-supported uses that will be covered:<br>  ○ Niemann-Pick disease type C (NPC) |
| **Required Medical Information:** | **Gaucher Disease**<br>• Diagnosis of Gaucher disease confirmed by ONE of the following:<br>  ○ An enzyme assay demonstrating a deficiency of betaglucocerebrosidase enzyme activity<br>  ○ Detection of biallelic pathogenic variants in the GBA gene by molecular genetic testing<br>• Enzyme replacement therapy is not a therapeutic option (e.g., due to allergy, hypersensitivity, or poor venous access)<br><br>**NPC**<br>• Diagnosis of NPC confirmed by genetic testing showing biallelic pathogenic variants in either the NPC1 gene or NPC2 gene<br>• Documentation of at least one neurological symptom of Niemann-Pick disease type C, such as:<br>  ○ Loss of motor function<br>  ○ Problems with swallowing or speech<br>  ○ Cognitive impairment<br>• Documentation of being ambulatory without needing an assistive device such as a wheelchair, walker, or cane<br>• Documentation of baseline signs and symptoms of NPC |
| **Appropriate Treatment Regimen & Other Criteria:** | **Gaucher Disease:**<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy<br><br>**NPC:**<br><br>**Reauthorization** requires:<br>• Documentation of treatment success defined as stability or improvement of Niemann-Pick disease type C signs and symptoms<br>• Documentation that patient is still ambulatory |
| **Exclusion Criteria:** | • Female of childbearing potential who is pregnant or planning a pregnancy |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, one of the following: |

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| | ○ A specialist in the management of Gaucher disease (hematologist, oncologist, hepatologist, geneticist or orthopedic specialist)<br>○ A specialist in the management of NPC (such as a geneticist, endocrinologist, metabolic disorder subspecialist, or neurologist)<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**MILTEFOSINE**
Affected Medications: IMPAVIDO (miltefosine)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of the following in adults and pediatric patients 12 years of age and older weighing greater than or equal to 30 kg (66 lbs):<br>  ▪ Visceral leishmaniasis caused by *Leishmania donovani*<br>  ▪ Cutaneous leishmaniasis caused by *Leishmania braziliensis*, *Leishmania guyanensis*, and *Leishmania panamensis*<br>  ▪ Mucosal leishmaniasis caused by *Leishmania braziliensis* |
| **Required Medical Information:** | **<u>All Indications</u>**<br>• Current weight<br><br>**<u>Visceral Leishmaniasis</u>**<br>• Documentation of diagnosis confirmed by smear or culture in tissue (usually bone marrow or spleen)<br><br>**<u>Cutaneous and Mucosal Leishmaniasis</u>**<br>• Documentation of diagnosis confirmed by histology, culture, or molecular analysis via polymerase chain reaction (PCR) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Dosing:</u>**<br>• 30 to 44 kg: 50 mg twice daily for 28 days<br>• 45 kg or greater: 50 mg three times daily for 28 days |
| **Exclusion Criteria:** | • Pregnancy<br>• Sjögren-Larsson syndrome<br>• Weight less than 30 kg (66 lbs)<br>• Treatment of leishmaniasis outside of the visceral, cutaneous, or mucosal settings<br>• Treatment of other *Leishmania* species |
| **Age Restriction:** | • 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an infectious disease specialist<br>• All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 1 month, unless otherwise specified |

POLICY NAME:
**MITAPIVAT**
Affected Medications: PYRUKYND (mitapivat tablet)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Hemolytic anemia due to pyruvate kinase deficiency (PKD) - Pyrukynd only<br>  ○ Treatment of anemia due to alpha- or beta-thalassemia – Aqvesme only |
| **Required Medical Information:** | **<u>Pyrukynd</u>**<br>• Documented diagnosis of pyruvate kinase deficiency (PKD), confirmed by BOTH of the following:<br>  ○ Presence of at least 2 variant alleles in the pyruvate kinase liver and red blood cell (PLKR) gene<br>  ○ At least one variant allele is a missense mutation<br><br>• Documentation of ONE of the following:<br>  ○ Regularly receiving red blood cell (RBC) transfusions, defined as 6 or more transfusions in the previous 12 months<br>  ○ Baseline hemoglobin (Hb) level of less than or equal to 10 g/dL with a history of no more than 4 transfusions in the previous 12 months<br><br>**<u>Aqvesme</u>**<br>• Documented diagnosis of alpha- or beta-thalassemia based on hemoglobin (Hb) electrophoresis, Hb high-performance liquid chromatography (HPLC), and/or deoxyribonucleic acid (DNA) analysis<br><br>• Documentation of ONE of the following:<br>  ○ Regularly receiving red blood cell (RBC) transfusions, defined as 6 or more transfusions in the previous 12 months<br>  ○ Baseline hemoglobin (Hb) level of less than or equal to 10 g/dL with a history of no more than 4 transfusions in the previous 6 months<br>AND<br>  ○ Documentation of inadequate clinical response or intolerance to Reblozyl (luspatercept-aamt)<br><br>**<u>All Indications</u>**<br>• Documentation of baseline transfusion count, including dates and number of units transfused |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Reauthorization</u>** requires documentation of treatment success and a clinically significant response to therapy, defined as:<br>• <u>For patients receiving regular transfusions at baseline</u>: must document greater than or equal to a 33% reduction in RBC units transfused compared to baseline<br>• <u>For patients not receiving regular transfusions at baseline</u>: must document greater than or equal to a 1.5 g/dL (for Pyrukynd) or 1.0 g/dL (for Aqvesme) increase in Hb from baseline sustained at 2 or more scheduled visits AND no transfusions were needed |
| **Exclusion** | • Splenectomy scheduled during treatment or have undergone within the 12-month period |

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| **Criteria:** | prior to starting treatment<br>• Previous bone marrow or stem cell transplant<br>• Previous gene therapy for thalassemia |
| **Age Restriction:** | • Must be 18 years or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**MOLLUSCUM CONTAGIOSUM AGENTS**
Affected Medications: YCANTH, ZELSUVMI

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Molluscum contagiosum (MC) |
| **Required Medical Information:** | • Diagnosis of MC confirmed by one of the following:<br>  ○ Presence of lesions that are consistent with MC (small, firm, pearly, with pitted centers, 2-5 millimeters in diameter, not associated with systemic symptoms such as fever)<br>  ○ For lesions with unclear cause or otherwise not consistent with MC, confirmation of diagnosis using dermoscopy, microscopy, histological examination, or biopsy<br>• Documentation of persistent itching or pain AND one of the following:<br>  ○ Concomitant bacterial infection of the lesion<br>  ○ Concomitant atopic dermatitis<br>  ○ Significant concern for contagion (such as daycare setting) and prevention cannot be reasonably prevented through good hygiene and covering lesions with bandages or clothing<br>  ○ Continued presence of lesions after 12 months |
| **Appropriate Treatment Regimen & Other Criteria:** | • Trial of at least two cycles of one of the following procedures for the removal of MC lesions:<br>  ○ Cryotherapy<br>  ○ Curettage<br>  ○ Laser therapy<br>• Adequate trial and failure of one additional treatment for MC that has evidence supporting use, such as:<br>  ○ Topical podofilox for at least 1 month<br>  ○ Oral cimetidine for at least 2 months<br>• **Dosing:**<br>  ○ Ycanth: Two applicators per treatment every 21 days, limit to 4 total treatments<br>  ○ Zelsuvmi: One kit for 12 weeks |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • Ycanth: 2 years of age and older<br>• Zelsuvmi: 1 year of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a dermatologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 3 months, unless otherwise specified |

POLICY NAME:
**MOMETASONE SINUS IMPLANT**
Affected Medications: SINUVA (mometasone sinus implant)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of chronic rhinosinusitis with nasal polyps in patients who have had ethmoid sinus surgery |
| **Required Medical Information:** | • Documented diagnosis of chronic rhinosinusitis with nasal polyps<br>• History of bilateral total ethmoidectomy<br>• Documentation of both of the following:<br>  ○ Presence of bilateral nasal polyps<br>  ○ Symptoms of sinonasal obstruction/congestion for over 12 weeks (decreased/absent sense of smell, facial pressure/pain, rhinorrhea/postnasal drip) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with at least 3 months of two intranasal corticosteroids after ethmoidectomy<br><br>**Reauthorization:** documentation of treatment success (reduction in symptoms, polyp size/obstruction, etc.), at least 9 months after previous treatment with Sinuva |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an otolaryngologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 1 month, unless otherwise specified |

POLICY NAME:
**MOTIXAFORTIDE**
Affected Medications: APHEXDA (motixafortide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan<br>  ○ In combination with filgrastim (granulocyte colony-stimulating factor [G-CSF]) to mobilize hematopoietic stem cells (HSCs) to the peripheral blood circulation to facilitate their collection for subsequent autologous stem cell transplantation (ASCT) in patients with multiple myeloma (MM)<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better (autologous HSCT must be NCCN recommended) |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• Documentation of diagnosis of multiple myeloma in first or second remission<br>• Eligible for Autologous stem cell transplantation (ASCT)<br>• At least 7 days from most recent high dose induction therapy<br>• No single agent chemotherapy or maintenance therapy within 7 days<br>• Eastern Cooperative Oncology Group (ECOG) performance status (PS) of 0 or 1 |
| **Appropriate Treatment Regimen & Other Criteria:** | • Inadequate stem cell collection amount despite previous trial with ALL the following:<br>  ○ Single agent granulocyte colony stimulating factor (G-CSF)<br>  ○ G-CSF in combination with plerixafor<br><br>**<u>No reauthorization</u>** |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or Eastern Cooperative Oncology Group (ECOG) performance status (PS) of 2 or greater |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 2 months unless otherwise specified |

POLICY NAME:
**MUCOPOLYSACCHARIDOSIS (MPS) AGENTS**
Affected Medications: VIMIZIM (elosulfase alfa), NAGLAZYME (galsulfase), MEPSEVII (vestronidase alfa-vjbk), ALDURAZYME (laronidase), ELAPRASE (idursulfase)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Vimizim: Mucopolysaccharidosis type IVA (MPS IVA; Morquio A syndrome)<br>  ○ Naglazyme: Mucopolysaccharidosis type VI (MPS VI, Maroteaux-Lamy syndrome)<br>  ○ Mepsevii: Mucopolysaccharidosis VII (MPS VII; Sly Syndrome)<br>  ○ Aldurazyme:<br>  ▪ Hurler Mucopolysaccharidosis type I (MPS I H)<br>  ▪ Herler-Scheie Mucopolysaccharidosis type I (MPS I H/S)<br>  ▪ Scheie form of Mucopolysaccharidosis (MPS I S) with moderate to severe symptoms<br>  ○ Elaprase: Mucopolysaccharidosis type II (MPS II; Hunters syndrome) |
| **Required Medical Information:** | • Diagnosis of specific MPS type confirmed by enzyme assay showing deficient activity of the relevant enzyme **OR** detection of pathogenic mutations in the relevant gene by molecular genetic testing, as follows:<br>  ○ For Vimizim: N-acetylgalactosamine 6-sulfatase (GALNS) enzyme or GALNS gene<br>  ○ For Naglazyme: N-acetylgalactosamine 4-sulfatase (ASB) enzyme or Arylsulfatase B (ARSB) gene<br>  ○ For Mepsevii: beta-glucuronidase (GUSB) enzyme or GUSB gene<br>  ○ For Aldurazyme: alpha-L-iduronidase (IDUA) enzyme or IDUA gene<br>  ○ For Elaprase: iduronate 2-sulfatase (I2S or IDS) enzyme or IDS gene<br>• Documented clinical signs and symptoms of MPS, such as soft tissue abnormality, skeletal abnormality, joint abnormality, respiratory disease, gait abnormality, motor issues, or cardiac abnormality<br>• Baseline value for one or more of the following:<br>  ○ Function test such as the Bruininks-Oseretsky Test of Motor Proficiency (BOT-2), 6-minute walk test (6MWT), three-minute stair climb test (3-MSCT), or pulmonary function tests (PFTs)<br>  ○ Liver and/or spleen volume<br>  ○ Urinary glycosaminoglycan (GAGs) level |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose does not exceed the recommended dosing according to the FDA label<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**<u>Reauthorization</u>** requires documentation of treatment success defined as **ONE** or more of the following:<br>• Stability or improvement in function tests such as BOT-2, 6MWT, 3-MSCT, **<u>or</u>** PFTs<br>• Reduction in liver and/or spleen volume<br>• Reduction in urinary GAG level<br>• Other clinically significant improvement in MPS signs and symptoms |

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| **Exclusion Criteria:** | • Treatment of central nervous system manifestation of the disorder<br>• Severe, irreversible cognitive impairment |
| **Age Restriction:** | • Vimizim and Naglazyme: 5 years of age and older<br>• Elaprase: 16 months of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist in the treatment of inherited metabolic disorders, such as a geneticist or endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**MUSCULAR DYSTROPHY RNA THERAPY**
Affected Medications: AMONDYS 45 (casimersen), EXONDYS 51 (eteplirsen), VYONDYS 53 (golodirsen), VILTEPSO (viltolarsen)

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| **Covered Uses:** | • Casimersen (Amondys 45), eteplirsen (Exondys 51), golodirsen (Vyondys 53), and viltolarsen (Viltepso) are not considered medically necessary due to insufficient evidence of therapeutic value. |
| **Required Medical Information:** | |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | |
| **Coverage Duration:** | |

POLICY NAME:
**MYELOID GROWTH FACTORS**
Affected Medications: UDENYCA (pegfilgrastim-cbqv), FULPHILA (pegfilgrastim-jmdb), NEULASTA (pegfilgrastim), ZIEXTENZO (pegfilgrastim-bmez), NYVEPRIA (pegfilgrastim-apgf), NEUPOGEN (filgrastim), ZARXIO (filgrastim-sndz), GRANIX (tbo-filgrastim), LEUKINE (sargramostim), NIVESTYM (filgrastim-aafi), RELEUKO (filgrastim-ayow), FYLNETRA (pegfilrastim-pbbk), ROLVEDON (eflapegrastim-xnst), STIMUFEND (pegfilgrastim-fpgk), NYPOZI (filgrastim-txid), RYZNEUTA (efbemalenograstim alfa)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br><br>**Neupogen, Nivestym, Nypozi, Releuko and Zarxio**<br><br>**Patients with Cancer Receiving Myelosuppressive Chemotherapy**<br>• Indicated to decrease the incidence of infection, as manifested by febrile neutropenia, in patients with non-myeloid malignancies receiving myelosuppressive anti-cancer drugs associated with a significant incidence of severe neutropenia with fever.<br><br>**Patients With Acute Myeloid Leukemia Receiving Induction or Consolidation Chemotherapy**<br>• Indicated for reducing the time to neutrophil recovery and the duration of fever, following induction or consolidation chemotherapy treatment of adults with acute myeloid leukemia.<br><br>**Patients with Cancer Receiving Bone Marrow Transplant**<br>• Indicated to reduce the duration of neutropenia and neutropenia-related clinical sequelae, (e.g., febrile neutropenia) in patients with non-myeloid malignancies undergoing myeloablative chemotherapy followed by marrow transplantation.<br><br>**Patients Undergoing Autologous Peripheral Blood Progenitor Cell Collection and Therapy (Neupogen, Nivestym, Nypozi, Zarxio)**<br>• Indicated for the mobilization of autologous hematopoietic progenitor cells into the peripheral blood for collection by leukapheresis.<br><br>**Patients With Severe Chronic Neutropenia**<br>• Indicated for chronic administration to reduce the incidence and duration of sequelae of neutropenia (e.g., fever, infections, oropharyngeal ulcers) in symptomatic patients with congenital neutropenia, cyclic neutropenia, or idiopathic neutropenia.<br><br>**Patients Acutely Exposed to Myelosuppressive Doses of Radiation (Hematopoietic Syndrome of Acute Radiation Syndrome) (Neupogen)**<br>• Indicated to increase survival in patients acutely exposed to myelosuppressive doses of radiation.<br><br>**Leukine**<br><br>**Use Following Induction Chemotherapy in Acute Myelogenous Leukemia**<br>• Indicated for use following induction chemotherapy in older adult patients with acute myelogenous leukemia to shorten time to neutrophil recovery and to reduce the incidence of severe and life-threatening infections and infections resulting in death. |

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| | **<u>Use in Mobilization and Following Transplantation of Autologous Peripheral Blood Progenitor Cells</u>**<br>• Indicated for the mobilization of hematopoietic progenitor cells into peripheral blood for collection by leukapheresis.<br><br>**<u>Use in Myeloid Reconstitution After Autologous Bone Marrow Transplantation</u>**<br>• Indicated for acceleration of myeloid recovery in patients with non-Hodgkin's lymphoma (NHL), acute lymphoblastic leukemia (ALL) and Hodgkin's disease undergoing autologous bone marrow transplantation (BMT).<br><br>**<u>Use in Myeloid Reconstitution After Allogeneic Bone Marrow Transplantation</u>**<br>• Indicated for acceleration of myeloid recovery in patients undergoing allogeneic BMT from human leukocyte antigen (HLA)-matched related donors.<br><br>**<u>Use in Bone Marrow Transplantation Failure or Engraftment Delay</u>**<br>• Indicated in patients who have undergone allogeneic or autologous BMT in whom engraftment is delayed or has failed.<br><br>**Neulasta, Fulphila, Udenyca, Ziextenzo, Nyvepria, Fylnetra, Stimufend, Ryzneuta, and Rolvedon**<br><br>**<u>Patients with Cancer Receiving Myelosuppressive Chemotherapy</u>**<br>• Indicated to decrease the incidence of infection, as manifested by febrile neutropenia, in patients with non-myeloid malignancies receiving myelosuppressive anti-cancer drugs associated with a significant incidence of severe neutropenia with fever.<br><br>**<u>Patients with Hematopoietic Subsyndrome of Acute Radiation Syndrome (Neulasta, Udenyca, Ziextenzo)</u>**<br>• Indicated to increase survival in patients acutely exposed to myelosuppressive doses of radiation<br><br>**Granix**<br>• Indicated to reduce the duration of severe neutropenia in patients with non-myeloid malignancies receiving myelosuppressive anti-cancer drugs associated with a clinically significant incidence of febrile neutropenia.<br><br>**Compendia supported uses that will be covered (if applicable)**<br>**Neupogen/Granix/Zarxio/Nypozi/Nivestym/Leukine:**<br>• Treatment of chemotherapy-induced febrile neutropenia in patients with non-myeloid malignancies<br>• Treatment of anemia in patients with myelodysplastic syndromes (MDS)<br>• Treatment of neutropenia in patients with MDS<br>• Following chemotherapy for acute lymphocytic leukemia (ALL)<br>• Stem cell transplantation-related indications<br>• Agranulocytosis<br>• Aplastic anemia<br>• Neutropenia related to human immunodeficiency virus (HIV)<br>• Neutropenia related to renal transplantation |
| **Required Medical Information:** | • Complete blood counts with differential and platelet counts will be monitored at baseline and regularly throughout therapy |

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| | • Documentation of therapy intention (curative, palliative) for prophylaxis of febrile neutropenia<br>• Documentation of patient specific risk factors for febrile neutropenia<br>• Documentation of febrile neutropenia risk associated with the chemotherapy regimen<br>• Documentation of planned treatment course<br>• Documentation of current patient weight |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Filgrastim products: Neupogen, Nivestym, Releuko, Zarxio, Granix, Nypozi</u>**<br><br>**When requested via the MEDICAL benefit:**<br>Coverage for the non-preferred products, Neupogen, Nypozi, Releuko and Granix, is provided when the member meets the following criteria:<br>• Documented treatment failure or intolerable adverse event to **Zarxio and Nivestym**<br><br>**When requested through the specialty PHARMACY benefit:**<br>Coverage for the non-preferred products, Neupogen, Nypozi, Zarxio, Releuko and Granix, is provided when the member meets the following criteria:<br>• Documented treatment failure or intolerable adverse event to **Nivestym**<br><br>**<u>Sargramostim product: Leukine</u>**<br><br>Coverage for the non-preferred product, Leukine, is provided when the member meets one of the following criteria:<br>• Leukine will be used for myeloid reconstitution after autologous or allogenic bone marrow transplant or bone marrow transplant engraftment delay or failure<br>• A documented treatment failure or intolerable adverse event to preferred products listed above<br><br>**<u>Pegfilgrastim products: Neulasta, Fulphila, Udenyca, Ziextenzo, Nyvepria, Fylnetra, Stimufend, Rolvedon</u>**<br><br>Coverage for the non-preferred products, Neulasta, Fylnetra, Rolvedon, Stimufend, Ziextenzo and Nyvepria is provided when the member meets the following criteria:<br>• Documented treatment failure or intolerable adverse event to **Fulphila and Udenyca**<br><br>**<u>Eflapegrastim product: Rolvedon and Efbemalenograstim product: Ryzneuta</u>**<br>Coverage for the non-preferred products, Rolvedon and Ryzneuta, is provided when the member meets the following criteria:<br>• Documented treatment failure or intolerable adverse event to the preferred pegfilgrastim products **Fulphila and Udenyca**<br><br>**<u>For prophylaxis of febrile neutropenia (FN) or other dose-limiting neutropenic events for patients receiving myelosuppressive anticancer drugs:</u>**<br>Meets **ONE** of the following:<br>• **Curative Therapy:**<br>  ○ High risk (greater than 20% risk) for febrile neutropenia based on chemotherapy regimen **OR**<br>  ○ Intermediate risk (10-20% risk) for febrile neutropenia based on chemotherapy regimen with documentation of significant patient risk factors for serious medical consequences **OR**<br>  ○ Has experienced a dose-limiting neutropenic event on a previous cycle of current |

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| | chemotherapy to be continued<br>• **Palliative Therapy:**<br>○ Myeloid growth factors will not be approved upfront for prophylaxis of febrile neutropenia in the palliative setting. Per the NCCN (National Comprehensive Cancer Network), chemotherapy regimens with a 20% or greater risk of neutropenic events should not be used. If however, a dose limiting neutropenic event occurs on a previous cycle of chemotherapy, and the effectiveness of chemotherapy will be reduced with dose reduction, growth factor will be approved for secondary prophylaxis on a case by case basis.<br><br>**<u>For Treatment of Severe Chronic Neutropenia</u>**<br>Must meet **<u>ALL</u>** the following:<br>• Congenital neutropenia, cyclic neutropenia, OR idiopathic neutropenia<br>• Current documentation of absolute neutrophil count (ANC) less than 500 cells/microliter<br>• Neutropenia symptoms (fever, infections, oropharyngeal ulcers) |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist or hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 6 months, unless otherwise specified |

POLICY NAME:
**NAFARELIN**
Affected Medications: SYNAREL (nafarelin)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Central Precocious Puberty (CPP)<br>  ○ Endometriosis |
| **Required Medical Information:** | **<u>Central Precocious Puberty:</u>**<br><br>• Documentation of CPP confirmed by basal luteinizing hormone (LH), follicle-stimulating hormone (FSH), and either estradiol or testosterone concentrations<br><br>**<u>Endometriosis:</u>**<br><br>• Documentation of moderate to severe pain due to endometriosis |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Endometriosis:</u>**<br><br>• Documentation of a trial and inadequate relief (or contraindication) after at least 3 months of both of the following first-line therapies:<br>  ○ Nonsteroidal anti-inflammatory drugs (NSAIDs)<br>  ○ Continuous (no placebo pills) hormonal contraceptives<br>• Maximum treatment duration 6 months total |
| **Exclusion Criteria:** | • Use for infertility (if benefit exclusion)<br>• Undiagnosed abnormal vaginal bleeding |
| **Age Restriction:** | • Endometriosis: 18 years of age and older<br>• Central precocious puberty (CPP): 11 years of age or younger (females), 12 years of age or younger (males) |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist or gynecologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Endometriosis: 6 months (no reauthorization), unless otherwise specified<br>• CPP: 12 months, unless otherwise specified |

POLICY NAME:
**NALOXEGOL**
Affected Medications: MOVANTIK (naloxegol)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ Opioid-induced constipation |
| **Required Medical Information:** | • Documentation supporting a diagnosis of opioid-induced constipation in a patient with chronic, non-cancer pain that has been taking opioids for at least 4 weeks |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure or intolerable adverse event to polyethylene glycol 3350 (PEG 3350) and one other laxative (such as lactulose)<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy, AND documented continued use of opioid pain medication |
| **Exclusion Criteria:** | • Known or suspected mechanical gastrointestinal obstruction. |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

**POLICY NAME:**
**NARSOPLIMAB-WUUQ**
**Affected Medications:** Yartemlea

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Diagnosis of hematopoietic stem cell transplant-associated thrombotic microangiopathy (TA-TMA) |
| **Appropriate Treatment Regimen & Other Criteria:** | If requesting twice weekly dosing:<br>• Documentation of inadequate treatment response to once-weekly dosing as assessed by treating provider<br><br>**Reauthorization** requires documentation of treatment response and rationale for additional doses. Treatment response defined as any of the following:<br>• Improvement or stabilization of organ function<br>• Reduction of red blood cell or platelet transfusion requirements<br>• Improvement in TMA markers (examples include platelet count, LDH)<br><br>Treating provider attestation |
| **Exclusion Criteria:** | • |
| **Age Restriction:** | |
| **Prescriber Restrictions:** | • Prescribed by, or in consultation with, a hematologist or transplant specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 8 doses (up to 8 weeks), unless otherwise specified<br>• Reauthorization: 2 doses (2 weeks), unless otherwise specified |

POLICY NAME:
**NATALIZUMAB**
Affected Medications: TYSABRI (natalizumab), TYRUKO (natalizumab-sztn)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:<br>  ▪ Clinically isolated syndrome (CIS)<br>  ▪ Relapsing-remitting multiple sclerosis (RRMS)<br>  ▪ Active secondary progressive multiple sclerosis (SPMS)<br>  ○ Crohn's disease (CD) |
| **Required Medical Information:** | • Screening for anti-JC virus (JCV) antibodies prior to initiating therapy with natalizumab<br><br>**<u>Relapsing Forms of MS</u>**<br>• Diagnosis confirmed with magnetic resonance imaging (MRI) per revised McDonald diagnostic criteria for MS<br>  ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS<br><br>**<u>Crohn's disease</u>**<br>• Moderate to severely active disease despite current treatment |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Relapsing Forms of MS</u>**<br>• Documentation of treatment failure (or documented intolerable adverse event) to:<br>  ○ Rituximab (preferred biosimilar products: Riabni and Ruxience) **OR**<br>  ○ Ocrevus (ocrelizumab) if previously established on treatment **OR**<br>  ○ Documentation of pregnancy and severe disease<br><br>**<u>Crohn's disease</u>**<br>• Documented treatment failure or intolerable adverse event with at least 12 weeks of TWO oral treatments: corticosteroids, azathioprine, 6-mercaptopurine, sulfasalazine, balsalazide, or methotrexate<br>**AND**<br>• Documented clinical failure with at least 12 weeks of infliximab (preferred biosimilar products: Inflectra and Renflexis)<br><br>**<u>Reauthorization:</u>**<br>• **Anti-JCV antibody <u>negative</u>**: documentation of positive clinical response to therapy<br>• **Anti-JCV antibody <u>positive</u>**: documentation of positive clinical response to therapy and periodic MRI to monitor for progressive multifocal leukoencephalopathy (PML) |
| **Exclusion Criteria:** | • Current or prior history of PML<br>• MS: concurrent use of other disease-modifying medications indicated for the treatment of multiple sclerosis<br>• CD: concurrent use of other targeted immune modulators for the treatment of Crohn's disease |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • MS: prescribed by, or in consultation with, a neurologist or a MS specialist<br>• CD: prescribed by, or in consultation with, a gastroenterologist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | **<u>Relapsing Forms of MS:</u>**<br>• Authorization: 12 months, unless otherwise specified<br><br>**<u>Crohn's Disease:</u>**<br>• Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**NEMOLIZUMAB-ILTO**
Affected Medications: NEMLUVIO (nemolizumab-ilto)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Prurigo nodularis (PN)<br>  ○ Atopic dermatitis (AD) |
| **Required Medical Information:** | **PN**<br>• Documentation of all the following:<br>  ○ Diagnosis confirmed by skin biopsy<br>  ○ Presence of at least 20 PN lesions for at least 3 months<br>  ○ Severe itching<br>**AD**<br>• Documentation of severe inflammatory skin disease defined as functional impairment (inability to use hands or feet for activities of daily living or significant facial involvement preventing normal social interaction) AND<br>• Body Surface Area (BSA) of at least 10% OR<br>• Hand, foot or mucous membrane involvement |
| **Appropriate Treatment Regimen & Other Criteria:** | **PN**<br>• Documented treatment failure with at least 2 weeks of a super high potency topical corticosteroid (such as clobetasol propionate 0.05%, halobetasol propionate 0.05%)<br>• Documentation of treatment failure with at least 12 weeks of one of the following: phototherapy, methotrexate, cyclosporine<br>• Documented treatment failure with at least 12 weeks of Dupixent (dupilumab)<br><br>**AD**<br>• Documentation of treatment failure with at least 6 weeks of one of the following: tacrolimus ointment, pimecrolimus cream, Eucrisa<br>• Documentation of treatment failure with at least 12 weeks of one of the following: phototherapy, methotrexate, cyclosporine<br>• Documented treatment failure with at least 12 weeks of Dupixent (dupilumab) |
| **Exclusion Criteria:** | • Concurrent use with another therapeutic immunomodulator agent |
| **Age Restriction:** | • PN: 18 years of age and older<br>• AD: 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a dermatologist, allergist, or immunologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**NEONATAL FC RECEPTOR ANTAGONISTS**
Affected Medications: VYVGART (efgartigimod alfa), VYVGART HYTRULO (efgartigimod alfa and hyaluronidase vial), VYVGART HYTRULO PFS (efgartigimod alfa and hyaluronidase prefilled syringe), RYSTIGGO (rozanolixizumab), IMAAVY (nipocalimab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>**Vyvgart**<br>• Generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive<br>**Rystiggo**<br>• Generalized myasthenia gravis (gMG) in adult patients who are AChR or anti-muscle-specific tyrosine kinase (MuSK) antibody positive<br>**Vyvgart Hytrulo**<br>• Generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive<br>• Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)<br>**Imaavy**<br>• Generalized myasthenia gravis (gMG) in adult and pediatric patients 12 years of age and older who are anti-acetylcholine receptor (AChR) or anti muscle-specific tyrosine kinase (MuSK) antibody positive |
| **Required Medical Information:** | **<u>Myasthenia Gravis</u>**<br>• Diagnosis of generalized myasthenia gravis (gMG) confirmed by one of the following:<br>  ○ A history of abnormal neuromuscular transmission test<br>  ○ A positive edrophonium chloride test<br>  ○ Improvement in gMG signs or symptoms with an acetylcholinesterase inhibitor<br>• Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV<br>• Documentation of **ONE** of the following:<br>  ○ MG-Activities of Daily Living (MG-ADL) total score of 6 or greater<br>  ○ Quantitative Myasthenia Gravis (QMG) total score of 12 or greater<br>• For Rystiggo and Imaavy: Positive serologic test for AChR or MuSK antibodies<br><br>**<u>CIDP (Vyvgart Hytrulo only)</u>**<br>• Documented baseline in strength/weakness using an objective clinical measuring tool (INCAT, Medical Research Council (MRC) muscle strength, 6 Minute Walk Test, Rankin, Modified Rankin)<br>• Documented disease course is progressive or relapsing and remitting for 2 months or longer<br>• Abnormal or absent deep tendon reflexes in upper or lower limbs<br>• Electrodiagnostic evidence of demyelination indicated by one of the following:<br>  ○ Motor distal latency prolongation in 2 nerves<br>  ○ Reduction of motor conduction velocity in 2 nerves<br>  ○ Prolongation of F-wave latency in 2 nerves<br>  ○ Absence of F-waves in at least 1 nerve<br>  ○ Partial motor conduction block of at least 1 motor nerve<br>  ○ Abnormal temporal dispersion in at least 2 nerves<br>  ○ Distal CMAP duration increase in at least 1 nerve |

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| | • Cerebrospinal fluid (CSF) analysis indicates all of the following (if electrophysiologic findings are non-diagnostic):<br>  ○ CSF white cell count of less than 10 cells/$mm^3$<br>  ○ CSF protein is elevated (greater than or equal to 45mg/dL) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Myasthenia Gravis</u>**<br>• Currently on a stable dose of at least one gMG therapy (acetylcholinesterase inhibitor, corticosteroid, or non-steroidal immunosuppressive therapy (NSIST)) that will be continued during initial treatment with Vyvgart, Vyvgart Hytrulo, Imaavy or Rystiggo<br>• Documentation of **ONE** of the following:<br>  ○ Treatment failure with an adequate trial (one year or more) of at least 2 immunosuppressive therapies (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)<br>  ○ Need for ongoing rescue therapy (at least 3 courses in the past 12 months) with plasmapheresis, plasma exchange, or intravenous immunoglobulin (IVIG) while consistently taking immunosuppressive therapy (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)<br><br>Coverage for **Rystiggo or Imaavy** is provided when one of the following is met:<br>• Currently receiving treatment with Rystiggo or Imaavy, excluding when the product is obtained as samples or via manufacturer's patient assistance programs<br>• Documented treatment failure or intolerable adverse event with Vyvgart for AChR antibody positive gMG, if age appropriate<br>• Documented treatment failure with rituximab for MuSK antibody positive gMG (preferred products: Riabni, Ruxience), if age appropriate<br><br>Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**<u>Reauthorization</u>:**<br>• Documentation of treatment success and a clinically significant response to therapy defined as:<br>  ○ A minimum 2-point reduction in MG-ADL score from baseline or improvement in QMG total score<br>  ○ Absent or reduced need for rescue therapy compared to baseline<br>• Documentation that the patient requires continuous treatment, after an initial beneficial response, due to new or worsening disease activity<br><br>**Note**: a minimum of 50 days for Vyvgart/Vyvgart Hytrulo or 63 days for Rystiggo must have elapsed from the start of the previous treatment cycle<br><br>**<u>CIDP (Vyvgart Hytrulo only)</u>**<br>• Documented trial and failure of at least 3 months of intravenous or subcutaneous immune globulin<br><br>**<u>Reauthorization</u>:**<br>• Documentation of a clinical response to therapy based on an objective clinical measuring tool (e.g., INCAT, Medical Research Council (MRC) muscle strength, 6-Minute walk test, Rankin, Modified Rankin) |
| **Exclusion Criteria:** | • Immunoglobulin G (IgG) levels less than 600 mg/dL at baseline<br>• Concurrent use with other disease-modifying biologics for the treatment of gMG |

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| **Age Restriction:** | • Vyvgart, Vyvgart Hytrulo, and Rystiggo: 18 years of age and older<br>• Imaavy: 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**NIEMANN-PICK DISEASE TYPE C (NPC) AGENTS**
Affected Medications: MIPLYFFA (arimoclomol citrate), AQNEURSA (levacetylleucine)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Niemann-Pick disease type C (NPC) |
| **Required Medical Information:** | • Diagnosis of NPC confirmed by genetic testing showing biallelic pathogenic variants in either the NPC1 gene or NPC2 gene<br>• Documentation of at least one neurological symptom of Niemann-Pick disease type C, such as:<br>  ○ Loss of motor function<br>  ○ Problems with swallowing or speech<br>  ○ Cognitive impairment<br>• Documentation of being ambulatory without needing an assistive device such as a wheelchair, walker, or cane<br>• Documentation of baseline signs and symptoms of NPC |
| **Appropriate Treatment Regimen & Other Criteria:** | **For Miplyffa:**<br>• Documentation that patient has been receiving miglustat with a stable dose for at least the past 6 consecutive months<br>• Documentation that Miplyffa will be taken in combination with miglustat<br><br>**Reauthorization** requires:<br>• Documentation of treatment success defined as stability or improvement of Niemann-Pick disease type C signs and symptoms<br>• Documentation that patient is still ambulatory<br>• For Miplyffa: that the drug continues to be used in combination with miglustat |
| **Exclusion Criteria:** | • Use of Miplyffa and Aqneursa in combination |
| **Age Restriction:** | • Miplyffa: 2 years of age and older<br>• Aqneursa: Adults and pediatric patients weighing 15 kilograms or greater |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist in the management of NPC (such as a geneticist, endocrinologist, metabolic disorder subspecialist, or neurologist)<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**NIROGACESTAT**
Affected Medications: OGSIVEO (nirogacestat)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Progressive desmoid tumor(s) requiring systemic therapy<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• Diagnosis of biopsy proven desmoid tumor/aggressive fibromatosis (DT/AF) with documentation of tumor progression (tumor growth causing chronic pain, disfigurement, internal bleeding, and/or impaired range of motion) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of clinical failure with sorafenib<br><br>**Reauthorization:** documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**NON-Preferred HYALURONIC ACID DERIVATIVES**
Affected Medications: DUROLANE (hyaluronic acid), EUFLEXXA (1% sodium hyaluronate), GEL-ONE (cross-linked hyaluronate), GELSYN-3 (sodium hyaluronate 0.84%), GENVISC 850 (sodium hyaluronate), HYALGAN (sodium hyaluronate), HYMOVIS (high molecular weight viscoelastic hyaluronan), MONOVISC (high molecular weight hyaluronan), SUPARTZ (sodium hyaluronate), SYNOJOYNT (sodium hyaluronate), TRILURON (sodium hyaluronate), TRIVISC (sodium hyaluronate), VISCO-3 (sodium hyaluronate)

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| 1. Is this the first time a Hyaluronic Acid (HA) derivative product is being used in this member for this indication? | Yes – Go to #2 | No – Document date of last use and go to Renewal criteria |
| 2. Is the request for a Food and Drug Administration (FDA)-approved indication: Treatment of osteoarthritis pain of the knee? | Yes – Go to #3 | No – Criteria not met |
| 3. Is there documented failure to respond to conservative non-pharmacologic therapy (such as ice, physical therapy) and simple analgesics (such as acetaminophen)? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Has there been a documented intolerable adverse event to Synvisc, Synvisc-One, and Orthovisc with date and description of reactions? | Yes – Go to #6 | No – Go to #5 |
| 5. Is the member currently undergoing treatment and coverage is required to complete the current course of treatment? | Yes – Document and go to #6 | No – Criteria not met |
| 6. Is the requested dose within the Food and Drug Administration (FDA)-approved label and PacificSource quantity limitations? | Yes – Document and approve up to 6 months | No – Criteria not met |
| **Renewal for hyaluronic acid (HA) after previous administration of HA product** | | |
| 1. Is there documentation of treatment success that lasted at least 6 months from date of previous HA administration AND documented intolerable adverse event to Synvisc, Synvisc-One, and Orthovisc with date and description of reactions? | Yes – Go to #2 | No – Criteria not met |
| 2. Is the requested dose within the Food and Drug Administration (FDA)-approved label and PacificSource quantity limitations? | Yes – Approve up to 6 months | No – Criteria not met |
| **Quantity Limitations** | | |

- Durolane: 1 injection per course
- Euflexxa: 3 injections per course
- Gel-One: 1 injection per course
- Gelsyn-3: 3 injections per course
- GenVisc 850: 3 to 5 injections per course
- Hyalgan: 5 injections per course
- Hymovis: 2 injections per course
- Monovisc: 1 injection per course
- Supartz: 3 to 5 injections per course
- Synojoynt: 3 injections per course
- Triluron: 3 injections per course
- Trivisc: 3 injections per course
- Visco-3: 3 injections per course

POLICY NAME:
**NON-PREFERRED MEDICAL DRUG CODES**
Affected Medications: BORTEZOMIB, PEMETREXED, LEUCOVORIN CALCIUM

<table>
<tr><td>Covered Uses:</td><td>● All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>● For oncology indications: National Comprehensive Cancer Network (NCCN) indications with evidence level of 2A or higher</td></tr>
<tr><td>Required Medical Information:</td><td></td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td>● Approval of a non-preferred medical drug listed below requires documentation of an intolerable adverse event to all the preferred alternatives, and the adverse event was not an expected adverse event attributed to the active ingredient<br><br>
<table>
<tr><th>Drug</th><th>Non-Preferred code</th><th>Preferred Alternatives</th></tr>
<tr><td>Bortezomib (Boruzu, Velcade)</td><td>J9046, J9054</td><td>J9041, J9048, J9049</td></tr>
<tr><td>Pemetrexed (Pemfexy, Alimta, Pemrydi RTU, Axtle)</td><td>J9304, J9292</td><td>J9294, J9296, J9297, J9305, J9314, J9324</td></tr>
<tr><td>Leucovorin calcium (Vykoura)</td><td>J9999, J3490</td><td>J0640, J0641, J0642</td></tr>
</table>
<br><u>Reauthorization</u> requires documentation of disease responsiveness to therapy</td></tr>
<tr><td>Exclusion Criteria:</td><td></td></tr>
<tr><td>Age Restriction:</td><td></td></tr>
<tr><td>Prescriber/Site of Care Restrictions:</td><td>● All approvals are subject to utilization of the most cost-effective site of care</td></tr>
<tr><td>Coverage Duration:</td><td>● Authorization: 12 months, unless otherwise specified</td></tr>
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POLICY NAME:
**NUEDEXTA**
Affected Medications: NUEDEXTA (dextromethorphan hydrobromide/quinidine sulfate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ Treatment of pseudobulbar affect (PBA) |
| **Required Medical Information:** | • Documentation of at least ONE underlying neurological condition associated with PBA such as:<br>  ○ amyotrophic lateral sclerosis (ALS)<br>  ○ extrapyramidal and cerebellar disorders (Parkinson's disease, multiple system atrophy, progressive supranuclear palsy)<br>  ○ multiple sclerosis (MS)<br>  ○ traumatic brain injury<br>  ○ Alzheimer's disease and other dementias<br>  ○ stroke.<br>• Baseline Center for Neurologic Study-Lability Scale (CNS-LS) score of 13 or greater<br>• Documentation of treatment failure to a 30-day trial of each of the following:<br>  ○ serotonin reuptake inhibitor (SSRI)<br>  ○ tricyclic antidepressant (TCA) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Reauthorization requires documentation of treatment success defined as decreased frequency of pseudobulbar affect (PBA) episodes. |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**NULIBRY**
Affected Medications: NULIBRY (fosdenopterin)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ To reduce the risk of mortality in patients with molybdenum cofactor deficiency (MoCD) Type A |
| **Required Medical Information:** | • Documentation of presumptive or genetically confirmed molybdenum cofactor deficiency (MoCD) Type A diagnosis<br><br>**<u>Presumptive diagnosis of Molybdenum cofactor deficiency (MoCD) Type A</u>**<br>• Documentation of family history meeting **ONE** of the following:<br>  ○ Affected sibling(s) with confirmed MoCD Type A; or a history of deceased sibling(s) with classic MoCD presentation<br>  ○ One or both parents are known to carry a copy of the mutated gene [Molybdenum Cofactor Synthesis 1 (MOCS1)]<br>  ○ Child has consanguineous parents with a family history of MoCD<br>• Onset of clinical and/or laboratory signs and symptoms consistent with MoCD Type A, such as:<br>  ○ Clinical presentation: intractable seizures, exaggerated startle response, high-pitched cry, axial hypotonia, limb hypertonia, feeding difficulties<br>  ○ Biochemical findings: elevated urinary sulfite and/or S-sulfocysteine (SSC), elevated xanthine in urine or blood, or low/absent uric acid in the urine or blood<br>• Genetic testing to confirm diagnosis of MoCD Type A is scheduled or in progress<br><br>**<u>Confirmed diagnosis of MoCD Type A</u>**:<br>• Diagnosis of MoCD Type A confirmed by genetic testing showing the presence of mutation in molybdenum cofactor synthesis gene 1 (MOSC1) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Reauthorization</u>**:<br>• Documentation of clinically significant response to therapy as determined by prescribing provider<br>• Documentation of genetically confirmed MoCD Type A (MOCS1 mutation) if initially approved for presumptive diagnosis |
| **Exclusion Criteria:** | • Molybdenum cofactor deficiency (MoCD) Type B (MOCS2 mutation)<br>• MoCD Type C (gephyrin or GPHN mutation) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neonatologist, pediatrician, pediatric neurologist, neonatal neurologist, or geneticist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **<u>Presumptive diagnosis:</u>**<br>• Authorization: 1 month, unless otherwise specified. Must have confirmed diagnosis for continued approval. |

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| | **Confirmed diagnosis:**<br>• Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**NUPLAZID**
Affected Medications: NUPLAZID (pimavanserin tartrate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of hallucinations and delusions associated with Parkinson's disease (PD) psychosis |
| **Required Medical Information:** | • Diagnosis of Parkinson's disease (PD)<br>• Presence of psychotic symptoms: hallucinations and/or delusions described as severe and frequent that started after the PD diagnosis |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of treatment failure or contraindication to a 30-day trial of quetiapine<br><br>**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**NUSINERSEN**
Affected Medications: SPINRAZA (nusinersen)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Spinal muscular atrophy (SMA) |
| **Required Medical Information:** | • Diagnosis of SMA type 1, 2, or 3 confirmed by genetic testing of chromosome 5q13.2 demonstrating ONE of the following:<br>  ○ Homozygous gene deletion of SMN1 (survival motor neuron 1)<br>  ○ Homozygous gene mutation of SMN1<br>  ○ Compound heterozygous gene mutation of SMN1<br>• Documentation of 2 or more copies of the SMN2 (survival motor neuron 2) gene<br>• Documentation of previous treatment history<br>• Documentation of one of the following baseline motor assessments appropriate for patient age and motor function:<br>  ○ Hammersmith Infant Neurological Examination (HINE-2)<br>  ○ Hammersmith Functional Motor Scale (HFSME)<br>  ○ Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)<br>  ○ Upper Limb Module (ULM) test<br>  ○ 6-Minute Walk Test (6MWT)<br>• Documentation of ventilator use status<br>  ○ Patient is NOT ventilator-dependent (defined as using a ventilator at least 16 hours per day on at least 21 of the last 30 days)<br>  ○ This does not apply to patients who require non-invasive ventilator assistance<br>• Planned treatment regimen |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with or intolerable adverse event on Evrysdi<br><br>**Reauthorization** requires documentation of improvement in baseline motor assessment score, clinically meaningful stabilization, or delayed progression of SMA-associated signs and symptoms |
| **Exclusion Criteria:** | • SMA type 4<br>• Advanced SMA at baseline (complete paralysis of limbs, permanent ventilation support)<br>• Prior treatment with SMA gene therapy (i.e., onasemnogene abeparvovec-xioi, onasemnogene abeparvovec-brve)<br>• Will not be used in combination with other agents for SMA (e.g., onasemnogene abeparvovec-xioi, onasemnogene abeparvovec-brve, risdiplam, etc.) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or provider who is experienced in treatment of spinal muscular atrophy<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 8 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**OBINUTUZUMAB**
Affected Medications: GAZYVA (obinutuzumab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Lupus Nephritis (LN)<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | **Lupus Nephritis**:<br>• Documentation of biopsy-proven active Class III, IV, and/or V disease<br>• Baseline measurement of one or more of the following: urine protein-creatinine ratio (uPCR), urine protein, estimated glomerular filtration rate (eGFR), or frequency of flares or corticosteroid use<br><br>**Oncology Uses**<br>• Documentation of disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | **Lupus Nephritis:**<br>• No dialysis in the past 12 months AND estimated glomerular filtration rate (eGFR) equal to or above 30 mL/min/1.73m$^2$<br>• UPCR ≥ 1 gram of protein per gram of creatinine **(**1 g/g)<br>• Failure of at least 12 weeks of mycophenolate mofetil AND cyclophosphamide<br><br>**Reauthorization** requires documentation of treatment success defined as ONE of the following:<br>• Improvement in eGFR<br>• Reduction in urinary protein-creatinine ratio or urine protein<br>• Decrease in flares or corticosteroid use<br><br>**Oncology Uses**<br>• Documentation of ECOG performance status of 1 or 2 OR Karnofsky performance score greater than 50%<br>• A Food and Drug Administration (FDA)-approved, or compendia supported dose, frequency, and duration of therapy<br>• Documented treatment failure of first line recommended and conventional therapies<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Use in combination with other biologic therapies for LN |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • **Lupus Nephritis**: Prescribed by or in consultation with nephrologist or rheumatologist<br>• **Oncologic Indications**: Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Initial Authorization:<br>  ○ Oncology – 4 months, unless otherwise specified<br>  ○ Lupus Nephritis - 12 months unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**OCRELIZUMAB**
Affected Medications: OCREVUS (ocrelizumab), OCREVUS ZUNOVO (ocrelizumab hyaluronidase)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ Primary progressive multiple sclerosis (PPMS)<br>  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:<br>  ▪ Clinically isolated syndrome (CIS)<br>  ▪ Relapsing-remitting multiple sclerosis (RRMS)<br>  ▪ Active secondary progressive disease (SPMS) |
| **Required Medical Information:** | **<u>All Indications:</u>**<br>• Diagnosis confirmed with magnetic resonance imaging (MRI) per revised McDonald diagnostic criteria for MS<br>  ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS<br><br>**<u>Primary Progressive MS:</u>**<br>• Documentation of at least one year of disease progression and baseline Expanded Disability Status Scale (EDSS) of 3.0 to 6.5 |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Relapsing forms of MS:</u>**<br>• Coverage of Ocrevus (ocrelizumab) or Ocrevus Zunovo (ocrelizumab hyaluronidase) requires documentation of one of the following:<br>  ○ Documented disease progression or intolerable adverse event with rituximab (biosimilar products, Riabni and Ruxience, preferred)<br>  ○ Currently receiving treatment with Ocrevus (ocrelizumab) or Ocrevus Zunovo (ocrelizumab hyaluronidase), excluding via samples or manufacturer's patient assistance program<br><br>**<u>Reauthorization</u>** requires documentation of treatment success |
| **Exclusion Criteria:** | • Active hepatitis B infection<br>• Concurrent use of other disease-modifying medications indicated for the treatment of MS |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or MS specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**OFEV**
Affected Medications: OFEV (nintedanib esylate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Idiopathic pulmonary fibrosis (IPF)<br>  ○ Chronic fibrosing interstitial lung disease (ILD) with a progressive phenotype<br>  ○ Systemic sclerosis-associated interstitial lung disease (SSc-ILD) |
| **Required Medical Information:** | **<u>Idiopathic Pulmonary Fibrosis (IPF)</u>**<br>• Documented diagnosis of idiopathic pulmonary fibrosis (IPF) confirmed by **ONE** of the following:<br>  ○ Usual interstitial pneumonia (UIP) pattern demonstrated on high-resolution computed tomography (HRCT)<br>  ○ UIP pattern demonstrated on surgical lung biopsy<br>  ○ Probable UIP pattern demonstrated on **BOTH** HRCT and surgical lung biopsy<br>• Documentation confirming known causes of interstitial lung disease have been ruled out (e.g., rheumatic disease, environmental exposure, drug toxicity)<br>• Documentation of **BOTH** of the following:<br>  ○ Baseline forced vital capacity (FVC) greater than or equal to 50 percent predicted<br>  ○ Baseline diffusing capacity for carbon monoxide (DLCO) greater than or equal to 30 percent predicted<br><br>**<u>Systemic Sclerosis-Associated Interstitial Lung Disease (SSc-ILD)</u>**<br>• Documented diagnosis of SSc-ILD<br>• Documentation of greater than or equal to 10% fibrosis on a chest high resolution computed tomography (HRCT) scan conducted within the previous 12 months<br>• Documentation of baseline FVC greater than or equal to 40% of predicted<br>• Documentation of predicted DLCO 30-89% of predicted<br><br>**<u>Chronic Fibrosing Interstitial Lung Disease (ILD) with a Progressive Phenotype</u>**<br>• Documented diagnosis of chronic fibrosing ILD with a progressive phenotype (aka progressive pulmonary fibrosis), confirmed by at least two of the following:<br>  ○ Worsening respiratory symptoms<br>  ○ Physiological evidence of disease progression (defined as DLCO reduced by 10% or greater **OR** FVC reduced by 5% or greater)<br>  ○ Radiological evidence of disease progression (eg, increased traction bronchiectasis, new ground-glass opacity or fine reticulation, new/increased honeycombing)<br>• Documentation of relevant fibrosis (greater than 10% fibrotic features) on chest HRCT scan<br>• Baseline FVC greater than or equal to 45% of predicted<br>• Baseline DLCO 30% to less than 80% of predicted |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>IPF:</u>**<br>• Documented treatment failure, contraindication, or intolerance to pirfenidone<br><br>**<u>SSc-ILD:</u>**<br>• Documented treatment failure with mycophenolate (MMF) |

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| | **<u>Reauthorization</u>** requires documentation of treatment success |
| **Exclusion Criteria:** | • Documentation of airway obstruction (such as pre-bronchodilator FEV/FVC less than 0.7)<br>• Combined use with pirfenidone (Esbriet) |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a pulmonologist or rheumatologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**OLEZARSEN**
Affected Medications: TRYNGOLZA (olezarsen sodium)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Reduce triglycerides as an adjunct to diet in adults with familial chylomicronemia syndrome (FCS) |
| **Required Medical Information:** | • Diagnosis of FCS (type 1 hyperlipoproteinemia) confirmed by genetic testing showing a pathogenic gene mutation in LPL, APOC2, APOA5, GPIHBP1 or LMF1 genes<br>• Fasting triglyceride level of at least 880 mg/dL<br>• Will be used as an adjunct to diet |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of following a low-fat diet with less than 20 grams of fat per day<br><br>**Reauthorization** requires documentation of treatment success defined as a decrease in triglycerides since starting therapy |
| **Exclusion Criteria:** | • History of acute coronary syndrome |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**OLIPUDASE ALFA**
Affected Medications: XENPOZYME (olipudase alfa-rpcp)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of non-central nervous system manifestations of acid sphingomyelinase deficiency (ASMD) in adult and pediatric patients |
| **Required Medical Information:** | • Documentation of acid sphingomyelinase deficiency as evidenced by one of the following:<br>  ○ Enzyme assay showing diminished (less than 10% of controls) or absent acid sphingomyelinase (ASM) activity<br>  ○ Gene sequencing showing biallelic pathogenic sphingomyelin phosphodiesterase-1 (SMPD1) mutation<br>• Documentation of clinical presentation outside the central nervous system (e.g., hepatosplenomegaly, interstitial lung disease, liver fibrosis, growth restriction of childhood)<br>• Documentation of current body mass index (BMI), weight, and height<br>• For adults 18 years of age and older, documentation of both of the following:<br>  ○ Diffusion capacity of lungs (DLCO) is less than or equal to 70% of the predicted normal value<br>  ○ Spleen volume greater than or equal to 6 multiples of normal (MN) measured by magnetic resonance imaging (MRI)<br>• For pediatrics 18 years of age and younger, documentation of both of the following:<br>  ○ Spleen volume greater than or equal to 5 MN measured by MRI<br>  ○ Height Z-score -1 or lower |
| **Appropriate Treatment Regimen & Other Criteria:** | **Dosing:** Dosed every two weeks based on FDA label<br>• Body mass index (BMI) less than or equal 30, the dosage is based on actual body weight (kg)<br>• BMI of greater than 30 is dosed based on adjusted body weight<br>• Adjusted body weight = (height in $m^2$) x 30<br><br>• Availability: 20 mg single-dose vials<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization** requires documentation of improvement in patient specific disease presentation such as:<br>• Improvement in PFT or DLCO<br>• Improvement in spleen and/or liver volume or function<br>• Improvement/stability in platelet counts<br>• Improvement in linear growth progression (pediatric) |
| **Exclusion Criteria:** | • Exclusive central nervous system manifestations |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a metabolic specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**OMAVELOXOLONE**
Affected Medications: Skyclarys (omaveloxolone)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of Friedreich's ataxia in adults and adolescents aged 16 years and older |
| **Required Medical Information:** | • Genetically confirmed diagnosis of Friedreich's Ataxia<br>• Documentation of baseline modified Friedreich's Ataxia Rating Scale (mFARS) score under 81<br>• Documentation that the patient is still ambulatory or retains enough activity to assist in activities of daily living |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** will require documentation of treatment success, such as a reduction in the rate of decline, as determined by prescriber |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 16 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**OMIDUBICEL**
Affected Medications: OMISIRGE (Omidubicel)

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better<br>• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• Documented diagnosis of a hematologic malignancy<br>• Clinically stable and eligible for umbilical cord blood transplantation (UCBT) following myeloablative conditioning<br>OR<br>• Diagnosis of severe aplastic anemia (SAA) documented by one of the following:<br>  ○ Bone marrow cellularity less than 30% with associated red blood cell (RBC) or platelet transfusion dependence<br>  ○ An absolute neutrophil count (ANC) of 1000 cells/μL or less<br>• Clinically stable and eligible for umbilical cord blood transplantation (UCBT) following reduced intensity conditioning |
| **Appropriate Treatment Regimen & Other Criteria:** | • Must **NOT** have a matched related donor (MRD), matched unrelated donor (MUD), mismatched unrelated donor (MMUD), or haploidentical donor readily available.<br>• Documentation that **NONE** of the following are present:<br>  ○ Other active malignancy<br>  ○ Active or uncontrolled infection<br>  ○ Active central nervous system (CNS) disease<br><br>**Severe Aplastic Anemia only**<br>• Trial and failure, contraindication, or intolerance to immunosuppressive therapy (Examples of immunosuppressant therapies include cyclosporine, Atgam, mycophenolate, or sirolimus)<br><br>**Reauthorization**: None - Omisirge will be used as a one-time treatment |
| **Exclusion Criteria:** | • Karnofsky Performance Status (KPS) of 50% or less or Eastern Cooperative Oncology Group (ECOG) score of 3 or greater<br>• HLA (human leukocyte antigen)-matched donor able to donate<br>• Prior allo-HSCT (hematopoietic stem cell transplantation)<br>• Pregnancy or lactation |
| **Age Restriction:** | • 12 years of age or older (hematologic malignancy)<br>• 6 years of age or older (SAA) |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist or hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 2 months for 1 time administration, unless otherwise specified |

# ONASEMNOGENE ABEPARVOVEC

**Included Products:** ZOLGENSMA (onasemnogene abeparvovec xioi), ITVISMA (onasemnogene abeparvovec-brve)

## Scope & Exclusions

### Included Indications:
All Food and Drug Administration (FDA)–approved indications not otherwise excluded by plan design.

### Exclusions:
Prior gene therapy use

### Prescriber Limits:
Prescribed by, or in consultation with, a pediatric neurologist or provider who is experienced in treatment of spinal muscular atrophy

## Authorization Criteria

### Required Medical Information:

1. All indications must be FDA-supported for the requested product.
2. Requested dosing must be according to the FDA label based on diagnosis, age, and weight.
3. Documentation of anti-adeno-associated virus (AAV) serotype 9 antibody titer less than or equal 1:50
4. Patient is NOT ventilator-dependent (defined as using a ventilator at least 16 hours per day on at least 21 of the last 30 days). This does not apply to patients who require non-invasive ventilator assistance.

### Zolgensma

1. Age less than 2 years
2. Diagnosis of SMA type 1 confirmed by genetic testing of chromosome 5q13.2 demonstrating ONE of the following:
    a. Homozygous gene deletion of SMN1 (survival motor neuron 1)
    b. Homozygous gene mutation of SMN1
    c. Compound heterozygous gene mutation of SMN1
3. Documentation of 2 or fewer copies of the SMN2 (survival motor neuron 2) gene

### Itvisma

1. Age 2 to less than 18 years of age
2. Functional status: patients are able to sit but have never been able to walk independently
3. If current or previous treatment with Spinraza or Evrysdi, documentation of intolerance or functional decline while on treatment and must stop treatment for 15 days (Evrysdi) or 4 months (Spinraza) prior to Itvisma use.

## Duration of Approval

Approved for one dose only per lifetime, unless otherwise specified

# ONCOLOGY AGENTS

Included Products: ABECMA, ABRAXANE, AUCATZYL, ADCETRIS, ADSTILADRIN, AKEEGA, ALECENSA, ALIQOPA, ALKERAN, ALUNBRIG 180MG ORAL TABLET, ANKTIVA, ARZERRA, ASPARLAS, AUGTYRO, AVMAPKI FAKZYNJA CO-PACK, AYVAKIT, AZEDRA, BALVERSA, BAVENCIO, BEIZRAY, BELEODAQ, BELRAPZO, BENDAMUSTINE, BENDEKA, BESPONSA, BIZENGRI, BLENREP, BLINCYTO, BRAFTOVI, BREYANZI, BRUKINSA, CABOMETYX, CALQUENCE, CAPRELSA, CARVYKTI, CLOFARABINE, CLOLAR, COLUMVI, COMETRIQ, COPIKTRA, COSELA, COTELLIC, CYRAMZA, DACOGEN, DANYELZA, DARZALEX, DARZALEX FASPRO, DATROWAY, DAURISMO, DOXIL, DOXORUBICIN LIPOSOMAL, ELAHERE, ELREXFIO, EMPLICITI, EMRELIS, ENHERTU, ENSACOVE, EPKINLY, ERBITUX, ERIVEDGE, ERLEADA, ERLOTINIB, ERWINAZE, EVOMELA, FOTIVDA, FRUZAQLA, GAZYVA, GAVRETO, GEFITINIB, GILOTRIF, HEPZATO, HERCEPTIN, HERCEPTIN HYLECTA, HERCESSI, HERNEXEOS, HERZUMA, HYCAMTIN, IBRANCE, IBRUTINIB, IBTROZI, ICLUSIG, IDHIFA, IMBRUVICA, IMDELLTRA, IMFINZI, IMJUDO, IMLYGIC IRESSA, INLEXZO, INLURIYO, INLYTA, INQOVI, INREBIC, IOBENGUANE I-131, ISTODAX, ITOVEBI, IWILFIN, IXEMPRA, JAKAFI, JAYPIRCA, JELMYTO, JEMPERLI, JEVTANA, KADCYLA, KANJINTI, KEYTRUDA, KEYTRUDA QLEX, KIMMTRAK, KRAZATI, KYMRIAH, KYPROLIS, LAPATINIB, LARTRUVO, LENALIDOMIDE, LENVIMA, LIBTAYO, LONSURF, LOQTORZI, LORBRENA, LUMAKRAS, LUMOXITI, LUNSUMIO, LUNSUMIO VELO, LUTATHERA, LYNOZYFIC, LYNPARZA, LYTGOBI, MARGENZA, MARQIBO, MATULANE, MEKINIST, MEKTOVI, MELPHALAN, MODEYSO, MONJUVI, MYLOTARG, NAB-PACLITAXEL, NEXAVAR, NERLYNX, NILANDRON, NINLARO, NIVOLUMAB, NUBEQA, ODOMZO, OJEMDA, OJJAARA, ONCASPAR, ONIVYDE, ONTRUZANT, ONUREG, OPDIVO, OPDIVO QVANTIG, OPDUALAG, ORSERDU, PADCEV, PAZOPANIB, PEMAZYRE, PEPAXTO, PERJETA, PHESGO, PHOTOFRIN, PIQRAY, PLUVICTO, POLIVY, POMALYST, POTELIGEO, PROLEUKIN, PROVENGE, QINLOCK, RETEVMO, REVLIMID, REVUFORJ, REZLIDHIA, REZUROCK, ROMIDEPSIN, ROMVIMZA, ROZLYTREK, RUBRACA, RYBREVANT, RYDAPT, RYLAZE, RYTELO, SARCLISA, SORAFENIB, STIVARGA, SUNITINIB, SUTENT, SYNRIBO, TABRECTA, TAFINLAR, TAGRISSO, TALVEY, TALZENNA, TARCEVA, TECARTUS, TECELRA, TECENTRIQ, TECENTRIQ HYBREZA, TECVAYLI, TEMODAR, TEMOZOLOMIDE, TEPADINA, TEPMETKO, TEVIMBRA, TIBSOVO, TIVDAK, TORISEL, TRASTUZUMAB, TRAZIMERA, TREANDA, TRODELVY, TRUQAP, TURALIO, TYKERB, UNITUXIN, UNLOXCYT, VANFLYTA, VECTIBIX, VENCLEXTA, VERZENIO, VIDAZA, VIVIMUSTA, VIZIMPRO, VONJO, VORANIGO, VOTRIENT, VYLOY, VYXEOS, XALKORI, XOFIGO, XOSPATA, XPOVIO, XTANDI, YERVOY, YESCARTA, YONDELIS, ZALTRAP, ZEGFROVY, ZEJULA TABLETS, ZELBORAF, ZEPZELCA, ZOLINZA, ZYDELIG, ZYKADIA, ZYNLONTA, ZYNYZ

## SCOPE & EXCLUSIONS

Included Indications: NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher

Exclusions: Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater if noted by NCCN or FDA label

Age Limits: As defined by FDA labeling for the requested indication.

Prescriber Limits: Prescribed by, or in consultation with, an oncologist

Other Limits: All approvals are subject to utilization of the most cost-effective site of care

# AUTHORIZATION CRITERIA

## Required Medical Information:

1. Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course
2. Requested dosing must be according to the FDA label based on diagnosis, age, and weight.
3. Requests for non-preferred products require inadequate response, intolerance, or an FDA-labeled and patient-specific contraindication to all preferred products as outlined within indication-specific criteria. FDA-labeled contraindications are defined as those listed within the package insert. Allergic reactions may be accepted as contraindicated when an ingredient found only in the preferred product is established as the causative agent.

**Preferred Drugs:** Keytruda (pembrolizumab), Opdivo (nivolumab), Tecentriq (atezolizumab), Kanjinti and Ogivri (trastuzumab), Perjeta (pertuzumab), Lunsumio (mosunetuzumab)

**Nonpreferred Drugs:** Keytruda Qlex, Opdivo Qvantig, Tecentriq Hybreza, Trazimera, Herzuma, Ontruzant, Herceptin, Hercessi, Herceptin Hylecta, Phesgo, Lunsumio Velo

# RENEWAL CRITERIA

Reauthorization will be approved based on disease response to therapy and FDA approved and NCCN guidelines allowed length of therapy

# DURATION OF APPROVAL

## Initial:

- 4 months for therapy to be continued until disease progression
- Neoadjuvant or fixed dose regimens to be approved for entire treatment cycle up to 12 months

## Renewal: 12 months, unless otherwise specified

POLICY NAME:
**OPIOID QUANTITY ABOVE 90 MORPHINE MILLIGRAM EQUIVALENTS (MME)**
Affected Medications: ALL OPIOIDS

<table>
<tr><td>Covered Uses:</td><td>• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design</td></tr>
<tr><td>Required Medical Information:</td><td>• Short term use of opioids with an MME per day greater than 90 MME requires one of the following:<br>○ Recent surgery<br>○ Acute injury<br><br>• Chronic use of opioids with a Morphine Milligram Equivalents (MME) per day greater than 90 MME requires:<br>○ A comprehensive individual treatment plan including attestation of a pain management agreement between the prescriber and patient<br>○ Continued assessment and documentation of risk of abuse<br>○ Documentation that previous tapers have been attempted or documentation of a taper plan or rationale for avoidance of taper initiation</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td><u>Calculating morphine milligram equivalents (MME)</u><br><br>
<table>
<tr><th>Opioid</th><th>Factor</th></tr>
<tr><td>Methadone</td><td>4.7</td></tr>
<tr><td>Codeine</td><td>0.15</td></tr>
<tr><td>Fentanyl transdermal (mcg/hr)</td><td>2.4</td></tr>
<tr><td>Hydrocodone</td><td>1</td></tr>
<tr><td>Hydromorphone</td><td>5</td></tr>
<tr><td>Morphine</td><td>1</td></tr>
<tr><td>Oxycodone (Roxicodone, Oxycontin)</td><td>1.5</td></tr>
<tr><td>Oxymorphone</td><td>3</td></tr>
<tr><td>Tramadol</td><td>0.2</td></tr>
<tr><td>Buprenorphine patch</td><td>**</td></tr>
<tr><td>Tapentadol</td><td>0.4</td></tr>
<tr><td>Oxycodone myristate</td><td>1.67</td></tr>
</table>
** The MME conversion factor for buprenorphine patches is based on the assumption that:<br>• One milligram of parenteral buprenorphine is equivalent to 75 milligrams of oral morphine and<br>• One patch delivers the dispensed micrograms (mcg) per hour over a 24-hour day.<br><br>Example:<br>5 mcg/hr buprenorphine patch X 24 hrs = 120 mcg/day buprenorphine = 0.12 mg/day<br>0.12 mg per day X 75 (1 mg buprenorphine=75 mg morphine) = 9 mg/day oral MME.<br>In other words, the conversion factor not accounting for days of use would be 9/5 or 1.8.</td></tr>
</table>

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| | • Since the buprenorphine patch remains in place for 7 days, we have multiplied the conversion factor by 7 (1.8 X 7 = 12.6). In this example, MME/day for four 5 mcg/hr buprenorphine patches dispensed for use over 28 days would work out as follows:<br><br>**Example**:<br>5 mcg/hr buprenorphine patch X (4 patches/28 days) X 12.6 = 9 MME/day.<br><br>Please note that because this allowance has been made based on the typical dosage of one buprenorphine patch per 7 days. You should first change all days supply in your prescription data to follow this standard, i.e., days supply for buprenorphine patches= # of patches x 7 |
| **Exclusion Criteria:** | • Pain related to current active cancer<br>• Chronic pain related to sickle cell disease<br>• Pain related to hospice care<br>• Surgery or documented acute injury – 1 month approval |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**ORAL-INTRANASAL FENTANYL**
Affected Medications: ABSTRAL, ACTIQ, FENTORA, FENTANYL CITRATE BUCCAL TABLET, LAZANDA, SUBSYS

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Management of breakthrough pain in cancer patients who are already receiving and who are tolerant to around-the-clock opioid therapy for their underlying persistent cancer pain |
| **Required Medical Information:** | • Documentation of ALL of the following:<br>  ○ This drug is being prescribed for breakthrough cancer-related pain<br>  ○ The patient is currently receiving, and will continue to receive, around-the-clock opioid therapy for underlying persistent cancer pain<br>  ○ The patient is opioid tolerant, defined as taking one of the following for one week or longer:<br>  ▪ At least 60 mg of oral morphine per day<br>  ▪ At least 25 mcg of transdermal fentanyl per hour<br>  ▪ At least 30 mg of oral oxycodone per day<br>  ▪ At least 8 mg of oral hydromorphone per day<br>  ▪ At least 25 mg of oral oxymorphone per day<br>  ▪ At least 60 mg of oral hydrocodone per day<br>  ▪ An equianalgesic dose of another opioid |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of **ONE** of the following:<br>  ○ The patient is unable to swallow, or has dysphagia, esophagitis, mucositis, or uncontrollable nausea/vomiting<br>  ○ The patient has documented intolerance or allergies to two other short-acting narcotics (such as oxycodone, morphine sulfate, hydromorphone, etc.)<br><br>**PDL only:** Actiq requests will require documentation of clinical trial and failure with fentanyl citrate lozenge on a handle<br><br>**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist or specialist in the treatment of cancer-related pain<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**ORENITRAM**
Affected Medications: ORENITRAM (Treprostinil oral)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1 |
| **Required Medical Information:** | **<u>Pulmonary Arterial Hypertension (PAH) WHO Group 1</u>**<br>• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:<br>  ○ Mean pulmonary artery pressure of at least 20 mm Hg<br>  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg<br>  ○ Pulmonary vascular resistance of at least 2.0 Wood units<br>• Etiology of PAH: idiopathic, heritable, or associated with connective tissue disease<br>• PAH secondary to one of the following conditions:<br>  ○ Connective tissue disease<br>  ○ Human immunodeficiency virus (HIV) infection<br>  ○ Cirrhosis<br>  ○ Anorexigens<br>  ○ Congenital left to right shunts<br>  ○ Schistosomiasis<br>  ○ Drugs and toxins<br>  ○ Portal hypertension<br>• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class II or higher symptoms<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blockers) unless there are contraindications:<br>  ○ Low systemic blood pressure (systolic blood pressure less than 90)<br>  ○ Low cardiac index<br>  OR<br>  ○ Presence of severe symptoms (functional class IV) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of failure with Remodulin<br>• The pulmonary hypertension has progressed despite maximal medical and/or surgical treatment of the identified condition<br>• Documentation that treprostinil is used as a single route of administration (Remodulin, Tyvaso, Orenitram should not be used in combination)<br>• Not recommended for PAH secondary to pulmonary venous hypertension (e.g., left sided atrial or ventricular disease, left sided valvular heart disease, etc) or disorders of the respiratory system (e.g., chronic obstructive pulmonary disease, interstitial lung disease, obstructive sleep apnea or other sleep disordered breathing, alveolar hypoventilation disorders, etc.)<br><br>**<u>Reauthorization</u>** requires documentation of treatment success defined as one or more of the following:<br>• Improvement in walking distance<br>• Improvement in exercise ability<br>• Improvement in pulmonary function<br>• Improvement or stability in WHO functional class |

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| **Exclusion Criteria:** | • Severe hepatic impairment (Child Pugh Class C) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified. |

POLICY NAME:
**ORGOVYX**
Affected Medications: ORGOVYX (relugolix)

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | |
| **Appropriate Treatment Regimen & Other Criteria:** | **Prostate Cancer**<br>• Documented treatment failure or intolerable adverse event with leuprolide or degarelix<br><br>**Reauthorization**: documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**OSILODROSTAT**
Affected Medications: ISTURISA (osilodrostat)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Cushing's syndrome |
| **Required Medical Information:** | • Diagnosis of Cushing's syndrome due to one of the following:<br>  ○ Adrenocorticotropic hormone (ACTH)-secreting pituitary adenoma (Cushing's disease)<br>  ○ Ectopic ACTH section (EAS) by a non-pituitary tumor<br>  ○ Cortisol secretion by an adrenal adenoma<br>• Documentation of at least **TWO** of the following:<br>  ○ Mean (at least two measurements) 24-hour urine free cortisol (mUFC) greater than 1.5 times the upper limit of normal (ULN) for the assay<br>  ○ Bedtime salivary cortisol (at least two measurements) greater than 145 ng/dL<br>  ○ Overnight dexamethasone suppression test (DST) with a serum cortisol greater than 1.8 mcg/dL |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation confirming surgery is not an option **OR** previous surgery has not been curative<br><br>**Reauthorization** requires documentation of treatment success defined as mUFC normalization (i.e., less than or equal to the ULN) |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist, neurologist, or adrenal surgeon<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**OTESECONAZOLE**
Affected Medications: VIVJOA (oteseconazole)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ To reduce the incidence of recurrent vulvovaginal candidiasis (RVVC) in females with a history of RVVC who are **not** of reproductive potential, alone or in combination with fluconazole |
| **Required Medical Information:** | • Diagnosis of RVVC defined as three or more episodes of symptomatic vulvovaginal candidiasis infection within the past 12 months<br>• Documented presence of signs/symptoms of current acute vulvovaginal candidiasis with a positive potassium hydroxide (KOH) test<br>• Documentation confirming that the patient is permanently infertile (e.g., due to tubal ligation, hysterectomy, salpingo-oophorectomy) or postmenopausal |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented disease recurrence following maintenance therapy with fluconazole 150 mg once per week for 6 months<br>• **Not to exceed one treatment course per year**<br><br>**Reauthorization** requires documentation of treatment success defined as a reduction in symptomatic vulvovaginal candidiasis episodes, and documentation supporting the need for additional treatment |
| **Exclusion Criteria:** | • Women of reproductive potential or who are pregnant or breastfeeding |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 3 months, unless otherwise specified |

POLICY NAME:
**OXERVATE**
Affected Medications: OXERVATE (cenegermin-bkbj)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of neurotrophic keratitis |
| **Required Medical Information:** | • Documentation of decreased corneal sensitivity (≤ 4 cm using the Cochet-Bonnet [CB] aesthesiometer) within the area of the recurrent/persistent epithelial defect (PED) or corneal ulcer **AND** outside of the area of the defect in at least one corneal quadrant<br>• Documentation of one of the following:<br>  ○ Stage 2 neurotrophic keratitis, confirmed by presence of recurrent or persistent corneal epithelial defect<br>  ○ Stage 3 neurotrophic keratitis, confirmed by presence of corneal ulceration (with or without stromal melting and perforation) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of treatment failure (e.g., persistent epithelial defects or corneal ulceration) with preservative-free artificial tears/ointments and **TWO** of the following:<br>  ○ Therapeutic contact lenses (TCLs) (e.g., corneal or scleral contact lenses, soft bandage contact lenses)<br>  ○ Amniotic membrane transplantation<br>  ○ Tarsorrhaphy<br>  ○ Conjunctival flap surgery<br>• Dose may not exceed more than 1 vial per eye per day<br><br>**Reauthorization** requires documentation of treatment response, as shown by a reduction in corneal staining with fluorescein |
| **Exclusion Criteria:** | • Active or suspected ocular or periocular infections |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 8 weeks, unless otherwise specified<br>• Reauthorization: 8 weeks, unless otherwise specified<br>• Lifetime Limit: 16 weeks (per affected eye) |

POLICY NAME:
**OXYBATES**
Affected Medications: LUMRYZ (sodium oxybate extended release), XYREM (sodium oxybate), XYWAV (oxybate salts), SODIUM OXYBATE

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Narcolepsy with cataplexy<br>  ○ Narcolepsy with excessive daytime sleepiness (EDS)<br>  ○ Idiopathic Hypersomnia (IH) (Xywav only) |
| **Required Medical Information:** | • Diagnosis confirmed by polysomnography and multiple sleep latency test<br>• Other causes of sleepiness have been ruled out or treated (including but not limited to obstructive sleep apnea, insufficient sleep syndrome, shift work, the effects of substances or medications, or other sleep disorders)<br><br>**Narcolepsy with cataplexy**:<br>• Documentation of cataplexy episodes defined as more than one episode of sudden loss of muscle tone with retained consciousness<br><br>**Narcolepsy with EDS or IH:**<br>• Current evaluation of symptoms and Epworth Sleepiness Scale (ESS) score of more than 10 despite treatment |
| **Appropriate Treatment Regimen & Other Criteria:** | **Narcolepsy with cataplexy:**<br>• Documented treatment failure with TWO of the following for at least 1 month each:<br>  ○ Venlafaxine<br>  ○ Fluoxetine<br>  ○ Duloxetine<br>  ○ Tricyclic antidepressant (such as clomipramine, protriptyline)<br><br>**Narcolepsy or IH, with EDS:**<br>• Documented treatment failure to all of the following (1 in each category required) for at least 1 month each:<br>  ○ Modafinil or armodafinil<br>  ○ Methylphenidate, or dextroamphetamine, or lisdexamfetamine<br>  ○ Sunosi (Narcolepsy with EDS only)<br><br>**Reauthorization:**<br>• Narcolepsy with cataplexy: requires clinically significant reduction in cataplexy episodes<br>• Narcolepsy or IH, with EDS: requires clinically significant improvement in activities of daily living and in Epworth Sleepiness Scale (ESS) score |
| **Exclusion Criteria:** | • Concurrent use of alcohol, sedative/hypnotic drugs, or other central nervous system depressants.<br>• Use for other untreated causes of sleepiness |
| **Age Restriction:** | • 7 years of age and older for cataplexy or EDS due to narcolepsy<br>• 18 years of age and older for EDS due to IH |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a sleep specialist or neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PALFORZIA**
Affected Medications: PALFORZIA (peanut allergen powder)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Mitigation of allergic reactions, including anaphylaxis, that may occur with accidental exposure to peanut |
| **Required Medical Information:** | • Documented treatment plan, including dose and frequency<br>• Diagnosis of peanut allergy confirmed by one of the following:<br>  ○ A positive skin prick test (SPT) response to peanut with a wheal diameter at least 3 mm larger than the control<br>  ○ Serum peanut-specific IgE level greater than or equal to 0.35 kUA/L<br>• Documented history of an allergic reaction to peanut with all of the following:<br>  ○ Signs and symptoms of a significant systemic allergic reaction to peanut (e.g., hives, swelling, wheezing, hypotension, gastrointestinal symptoms)<br>  ○ The reaction occurred within a short period of time following a known ingestion of peanut or peanut-containing food<br>  ○ The reaction was severe enough to warrant a prescription for an epinephrine injection<br>• Documentation indicating a significant impact on quality of life due to peanut allergies |
| **Appropriate Treatment Regimen & Other Criteria:** | **Dosing:**<br>• Requests for initial dose escalation: must be between 1 and 17 years of age<br>• Requests for up-dosing and maintenance phase: 1 year of age and older<br><br>**Reauthorization** requires documentation of completion of the appropriate initial dose escalation and up-dosing phases prior to moving on to the maintenance phase AND documentation of treatment success and a clinically significant response to therapy, defined by one or more of the following:<br>• Improvement in quality of life<br>• Reduction in severe allergic reactions<br>• Reduction in epinephrine use<br>• Reduction in physician office visits, ER visits, or hospitalizations due to peanut allergy |
| **Exclusion Criteria:** | • Use for the emergency treatment of allergic reactions, including anaphylaxis<br>• Uncontrolled asthma<br>• History of eosinophilic esophagitis (EoE) and other eosinophilic gastrointestinal disease<br>• History of cardiovascular disease, including uncontrolled or inadequately controlled hypertension<br>• History of a mast cell disorder, including mastocytosis, urticarial pigmentosa, and hereditary or idiopathic angioedema |
| **Age Restriction:** | • 1 year of age and older (see Appropriate Treatment Regimen & Other Criteria for specific age-related dosing requirements) |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an allergist or immunologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified |

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| | • Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PALIVIZUMAB**
Affected Medications: SYNAGIS (palivizumab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | **Congenital Heart Disease (CHD)**<br>• **12 months of age and younger:** Documentation of one of the following:<br>  ○ Pharmacologically treated acyanotic heart disease that will require surgical intervention<br>  ○ Cyanotic heart defects<br>  ○ Moderate to severe pulmonary hypertension<br>• **24 months of age and younger:** Receipt of cardiac transplantation during the RSV season<br><br>**Chronic Lung Disease (CLD) of Prematurity**<br>• Gestational age less than 32 weeks and 0 days<br>• **12 months of age and younger:** Required supplemental oxygen for at least the first 28 days after birth<br>• **24 months of age and younger:** Documentation of **both** of the following:<br>  ○ Required supplemental oxygen for at least the first 28 days after birth<br>  ○ Required continued medical support during the 6-month period prior to RSV season (chronic corticosteroids, diuretics, supplemental oxygen)<br><br>**Cystic Fibrosis (CF)**<br>• Documented diagnosis of cystic fibrosis<br>• **12 months of age and younger:** Clinical evidence of chronic lung disease and/or nutritional compromise<br>• **24 months of age and younger:** Documentation of **ONE** of the following:<br>  ○ Manifestations of severe lung disease (prior hospitalization for pulmonary exacerbation in the first year of life, abnormalities on chest X-ray or computed tomography that persist when stable)<br>  ○ Weight for length less than the 10$^{th}$ percentile<br><br>**Pulmonary Abnormalities/Neuromuscular Disorders**<br>• 12 months of age and younger<br>• Documentation of congenital anomaly or neuromuscular disease resulting in ineffective cough and impaired ability to clear the upper airway of secretions (excluding cystic fibrosis)<br><br>**Premature Infants**<br>• Gestational age less than 29 weeks and 0 days<br>• 12 months of age and younger |
| **Appropriate Treatment Regimen &** | **RSV Season**<br>• Not to exceed 5 monthly doses (15 mg/kg per dose) during the RSV season, with first |

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| **Other Criteria:** | dose administered prior to commencement of the RSV season<br>  ○ If hospitalized at the start of RSV season, administer first dose 48-72 hours prior to discharge<br>• Discontinue monthly prophylaxis if hospitalized for breakthrough RSV<br><br>**Off Season**<br>• Approvable for one 15 mg/kg dose when RSV activity is 10% or greater for the region, per the CDC |
| **Exclusion Criteria:** | • Administration of nirsevimab (Beyfortus) during the current RSV season<br>• For use in the treatment of RSV |
| **Age Restriction:** | • See Required Medical Information |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • **RSV Season:** 5 months (not to exceed end of RSV season), unless otherwise specified<br>• **Off Season:** 1 month, unless otherwise specified |

POLICY NAME:
**PALOVAROTENE**
Affected Medications: SOHONOS (palovarotene)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ To reduce the volume of new heterotopic ossification in patients with fibrodysplasia ossificans progressiva (FOP) |
| **Required Medical Information:** | • Documentation of genetic testing confirming a diagnosis of FOP with an *activin receptor type 1* (*ACVR1*) R206H mutation<br>• Radiographic testing has confirmed the presence of both of the following:<br>  ○ Heterotopic ossification (HO)<br>  ○ Joint malformations (such as hallux valgus deformity, malformed first metatarsal, absent or fused interphalangeal joint)<br>• Documentation of at least two flare-ups in the past 12 months requiring prescription strength non-steroidal anti-inflammatory drugs (NSAIDs) and oral glucocorticoids (e.g., prednisone) |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires documentation of treatment success defined as a decrease in HO volume or number of flare-ups compared to baseline |
| **Exclusion Criteria:** | • Patients weighing less than 10 kg<br>• Pregnancy |
| **Age Restriction:** | • Females: 8 years of age and older<br>• Males: 10 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist in rare connective tissue diseases (e.g., endocrinologist, geneticist, orthopedist, rheumatologist)<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PALYNZIQ**
Affected Medications: PALYNZIQ (pegvaliase-pqpz)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Documentation of a diagnosis of PKU<br>• Documentation of treatment failure with dual therapy of sapropterin and a Phe restricted diet as shown by a blood Phe level greater than 600 micromol/L (10 mg/dL) despite treatment |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation that Palynziq will not be used in combination with sapropterin<br><br>**Reauthorization** requires documentation of one of the following:<br>• Reduction in baseline Phe levels by 20 percent<br>• Increase in dietary Phe tolerance<br>• Improvement in clinical symptoms |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist in metabolic disorders or an endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PARATHYROID HORMONE**
Affected Medications: YORVIPATH (palopegteriparatide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of hypoparathyroidism |
| **Required Medical Information:** | • Documentation of the following lab values while on standard of care calcium and active vitamin D treatment:<br>  ○ 25-hydroxyvitamin D levels between 20-80 ng/mL<br>  ○ Total serum calcium (albumin-corrected) greater than 7.8 mg/dL |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented failure with at least 12 weeks of a consistent supplementation regimen as follows:<br>  ○ Calcium 1000-2000 mg (elemental) daily<br>  ○ Vitamin D metabolite (calcitriol) OR vitamin D analog<br><br>**Reauthorization** will require documentation of treatment success defined as total serum calcium (albumin-corrected) within the lower half of the normal range (approximately 8-9 mg/dL) |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist or nephrologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PARATHYROID HORMONE ANALOGS**
Affected Medications: TERIPARATIDE, TYMLOS (abaloparatide), FORTEO (teriparatide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of osteoporosis in men and postmenopausal women at high risk for fracture (teriparatide, Tymlos, and Forteo)<br>  ○ Treatment of glucocorticoid-induced osteoporosis in men and women at high risk for fracture (teriparatide and Forteo only) |
| **Required Medical Information:** | • Diagnosis of osteoporosis as defined by at least **one** of the following:<br>  ○ T-score –2.5 or lower (current or past) at the lumbar spine, femoral neck, total hip, or 1/3 radius site<br>  ○ T-score between –1.0 and –2.5 at the lumbar spine, femoral neck, total hip, or 1/3 radius site **AND** increased risk of fracture as defined by at least one of the following Fracture Risk Assessment Tool (FRAX) scores:<br>  ▪ FRAX 10-year probability of major osteoporotic fracture is 20% or greater<br>  ▪ FRAX 10-year probability of hip fracture is 3% or greater<br>  ○ History of non-traumatic fractures in the absence of other metabolic bone disorders (postmenopausal women with osteoporosis only)<br>• For glucocorticoid-induced osteoporosis, in addition to the above, must also provide documentation of the following:<br>  ○ Treatment with 5 mg or higher of prednisone (or equivalent) per day for at least 3 months |
| **Appropriate Treatment Regimen & Other Criteria:** | Documentation of **ONE** of the following:<br>• Treatment failure (new fracture or worsening T-score despite adherence to an adequate trial of therapy), contraindication, or intolerance to the following:<br>  ○ Oral or intravenous bisphosphonate (such as, alendronate, risedronate, zoledronic acid or ibandronate)<br>• High risk of fracture defined as T-score -3.5 or lower, **OR** T-score -2.5 or lower with a history of fragility fractures<br><br>For **Forteo** requests: Documented treatment failure with Tymlos and teriparatide<br><br>**<u>Total duration of therapy with parathyroid hormone analogs should not exceed 2 years in a lifetime</u>**<br>• Forteo or teriparatide may be reauthorized for up to one additional year beyond two years of parathyroid hormone analog use (maximum of 3 total years) if meeting the following criteria:<br>  ○ Documentation of treatment success with parathyroid hormone use, defined as reduced frequency of fragility fractures or stable T-score while on Forteo or teriparatide<br>  ○ Documentation that after 24 months of parathyroid hormone analog use, the patient remains at or has returned to having a high risk for fracture as evidenced by new fragility fracture or decline in T-score |
| **Exclusion Criteria:** | • Paget's Disease<br>• Open epiphyses (such as, pediatric or young adult patient)<br>• Bone metastases or skeletal malignancies<br>• Hereditary disorders predisposing to osteosarcoma |

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| | • Prior external beam or implant radiation therapy involving the skeleton<br>• Concurrent use of bisphosphonates, other parathyroid hormone analogs, or RANK ligand inhibitors<br>• Preexisting hypercalcemia<br>• Pregnancy |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 24 months (no reauthorization), unless otherwise specified |

POLICY NAME:
**PEDMARK**
Affected Medications: PEDMARK (sodium thiosulfate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ To reduce the risk of ototoxicity associated with cisplatin in pediatric patients 1 month of age and older with localized, non-metastatic solid tumors. |
| **Required Medical Information:** | • Documentation of a treatment plan that is a cisplatin-based regimen treating a localized, non-metastatic solid tumor |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | • Metastatic disease |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 6 months or duration of cisplatin regimen, unless otherwise specified |

POLICY NAME:
**PEGASYS**
Affected Medications: PEGASYS (peginterferon alfa-2a)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved and compendia-supported indications not otherwise excluded by plan design<br>  ○ Chronic hepatitis B (CHB)<br>  ▪ Treatment of adults with HBeAg-positive and HBeAg-negative CHB infection who have compensated liver disease, evidence of viral replication, and liver inflammation<br>  ▪ Treatment of HBeAg-positive CHB in non-cirrhotic pediatric patients 3 years of age and older with evidence of viral replication and elevations in serum alanine aminotransferase (ALT)<br>  ○ Polycythemia vera<br>  ○ Essential thrombocythemia |
| **Required Medical Information:** | • Documentation of anticipated treatment course, to include full antiviral regimen, and duration of therapy<br><br>**<u>CHB – Compensated Cirrhosis</u>**<br>• Documentation of compensated cirrhosis<br>• Documented HBV DNA level greater than 2,000 IU/mL<br><br>**<u>CHB – Non-cirrhotic</u>**<br>• Documentation of HBeAg-positive non-cirrhotic disease<br>• Documented HBV DNA level greater than 20,000 IU/mL<br>• Current (within 12 weeks) serum ALT level ≥ 2 times the upper limit of normal<br><br>**<u>Polycythemia Vera (PV)</u>**<br>• Diagnosis of polycythemia vera confirmed by all major criteria (1-3) OR the first 2 major criteria (1-2) plus the minor criterion:<br>  ○ **Major criteria:**<br>  (1) Elevated hemoglobin concentration (greater than 16 g/dL), elevated hematocrit (greater than 48 percent), or increased red blood cell mass (greater than 25% above mean normal predicted value)<br>  (2) Presence of *JAK2* V617F **or** *JAK2* exon 12 mutation<br>  (3) Bone marrow biopsy showing age-adjusted hypercellularity with trilineage proliferation (panmyelosis), including prominent erythroid, granulocytic, and increase in pleomorphic, mature megakaryocytes without atypia. May not be required in patients with sustained absolute erythrocytosis (hemoglobin over 18.5 g/dL and hematocrit over 55.5 percent in men; hemoglobin over 16.5 g/dL and hematocrit over 49.5 percent in women) with presence of a *JAK2* V617F or *JAK2* exon 12 mutation<br>  ○ **Minor criterion:** Subnormal serum erythropoietin level<br><br>**<u>Essential Thrombocythemia (ET)</u>**<br>• Diagnosis of essential thrombocythemia confirmed by all major criteria (1-4) **OR** the first 3 major criteria (1-3) plus the minor criterion:<br>  ○ **Major criteria:** |

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| | (1) Platelet count greater than or equal to 450,000 cells/mcL<br>(2) Bone marrow biopsy showing proliferation mainly of the megakaryocytic lineage, with hyperlobulated staghorn-like nuclei, infrequently dense clusters; no significant increase or left shift in neutrophil granulopoiesis or erythropoiesis; no relevant bone marrow fibrosis<br>(3) Diagnostic criteria for BCR::ABL1-positive chronic myeloid leukemia, polycythemia vera, primary myelofibrosis, or other neoplasms are not met.<br>(4) Presence of *JAK2*, *CALR*, or *MPL* mutation<br>○ **Minor criterion:** Presence of another clonal marker (e.g., *ASXL1*, *EZH2*, *TET2*, *IDH1/IDH2*, *SRSF2*, or *SRF3B1* mutation) **OR** no identifiable cause for thrombocytosis (such as iron deficiency, chronic infection, chronic inflammatory disease, prior splenectomy) |
| **Appropriate Treatment Regimen & Other Criteria:** | **PV and ET**<br>• Documented treatment failure, intolerance, or contraindication to hydroxyurea |
| **Exclusion Criteria:** | • Autoimmune hepatitis<br>• Hepatic decompensation (Child-Pugh score greater than 6) |
| **Age Restriction:** | • CHB with compensated cirrhosis: 18 years of age and older<br>• CHB without cirrhosis: 3 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a gastroenterologist, hematologist, hepatologist, oncologist, or infectious disease specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **CHB:**<br>• Authorization: 12 months, unless otherwise specified<br>**PV, ET:**<br>• Initial Authorization: 4 months, unless otherwise specified.<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PEGLOTICASE**
Affected Medications: KRYSTEXXA (pegloticase)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Chronic gout in adults refractory to conventional therapy |
| **Required Medical Information:** | • Baseline serum uric acid (SUA) level greater than 8 mg/dL<br>• Documentation of **ONE** of the following:<br>  ○ 2 or more gout flares per year that were inadequately controlled by colchicine and/or nonsteroidal anti-inflammatory drugs (NSAIDS) or oral/injectable corticosteroids<br>  ○ At least 1 non-resolving subcutaneous gouty tophus<br>  ○ Chronic gouty arthritis (defined clinically or radiographically as joint damage due to gout) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented contraindication, intolerance or clinical failure (defined as inability to reduce SUA level to less than 6 mg/dL) following a 12-week trial at maximum tolerated dose to BOTH:<br>  ○ Xanthine oxidase inhibitor (allopurinol or febuxostat)<br>  ○ Combination of a xanthine oxidase inhibitor AND a uricosuric agent (such as probenecid). If xanthine oxidase inhibitor is contraindicated, trial with uricosuric agent required.<br>• Documentation Krystexxa will be used in combination with oral methotrexate 15 mg weekly unless contraindicated<br><br>**<u>Reauthorization</u>** will require **ALL** of the following:<br>• Documentation of SUA less than 6 mg/dL prior to next scheduled Krystexxa dose<br>• Documentation of response to treatment such as reduced size of tophi or number of flares or affected joints<br>• Rationale to continue treatment after resolution of tophi or reduction in symptoms |
| **Exclusion Criteria:** | • Concurrent use with oral urate-lowering therapies |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a nephrologist or rheumatologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 6 months, unless otherwise specified |

# PEGZILARGINASE

Included Products: Loargys

## SCOPE & EXCLUSIONS

Included Indications: All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. Drug compendia supported indications may be covered. Covered Food and Drug Administration (FDA)-approved uses:

- Hyperargininemia in adult and pediatric with Arginase 1 Deficiency (ARG1-D)

Exclusions: Extreme mobility impairment, prior liver or hematopoietic transplant

Age Limits: As defined by FDA labeling for the requested indication.

Prescriber Limits: Prescribed by, or in consultation with an appropriate specialist experienced in urea cycle disorders.

Other Limits: All approvals are subject to utilization of the most cost-effective site of care

## INITIAL AUTHORIZATION CRITERIA

### Required Medical Information:

1. All Indications must be FDA-supported for the requested product [or strongly supported in drug compendia (ex. Lexi Drug, Drugdex)]
2. Requested dosing must be according to the FDA label based on diagnosis, age, and weight.

### Arginase 1 Deficiency (ARG1-D)

1. Documented ARG1-D diagnosis, as evidenced by either biochemical evidence of the disease with persistently elevated pArg levels, pathogenic variants in ARG1, or diminished erythrocyte ARG1 activity
2. Documentation of high plasma arginine (pArg) of 250 µM/L or more at baseline
3. Impairment on any secondary functional mobility assessment
4. Documentation that drug will be used in conjunction with a protein-restricted diet
5. Documentation of failure with use of oral sodium phenylbutyrate in combination with a protein-restricted diet (such as high pArg despite treatment)

## RENEWAL CRITERIA

Documented treatment response as evidenced by plasma arginine (pArg) levels maintained at 40 µmol/L–115 µmol/L

## DURATION OF APPROVAL

Initial: 6 months, unless otherwise specified

Renewal: 12 months, unless otherwise specified

POLICY NAME:
**PEMIVIBART**
Affected Medications: PEMGARDA (pemivibart)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) or compendia supported indications not otherwise excluded by plan design<br>  ○ Pre-exposure prophylaxis of coronavirus disease 2019 (COVID-19) in adults and adolescents with moderate-to-severe immune compromise due to a medical condition or receipt of immunosuppressive medications/treatments **and** are unlikely to mount an adequate immune response to COVID-19 vaccination |
| **Required Medical Information:** | • Documentation of moderate-to-severe immune compromise due to a medical condition or receipt of immunosuppressive medications or treatments, and are unlikely to mount an adequate response to COVID-19 vaccination, meeting one of the following:<br>  ○ Active treatment for solid tumor and hematologic malignancies<br>  ○ Hematologic malignancies associated with poor responses to COVID-19 vaccines regardless of current treatment status (e.g., chronic lymphocytic leukemia, non-Hodgkin lymphoma, multiple myeloma, acute leukemia)<br>  ○ Receipt of solid-organ transplant or an islet transplant and taking immunosuppressive therapy<br>  ○ Receipt of chimeric antigen receptor (CAR)-T-cell or hematopoietic stem cell transplant (within 2 years of transplantation or taking immunosuppressive therapy)<br>  ○ Moderate or severe primary immunodeficiency (e.g., common variable immunodeficiency disease, severe combined immunodeficiency, DiGeorge syndrome, Wiskott-Aldrich syndrome)<br>  ○ Advanced or untreated human immunodeficiency viruses (HIV) infection (people with HIV and CD4 cell counts less than 200/$mm^3$, history of an AIDS-defining illness without immune reconstitution, or clinical manifestations of symptomatic HIV)<br>  ○ Active treatment with high-dose corticosteroids (at least 20 mg prednisone or equivalent per day when administered for 2 or more weeks), alkylating agents, antimetabolites, transplant-related immunosuppressive drugs, cancer chemotherapeutic agents classified as severely immunosuppressive, and biologic agents that are immunosuppressive or immunomodulatory (such as B-cell depleting agents)<br>• Documentation of prophylactic use<br>• Weight of 40 kg or more |
| **Appropriate Treatment Regimen & Other Criteria:** | • **Dosing** is in accordance with FDA labeling and does not exceed 4500 mg once every 3 months<br><br>**Reauthorization** requires documentation of continued immune compromise |
| **Exclusion Criteria:** | • Positive SARS-CoV-2 antigen test or PCR test within the last 3 months<br>• Received COVID-19 vaccine within the last 2 weeks |
| **Age Restriction:** | • 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 3 months, unless otherwise specified |

POLICY NAME:
**PENICILLAMINE**
Affected Medications: PENICILLAMINE CAPSULE

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Cystinuria<br>  ○ Wilson's Disease<br>  ○ Rheumatoid arthritis<br>  ○ Copper measurement in urine |
| **Required Medical Information:** | • Documented treatment plan including routine urinalysis, WBCs, hemoglobin, platelet count, liver function tests, renal function tests due to risk of fatalities due to aplastic anemia, agranulocytosis, thrombocytopenia, myasthenia gravis, and Goodpasture's Syndrome<br><br>**<u>Wilson's Disease</u>**<br>• Diagnosis confirmed by **ONE** of the following:<br>  ○ Genetic testing results confirming biallelic pathogenic *ATP7B* mutations (in either symptomatic or asymptomatic individuals)<br>  ○ Liver biopsy findings consistent with Wilson's disease<br>  ○ Presence of Kayser-Fleischer (KF) rings **AND** serum ceruloplasmin level less than 20 mg/dL **AND** 24-hour urinary copper excretion greater than 40 mcg<br>  ○ Presence of Kayser-Fleischer (KF) rings **AND** 24-hour urinary copper excretion greater than 100 mcg<br>  ○ Absence of KF rings with serum ceruloplasmin level less than 10 mg/dL **AND** 24-hour urinary copper excretion greater than 100 mcg<br><br>**<u>Rheumatoid arthritis</u>**<br>• Documentation of severe, active disease defined by one of the following:<br>  ○ The Disease Activity Score derivative for 28 joints (DAS-28) greater than 3.2<br>  ○ The Simplified Disease Activity Index (SDAI) greater than 11<br>  ○ The Clinical Disease Activity Index (CDAI) greater than 10<br>  ○ Weighted Routine Assessment of Patient Index Data 3 (RAPID3) of at least 2.3 |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Rheumatoid arthritis</u>**<br>• Has failed to respond to an adequate trial of conventional therapies (such as methotrexate, sulfasalazine, hydroxychloroquine, leflunomide, Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Xeljanz, Rinvoq, and Inflectra)<br><br>**<u>Reauthorization</u>** requires documentation of disease responsiveness to therapy<br>• For Wilson's disease, must have normalization of free serum copper (non-ceruloplasmin bound copper) to less than 15 mcg/dL and 24-hour urinary copper in the range of 200 to 500 mcg |
| **Exclusion Criteria:** | • Use of penicillamine during pregnancy (except for treatment of Wilson's disease or cystinuria) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist familiar with the toxicity and dosage considerations (such as a hepatologist, gastroenterologist, or liver transplant physician for Wilson's Disease) |

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| | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PHENOXYBENZAMINE**
Affected Medications: PHENOXYBENZAMINE, DIBENZYLINE (phenoxybenzamine)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of sweating and hypertension associated with pheochromocytoma |
| **Required Medical Information:** | • Documented diagnosis of pheochromocytoma that requires treatment to control episodes of hypertension and sweating<br>• This drug will be used for one of the following:<br>  ○ Preoperative preparation for a scheduled surgical resection<br>  ○ Chronic treatment of pheochromocytoma that is not amenable to surgery |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of treatment failure, intolerance, or contraindication to a selective alpha-1 adrenergic receptor blocker (e.g., doxazosin, terazosin, prazosin)<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist or a specialist with experience in the management of pheochromocytoma<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Preoperative preparation: 1 month, unless otherwise specified<br>• Chronic treatment: 12 months, unless otherwise specified |

POLICY NAME:
**PHOSPHODIESTERASE-5 (PDE-5) ENZYME INHIBITORS FOR PULMONARY ARTERIAL HYPERTENSION**
Affected Medications: ALYQ (tadalafil 20 mg tablet), TADALAFIL (PAH) 20 MG TABLET, TADLIQ (tadalafil 20 mg/5 ml suspension), SILDENAFIL 20 MG TABLET, SILDENAFIL 10 MG/ML SUSPENSION, LIQREV (sildenafil 10 mg/mL suspension)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1 |
| **Required Medical Information:** | • Diagnosis of World Health Organization (WHO) Group 1 PAH confirmed by right heart catheterization meeting the following criteria:<br>  ○ Mean pulmonary artery pressure of at least 20 mm Hg<br>  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg<br>  ○ Pulmonary vascular resistance of at least 2.0 Wood units<br>• New York Heart Association (NYHA)/WHO Functional Class II or higher symptoms<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blocker) unless there are contraindications:<br>  ○ Low systemic blood pressure (systolic blood pressure less than 90)<br>  ○ Low cardiac index<br>  ○ Presence of severe symptoms (functional class IV) |
| **Appropriate Treatment Regimen & Other Criteria:** | • For all brand requests: Documented inadequate response or intolerance to sildenafil citrate 20 mg tablets and tadalafil 20 mg tablets<br>• Requests for oral suspension must have documented inability to swallow tablets<br><br>**Reauthorization** requires documentation of treatment success defined as one or more of the following:<br>• Improvement in walking distance<br>• Improvement in exercise ability<br>• Improvement in pulmonary function<br>• Improvement or stability in WHO functional class |
| **Exclusion Criteria:** | • Concomitant nitrate therapy on a regular or intermittent basis<br>• Concomitant use of a guanylate cyclase stimulator (such as riociguat or vericiguat)<br>• Use for erectile dysfunction |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**PIRFENIDONE**
Affected Medications: PIRFENIDONE (267 and 801 mg)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Idiopathic Pulmonary Fibrosis (IPF) |
| **Required Medical Information:** | • Documented diagnosis of idiopathic pulmonary fibrosis (IPF) confirmed by **ONE** of the following:<br>  ○ Usual interstitial pneumonia (UIP) pattern demonstrated on high-resolution computed tomography (HRCT)<br>  ○ UIP pattern demonstrated on surgical lung biopsy<br>  ○ Probable UIP pattern demonstrated on **BOTH** HRCT and surgical lung biopsy<br>• Documentation confirming known causes of interstitial lung disease have been ruled out (e.g., rheumatic disease, environmental exposure, drug toxicity)<br>• Documentation of **BOTH** of the following:<br>  ○ Baseline forced vital capacity (FVC) greater than or equal to 50 percent predicted<br>  ○ Baseline diffusing capacity for carbon monoxide (DLCO) greater than or equal to 30 percent predicted |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires documentation of treatment success |
| **Exclusion Criteria:** | • Combined use with nintedanib (Ofev) |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**POMBILITI and OPFOLDA**
Affected Medications: POMBILITI (cipaglucosidase alfa-atga intravenous injection), OPFOLDA (miglustat oral capsule)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Late-onset Pompe disease for patients weighing 40 kg or more and who are not improving on their current enzyme replacement therapy (ERT) |
| **Required Medical Information:** | • Diagnosis of late-onset Pompe disease confirmed by one of the following:<br>  ○ Enzyme assay demonstrating a deficiency of acid alpha-glucosidase (GAA) enzyme activity<br>  ○ DNA testing that identifies mutations in the GAA gene<br>• One or more clinical signs or symptoms of late-onset Pompe disease:<br>  ○ Progressive proximal weakness in a limb-girdle distribution<br>  ○ Delayed gross-motor development in childhood<br>  ○ Involvement of respiratory muscles causing respiratory difficulty (such as reduced forced vital capacity [FVC] or sleep disordered breathing)<br>  ○ Skeletal abnormalities (such as scoliosis or scapula alata)<br>  ○ Low/absent reflexes<br>• Documentation that patient has a 6-minute walk test (6MWT) of 75 meters or more<br>• Documentation of a sitting percent predicted forced vital capacity (FVC) of 30% or more<br>• Patient weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of planned treatment regimen for both Pombiliti and Opfolda which are within FDA-labeling<br>• Documentation that patient is no longer improving after at least one year of current enzyme replacement therapy (ERT) with Lumizyme (alglucosidase alfa) or Nexviazyme (avalglucosidase alfa-ngpt)<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy as evidenced by an improvement, stabilization, or slowing of progression in percent-predicted FVC and/or 6MWT |
| **Exclusion Criteria:** | • Pregnancy or, if female of reproductive potential, not using effective contraception during treatment<br>• Use of invasive or noninvasive ventilation support for more than 6 hours a day while awake<br>• Diagnosis of infantile-onset Pompe disease<br>• Concurrent treatment with Lumizyme or Nexviazyme<br>• Pombiliti or Opfolda as monotherapy<br>• Use of Opfolda for Gaucher disease |
| **Age Restriction:** | • 18 years of age or older |

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| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a metabolic specialist, endocrinologist, biochemical geneticist, or provider experienced in the management of Pompe disease<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**POSACONAZOLE**
Affected Medications: POSACONAZOLE

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of oropharyngeal candidiasis (including infections refractory to itraconazole and/or fluconazole)<br>  ○ Treatment of invasive aspergillosis<br>  ○ Prophylaxis of invasive *Aspergillus* and *Candida* infections |
| **Required Medical Information:** | • Pediatric requests: Current body weight<br><br>**<u>Treatment of Oropharyngeal Candidiasis</u>**<br>• Documentation of oropharyngeal candidiasis<br>• Susceptibility cultures confirm posaconazole activity<br><br>**<u>Invasive Aspergillosis</u>**<br>• Documentation of invasive aspergillosis<br><br>**<u>Prophylaxis of Invasive *Aspergillus* and *Candida* Infections</u>**<br>• Documentation of severely immunocompromised state (such as hematopoietic stem cell transplant [HSCT] with graft-versus-host disease [GVHD], hematologic malignancy with prolonged neutropenia due to chemotherapy) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Treatment of Oropharyngeal Candidiasis</u>**<br>• Documented treatment failure (defined as no response to therapy) with both of the following:<br>  ○ Fluconazole<br>  ○ Itraconazole oral solution<br><br>**<u>Treatment of Invasive Aspergillosis</u>**<br>• Documented treatment failure or intolerable adverse event with voriconazole<br><br>**<u>Prophylaxis of Invasive *Aspergillus* and *Candida* Infections</u>**<br>• Documented treatment failure or intolerable adverse event with at least one systemic agent (e.g., fluconazole for *Candida* infections; voriconazole, amphotericin B, or itraconazole for *Aspergillus* infections). |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an infectious disease specialist, transplant physician, or oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 6 months, unless otherwise specified. |

POLICY NAME:
**POZELIMAB**
Affected Medications: VEOPOZ (pozelimab-bbfg)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of CD55-deficient protein-losing enteropathy (PLE) or CHAPLE disease |
| **Required Medical Information:** | • Diagnosis of CD-55-deficient PLE confirmed by biallelic CD55 loss-of-function mutation using molecular genetic testing<br>• Documentation of hypoalbuminemia (serum albumin of 3.2 g/dL or less)<br>• Clinical signs and features of active PLE including abdominal pain, diarrhea, peripheral edema, or facial edema<br>• Documentation of at least two albumin transfusions or hospitalizations in the past year |
| **Appropriate Treatment Regimen & Other Criteria:** | • **Dosing** is in accordance with FDA labeling and does not exceed the following:<br>  ○ Loading Dose: 30 mg/kg by intravenous infusion for 1 dose<br>  ○ Maintenance Dose: Starting on day 8;<br>  10 mg/kg as a subcutaneous injection once weekly<br>  May be increased to 12 mg/kg starting week 4<br>  ○ Maximum maintenance dosage of 800 mg once weekly<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization** requires documentation of positive clinical response with all the following:<br>• Improvement or stabilization of clinical symptoms<br>• Improvement or normalization of serum albumin concentrations<br>• Reduction in albumin transfusion requirements and/or hospitalizations |
| **Exclusion Criteria:** | • Receiving concurrent therapy with Soliris (eculizumab)<br>• Unresolved Neisseria meningitidis, Streptococcus pneumoniae, or Haemophilus influenzae type b (Hib) infection |
| **Age Restriction:** | • 1 year of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist, gastroenterologist, or provider that specializes in rare genetic hematologic diseases<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PRADEMAGENE ZAMIKERACEL**
Affected Medications: ZEVASKYN (prademagene zamikeracel)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Recessive Dystrophic Epidermolysis Bullosa (RDEB) |
| **Required Medical Information:** | • Complete description of the site(s) of application<br>• Diagnosis of RDEB confirmed by both of the following:<br>  ○ Skin biopsy of an induced blister with immunofluorescence mapping (IFM) and/or transmission electron microscopy (TEM)<br>  ○ Genetic test results documenting mutations in the COL7A1 gene<br>• Presence of partial-thickness RDEB wounds that are open and meet all the following:<br>  ○ Area must be at least 20 $cm^2$<br>  ○ Present for at least 6 months<br>  ○ Classified as a stage 2 wound, defined as partial thickness loss of dermis presenting as a shallow open ulcer with a pink or red wound bed, without slough or bruising |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of receiving standard of care preventative or treatment therapies for wound care, control of infection, nutritional support.<br>• Documented treatment failure with all the following: Filsuvez, Vyjuvek<br>• Dosing is in accordance with FDA labeling and does not exceed 12 sheets per one-time surgical application |
| **Exclusion Criteria:** | • Concurrent use with Vyjuvek (beremagene geperpavec-svdt) or Filsuvez (birch triterpenes)<br>• History of squamous cell carcinoma or active infection in the affected wound(s)<br>• Administered to wound(s) previously treated with Zevaskyn<br>• Administered to wound(s) that are currently healed |
| **Age Restriction:** | • 6 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a dermatologist or a specialist experienced in the treatment of epidermolysis bullosa<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 1 month, unless otherwise specified (one treatment per wound per lifetime) |

POLICY NAME:
**PROSTAGLANDIN IMPLANTS**
Affected Medications: Durysta (bimatoprost intracameral implant), iDose TR (travoprost intracameral implant)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Reduction of intraocular pressure (IOP) in patients with open angle glaucoma (OAG) or ocular hypertension (OHT) |
| **Required Medical Information:** | • Diagnosis of OAG or OHT with a baseline IOP of at least 22 mmHg<br>• Documentation of clinical justification for inability to manage routine topical therapy (e.g., due to progression of glaucoma, aging, comorbidities, and administration difficulties that cannot be addressed through instruction and technique) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure or intolerable adverse event with at least two IOP-lowering agents with different mechanisms of action, (used concurrently), one of which must include a prostaglandin analog such as latanoprost, bimatoprost, tafluprost, travoprost<br>• For iDose TR requests:<br>  ○ Documented treatment failure to the preferred product Durysta |
| **Exclusion Criteria:** | • Repeat implantation with the same prostaglandin implant<br>• Diagnosis of corneal endothelial cell dystrophy (e.g., Fuchs' Dystrophy)<br>• Prior corneal or endothelial cell transplantation (e.g., Descemet's Stripping Automated Endothelial Keratoplasty [DSAEK])<br>• Active or suspected ocular or periocular infections<br>• Absent or ruptured posterior lens capsule (Durysta) |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 1 month (one implant per impacted eye), unless otherwise specified |

POLICY NAME:
**PROXIMAL COMPLEMENT INHIBITOR**
Affected Medications: EMPAVELI (pegcetacoplan), FABHALTA (iptacopan)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of adults with paroxysmal nocturnal hemoglobinuria (PNH)<br>  ○ Reduce proteinuria in adults with primary immunoglobulin A nephropathy (IgAN) at risk of rapid disease progression, generally a urine protein-to-creatinine ratio (UPCR) ≥ 1.5 g/g (Fabhalta)<br>  ○ Treatment of complement 3 glomerulopathy (C3G) to reduce proteinuria<br>  ○ Treatment of primary immune-complex membranoproliferative glomerulonephritis (IC-MPGN), to reduce proteinuria (Empaveli) |
| **Required Medical Information:** | • Patients must be administered a meningococcal vaccine at least two weeks prior to initiation of the requested therapy and revaccinated according to current Advisory Committee on Immunization Practices (ACIP) guidelines<br><br>**PNH**<br>• Detection of PNH clones of at least 5% by flow cytometry diagnostic testing<br>  ○ Presence of at least 2 different glycosylphosphatidylinositol (GPI) protein deficiencies (e.g., CD55, CD59, etc.) within at least 2 different cell lines (e.g., granulocytes, monocytes, erythrocytes)<br>• Baseline lactate dehydrogenase (LDH) levels greater than or equal to 1.5 times the upper limit of normal range.<br>• One of the following PNH-associated clinical findings:<br>  ○ Presence of a thrombotic event<br>  ○ Presence of organ damage secondary to chronic hemolysis<br>  ○ History of 4 or more blood transfusions required in the previous 12 months<br><br>**IgAN (Fabhalta)**<br>• Diagnosis of IgAN confirmed with biopsy<br>• Documentation of one of the following (with labs current within 30 days of request):<br>  ○ Proteinuria defined as 0.5 g/day or greater<br>  ○ UPCR greater than 1.5 g/g<br><br>**C3G**<br>• Biopsy proven diagnosis of C3G<br>• UPCR of equal or greater than 1 g/g<br>• Estimated glomerular rate (eGFR) of 30 mL/min/1.73 $m^2$ or greater<br><br>**IC-MPGN (Empaveli)**<br>• Biopsy proven diagnosis of IC-MPGN<br>• UPCR greater than or equal to 1g/g<br>• Estimated glomerular rate (eGFR) of 30 mL/min/1.73$m^2$ or greater |
| **Appropriate Treatment Regimen & Other Criteria:** | **PNH**<br>• For Empaveli: documented inadequate response, contraindication, or intolerance to ravulizumab (Ultomiris)<br>• For Fabhalta: documented inadequate response, contraindication, or intolerance to another complement inhibitor such as ravulizumab (Ultomiris) or Empaveli |

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| | **<u>Reauthorization</u>** requires documentation of treatment success defined as a decrease in serum LDH, stabilized/improved hemoglobin, decreased transfusion requirement, and reduction in thromboembolic events compared to baseline<br><br>**<u>IgAN (Fabhalta)</u>**<br>• Documented treatment failure (defined as proteinuria equal to or greater than 0.5 g/day OR UPCR greater than 1.5 g/g) with a minimum of 12 weeks of all of the following:<br>  ○ Maximum tolerated dose of an angiotensin-converting enzyme (ACE) inhibitor or angiotensin receptor blocker (ARB)<br>  ○ Glucocorticoid therapy such as oral prednisone or methylprednisolone (or an adverse effect to two or more glucocorticoid therapies that is not associated with the corticosteroid class)<br><br>**<u>Reauthorization</u>** requires documentation of treatment success defined as reduction in UPCR or proteinuria from baseline<br><br>**<u>C3G</u>**<br>• Documented inadequate response to all the following:<br>  ○ Maximally tolerated renin-angiotensin system (RAS) inhibitor<br>  ○ Mycophenolate mofetil or mycophenolate sodium<br><br>**<u>IC-MPGN (Empaveli)</u>**<br>• Documented inadequate response to all of the following:<br>  ○ Maximally tolerated renin-angiotensin system (RAS) inhibitor<br>  ○ Mycophenolate mofetil or mycophenolate sodium<br><br>**<u>Reauthorization</u>** requires documentation of treatment success defined as reduction in UPCR or proteinuria from baseline |
| **Exclusion Criteria:** | • Concurrent use with other biologics for PNH (Soliris, Ultomiris, Empaveli, or Fabhalta) except when cross tapering according to FDA approved dosing<br>• Current meningitis infection or other unresolved serious infection caused by encapsulated bacteria<br>• Fabhalta should not be administered in combination with other medications indicated for immunoglobulin A nephropathy due to lack of clinical trial data supporting additive efficacy |
| **Age Restriction:** | • **PNH**: 18 years of age and older<br>• **C3G**:<br>  ○ Fabhalta: 18 years of age and older<br>  ○ Empaveli: 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist or a nephrologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PRIMARY BILIARY CHOLANGITIS AGENTS**
Affected Medications: OCALIVA (obeticholic acid), IQIRVO (elafibranor), LIVDELZI (seladelpar)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Primary biliary cholangitis (PBC) |
| **Required Medical Information:** | • Liver function tests (including alkaline phosphatase and bilirubin)<br>• Child-Pugh score |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation that after at least 12 months of adherent therapy with ursodiol or clinical inability to tolerate ursodiol, the patient has **ONE** of the following:<br>  ○ Alkaline phosphatase level (ALP) at least 1.67 times the upper limit of normal (ULN) of the reference lab<br>  ○ Total bilirubin above the ULN of the reference lab<br><br>**Reauthorization** will require documentation of treatment success defined as a significant reduction in alkaline phosphatase (ALP) and/or bilirubin levels |
| **Exclusion Criteria:** | • Complete biliary obstruction<br>• Decompensated cirrhosis (e.g., Child-Pugh Class B or C) or a prior decompensation event<br>• For Ocaliva: Compensated cirrhosis with evidence of portal hypertension (e.g., ascites, gastroesophageal varices, persistent thrombocytopenia)<br>• Use in combination with another drug on this policy (Ocaliva, Iqirvo, Livdelzi) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hepatologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PYRIMETHAMINE**
Affected Medications: Daraprim, pyrimethamine

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Toxoplasmosis |
| **Required Medical Information:** | • Documentation of recent *Toxoplasma* infection<br>• Documentation of one of the following:<br>  ○ Severe symptoms (pneumonitis, myocarditis, etc) or prolonged symptoms greater than 4 weeks with significant impact on quality of life<br>  ○ Immunocompromised status |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing Regimen (adult):<br>  ○ Day 1: Pyrimethamine 100 mg, sulfadiazine 2-4 gm divided four times daily, leucovorin 5-25 mg<br>  ○ Day 2: Pyrimethamine 25-50 mg, sulfadiazine 2-4 gm divided four times daily, leucovorin 5-25 mg<br>  ○ Day 3 and beyond: Pyrimethamine 25-50 mg, sulfadiazine 500 mg-1 gm divided four times daily, leucovorin 5-25 mg |
| **Exclusion Criteria:** | • Treatment regimen does not contain leucovorin and a sulfonamide (or alternative if allergic to sulfa) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: Up to 6 weeks, with no reauthorization unless otherwise specified |

POLICY NAME:
**RAVICTI**
Affected Medications: RAVICTI (glycerol phenylbutyrate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Chronic management of patients with urea cycle disorders (UCDs) who cannot be managed by dietary protein restriction and/or amino acid supplementation alone |
| **Required Medical Information:** | • Diagnosis confirmed by enzymatic, biochemical, or genetic testing |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with dietary protein restriction and/or amino acid supplementation alone<br>• For UCDs involving deficiencies in carbamoylphosphate synthetase (CPS), ornithine transcarbamylase (OTC), or argininosuccinic acid synthetase (AS), documentation of ONE of the following:<br>  ○ Treatment failure (or intolerable adverse event) with sodium phenylbutyrate<br>  ○ Comorbid condition with high risk of sodium-induced fluid retention (such as heart failure, renal impairment, edema)<br>• Must be used in combination with dietary protein restriction<br><br>**Reauthorization** will require **BOTH** of the following:<br>• Documented treatment success defined as ammonia levels maintained within normal limits<br>• That this drug continues to be used in combination with dietary protein restriction |
| **Exclusion Criteria:** | • Known hypersensitivity to phenylbutyrate<br>• Use for treatment of acute hyperammonemia or N-acetylglutamate synthase (NAGS) deficiency |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist experienced in the treatment of metabolic diseases<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**RAVULIZUMAB-CWVZ**
Affected Medications: ULTOMIRIS (ravulizumab-cwvz)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis<br>  ○ Atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy<br>  ○ Generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive<br>  ○ Neuromyelitis optica spectrum disorder (NMOSD) who are anti-aquaporin-4 (AQP4) antibody positive for adult patients |
| **Required Medical Information:** | **PNH**<br>• Detection of PNH clones of at least 5% by flow cytometry diagnostic testing<br>  ○ Presence of at least 2 different glycosylphosphatidylinositol (GPI) protein deficiencies (e.g., CD55, CD59, etc.) within at least 2 different cell lines (e.g., granulocytes, monocytes, erythrocytes)<br>• Baseline lactate dehydrogenase (LDH) levels greater than or equal to 1.5 times the upper limit of normal range.<br>• One of the following PNH-associated clinical findings:<br>  ○ Presence of a thrombotic event<br>  ○ Presence of organ damage secondary to chronic hemolysis<br>  ○ History of 4 or more blood transfusions required in the previous 12 months<br><br>**aHUS**<br>• Clinical presentation of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury<br>• Patient shows signs of thrombotic microangiopathy (TMA) (e.g., changes in mental status, seizures, angina, dyspnea, thrombosis, increasing blood pressure, decreased platelet count, increased serum creatinine, increased LDH, etc.)<br>• ADAMTS13 activity level greater than or equal to 10%<br>• Shiga toxin E. coli related hemolytic uremic syndrome (ST-HUS) has been ruled out<br>• History of 4 or more blood transfusions required in the previous 12 months<br><br>**gMG**<br>• Diagnosis of gMG confirmed by **ONE** of the following:<br>  ○ A history of abnormal neuromuscular transmission test<br>  ○ A positive edrophonium chloride test<br>  ○ Improvement in gMG signs or symptoms with an acetylcholinesterase inhibitor<br>• Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV<br>• Positive serologic test for AChR antibodies<br>• Documentation of **ONE** of the following:<br>  ○ MG-Activities of Daily Living (MG-ADL) total score of 6 or greater<br>  ○ Quantitative Myasthenia Gravis (QMG) total score of 12 or greater<br><br>**NMOSD**<br>• Diagnosis of NMOSD with aquaporin-4 immunoglobulin G (AQP4- IgG) antibody positive disease confirmed by all of the following:<br>  ○ Documentation of positive test for AQP4-IgG antibodies via cell-based assay |

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<tr><td></td><td>
<ul>
<li>Exclusion of alternative diagnoses (such as multiple sclerosis)</li>
<li>At least ONE core clinical characteristic:
<ul>
<li>Acute optic neuritis</li>
<li>Acute myelitis</li>
<li>Area postrema syndrome (episode of otherwise unexplained hiccups or nausea/vomiting)</li>
<li>Acute brainstem syndrome</li>
<li>Symptomatic narcolepsy OR acute diencephalic clinical syndrome with NMSOD-typical diencephalic MRI lesions</li>
<li>Symptomatic cerebral syndrome with NMOSD-typical lesion on magnetic resonance imaging (MRI) [see table below]</li>
<li>Acute cerebral syndrome with NMOSD-typical brain lesion on MRI [see table below]</li>
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</ul>
<table>
<tr><th>Clinical presentation</th><th>Possible MRI findings</th></tr>
<tr><td>Diencephalic syndrome</td><td>• Periependymal lesion<br>• Hypothalamic/thalamic lesion</td></tr>
<tr><td>Acute cerebral syndrome</td><td>• Extensive periependymal lesion<br>• Long, diffuse, heterogenous, or edematous corpus callosum lesion<br>• Long corticospinal tract lesion<br>• Large, confluent subcortical or deep white matter lesion</td></tr>
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</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td>
<u>aHUS</u>
<ul>
<li>Failure to respond to plasma therapy within 10 days
<ul>
<li>Trial of plasma therapy not required if one of the following is present:
<ul>
<li>Life-threatening complications of HUS such as seizures, coma, or heart failure</li>
<li>Confirmed presence of a high-risk complement genetic variant (e.g., CFH or CFI)</li>
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<u>gMG</u>
<ul>
<li>Documentation of one of the following:
<ul>
<li>Treatment failure with an adequate trial (one year or more) of at least 2 immunosuppressive therapies (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)</li>
<li>Has required three or more courses of rescue therapy (plasmapheresis/plasma exchange and/or intravenous immunoglobulin), while on at least one immunosuppressive therapy, over the last 12 months</li>
</ul></li>
<li>Documented inadequate response, contraindication, or intolerance to efgartigimod-alfa (Vyvgart)</li>
</ul>
<u>NMOSD</u>
<ul>
<li>Documented inadequate response, contraindication, or intolerance to ALL of the following:
<ul>
<li>Rituximab (preferred products: Riabni, Ruxience)</li>
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| | ○ Satralizumab-mwge (Enspryng)<br>○ Inebilizumab-cdon (Uplizna)<br><br>**Reauthorization** requires:<br>• gMG: documentation of treatment success defined as an improvement in MG-ADL and QMG scores from baseline<br>• PNH: documentation of treatment success defined as a decrease in serum LDH, stabilized/improved hemoglobin, decreased transfusion requirement, and reduction in thromboembolic events compared to baseline<br>• aHUS: documentation of treatment success defined as a decrease in serum LDH, stabilized/improved serum creatinine, increased platelet count, and decreased plasma exchange/infusion requirement compared to baseline<br>• NMOSD: documentation of treatment success defined as the stabilization or improvement in neurological symptoms as evidenced by a decrease in acute relapses, Expanded Disability Status Scale (EDSS) score, hospitalizations, or plasma exchange treatments |
| **Exclusion Criteria:** | • Current meningitis infection<br>• Concurrent use with other disease-modifying biologics for requested indication, unless otherwise specified |
| **Age Restriction:** | • PNH, aHUS: 1 month of age and older<br>• gMG: 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist<br>○ PNH: hematologist<br>○ aHUS: hematologist or nephrologist<br>○ gMG: neurologist<br>○ NMOSD: neurologist or neuro-ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**RELYVRIO**
Affected Medications: RELYVRIO (sodium phenylbutyrate-taurursodiol)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Amyotrophic lateral sclerosis (ALS) |
| **Required Medical Information:** | • Definite or probable Amyotrophic lateral sclerosis (ALS) based on El Escorial revised (Airlie House) criteria<br>• Symptom onset within 18 months<br>• Slow vital capacity (SVC) of at least 60 percent<br>• Patient currently retains most activities of daily living defined as at least 2 points on all 12 items of the ALS functional rating scale-revised (ALSFRS-R) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of one of the following:<br>  ○ Member is stable on riluzole<br>  ○ Prescriber has indicated clinical inappropriateness of riluzole<br><br>**Reauthorization**: Documentation of treatment success as determined by prescriber including retaining most activities of daily living |
| **Exclusion Criteria:** | • Presence of a tracheostomy<br>• Use of permanent assisted ventilation |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**REMESTEMCEL**
Affected Medications: RYONCIL (remestemcel-L-rknd)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>• Compendia-supported uses that will be covered (if applicable) |
| **Required Medical Information:** | • Diagnosis of grade B through D acute graft-versus-host disease; (aGVHD) with symptoms involving skin, liver, and/or GI tract<br>• Steroid resistance defined as consecutive treatment with 2 mg/kg/day of methylprednisolone (or equivalent) resulting in:<br>  ○ Progression within 3 days<br>  OR<br>  ○ No improvement in 7 days |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure or intolerance to:<br>  ○ At least one other second-line therapy (such as calcineurin inhibitors, mycophenolate, everolimus, sirolimus, etanercept, infliximab, anti-thymocyte globulin, extracorporeal photopheresis)<br>  ○ Jakafi (if 12 years of age and older)<br><br>**<u>Reauthorization</u>:**<br>• Partial Response (PR) defined as organ improvement of at least 1 stage without worsening of any other organ<br>  OR<br>• Mixed Response (MR) defined as improvement in at least 1 evaluable organ stage with worsening in another<br>  OR<br>• aGVHD flare defined as grade B through D progression after achieving initial complete response (CR)<br>  AND<br>• Documentation showing symptom improvement while on therapy |
| **Exclusion Criteria:** | • Grade B acute graft-versus-host disease (aGVHD) involving skin only |
| **Age Restriction:** | • 2 months to 17 years of age |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist, hematologist, bone marrow transplant specialist, or specialist experienced in the treatment of aGVHD<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 weeks, unless otherwise specified<br>• Reauthorization: 4 weeks, unless otherwise specified |

POLICY NAME:
**REMODULIN**
Affected Medications: REMODULIN INJECTION (treprostinil)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1<br>  ○ Pulmonary Arterial Hypertension in patients requiring transition from epoprostenol |
| **Required Medical Information:** | **<u>Pulmonary Arterial Hypertension (PAH) WHO Group 1</u>**<br>• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:<br>  ○ Mean pulmonary artery pressure of at least 20 mm Hg<br>  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg<br>  ○ Pulmonary vascular resistance of at least 2.0 Wood units<br>• Etiology of PAH: idiopathic PAH, hereditary PAH, OR<br>• PAH secondary to one of the following conditions:<br>  ○ Connective tissue disease<br>  ○ Human immunodeficiency virus (HIV) infection<br>  ○ Cirrhosis<br>  ○ Anorexigens<br>  ○ Congenital left to right shunts<br>  ○ Schistosomiasis<br>  ○ Drugs and toxins<br>  ○ Portal hypertension<br>• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class II or higher symptoms<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blockers) unless there are contraindications:<br>  ○ Low systemic blood pressure (systolic blood pressure less than 90)<br>  ○ Low cardiac index<br>  OR<br>  ○ Presence of severe symptoms (functional class IV) |
| **Appropriate Treatment Regimen & Other Criteria:** | • The pulmonary hypertension has progressed despite maximal medical and/or surgical treatment of the identified condition<br>• Documentation that treprostinil is used as a single route of administration (Remodulin, Tyvaso, Orenitram should not be used in combination)<br>• Treatment with oral calcium channel blocking agents has been tried and failed, or has been considered and ruled out<br>• Treatment with combination of endothelin receptor antagonist (ERA) and phosphodiesterase 5 inhibitor (PDE5I) has been tried and failed for WHO functional class II and III<br><br>**<u>Reauthorization</u>** requires documentation of treatment success defined as one or more of the following:<br>• Improvement in walking distance<br>• Improvement in exercise ability<br>• Improvement in pulmonary function<br>• Improvement or stability in WHO functional class |

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| **Exclusion Criteria:** | • PAH secondary to pulmonary venous hypertension (e.g., left sided atrial or ventricular disease, left sided valvular heart disease, etc) or disorders of the respiratory system (e.g., chronic obstructive pulmonary disease, interstitial lung disease, obstructive sleep apnea or other sleep disordered breathing, alveolar hypoventilation disorders, etc.) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**RESMETIROM**
Affected Medications: REZDIFFRA (resmetirom)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of adults with noncirrhotic nonalcoholic steatohepatitis (NASH) with moderate to advanced liver fibrosis (consistent with stages F2 to F3 fibrosis), in conjunction with diet and exercise |
| **Required Medical Information:** | • Diagnosis of NASH or metabolic dysfunction–associated steatohepatitis (MASH) with moderate to advanced (F2 to F3) liver fibrosis confirmed by **ONE** of the following:<br>  ○ Conclusive result from a well-validated non-invasive test such as:<br>  ▪ Fibroscan-AST (FAST) score<br>  ▪ MAST (score from MRI–proton density fat fraction, Magnetic resonance elastography [MRE], and serum AST)<br>  ▪ MEFIB (Fibrosis-4 Index ≥1.6 and MRE ≥3.3 kPa)<br>  ○ Liver biopsy (also required if non-invasive testing is inconclusive or other causes for liver disease have not been ruled out)<br>• Other causes for liver steatosis have been ruled out (such as alcohol-associated liver disease, chronic hepatitis C, Wilson disease, drug-induced liver disease)<br>• Baseline lab values for AST and ALT |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of abstinence from alcohol consumption<br>• Documentation of comprehensive comorbidity management being undertaken, including all the following:<br>  ○ Use of diet and exercise for weight management<br>  ○ Medications to manage associated comorbid conditions, such as thyroid disease (must not have active disease), diabetes, dyslipidemia, hypertension, or cardiovascular conditions<br>• Documented treatment failure or intolerable adverse event with a glucagon-like peptide-1 (GLP-1) receptor agonist<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy based on improvements or stability in laboratory results, such as ALT and AST, or fibrosis as evaluated by a non-invasive test |
| **Exclusion Criteria:** | • History of excessive alcohol use or alcohol-associated liver disease<br>• Current excessive alcohol use<br>• Continued use of medications associated with liver steatosis<br>• Stage 4 liver disease or cirrhosis<br>• Use for other liver disease<br>• Active or untreated thyroid disease |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hepatologist or gastroenterologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**RETHYMIC**
Affected Medications: RETHYMIC (allogeneic processed thymus tissue-agdc)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Immune reconstitution in pediatric patients with congenital athymia |
| **Required Medical Information:** | • Documentation of congenital athymia associated with one of the following:<br>  ○ Complete DiGeorge Syndrome (cDGS)<br>  ○ Forkhead Box N1 (*FOXN1*) deficiency<br>  ○ 22q11.2 deletion<br>  ○ CHARGE Syndrome (Coloboma, Heart defects, Atresia of the nasal choanae, Retardation of growth and development, Genitourinary anomalies, Ear anomalies)<br>  ○ CHD7 mutation<br>  ○ 10p13-p14 deletion |
| **Appropriate Treatment Regimen & Other Criteria:** | • Congenital athymia confirmed by flow cytometry that demonstrates:<br>  ○ Fewer than 50 naïve T cells/mm3 in the peripheral blood OR<br>  ○ Less than 5% of total T cells being naïve T cells |
| **Exclusion Criteria:** | • Treatment of patients with severe combined immunodeficiency (SCID)<br>• Prior thymus transplant |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a pediatric immunologist or prescriber experienced in the treatment of congenital athymia<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 1 month (1 treatment only), unless otherwise specified |

POLICY NAME:
**REVAKINAGENE TARORETCEL-LWEY**
Affected Medications: ENCELTO (revakinagene taroretcel-lwey intravitreal implant) - Available on Medical Benefit only

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Idiopathic macular telangiectasia type 2 (MacTel). |
| **Required Medical Information:** | • Documented diagnosis of MacTel type 2 with evidence of fluorescein leakage and at least one of these features:<br>  ○ Hyperpigmentation outside of a 500 micron radius from the center of the fovea, retinal opacification, crystalline deposits, right-angle vessels<br>• Inner Segment/Outer Segment (IS/OS) photoreceptor (PR) break/loss in ellipsoid zone (EZ) between 0.16 and 2 $mm^2$ measured by Spectral Domain Optical Coherence Tomography (SD-OCT)<br>• Best-corrected visual acuity (BCVA) score of 54 letters or better (20/80 Snellen equivalent) |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | • Evidence of neovascular MacTel type 2<br>• MacTel type 1 |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an ophthalmologist or surgeon<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 1 month (1 injection per eye per lifetime), unless otherwise specified |

POLICY NAME:
**RIBOCICLIB**
Affected Medications: KISQALI (ribociclib), KISQALI FEMARA (ribociclib/letrozole)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• Documentation of hormone receptor (HR)-positive, human epidermal growth factor receptor 2 (*HER2*)-negative breast cancer |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of early, high risk breast cancer with any lymph node involvement (excluding microscopic nodal involvement)<br>OR<br>• If no nodal involvement either:<br>○ Tumor size over 5 cm<br>OR<br>○ Tumor size from 2 cm to 5 cm and Grade 2 disease with high genomic risk or Ki-67 ≥ 20%<br>OR<br>○ Tumor size from 2 cm to 5 cm and Grade 3 disease<br>OR<br>• Documentation of intolerable adverse event with abemaciclib or palbociclib in treatment of HR-positive, *HER2*-negative advanced or metastatic breast cancer<br>AND<br>• Will be used in combination with an aromatase inhibitor or fulvestrant as recommended by the NCCN guidelines.<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br>• Maximum treatment duration for ribociclib in early, high risk breast cancer is 3 years. Reauthorization not allowed after 3 years of treatment |

POLICY NAME:
**RILONACEPT**
Affected Medications: ARCALYST (rilonacept)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome (FCAS), and Muckle-Wells Syndrome (MWS) in adults and pediatric patients 12 years and older<br>  ○ The maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in adults and pediatric patients weighing at least 10 kg<br>  ○ Treatment of recurrent pericarditis (RP) and reduction in risk of recurrence in adults and pediatric patients 12 years and older |
| **Required Medical Information:** | Documentation confirming one of the following:<br>• Diagnosis of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome (FCAS), and Muckle-Wells Syndrome (MWS)<br>• Diagnosis of Deficiency of Interleukin-1 Receptor Antagonist (DIRA)<br>  ○ Must include genetic testing results which confirm the presence of homozygous mutations in the interleukin-1 receptor antagonist (IL1RN) gene<br>  ○ Disease must currently be in remission<br>• Diagnosis of Recurrent Pericarditis with an inflammatory phenotype shown by one of the following:<br>  ○ Fever, elevated C-Reactive protein (CRP), elevated white blood cell count, elevated erythrocyte sedimentation rate (ESR), pericardial late gadolinium enhancement (LGE) on cardiac magnetic resonance (CMR), or pericardial contrast enhancement on computed tomography (CT) scan |
| **Appropriate Treatment Regimen & Other Criteria:** | **All Indications:**<br>• Documented treatment failure or intolerable adverse event with trial of Kineret (anakinra)<br><br>**Recurrent Pericarditis:**<br>• Documented treatment failure or intolerable adverse event to triple therapy with all of the following:<br>  ○ Colchicine<br>  ○ Non-steroidal anti-inflammatory (NSAID) or aspirin<br>  ○ Glucocorticoid<br><br>**Reauthorization:**<br>• **All indications:** documentation of treatment success and a clinically significant response to therapy<br>• **Recurrent pericarditis:** documentation that the patient is unable to remain asymptomatic with normal CRP levels upon trial of an appropriate tapering regimen |
| **Exclusion Criteria:** | • Active or chronic infection<br>• Concurrent therapy with anakinra, tumor necrosis factor (TNF) inhibitors, or other biologics |
| **Age Restriction:** | • CAPS or Recurrent Pericarditis: 12 years of age and older |

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| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a rheumatologist, immunologist, cardiologist, or dermatologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**RILZABRUTINIB**
Affected Medications: WAYRILZ (rilzabrutinib)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Immune thrombocytopenia, persistent or chronic**:** treatment of persistent or chronic immune thrombocytopenia in adults who have had an insufficient response to a previous treatment |
| **Required Medical Information:** | **Thrombocytopenia in patients with chronic ITP**<br>• Documentation of ONE of the following:<br>  ○ Platelet count less than 20,000/microliter<br>  ○ Platelet count less than 30,000/microliter AND symptomatic bleeding<br>  ○ Platelet count less than 50,000/microliter AND increased risk for bleeding (such as peptic ulcer disease, use of antiplatelets or anticoagulants, history of bleeding at higher platelet count, need for surgery or invasive procedure) |
| **Appropriate Treatment Regimen & Other Criteria:** | **Thrombocytopenia in patients with chronic ITP**<br>• Documentation of inadequate response, defined as platelets did not increase to at least 50,000/microliter, to the following therapies:<br>  ○ For those not status post splenectomy, documented treatment failure (defined as failure to achieve a platelet count of at least 50,000/mcL) with at least TWO therapies for immune thrombocytopenia, including corticosteroids, rituximab, or immunoglobulin<br>  OR<br>  ○ For those status post splenectomy, documented corticosteroid dependence (defined as inability to maintain a platelet count of at least 50,000/mcL without continuous use of a corticosteroid).<br>  AND<br>  ○ Documented treatment failure with eltrombopag olamine<br><br>**Reauthorization:**<br>• Response to treatment with platelet count of at least 50,000/microliter or above (not to exceed 400,000/microliter) |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months unless otherwise specified |

POLICY NAME:
**RIOCIGUAT**
Affected Medications: ADEMPAS (riociguat)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1<br>  ○ Chronic-Thromboembolic Pulmonary Hypertension (WHO Group 4) |
| **Required Medical Information:** | **<u>Chronic Thromboembolic Pulmonary Hypertension (CTEPH)</u>**<br>• Documentation of CTEPH (WHO Group 4) meeting the following criteria:<br>  ○ Evidence of thromboembolic occlusion of proximal or distal pulmonary vasculature on CT/MRI or V/Q scan<br>  ○ Mean pulmonary arterial pressure greater than 20 mm Hg<br>  ○ PAWP less than 15 mm Hg<br>  ○ Elevated pulmonary vascular resistance over 2 Wood units<br><br>**<u>Pulmonary Arterial Hypertension (PAH)</u>**<br>• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:<br>  ○ Mean pulmonary artery pressure of at least 20 mm Hg<br>  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg<br>  ○ Pulmonary vascular resistance of at least 2.0 Wood units<br>• Etiology of PAH (idiopathic, heritable, or associated with connective tissue disease)<br>• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class II or higher symptoms<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blocker) unless there are contraindications:<br>  ○ Low systemic blood pressure (systolic blood pressure less than 90)<br>  ○ Low cardiac index<br>  OR<br>  ○ Presence of severe symptoms (functional class IV) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>CTEPH</u>**<br>• Documentation of failure of or inability to receive pulmonary endarterectomy surgery<br>• Current therapy with anticoagulants<br><br>**<u>PAH</u>**<br>• Documented failure to the following therapy classes: Phosphodiesterase type 5 (PDE5) inhibitors **AND** endothelin receptor antagonists<br><br>**<u>Reauthorization</u>** requires documentation of treatment success defined as one or more of the following:<br>• Improvement in walking distance<br>• Improvement in exercise ability<br>• Improvement in pulmonary function<br>• Improvement or stability in WHO functional class |
| **Exclusion Criteria:** | • Concomitant use with nitrates or nitric oxide donors (such as amyl nitrite)<br>• Concomitant use with specific PDE-5 inhibitors (such as sildenafil, tadalafil, or vardenafil) or non-specific PDE inhibitors (such as dipyridamole or theophylline) |

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| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or a pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**RISDIPLAM**
Affected Medications: EVRYSDI (risdiplam)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Spinal muscular atrophy (SMA) |
| **Required Medical Information:** | • Diagnosis of SMA type 1, 2, or 3 confirmed by genetic testing of chromosome 5q13.2 demonstrating ONE of the following:<br>  ○ Homozygous gene deletion of SMN1 (survival motor neuron 1)<br>  ○ Homozygous gene mutation of SMN1<br>  ○ Compound heterozygous gene mutation of SMN1<br>• Documentation of 4 or fewer copies of the SMN2 (survival motor neuron 2) gene<br>• Documentation of one of the following baseline motor assessments appropriate for patient age and motor function:<br>  ○ Hammersmith Infant Neurological Examination (HINE-2)<br>  ○ Hammersmith Functional Motor Scale (HFSME)<br>  ○ Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)<br>  ○ Upper Limb Module (ULM) test<br>  ○ 6-Minute Walk Test (6MWT)<br>• Documentation of previous treatment history<br>• Documentation of ventilator use status:<br>  ○ Patient is NOT ventilator-dependent (defined as using a ventilator at least 16 hours per day on at least 21 of the last 30 days)<br>  ○ This does not apply to patients who require non-invasive ventilator assistance<br>• Patient weight and planned treatment regimen |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires documentation of improvement in baseline motor assessment score, clinically meaningful stabilization, or delayed progression of SMA-associated signs and symptoms |
| **Exclusion Criteria:** | • SMA type 4<br>• Advanced SMA at baseline (complete paralysis of limbs, permanent ventilation support)<br>• Prior treatment with SMA gene therapy (i.e., onasemnogene abeparvovec-xioi, onasemnogene abeparvovec-brve)<br>• Will not be used in combination with other agents for SMA (e.g., onasemnogene abeparvovec-xioi, onasemnogene abeparvovec-brve, nusinersen, etc.) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or provider who is experienced in treatment of spinal muscular atrophy<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

# RITUXIMAB

## Included Products

**Preferred Rituximab Products:** Riabni, Ruxience, Truxima
**Non-Preferred Rituximab Products:** Rituxan, Rituxan Hycela

# SCOPE & EXCLUSIONS

## Included Indications

All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. Drug compendia supported indications may be covered. Covered Food and Drug Administration (FDA)-approved and compendia-supported uses:

- Rheumatoid arthritis (RA)
- Microscopic Polyangiitis (MPA)
- Granulomatosis with Polyangiitis (GPA)
- Eosinophilic granulomatosis with polyangiitis (EGPA)
- Relapsing forms of multiple sclerosis (MS)
    - Clinically isolated syndrome (CIS)
    - Relapsing-remitting multiple sclerosis (RRMS)
    - Active secondary progressive disease (SPMS)
- Neuromyelitis Optica Spectrum Disorder (NMOSD)
- Pemphigus Vulgaris (PV) and other autoimmune blistering skin diseases (such as but not limited to pemphigus foliaceus, bullous pemphigoid, cicatricial pemphigoid, epidermolysis bullosa acquisita, and paraneoplastic pemphigus)
- Thrombocytopenia in patients with immune thrombocytopenia (ITP)
- NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher

## Exclusions

The requested product will not be given concurrently with:

- Anti-CD20-directed therapy or other disease-modifying medications indicated for the treatment of MS
- Targeted immune modulators

**Age Limits:** As defined by FDA labeling or compendia for the requested indication.

**Prescriber Limits:** Prescribed by, or in consultation with, appropriate specialist for indication.

## Other Limits:

- All approvals are subject to utilization of the most cost-effective site of care.
- Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced

# INITIAL AUTHORIZATION CRITERIA

## Required Medical Information:

1. All Indications must be FDA-supported for the requested product or strongly supported in drug compendia (ex. Lexi Drug, Drugdex)

2. Requested dosing must be according to the FDA label or compendia based on diagnosis, age, and weight.
3. Requests for non-preferred products require inadequate response, intolerance, or an FDA-labeled and patient-specific contraindication to all preferred products as outlined at the beginning of the policy and within indication-specific criteria.
    a. FDA-labeled contraindications are defined as those listed within the package insert.
    b. Allergic reactions may be accepted as contraindicated when an ingredient found only in the preferred product is established as the causative agent.

## Oncology

1. Please refer to the ONCOLOGY AGENTS policy for criteria.

## Rheumatoid Arthritis (RA)

1. Diagnosis of moderate-to-severe Rheumatoid Arthritis.
2. Documented failure with two of the preferred pharmacy drugs (Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Xeljanz, Rinvoq)
3. Dosing:
    - Initial: Approvable for up to 2 doses of 1,000 mg given 2 weeks apart.
    - Repeat course: Approvable for 16 weeks or more after the first dose of the previous rituximab regimen and the patient has responded (e.g., less joint pain, morning stiffness, or fatigue, or improved mobility, or decreased soft tissue swelling in joints or tendon sheaths) as determined by the prescribing physician.

## Relapsing Forms of Multiple Sclerosis (MS)

1. Diagnosis of MS
2. Dosing:
    - Initial: Approvable for one-time induction dose (e.g., 1,000 mg once every 2 weeks for 2 doses)
    - Maintenance: approvable for up to 2,000 mg annually. Higher doses will require documentation to support

## Microscopic Polyangiitis (MPA) or Granulomatosis with Polyangiitis (GPA)

1. Documentation of active GPA or MPA
2. Dosing:
    - Initial: Approvable for one-time induction dose (e.g., 1,000 mg once every 2 weeks for 2 doses **or** 375 mg/m2 once weekly for 4 doses), to be used in combination with a systemic glucocorticoid
    - Maintenance: Approvable for up to 1,000 mg annually. Higher doses will require supportive documentation (e.g., positive ANCA titers, detection of CD19+ lymphocytes)

## Eosinophilic Granulomatosis with Polyangiitis (EGPA)

1. Documented diagnosis of active EGPA confirmed by:
    - Eosinophilia at baseline (blood eosinophil level over 10% or absolute count over 1,000 cells/mcL)
    - At least **TWO** of the following:
        - Asthma
        - Histopathological evidence of eosinophilic vasculitis, perivascular eosinophilic infiltration, or eosinophil-rich granulomatous inflammation
        - Peripheral neuropathy (not due to radiculopathy)
        - Pulmonary infiltrates
        - Sinonasal abnormality/obstruction
        - Cardiomyopathy (confirmed on imaging)

- Glomerulonephritis
- Alveolar hemorrhage
- Palpable purpura
- Antineutrophil cytoplasmic antibody (ANCA) positive (anti-MPO-ANCA or anti-PR3-ANCA)

2. **Non-severe disease** (respiratory/sinonasal disease, uncomplicated skin manifestations, arthralgias, mild systemic symptoms, etc.): Documented relapsed or refractory disease with systemic glucocorticoids **AND** one immunosuppressive therapy (azathioprine, methotrexate, mycophenolate)

## Neuromyelitis Optica Spectrum Disorder (NMOSD)

1. Diagnosis of seropositive aquaporin-4 immunoglobulin G (AQP4-IgG) NMOSD confirmed by all the following:
   - Documentation of AQP4-IgG-specific antibodies on cell-based assay
   - Exclusion of alternative diagnoses (such as multiple sclerosis)
   - At least **one** core clinical characteristic:
     - Acute optic neuritis
     - Acute myelitis
     - Acute area postrema syndrome (episode of otherwise unexplained hiccups or nausea/vomiting)
     - Acute brainstem syndrome
     - Symptomatic narcolepsy **OR** acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesion on magnetic resonance imaging (MRI) [*see table below*]
     - Acute cerebral syndrome with NMOSD-typical brain lesion on MRI [*see table below*]

| Clinical presentation | Possible MRI findings |
|---|---|
| Diencephalic syndrome | • Periependymal lesion<br>• Hypothalamic/thalamic lesion |
| Acute cerebral syndrome | • Extensive periependymal lesion<br>• Long, diffuse, heterogenous, or edematous corpus callosum lesion<br>• Long corticospinal tract lesion<br>• Large, confluent subcortical or deep white matter lesion |

2. Dosing:
   - Initial: Approvable for one-time induction dose (e.g., 1,000 mg once every 2 weeks for 2 doses)
   - Maintenance: Approvable up to 2,000 mg annually. Higher doses will require documentation to support (e.g., detection of CD19+ lymphocytes)

## Pemphigus Vulgaris (PV) and Other Autoimmune Blistering Skin Diseases

1. Diagnosis confirmed by biopsy
2. Documented severe or refractory disease with failure to conventional topical and oral systemic therapies
3. Documented treatment failure with 12 weeks of a corticosteroid AND
4. Documented treatment failure with 12 weeks of an immunosuppressant at an adequate dose (e.g., azathioprine, mycophenolate, methotrexate, etc.) or other appropriate corticosteroid-sparing therapy

## Thrombocytopenia in Patients with immune thrombocytopenia (ITP)

1. Platelet count less than 20,000/mcL
2. One of the following:

- Documented steroid dependence to maintain platelets/prevent bleeding for at least 3 months
- Lack of clinically meaningful response to corticosteroids (defined as inability to increase platelets to at least 50,000/mcL)

## RENEWAL CRITERIA

Documentation of disease responsiveness to therapy.

## DURATION OF APPROVAL

### Initial:

- PV, MPA, GPA, EGPA – 3 months, unless otherwise specified
- RA, MS, NMOSD – 6 months, unless otherwise specified

**Renewal:** Reauthorization will be approved until no longer eligible with the plan; subject to formulary or benefit changes.

POLICY NAME:
**RNA INTERFERENCE DRUGS FOR PRIMARY HYPEROXALURIA 1**
Affected Medications: OXLUMO (lumasiran), RIVFLOZA (nedosiran)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Primary hyperoxaluria type 1 (PH1) |
| **Required Medical Information:** | • A diagnosis of primary hyperoxaluria type 1 (PH1) confirmed by genetic testing confirming presence of AGXT gene mutation<br>• Metabolic testing demonstrating elevated urinary **oxalate** excretion<br>• Presence of clinical manifestations diagnostic of PH1 such as:<br>  ○ Metabolic testing demonstrating elevated urinary **glycolate** excretion<br>  ○ Normal levels of L-glyceric acid (elevation indicates PH type 2)<br>  ○ Normal levels of hydroxy-oxo-glutarate (elevation indicates PH type 3)<br>• For Rivfloza: eGFR of 30 or more |
| **Appropriate Treatment Regimen & Other Criteria:** | • For Rivfloza: Trial and failure or contraindication with Oxlumo<br><br>**Reauthorization** requires documentation of the following criteria related to treatment success:<br>• Reduction from baseline in urine or plasma oxalate levels<br>• Improvement, stabilization, or slowed worsening of one or more clinical manifestation of PH1 (i.e., nephrocalcinosis, renal stone events, renal impairment, systemic oxalosis) |
| **Exclusion Criteria:** | • Diagnosis of primary hyperoxaluria type 2 or type 3<br>• Secondary hyperoxaluria<br>• Concurrent use of another RNA interference drug for PH1 |
| **Age Restriction** | • For Rivfloza: age in accordance with FDA labeling |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a nephrologist, urologist, geneticist, or specialist in the treatment of PH1<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ROMIPLOSTIM**
Affected Medications: NPLATE (romiplostim)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Adult patients with immune thrombocytopenia (ITP) who have had an insufficient response to corticosteroids, immunoglobulins, or splenectomy<br>  ○ Pediatric patients 1 year of age and older with ITP for at least 6 months who have had an insufficient response to corticosteroids, immunoglobulins, or splenectomy<br>  ○ Adult and pediatric patients (including term neonates) with acute exposure to myelosuppressive radiation doses |
| **Required Medical Information:** | **Thrombocytopenia in patients with ITP**<br>• Documentation of **ONE** of the following:<br>  ○ Platelet count less than 20,000/microliter<br>  ○ Platelet count less than 30,000/microliter AND symptomatic bleeding<br>  ○ Platelet count less than 50,000/microliter AND increased risk for bleeding (such as peptic ulcer disease, use of antiplatelets or anticoagulants, history of bleeding at higher platelet count, need for surgery or invasive procedure)<br><br>**Hematopoietic syndrome of acute radiation syndrome**<br>• Suspected or confirmed exposure to radiation levels greater than 2 gray (Gy) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Current weight<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Thrombocytopenia in patients with ITP**<br>• Documentation of inadequate response, defined as platelets did not increase to at least 50,000/microliter, to the following therapies:<br>  ○ **ONE** of the following:<br>  ▪ Inadequate response with at least 2 therapies for immune thrombocytopenia, including corticosteroids, rituximab, or immunoglobulin<br>  ▪ Splenectomy<br>  ○ Eltrombopag olamine<br><br>**Reauthorization (ITP only):**<br>• Response to treatment with platelet count of at least 50,000/microliter (not to exceed 400,000/microliter)<br>**OR**<br>• The platelet counts have not increased to a level of at least 50,000/microliter and member has NOT been on the maximum dose for at least 4 weeks<br><br>**Hematopoietic syndrome of acute radiation syndrome**<br>• Approved for one-time single subcutaneous injection of 10 mcg/kg |
| **Exclusion Criteria:** | • Treatment of thrombocytopenia due to myelodysplastic syndrome (MDS)<br>• Use in combination with another thrombopoietin receptor agonist, spleen tyrosine kinase inhibitor, or similar treatments (Eltrombopag olamine, Nplate, Tavalisse) |
| **Age Restriction:** | |

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| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **Thrombocytopenia in patients with ITP**<br>• Initial Approval: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br><br>**Hematopoietic syndrome of acute radiation syndrome**<br>• Authorization: 1 month, unless otherwise specified |

POLICY NAME:
**ROMOSOZUMAB**
Affected Medications: EVENITY (romosozumab-aqqg)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of osteoporosis in postmenopausal women at high risk of fracture (defined as history of osteoporotic fracture or multiple risk factors for fracture) or have history of treatment failure or intolerance to other available osteoporosis therapy |
| **Required Medical Information:** | • Diagnosis of osteoporosis as defined by at least **one** of the following:<br>  ○ T-score less than or equal to –2.5 (current or past) at the lumbar spine, femoral neck, total hip, or 1/3 radius site<br>  ○ T-score between –1.0 and –2.5 at the lumbar spine, femoral neck, total hip, or 1/3 radius site **AND** increased risk of fracture as defined by at least one of the following Fracture Risk Assessment Tool (FRAX) scores:<br>  ▪ FRAX 10-year probability of major osteoporotic fracture is 20% or greater<br>  ▪ FRAX 10-year probability of hip fracture is 3% or greater<br>  ○ History of non-traumatic fractures in the absence of other metabolic bone disorders |
| **Appropriate Treatment Regimen & Other Criteria:** | • Treatment failure, contraindication, or intolerance to all of the following:<br>  ○ Intravenous bisphosphonate (zoledronic acid or ibandronate)<br>  ○ Prolia (denosumab)<br><br>**<u>Total duration of therapy with Evenity should not exceed 12 months in a lifetime</u>** |
| **Exclusion Criteria:** | • Heart attack or stroke event within the preceding year<br>• Concurrent use of bisphosphonates, parathyroid hormone analogs, or RANK ligand inhibitors<br>• Hypocalcemia that is uncorrected prior to initiating Evenity |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months (no reauthorization), unless otherwise specified |

POLICY NAME:
**RUFINAMIDE**
Affected Medications: BANZEL (rufinamide), RUFINAMIDE SUSPENSION, RUFINAMIDE TABLET

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Lennox-Gastaut Syndrome(LGS) |
| **Required Medical Information:** | **<u>All Indications</u>**<br>• Patient weight<br>• Documentation that rufinamide will be used as adjunctive therapy<br><br>**<u>Lennox-Gastaut Syndrome (LGS)</u>**<br>• Documentation of at least 8 drop seizures per month while on stable antiepileptic drug therapy<br>• Documented treatment and inadequate seizure control with at least three guideline directed therapies including:<br>  ○ Valproate **and**<br>  ○ Lamotrigine **and**<br>  ○ Topiramate, felbamate, **or** clobazam |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing: not to exceed 3200 mg daily<br><br>**<u>Reauthorization</u>** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Familial Short QT syndrome<br>• Use as monotherapy for seizure control |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**RYPLAZIM**
Affected Medications: RYPLAZIM

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Plasminogen Deficiency Type 1 |
| **Required Medical Information:** | • Diagnosis of symptomatic congenital plasminogen deficiency (C-PLGD) type 1, as evidenced by documentation of all of the following:<br>  ○ Clinical signs and symptoms of the disease (such as ligneous conjunctivitis, gingivitis, tonsillitis, abnormal wound healing)<br>  ○ Presence of (ligneous) pseudomembranous lesions with documentation of size, location, and total number of lesions<br>  ○ Baseline plasminogen activity level less than or equal to 45% of laboratory standard |
| **Appropriate Treatment Regimen & Other Criteria:** | **Dosing**<br>• Dosing may not exceed 6.6 mg/kg every 2 days.<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced.<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy, defined as the following:<br>• Trough plasminogen activity level (taken 72 hours after dose) increased by 10% or greater above baseline<br>• Improvement (reduction) in lesion number/size from baseline |
| **Exclusion Criteria:** | • Prior treatment failure with Ryplazim<br>• Treatment of idiopathic pulmonary fibrosis |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SACROSIDASE**
Affected Medications: SUCRAID (sacrosidase)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Oral replacement therapy for congenital sucrase-isomaltase deficiency (CSID) |
| **Required Medical Information:** | • Documentation of confirmed congenital sucrose-isomaltase deficiency, diagnosed by one of the following:<br>  ○ Small bowel biopsy<br>  ○ Sucrose breath test<br>  ○ Genetic test<br>• Documentation of current symptoms (e.g., diarrhea, abdominal pain or cramping, bloating, gas, loose stools, nausea, vomiting)<br><br>**Reauthorization:** requires documentation of treatment success and a clinically significant response to therapy (fewer stools, lower number of symptoms) |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 5 months of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a gastroenterologist or genetic specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SAPROPTERIN**
Affected Medications: KUVAN (sapropterin), SAPROPTERIN

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Reduce phenylalanine (Phe) levels in those that are one month of age and older with phenylketonuria (PKU) |
| **Required Medical Information:** | • Documentation of a diagnosis of PKU<br>• Baseline (pre-treatment) blood Phe level greater than or equal to 360 micromol/L (6 mg/dL)<br>• Documentation of failure to Phe restricted diet as monotherapy |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of continuation on a Phe restricted diet<br><br>**Reauthorization** requires documentation of one of the following:<br>• Reduction in baseline Phe levels by 30 percent or levels maintained between 120 - 360 micromol/L (2 - 6 mg/dL)<br>• Increase in dietary Phe tolerance<br>• Improvement in clinical symptoms |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist in metabolic disorders or an endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 2 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SATRALIZUMAB-MWGE**
Affected Medications: ENSPRYNG (satralizumab-mwge)

<table>
<tr><td>Covered Uses:</td><td>• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Neuromyelitis optica spectrum disorder (NMOSD) in adults who are anti-aquaporin-4 (AQP4) antibody positive</td></tr>
<tr><td>Required Medical Information:</td><td><u>NMOSD</u><br>• Diagnosis of seropositive aquaporin-4 immunoglobulin G (AQP4-IgG) NMOSD confirmed by all the following:<br>  ○ Documentation of AQP4-IgG-specific antibodies on cell-based assay<br>  ○ Exclusion of alternative diagnoses (such as multiple sclerosis)<br>  ○ At least one core clinical characteristic:<br>  ▪ Acute optic neuritis<br>  ▪ Acute myelitis<br>  ▪ Acute area postrema syndrome (episode of otherwise unexplained hiccups or nausea/vomiting)<br>  ▪ Acute brainstem syndrome<br>  ▪ Symptomatic narcolepsy OR acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesion on magnetic resonance imaging (MRI) [see table below]<br>  ▪ Acute cerebral syndrome with NMOSD-typical brain lesion on MRI [see table below]<br><br><table><tr><th>Clinical presentation</th><th>Possible MRI findings</th></tr><tr><td>Diencephalic syndrome</td><td>• Periependymal lesion<br>• Hypothalamic/thalamic lesion</td></tr><tr><td>Acute cerebral syndrome</td><td>• Extensive periependymal lesion<br>• Long, diffuse, heterogenous, or edematous corpus callosum lesion<br>• Long corticospinal tract lesion<br>• Large, confluent subcortical or deep white matter lesion</td></tr></table><br>• History of at least 1 attack in the past year, or at least 2 attacks in the past 2 years, requiring rescue therapy</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td>• Documented inadequate response, contraindication, or intolerance to rituximab (preferred agents Riabni and Ruxience)<br><br><u>Reauthorization</u> requires documentation of treatment success</td></tr>
<tr><td>Exclusion Criteria:</td><td>• Active Hepatitis B Virus (HBV) infection<br>• Active or untreated latent tuberculosis<br>• Concurrent with other disease-modifying biologics for requested indication</td></tr>
<tr><td>Age Restriction:</td><td>• 18 years of age and older</td></tr>
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| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or neuro-ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SEBELIPASE ALFA**
Affected Medications: KANUMA (sebelipase alfa)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of Lysosomal Acid Lipase (LAL) deficiency |
| **Required Medical Information:** | • Diagnosis of LAL deficiency or Rapidly Progressive LAL deficiency within the first 6 months of life confirmed by one of the following:<br>  ○ Absence or deficiency in lysosomal acid lipase activity<br>  ○ Mutation in the lipase A, lysosomal acid type (*LIPA*) gene<br>• Documentation of patient weight<br>• Documentation of prescribed treatment regimen (dose and frequency)<br>• Baseline fasting lipid panel including LDL-c prior to initiating therapy (not required for Rapidly Progressive LAL deficiency) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization**:<br>• Rapidly Progressive LAL deficiency: documentation of improvement in weight-for-age Z-score<br>• LAL deficiency: documentation of improvement in LDL-c |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 1 month of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist or metabolic specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SELF-ADMINISTERED DRUGS (SAD)**
Affected Medications: Please refer to package insert for directions on self-administration.

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|---|---|
| **Covered Uses:** | |
| **Required Medical Information:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Appropriate Treatment Regimen & Other Criteria:** | • Pharmaceuticals covered under your pharmacy benefit are in place of, not in addition to, those same covered supplies under the medical plan. Please refer to your benefit book for more information. |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | |
| **Coverage Duration:** | |

POLICY NAME:
**SEROSTIM**
Affected Medications: SEROSTIM (somatropin)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ HIV (human immunodeficiency virus)-associated wasting, cachexia |
| **Required Medical Information:** | • Documentation of current body mass index (BMI), actual body weight, and ideal body weight (IBW)<br>• Serostim is used in combination with antiretroviral therapy to which the patient has documented compliance<br>• Alternative causes of wasting (e.g., inadequate nutrition intake, malabsorption, opportunistic infections, hypogonadism) have been ruled out or treated appropriately<br>• Prior to somatropin, patient had a suboptimal response to at least 1 other therapy for wasting or cachexia (e.g., megestrol, dronabinol, cyproheptadine, or testosterone therapy if hypogonadal) unless contraindicated or not tolerated<br>• Diagnosis of HIV-association wasting syndrome or cachexia confirmed by **one** of the following:<br>  ○ Unintentional weight loss greater than or equal to 10% of body weight over prior 12 months<br>  ○ Unintentional weight loss greater than or equal to 5% of body weight over prior 6 months<br>  ○ BMI less than 20 $kg/m^2$<br>  ○ Weight is less than 90% of IBW |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Reauthorization</u>:**<br>• Documentation of treatment success and clinically significant response to therapy (e.g., improved or stabilized BMI, increased physical endurance compared to baseline, etc.)<br>• Documentation of continued compliance to antiretroviral regimen |
| **Exclusion Criteria:** | • Acute critical illness due to complications following open heart or abdominal surgery, multiple accidental trauma or acute respiratory failure<br>• Active malignancy<br>• Acute respiratory failure<br>• Active proliferative or severe non-proliferative diabetic retinopathy |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an infectious disease specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 8 months (maximum duration of therapy 48 weeks total) |

**POLICY NAME:**
**SIBEPRENLIMAB-SZSI**
**Affected Medications:** Voyxact (sibeprenlimab-szsi)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ To reduce proteinuria in adults with primary immunoglobulin A nephropathy (IgAN) at risk for disease progression |
| **Required Medical Information:** | • Diagnosis of IgAN confirmed with biopsy<br>• Documentation of one of the following (with labs current within 30 days of request):<br>  ○ Proteinuria defined as 0.5 g/day or greater<br>  ○ UPCR greater than 0.75 g/g |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure (defined as proteinuria equal to or greater than 0.5 g/day OR UPCR greater than 0.75 g/g) with a minimum of 12 weeks of all the following:<br>  ○ Maximum tolerated dose of an angiotensin-converting enzyme (ACE) inhibitor or angiotensin receptor blocker (ARB)<br>  ○ Glucocorticoid therapy such as oral prednisone or methylprednisolone (or an adverse effect to two or more glucocorticoid therapies that is not associated with the corticosteroid class)<br><br>**Reauthorization** requires documentation of treatment success defined as reduction in UPCR or proteinuria from baseline. |
| **Exclusion Criteria:** | • The requested medication should not be administered in combination with other medications indicated for immunoglobulin A nephropathy due to lack of clinical trial data supporting additive efficacy (e.g. sibeprenlimab-szsi and sparsentan). |
| **Age Restriction:** | |
| **Prescriber Restrictions:** | • Prescribed by, or in consultation with, a nephrologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SIGNIFOR**
Affected Medications: SIGNIFOR (pasireotide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Cushing's disease |
| **Required Medical Information:** | • Documented diagnosis of Cushing's disease<br>• Documentation of at least **TWO** of the following:<br>  ○ Mean 24-hour urine free cortisol (mUFC) greater than 1.5 times the upper limit of normal (ULN) for the assay (at least two measurements)<br>  ○ Bedtime salivary cortisol greater than 145 ng/dL (at least two measurements)<br>  ○ Overnight dexamethasone suppression test (DST) with a serum cortisol greater than 1.8 mcg/dL |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure or intolerable adverse event to ketoconazole and cabergoline<br>• Documentation confirming pituitary surgery is not an option **OR** previous surgery has not been curative<br><br>**Reauthorization** requires documentation of treatment success defined as mUFC normalization (i.e., less than or equal to the ULN) |
| **Exclusion Criteria:** | • Severe hepatic impairment (Child Pugh C) |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**SIGNIFOR LAR**
Affected Medications: SIGNIFOR LAR (pasireotide)

| | |
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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Acromegaly<br>  ○ Cushing's disease |
| **Required Medical Information:** | **<u>Acromegaly</u>**<br>• Documentation confirming clinical manifestations of disease<br>• Diagnosis of acromegaly confirmed by **ONE** of the following:<br>  ○ Elevated pre-treatment serum insulin-like growth factor-1 (IGF-1) level for age/gender<br>  ○ Serum growth hormone (GH) level of 1 microgram/mL or greater after an oral glucose tolerance test (OGTT)<br><br>**<u>Cushing's Disease</u>**<br>• Documented diagnosis of Cushing's disease<br>• Documentation of at least **TWO** of the following:<br>  ○ Mean 24-hour urine free cortisol (mUFC) greater than 1.5 times the upper limit of normal (ULN) for the assay (at least two measurements)<br>  ○ Bedtime salivary cortisol greater than 145 ng/dL (at least two measurements)<br>  ○ Overnight dexamethasone suppression test (DST) with a serum cortisol greater than 1.8 mcg/dL |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Acromegaly</u>**<br>• Documented treatment failure or intolerance to **ONE** of the following: lanreotide (Somatuline Depot), Sandostatin LAR, or pegvisomant (Somavert)<br>• Documentation confirming **ONE** of the following:<br>  ○ Inadequate response to surgery or radiotherapy<br>  ○ Not a candidate for surgical management or radiotherapy (e.g., medically unstable, high risk for complications under anesthesia, major systemic complications of acromegaly, severe hypertension, uncontrolled diabetes, etc.)<br>• **Dosing:** Not to exceed 60 mg every 4 weeks (after 3 months of 40 mg)<br><br>**<u>Reauthorization</u>** requires documentation of treatment success shown by decreased/normalized IGF-1 or GH levels<br><br>**<u>Cushing's Disease</u>**<br>• Documentation confirming pituitary surgery is not an option **OR** previous surgery has not been curative<br>• Documented treatment failure or intolerance to ketoconazole and cabergoline<br>• **Dosing:** Not to exceed 40 mg every 4 weeks (after 4 months of 10 mg)<br><br>**<u>Reauthorization</u>** requires documentation of treatment success defined as mUFC normalization (i.e., less than or equal to the ULN) |

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| **Exclusion Criteria:** | • Severe hepatic impairment (Child Pugh C) |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SILTUXIMAB**
Affected Medications: SYLVANT (siltuximab)

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|---|---|
| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of patients with multicentric Castleman's disease (MCD) who are human immunodeficiency virus (HIV) negative and human herpesvirus-8 (HHV-8) negative<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• The diagnosis was confirmed by biopsy of lymph gland<br>• Documented negative tests for HIV and HHV-8<br>• Patient weight |
| **Appropriate Treatment Regimen & Other Criteria:** | **Dosing**<br>• **MCD:** 11 mg/kg intravenous (IV) infusion once every 3 weeks until treatment failure<br>• **Cytokine release syndrome (CRS):** 11 mg/kg IV one time only<br>• Availability: 100 mg and 400 mg vials<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • **MCD:**<br>  ○ Initial Authorization: 4 months, unless otherwise specified<br>  ○ Reauthorization: 12 months, unless otherwise specified<br>• **CRS:** 1 month (1 dose only), unless otherwise specified |

POLICY NAME:
**SODIUM PHENYLBUTYRATE**
Affected Medications: SODIUM PHENYLBUTYRATE

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Adjunctive therapy in the chronic management of patients with urea cycle disorders (UCDs) involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC), or argininosuccinic acid synthetase (AS)<br>  ○ Neonatal-onset deficiency (complete enzymatic deficiency, presenting within the first 28 days of life)<br>  ○ Late-onset disease (partial enzymatic deficiency, presenting after the first month of life) with history of hyperammonemic encephalopathy |
| **Required Medical Information:** | • Diagnosis confirmed by blood, enzymatic, biochemical, or genetic testing |
| **Appropriate Treatment Regimen & Other Criteria:** | • Oral tablets require documented inability to use sodium phenylbutyrate powder<br>• Documented treatment failure with dietary protein restriction and/or amino acid supplementation alone<br>• Must be used in combination with dietary protein restriction<br><br>**Reauthorization** will require **BOTH** of the following:<br><br>• Documentation of treatment success defined as ammonia levels maintained within normal limits<br>• That this drug continues to be used in combination with dietary protein restriction |
| **Exclusion Criteria:** | • Use for management of acute hyperammonemia |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist experienced in the treatment of metabolic diseases<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**SOLRIAMFETOL**
Affected Medications: SUNOSI (solriamfetol)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Excessive daytime sleepiness associated with narcolepsy<br>  ○ Excessive daytime sleepiness associated with obstructive sleep apnea |
| **Required Medical Information:** | **Narcolepsy**<br>• Diagnosis confirmed by polysomnography and multiple sleep latency test<br>• Symptoms of excessive daytime sleepiness consistent with narcolepsy have been present for at least 3 months<br>• An Epworth Sleepiness Scale score of more than 10 despite treatment<br><br>**Obstructive Sleep Apnea (OSA)**<br>• Diagnosis confirmed by sleep study<br>• An Epworth Sleepiness Scale score of more than 10 despite drug treatment and current use of continuous positive airway pressure (CPAP) for at least 3 months<br>• Documentation that CPAP use will be continued during treatment with solriamfetol<br><br>**All indications:**<br>• Documentation that other causes of sleepiness have been treated or ruled out (including but not limited to insufficient sleep syndrome, shift work, the effects of substances or medications, or other sleep disorders) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented trial and failure or contraindication to modafinil OR armodafinil<br>• For narcolepsy only, documented trial and failure or contraindication to ONE of the following: methylphenidate, dextroamphetamine, lisdexamfetamine, amphetamine-dextroamphetamine<br><br>**Reauthorization** requires clinically significant improvement in activities of daily living and in Epworth Sleepiness Scale score |
| **Exclusion Criteria:** | • Use for other untreated causes of sleepiness<br>• Concurrent use of sedative/hypnotic drugs or other central nervous system depressants |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a sleep specialist or neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SOMATOSTATIN ANALOGS**
Affected Medications: OCTREOTIDE, SANDOSTATIN LAR, LANREOTIDE, SOMATULINE DEPOT (lanreotide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br><br>**Octreotide, Sandostatin LAR:**<br>○ Acromegaly<br>○ Symptomatic treatment of metastatic carcinoid tumors (carcinoid syndrome)<br>○ Symptomatic treatment of vasoactive intestinal peptide tumors (VIPomas)<br><br>**Lanreotide, Somatuline Depot:**<br>○ Acromegaly<br>○ Carcinoid syndrome (to reduce the frequency of short-acting somatostatin analog rescue therapy)<br>○ Unresectable, well- or moderately-differentiated, locally advanced or metastatic gastroenteropancreatic neuroendocrine tumors (GEP-NETs)<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | **Acromegaly**<br>• Documentation confirming clinical manifestations of disease<br>• Diagnosis of acromegaly confirmed by **ONE** of the following:<br>○ Elevated pre-treatment serum insulin-like growth factor-1 (IGF-1) level for age/gender<br>○ Serum growth hormone (GH) level of 1 microgram/mL or greater after an oral glucose tolerance test (OGTT)<br><br>**All other indications**<br>Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | **Acromegaly**<br>• Documentation confirming **ONE** of the following:<br>○ Inadequate response to surgery or radiotherapy<br>○ Not a candidate for surgical management or radiotherapy (e.g., medically unstable, high risk for complications under anesthesia, major systemic complications of acromegaly, severe hypertension, uncontrolled diabetes, etc.)<br><br>**Sandostatin LAR**<br>• Coverage for the non-preferred product **Sandostatin LAR** is provided when **ONE** of the following criteria is met:<br>○ Currently receiving treatment with Sandostatin LAR, excluding when the product is obtained as samples or via manufacturer's patient assistance programs<br>○ Documented inadequate response or intolerable adverse event with one of the following: Lanreotide, Somatuline Depot, OR Somavert (Note: Somavert indicated for acromegaly only)<br><br>**Lanreotide, Somatuline Depot** |

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| | • GEP-NETs must use 120 mg injection<br><br>**<u>Reauthorization:</u>**<br>• **Acromegaly:** requires documentation of treatment success shown by decreased/normalized IGF-1 or GH levels<br>• **All other indications:** requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist, endocrinologist, or gastroenterologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SOMAVERT**
Affected Medications: SOMAVERT (pegvisomant)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Acromegaly |
| **Required Medical Information:** | • Documentation confirming clinical manifestations of disease<br>• Diagnosis of acromegaly confirmed by **ONE** of the following:<br>  ○ Elevated pre-treatment serum insulin-like growth factor-1 (IGF-1) level for age/gender<br>  ○ Serum growth hormone (GH) level of 1 microgram/mL or greater after an oral glucose tolerance test (OGTT) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure or intolerance to octreotide or lanreotide (Somatuline Depot)<br>• Documentation confirming one of the following:<br>  ○ Inadequate response to surgery or radiotherapy<br>  ○ Not a candidate for surgical management or radiotherapy (e.g., medically unstable, high risk for complications under anesthesia, major systemic complications of acromegaly, severe hypertension, uncontrolled diabetes, etc.)<br>• **Dosing:** Not to exceed 30 mg daily<br><br>**Reauthorization** requires documentation of treatment success shown by decreased/normalized IGF-1 or GH levels |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SOTATERCEPT-CSRK**
Affected Medications: WINREVAIR (sotatercept-csrk)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1 |
| **Required Medical Information:** | • Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:<br>  ○ Mean pulmonary artery pressure of at least 20 mm Hg<br>  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg<br>  ○ Pulmonary vascular resistance of at least 5 Wood units<br>  ○<br>• Etiology of PAH: idiopathic PAH, hereditary PAH<br>**OR**<br>• PAH secondary to one of the following conditions:<br>  ○ Connective tissue disease<br>  ○ Simple, congenital systemic to pulmonary shunts at least 1 year following repair<br>  ○ Drugs and toxins<br>• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class II or III symptoms<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blockers) unless there are contraindications:<br>  ○ Low systemic blood pressure (systolic blood pressure less than 90)<br>  ○ Low cardiac index (cardiac index less than 2 L/min/$m^2$)<br>**OR**<br>  ○ Presence of severe symptoms (functional class IV)<br>• Baseline 6-minute walk test (6MWD) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation that drug will be used as an add-on treatment with all the following (one from each category) at therapeutic doses for at least 90 days:<br>  ○ Phosphodiesterase-5 (PDE-5) inhibitor: sildenafil, tadalafil<br>  ○ Endothelin Receptor Antagonist: ambrisentan, bosentan, Opsumit<br>  ○ Prostacyclin: treprostinil, epoprostenol, Ventavis<br>• Documentation of inadequate response or intolerance to oral calcium channel blocking agents (nifedipine, diltiazem) if positive Acute Vasoreactivity Test<br><br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**<u>Reauthorization</u>** requires documentation of treatment success defined as one or more of the following:<br>• Improvement in walking distance (6MWD)<br>• Improvement or stability in WHO functional class |
| **Exclusion Criteria:** | • Human immunodeficiency virus (HIV)-associated PAH<br>• PAH associated with portal hypertension<br>• Schistosomiasis-associated PAH<br>• Pulmonary veno occlusive disease<br>• Platelet count less than 50,000/$mm^3$ (50 x $10^9$/L)<br>• Hemoglobin (Hgb) at screening above gender-specific upper limit of normal (ULN) |

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| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SPESOLIMAB**
Affected Medications: SPEVIGO INTRAVENOUS (IV) SOLUTION

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Generalized pustular psoriasis flares (GPP, also called von Zumbusch psoriasis) |
| **Required Medical Information:** | • Diagnosis of generalized pustular psoriasis as confirmed by the following:<br>  ○ The presence of widespread sterile pustules arising on erythematous skin<br>  ○ Pustulation is not restricted to psoriatic plaques<br>• Signs and symptoms of an acute GPP flare of moderate-to-severe intensity as follows:<br>  ○ A Generalized Pustular Psoriasis Physician Global Assessment (GPPGA) total score of greater than or equal to 3<br>  ○ A GPPGA pustulation category subscore of greater than or equal to 2<br>  ○ Greater than or equal to 5% body surface area (BSA) covered with erythema and the presence of pustules |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure of acute disease flare (or documented intolerable adverse event) with:<br>  ○ A one-week trial of cyclosporine<br>  AND<br>  ○ Infliximab (preferred biosimilars Inflectra, Renflexis)<br>• Treatment for each flare is limited to two 900 mg infusions of Spevigo separated by 1 week |
| **Exclusion Criteria:** | • Previous use of Spevigo<br>• Erythrodermic plaque psoriasis without pustules or with pustules restricted to psoriatic plaques<br>• Synovitis-acne-pustulosis-hyperostosis-osteitis syndrome<br>• Drug-induced acute generalized exanthematous pustulosis |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a dermatologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 1 month with no reauthorization, unless otherwise specified |

POLICY NAME:
**SPHINGOSINE 1-PHOSPHATE (S1P) RECEPTOR MODULATORS**
Affected Medications: MAYZENT (siponimod), PONVORY (ponesimod), VELSIPITY (etrasimod), ZEPOSIA (ozanimod)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following **(Mayzent, Ponvory, Zeposia)**:<br>  ▪ Clinically isolated syndrome (CIS)<br>  ▪ Relapsing-remitting multiple sclerosis (RRMS)<br>  ▪ Active secondary progressive disease (SPMS)<br>  ○ Ulcerative colitis (UC) **(Velsipity, Zeposia)** |
| **Required Medical Information:** | **MS**<br>• Diagnosis confirmed with magnetic resonance imaging (MRI) per revised McDonald diagnostic criteria for MS<br>  ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS<br><br>**UC**<br>• Diagnosis supported by endoscopy/colonoscopy/sigmoidoscopy or biopsy<br>• Documentation of moderate to severely active disease despite current treatment |
| **Appropriate Treatment Regimen & Other Criteria:** | **MS**<br>• Coverage of Mayzent (siponimod), Ponvory (ponesimod), or Zeposia (ozanimod) requires documentation of **ONE** of the following:<br>  ○ Treatment failure with (or intolerance to) **TWO** of the following: dimethyl fumarate, fingolimod, teriflunomide<br>  ○ Currently receiving treatment with Mayzent (siponimod), Ponvory (ponesimod), or Zeposia (ozanimod), excluding via samples or manufacturer's patient assistance program<br><br>**UC**<br>• Documented failure with at least two oral treatments for a minimum of 12 weeks each: corticosteroids, sulfasalazine, azathioprine, mesalamine, balsalazide, cyclosporine, 6-mercaptopurine<br>• Documented treatment failure with or intolerable adverse event with all preferred pharmacy drugs: Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Xeljanz, Yesintek and Selarsdi, Rinvoq, Skyrizi, Tremfya<br>• **Zeposia**: Documentation of treatment failure with (or intolerance to) Velsipity<br><br>**Reauthorization** requires provider attestation of treatment success |
| **Exclusion Criteria:** | • Mayzent: CYP2C9*3/*3 genotype<br>• Concurrent use of other disease-modifying medications indicated for the treatment of MS<br>• Concurrent use with a JAK inhibitor or biologic medication for the treatment of UC |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • MS: Prescribed by, or in consultation with, a neurologist or MS specialist |

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| | • UC: Prescribed by, or in consultation with, a gastroenterologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization:<br>  ○ UC: 6 months, unless otherwise specified<br>  ○ MS: 24 months, unless otherwise specified<br>• Reauthorization: 24 months, unless otherwise specified |

POLICY NAME:
**SPRAVATO**
Affected Medications: SPRAVATO (esketamine nasal spray)

<table>
<tr><td>Covered Uses:</td><td>
<ul>
<li>All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
<ul>
<li>Indicated for the treatment of:
<ul>
<li>Treatment-resistant depression (TRD) in adults</li>
<li>Depressive symptoms in adults with major depressive disorder (MDD) with acute suicidal ideation or behavior in conjunction with an oral antidepressant</li>
</ul></li>
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</td></tr>
<tr><td>Required Medical Information:</td><td>
<u>Diagnosis of Treatment-Resistant Depression (TRD)</u>
<ul>
<li>Assessment of patient’s risk for abuse or misuse</li>
<li>Baseline Patient Health Questionnaire-9 (PHQ-9) score (or other standard rating scale)</li>
<li>Inventory of Depressive Symptomatology-Clinician (IDS-C30) score of 34 or greater, PHQ-9 score of 15 or greater (or other standard rating scale) indicating moderate to severe depression</li>
</ul>
<u>Diagnosis of Major Depressive Disorder (MDD) with acute suicidal ideation or behavior:</u>
<ul>
<li>Assessment of patient’s risk for abuse or misuse</li>
<li>Montgomery-Asberg Depression Rating Scale (MADRS) total score greater than 28, PHQ-9 score of 15 or greater, or other standard rating scale indicating severe depression</li>
</ul>
</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td>
<u>Treatment-Resistant Depression:</u>
<ul>
<li>Documented treatment failure (defined by less than 50% improvement in depression symptom severity using a standard rating scale such as a PHQ-9) to an adequate trial (at least 6 weeks each), or intolerance, of at least three antidepressants from at least two different classes, during the current depressive episode</li>
<li>Failure to respond to augmentation therapy such as:
<ul>
<li>Two antidepressants with different mechanisms of action used concurrently</li>
<li>An antidepressant and a second-generation antipsychotic used concurrently</li>
<li>An antidepressant and lithium used concurrently</li>
<li>An antidepressant and buspirone used concurrently</li>
<li>An antidepressant and thyroid hormone used concurrently</li>
</ul></li>
<li>Failure to respond to evidence-based psychotherapy such as Cognitive Behavioral Therapy (CBT) and/or Interpersonal Therapy as documented by an objective scale such as a PHQ-9</li>
<li>Dosing according to the approved label:</li>
</ul>
<table>
<tr><td colspan="2"></td><td>Adults</td></tr>
<tr><td>Induction Phase</td><td><u>Weeks 1 to 4</u></td><td></td></tr>
<tr><td></td><td>Administer twice per week</td><td>56 mg or 84 mg</td></tr>
<tr><td>Maintenance Phase</td><td><u>Weeks 5 to 8</u></td><td></td></tr>
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|---|---|
| | <table><tr><td></td><td>Administer once weekly</td><td>56 mg or 84 mg</td></tr><tr><td></td><td><u>Weeks 9 and after</u></td><td></td></tr><tr><td></td><td>Administer every 2 weeks or once weekly*</td><td>56 mg or 84 mg</td></tr></table><br>*Dosing frequency should be individualized to the least frequent dosing to maintain remission/response<br><br><u>Reauthorization (for TRD indication only)</u> requires:<br>• Documentation of treatment success defined as at least a 50% reduction in symptoms of depression compared to baseline using a standard rating scale that measures depressive symptoms<br><br><u>Major depressive disorder (MDD) with acute suicidal ideation or behavior:</u><br>• Documentation of current inpatient psychiatric hospitalization OR adequate documentation of why patient is not currently at inpatient level of care<br>• Spravato will be used in combination with an oral antidepressant<br>• Dosing: 84 mg twice weekly for 4 weeks maximum (No reauthorization unless requirements for TRD met) |
| **Exclusion Criteria:** | • Concomitant psychotic disorder<br>• Bipolar or related disorders<br>• History of substance use disorder<br>• Use as an anesthetic agent<br>• Pregnancy<br>• Aneurysmal vascular disease (including thoracic and abdominal aorta, intracranial, and peripheral arterial vessels) or arteriovenous malformation<br>• History of intracerebral hemorrhage<br>• Hypersensitivity to esketamine, ketamine, or any of the excipients |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a psychiatrist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | <u>Initial Authorization</u>:<br>• Major depressive disorder (MDD) with acute suicidal ideation or behavior: 1 month (limit #24 nasal spray devices in 28 days of treatment only), unless otherwise specified<br>• TRD: 2 months (Induction phase – maximum of 23 nasal spray devices in first 28 days followed by once weekly maintenance phase), unless otherwise specified<br><u>Reauthorization</u>: (TRD indication only): 6 months, unless otherwise specified |

POLICY NAME:
**STIRIPENTOL**
Affected Medications: DIACOMIT (stiripentol)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of seizures associated with Dravet syndrome (DS) |
| **Required Medical Information:** | • Current Weight<br>• Documentation that therapy is being used as adjunct to clobazam for seizures<br>• Documentation of at least 4 generalized clonic or tonic-clonic seizures in the last month while on stable antiepileptic drug therapy |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment and inadequate control of seizures with at least four guideline directed therapies including:<br>  ○ Valproate<br>  ○ Clobazam<br>  ○ Topiramate<br>  ○ Clonazepam, levetiracetam, or zonisamide<br><br>**Reauthorization** will require documentation of treatment success and a reduction in seizure severity, frequency, or duration |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 6 months of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**STRENSIQ**
Affected Medications: STRENSIQ (asfotase alfa)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Perinatal/infantile or Juvenile onset hypophosphatasia (HPP) |
| **Required Medical Information:** | **<u>Diagnosis of Perinatal/Infantile or Juvenile onset hypophosphatasia (HPP) with ALL of the following:</u>**<br>• Age of onset less than 18 years<br>• One of the following:<br>  ○ Clinical manifestations consistent with hypophosphatasia at onset prior to age 18, such as: vitamin B6 dependent seizures, respiratory insufficiency, failure to thrive, non-traumatic fracture, dental abnormalities, low score on 6-minute walk test, low bone density score<br>  ○ Skeletal abnormalities confirmed with radiographic imaging (such as flared and frayed metaphyses, widened growth plate, bowed arms or legs, rachitic chest deformity, craniosynostosis)<br>• Genetic test confirming mutation of tissue-non-specific alkaline phosphatase (TNSALP) gene<br>• Low level of serum alkaline phosphatase (ALP) evidenced by lab result below reference range for patient's age and gender<br>• Elevated levels of one of the following:<br>  ○ Urine or serum concentration of phosphoethanolamine (PEA)<br>  ○ Serum concentration of pyridoxal 5'-phosphate (PLP) in the absence of vitamin supplements within one week prior to the test<br>  ○ Urinary inorganic pyrophosphate (PPi) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br>  ○ Please note: the 80 mg/0.8 mL vial is for patients weighing greater than 40 kilograms only<br><br>**<u>Reauthorization</u>** requires documentation of:<br>• Laboratory results confirming a decrease in urine concentration of urine or serum phosphoethanolamine (PEA), serum concentration of pyridoxal 5'-phosphate (PLP), or urinary inorganic pyrophosphate (PPi)<br>• Improvement or stabilization in the clinical signs and symptoms of hypophosphatasia, such as:<br>  ○ Radiographic evidence of improvement in skeletal deformities or growth<br>  ○ Improvement in 6-minute walk test<br>  ○ Improved bone density<br>  ○ Reduction in fractures<br>  ○ Respiratory function/breathing<br>  ○ Improvement in developmental milestones |
| **Exclusion Criteria:** | • Other types of osteomalacia or hypophosphatasia, including adult onset hypophosphatasia |

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| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist or specialist experienced in the treatment of metabolic bone disorders<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SUBCUTANEOUS IMMUNE GLOBULIN**
Affected Medications: CUTAQUIG, CUVITRU, GAMUNEX-C, HIZENTRA, HYQVIA, XEMBIFY, GAMMAKED

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Primary immunodeficiency (PID)/Wiskott-Aldrich syndrome<br>  ▪ Such as: x-linked agammaglobulinemia, common variable immunodeficiency (CVID), transient hypogammaglobulinemia of infancy, immunoglobulin G (IgG) subclass deficiency with or without immunoglobulin A (IgA) deficiency, antibody deficiency with near normal immunoglobulin levels) and combined deficiencies (severe combined immunodeficiencies, ataxia-telangiectasia, x-linked lymphoproliferative syndrome) [list not all inclusive]<br>  ○ Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) – **HyQvia, Hizentra Gammaked, and Gamunex-C only** |
| **Required Medical Information:** | • Monthly intravenous immune globulin (IVIG) dose for those transitioning<br>• Patient weight<br><br>**<u>Primary Immunodeficiency (PID)</u>**<br>• Type of immunodeficiency<br>• Documentation of one of the following:<br>  ○ Recent IgG level less than 200<br>  ○ Low IgG levels (below the laboratory reference range lower limit of normal) AND a history of multiple hard to treat infections as indicated by at least one of the following:<br>  ▪ Four or more ear infections within 1 year<br>  ▪ Two or more serious sinus infections within 1 year<br>  ▪ Two or more months of antibiotics with little effect<br>  ▪ Two or more pneumonias within 1 year<br>  ▪ Recurrent or deep skin abscesses<br>  ▪ Need for intravenous antibiotics to clear infections<br>  ▪ Two or more deep-seated infections including septicemia<br>• Documentation showing a deficiency in producing antibodies in response to vaccination including all of the following:<br>  ○ Titers that were drawn before challenging with vaccination<br>  ○ Titers that were drawn between 4 and 8 weeks after vaccination<br><br>**<u>Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)</u>**<br>• Documented baseline in strength/weakness has been documented using an objective clinical measuring tool (INCAT, Medical Research Council (MRC) muscle strength, 6 Minute Walk Test, Rankin, Modified Rankin)<br>• Documented disease course is progressive or relapsing and remitting for 2 months or longer<br>• Abnormal or absent deep tendon reflexes in upper or lower limbs |

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| | • Electrodiagnostic evidence of demyelination indicated by one of the following:<br>  ○ Motor distal latency prolongation in 2 nerves<br>  ○ Reduction of motor conduction velocity in 2 nerves<br>  ○ Prolongation of F-wave latency in 2 nerves<br>  ○ Absence of F-waves in at least 1 nerve<br>  ○ Partial motor conduction block of at least 1 motor nerve<br>  ○ Abnormal temporal dispersion in at least 2 nerves<br>  ○ Distal CMAP duration increase in at least 1 nerve<br>• Cerebrospinal fluid (CSF) analysis indicates all of the following (if electrophysiologic findings are non-diagnostic):<br>  ○ CSF white cell count of less than 10 cells/$mm^3$<br>  ○ CSF protein is elevated (greater than or equal to 45mg/dL) |
| **Appropriate Treatment Regimen & Other Criteria:** | **PID**<br>• Documentation of at least 3 months of IVIG therapy<br><br>**CIDP**<br>• Refractory to or intolerant of corticosteroids (prednisolone, prednisone) given in therapeutic doses over at least three months<br><br>**Reauthorization:**<br>• PID: requires disease response defined as a decrease in the frequency or severity of infections<br>• CIDP: requires a beneficial clinical response to maintenance therapy, without relapses, based on an objective clinical measuring tool (e.g., INCAT, Medical Research Council (MRC) muscle strength, 6 Minute walk test, Rankin, Modified Rankin) |
| **Exclusion Criteria:** | • IgA deficiency with antibodies to IgA<br>• History of hypersensitivity to immune globulin or product components<br>• Hyperprolinemia type I or II |
| **Age Restriction:** | • PID: 2 years of age and older<br>• CIDP: 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • PID: prescribed by, or in consultation with, an immunologist<br>• CIDP: prescribed by, or in consultation with, a neurologist or neuromuscular specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | Initial Authorization:<br>• PID: 12 months, unless otherwise specified<br>• CIDP: 3 months, unless otherwise specified<br>Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SUTIMLIMAB**
Affected Medications: ENJAYMO (sutimlimab-jome)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of hemolysis in adults with cold agglutinin disease (CAD) |
| **Required Medical Information:** | **Cold Agglutinin Disease (CAD)**<br>• Documentation of current weight<br>• Diagnosis of CAD as confirmed by all of the following:<br>  ○ Chronic hemolysis as confirmed by hemoglobin level of 10 g/dL or less AND elevated indirect bilirubin level<br>  ○ Positive monospecific direct antiglobulin test (DAT) or Coombs test for C3d<br>  ○ A positive DAT or Coombs test for IgG of 1+ or less<br>  ○ Cold agglutinin titer of greater than or equal to 64 at 4°C |
| **Appropriate Treatment Regimen & Other Criteria:** | **Cold Agglutinin Disease (CAD)**<br>• Dosing:<br>  ○ 39 kg to less than 75 kg: 6,500 mg/dose<br>  ○ 75 kg or greater: 7,500 mg/dose<br>  ○ Administered weekly for the first two weeks, then every two weeks thereafter<br><br>**Reauthorization:** documentation of disease responsiveness to therapy (e.g., increased hemoglobin, normalized markers of hemolysis [bilirubin, lactate dehydrogenase, reticulocyte count], reduced blood transfusion requirements) |
| **Exclusion Criteria:** | • Disease secondary to infection, rheumatologic disease, systemic lupus erythematosus, or overt hematologic malignancy<br>• Concomitant use of rituximab with or without cytotoxic agents |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**TTR STABILIZERS**
Affected Medications: VYNDAQEL (tafamidis meglumine 20 mg), VYNDAMAX (tafamidis 61 mg), ATTRUBY (acoramidis hydrochloride)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of wild type or hereditary transthyretin amyloid cardiomyopathy (ATTR-CM) to reduce cardiovascular mortality and cardiovascular-related hospitalizations in adults |
| **Required Medical Information:** | • Diagnosis of ATTR-CM supported by **ONE** of the following (a, b, or c):<br>  d. Cardiac tissue biopsy confirms presence of ATTR amyloid deposits by immunohistochemistry (IHC) or mass spectrometry<br>  e. Documentation of **BOTH** of the following (i and ii):<br>  i. Noncardiac tissue biopsy confirms presence of ATTR amyloid deposits by IHC or mass spectrometry<br>  ii. Imaging consistent with cardiac amyloidosis (echocardiogram [ECG], cardiac magnetic resonance [CMR], or positron emission tomography [PET])<br>  f. Documentation of **ALL** the following (i, ii, and iii):<br>  i. Grade 2 to 3 uptake on cardiac scintigraphy (utilizing Tc-PYP, Tc-DPD, or Tc-HMDP radiotracers)<br>  ii. Normal serum kappa/lambda free light chain (sFLC) ratio, serum protein immunofixation, **AND** urine protein immunofixation<br>  iii. Imaging consistent with cardiac amyloidosis (ECG, CMR, or PET)<br>• Documentation of New York Heart Association (NYHA) Functional Class I to III |
| **Appropriate Treatment Regimen & Other Criteria:** | **Coverage for Vyndaqel or Vyndamax is provided when the following is met:**<br>• Documented treatment failure with Attruby (acoramidis)<br><br>**<u>Reauthorization</u>** requires documentation of disease responsiveness (improvement in symptoms, quality of life, or 6-Minute Walk Test; slowing or stabilization of disease progression; reduced cardiovascular-related hospitalizations, etc.) |
| **Exclusion Criteria:** | • NYHA Functional Class IV heart failure<br>• Presence of light-chain (primary) amyloidosis<br>• Prior liver or heart transplant<br>• Implanted cardiac mechanical assist device<br>• Combined use with another TTR stabilizer or TTR silencer (such as eplontersen, patisiran, vultrisiran) |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or specialist experienced in the treatment of amyloidosis<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**TAGRAXOFUSP-ERZS**
Affected Medications: ELZONRIS (tagraxofusp-erzs)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of blastic plasmacytoid dendritic cell neoplasm (BPDCN) in adults and in pediatric patients at least 2 years of age<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | • Diagnosis of BPDCN is confirmed by ALL of the following:<br>  ○ A biopsy showing the morphology of plasmacytoid dendritic blast cells<br>  ○ At least 3 of the following plasmacytoid dendritic cell (pDC) markers are expressed by immunohistochemistry (IHC) or flow cytometry:<br>  ▪ CD123<br>  ▪ CD4<br>  ▪ CD56<br>  ▪ TCF4<br>  ▪ TCL1<br>  ▪ CD303<br>  ▪ CD304<br>  ○ The following pDC markers are negative: CD3, CD14, CD19, CD34, lysozyme, myeloperoxidase<br>  ○ Diagnosis is made by a board-certified hematopathologist or dermatopathologist<br>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• Pregnancy |
| **Age Restriction:** | • 2 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a prescriber experienced in the treatment of BPDCN<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

# TARGETED IMMUNE MODULATORS

### Included Products:
Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Cosentyx SQ and IV, Otezla, Tremfya SQ and IV, Xeljanz, Skyrizi SQ and IV, Rinvoq, Dupixent, Adbry, Fasenra, Nucala, Tezspire, Selarsdi SQ and IV, Yesintek SQ and IV, Xolair Orencia IV, Inflectra, Renflexis, Simponi Aria, Tofidence IV, Tyenne IV, Inflectra, Renflexis,
Avsola, Actemra IV, Avtozma IV, Otulfi IV, Omvoh, Pyzchiva IV, Steqeyma IV, Wezlana IV, Imuldosa IV, Entyvio, Stelara IV, Starjemza IV, Remicade, Infliximab (J1745)

## Scope & Exclusions

### Included Indications:
All Food and Drug Administration (FDA)–approved indications not otherwise excluded by plan design. Drug Compendia supported indications may be covered.

### Exclusions:
The requested product will not be given concurrently with another targeted immune modulator product unless there is no product which covers all indications.

Requests for Xolair where weight and IgE levels are outside of recommended dosing schedule are considered experimental and are not covered.

## Initial Authorization Criteria

### Required Medical Information:
1. All indications must be FDA-supported for the requested product or strongly supported in drug compendia (e.g. Micromedex, LexiComp). Exception: biosimilar products may be covered for all FDA-approved indications that the innovator product has been granted.
2. Requested dosing must be according to the FDA label based on diagnosis, age, and weight.
3. Requests for non-preferred products require inadequate response, intolerance, or an FDA-labeled *and* patient-specific contraindication to all preferred products as outlined within indication-specific criteria.
   FDA-labeled contraindications are defined as those listed within the package insert. Allergic reactions may be accepted as contraindicated when an ingredient found only in the preferred product is established as the causative agent.
4. Requests for non-preferred medical drugs require clinical failure of one preferred pharmacy drug AND one preferred medical drug, when both categories are available.
5. Indication-specific criteria are met:
   a. Acute Graft Versus Host Disease (GVHD) Prophylaxis
   b. Allergic Asthma
   c. Allergic Fungal Rhinosinusitis (AFRS)
   d. Ankylosing Spondylitis (AS), Non-radiographic Axial Spondyloarthritis (nr-axSpA) & Psoriatic Arthritis with Axial Involvement
   e. Atopic Dermatitis (AD)
   f. Bullous Pemphigoid (BP)
   g. Chronic Obstructive Pulmonary Disorder (COPD)
   h. Chronic Rhinosinusitis with Nasal Polyposis (CRSwNP)
   i. Chronic Spontaneous Urticaria (CSU))

j. Crohn's Disease (CD))
k. Cytokine Release Syndrome (CRS)
l. Enthesitis-Related Arthritis (ERA)
m. Eosinophilic Granulomatosis with Polyangiitis (EGPA)
n. Eosinophilic Asthma
o. Eosinophilic Esophagitis (EoE)
p. Generalized Pustular Psoriasis (GPP) Flare
q. Giant Cell Arteritis (GCA)
r. Hypereosinophilic Syndrome (HES)
s. Hidradenitis Suppurativa (HS)
t. IgE-Mediated Food Allergy
u. Juvenile Idiopathic Arthritis (JIA)
v. Juvenile Psoriatic Arthritis (JPsA)
w. Oral Ulcers Associated with Behcet's Disease
x. Plaque Psoriasis (PP)
y. Prurigo Nodularis (PN)
z. Psoriatic Arthritis (PsA)
aa. Rheumatoid Arthritis (RA)
bb. Severe Asthma
cc. Ulcerative Colitis (UC)
dd. Uveitis

## Acute Graft Versus Host Disease (GVHD) Prophylaxis

**Preferred Medical Drugs:** Orencia IV

1. Documentation of planned hematopoietic stem cell transplant (HSCT)
2. No prior allogeneic hematopoietic stem cell transplant (HSCT)
3. No current human immunodeficiency virus infection (HIV) or uncontrolled active infection (viral, bacterial, fungal, protozoal)
4. Maximum approval 4 days of treatment with no reauthorization

## Allergic Asthma

**Preferred Pharmacy Drugs:** Xolair

1. Diagnosis of moderate to severe allergic asthma with both of the following:
    a. Positive skin test or in vitro reactivity to a perennial aeroallergen (e.g., house dust mite, animal dander [dog, cat], cockroach, feathers, mold spores)
    b. Continued symptoms despite use of high-dose inhaled corticosteroid (ICS) plus long-acting beta agonist (LABA)
2. Requested dose is within the Food and Drug Administration (FDA) approved label based on patient weight, age, and IgE level

## Allergic Fungal Rhinosinusitis (AFRS)

**Preferred Pharmacy Drugs:** Dupixent

1. Diagnosis with AFRS confirmed by all the following:
    a. Type I hypersensitivity (IgE-mediated inflammatory response to fungal hyphae as assessed by history, skin, or in vitro testing)
    b. Presence of polyps in the nasal passage

   c. Characteristic CT findings (such as heterogenous opacity or differential densities in the soft tissue)
   d. Eosinophilic mucin without fungal invasion into sinus tissue
2. History of sino-nasal surgery

## Ankylosing Spondylitis (AS), Non-radiographic Axial Spondyloarthritis (nr-axSpA) & Psoriatic Arthritis with Axial Involvement

**Preferred Pharmacy Drugs:** Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Cosentyx, Xeljanz, Rinvoq
**Preferred Medical Drugs:** Inflectra, Renflexis, Simponi Aria
**Non-preferred Medical Drugs:** Remicade, Infliximab (J1745), Avsola, Cosentyx IV

1. Diagnosis of Axial Spondyloarthritis
2. Documented treatment failure of two daily prescription strength nonsteroidal anti-inflammatory drugs (ibuprofen, naproxen, diclofenac, meloxicam, etc) OR locally administered parenteral glucocorticoid (for isolated sacroiliitis, enthesitis, peripheral arthritis)

## Atopic Dermatitis (AD)

**Preferred Pharmacy Drugs:** Rinvoq, Dupixent, Adbry

1. Diagnosis of moderate-to-severe Atopic Dermatitis
2. Documentation of severe, inflammatory skin disease defined as functional impairment (inability to use hands or feet for activities of daily living or significant facial involvement preventing normal social interaction)
3. Body surface area (BSA) involvement greater than or equal to 10% or hand, foot, or mucous membrane involvement
4. Documented treatment failure with any TWO of the following:
   a. Medium potency to super-high potency topical corticosteroid
   b. Topical calcineurin inhibitor
   c. Phototherapy, cyclosporine, azathioprine, methotrexate, mycophenolate

## Bullous Pemphigoid (BP)

**Preferred Pharmacy Drugs:** Dupixent

1. Documented diagnosis of bullous pemphigoid
2. Documented treatment failure with TWO of the following: high potency corticosteroid (e.g. clobetasol, betamethasone, halobetasol, fluocinonide), oral corticosteroid, oral doxycycline, azathioprine, mycophenolate, methotrexate

## Chronic Obstructive Pulmonary Disorder (COPD)

**Preferred Pharmacy Drugs:** Dupixent, Nucala

1. Diagnosis of COPD with moderate to severe airflow limitation
2. Baseline eosinophil count of at least 300 cells/mcL
3. Documented use of inhaled triple therapy consisting of a long-acting muscarinic antagonist (LAMA), long-acting beta agonist (LABA), and inhaled corticosteroid (ICS) with continued symptoms

## Chronic Rhinosinusitis with Nasal Polyposis (CRSwNP)

**Preferred Pharmacy Drugs:** Dupixent, Nucala, Tezspire, Xolair

1. Diagnosis of chronic rhinosinusitis with nasal polyps
2. Documentation of ONE of the following:
   a. History of sinus surgery
   b. Documentation of intranasal corticosteroid treatment failure

3. Dosing for Xolair requests must be within the Food and Drug Administration (FDA) approved label based on patient weight and IgE level

## Chronic Spontaneous Urticaria (CSU)

**Preferred Pharmacy Drugs:** Dupixent, Xolair

1. Diagnosis of Chronic Spontaneous Urticaria
2. Documented treatment failure with up to 4-fold standard dosing of one second generation H1-antihistamine product for at least one month: cetirizine, fexofenadine, loratadine, desloratadine, or levocetirizine
3. Treatment failure of add-on therapy with ONE of the following: H2-antagonist (famotidine or cimetidine), leukotriene antagonist (montelukast or zafirlukast), cyclosporine A

## Crohn's Disease (CD)

**Preferred Pharmacy Drugs:** Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Skyrizi, Rinvoq, Tremfya, Selarsdi, Yesintek

**Preferred Medical Drugs:** Inflectra, Renflexis, Skyrizi IV, Selarsdi IV, Yesintek IV, Tremfya IV

**Non-preferred Medical Drugs:** Remicade, Infliximab (J1745), Avsola, Entyvio, Otulfi IV, Omvoh, Pyzchiva IV, Steqeyma IV, Wezlana IV, Imuldosa IV, Stelara IV, Starjemza IV

1. Diagnosis of moderate-to-severe Crohn's Disease supported by endoscopy/colonoscopy/sigmoidoscopy or biopsy

## Cytokine Release Syndrome (CRS)

**Preferred Medical Drugs:** Tofidence IV, Tyenne IV

**Non-preferred Medical Drugs:** Actemra IV, Avtozma IV

1. Diagnosis of Cytokine Release Syndrome

## Enthesitis-Related Arthritis (ERA)

**Preferred Pharmacy Drugs**: Cosentyx

1. Diagnosis of Enthesitis-Related Arthritis
2. Documented treatment failure with a nonsteroidal anti-inflammatory drug (ibuprofen, naproxen, celecoxib, meloxicam, etc.)
3. Documented clinical failure of one disease modifying antirheumatic drug (DMARD): methotrexate, sulfasalazine, leflunomide

## Eosinophilic Granulomatosis with Polyangiitis (EGPA)

**Preferred Pharmacy Drugs:** Fasenra, Nucala

1. Diagnosis of EGPA defined by one of the following at baseline:
    a. Absolute eosinophil count over 1,000 cells/mcL
    b. Blood eosinophil level over 10% on while blood cell differential count
2. History or presence of asthma
3. Manifestations of EGPA are active and non-severe (respiratory/sinonasal disease, uncomplicated skin manifestations, arthralgias, mild systemic symptoms, etc.)
4. Inadequate response or contraindication to at least two oral immunosuppressant drugs (azathioprine, methotrexate, mycophenolate)
5. One of the following:
    a. Relapsing or refractory disease
    b. Maintenance of disease remission

## Eosinophilic Asthma

**Preferred Pharmacy Drugs**: Dupixent, Fasenra, Nucala
**Nonpreferred Medical Drugs**: Cinqair

1. Diagnosis of moderate-to-severe eosinophilic phenotype or oral corticosteroid dependent asthma
2. Baseline eosinophil count of at least 150 cells/mcL (If requesting Cinqair must have eosinophil count of at least 400 cells/mcL)
3. Documented use of high-dose corticosteroid (ICS) with a long-acting beta agonist (LABA) with continued symptoms

## Eosinophilic Esophagitis (EoE)

**Preferred Pharmacy Drugs:** Dupixent

1. Diagnosis of eosinophilic esophagitis confirmed by endoscopic biopsy
2. Documented clinical failure of both of the following: high-dose, twice-daily proton pump inhibitor and swallowed corticosteroid therapy (such as fluticasone or budesonide)

## Generalized Pustular Psoriasis (GPP) Flare

**Preferred Medical Drugs:** Inflectra, Renflexis
**Non-preferred Medical Drugs:** Remicade, Avsola, Infliximab (J1745)

1. Diagnosis of moderate-to-severe GPP flare
2. Documentation of 1 week treatment failure of cyclosporine
3. If request is for a non-preferred drug: documentation of treatment failure with one preferred medical drug

## Giant Cell Arteritis (GCA)

**Preferred Pharmacy Drugs:** Rinvoq
**Preferred Medical Drugs:** Tofidence IV, Tyenne IV
**Non-preferred Medical Drugs:** Actemra IV, Avtozma IV

1. Diagnosis of Giant Cell Arteritis (GCA) confirmed by biopsy or imaging
2. Documentation of disease refractory to glucocorticoids

## Hypereosinophilic Syndrome (HES)

**Preferred Pharmacy Drugs**: Nucala

1. Diagnosis of HES with baseline eosinophil count greater than or equal to 1,000 cells/mcL
2. Lab work showing Fip1-like1-platelet-derived growth factor receptor alpha (FIP1L1-PDGFRα) mutation negative disease
3. Documentation that disease is currently controlled on the highest tolerated glucocorticoid dose (defined as an improvement in clinical symptoms and a decrease in eosinophil count by at least 50% from baseline)
4. Treatment failure with hydroxyurea AND interferon alfa

## Hidradenitis Suppurativa (HS)

**Preferred Pharmacy Drugs:** Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Cosentyx
**Preferred Medical Drugs:** Inflectra, Renflexis
**Non-preferred Medical Drugs:** Remicade, Infliximab (J1745), Avsola

1. Diagnosis of moderate-to-severe Hidradenitis Suppurativa (HS)
2. Documented clinical failure of oral antibiotics (tetracycline, doxycycline, minocycline OR clindamycin with rifampin)
3. Documented clinical failure of isotretinoin OR acitretin

## IgE-Mediated Food Allergy

**Preferred Pharmacy Drugs:** Xolair

1. Diagnosis of IgE-mediated food anaphylactic allergy to three or more foods with documented positive skin prick test and positive serum IgE
2. History of past IgE-mediated food anaphylactic reactions requiring use of epinephrine despite avoidance of food allergen
3. Treatment failure of oral immunotherapy (OIT)
4. Dosing for Xolair requests must be within the Food and Drug Administration (FDA) approved label based on patient weight and IgE level

## Juvenile Idiopathic Arthritis (JIA)

**Preferred Pharmacy Drugs:** Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Xeljanz, Rinvoq
**Preferred Medical Drugs:** Simponi Aria, Tofidence IV, Tyenne IV
**Non-preferred Medical Drugs:** Orencia IV, Actemra IV, Avtozma IV

1. Diagnosis of Juvenile Idiopathic Arthritis
2. Documented treatment failure with glucocorticoid injections or oral corticosteroids
3. Documented treatment failure of methotrexate or leflunomide

## Juvenile Psoriatic Arthritis (JPsA)

**Preferred Pharmacy Drugs:** Cosentyx, Enbrel

1. Diagnosis of Juvenile Psoriatic Arthritis
2. Documented treatment failure with a nonsteroidal anti-inflammatory drug (ibuprofen, naproxen, celecoxib, meloxicam, etc)
3. Documented clinical failure of one disease modifying antirheumatic drug (DMARD): methotrexate, sulfasalazine, leflunomide

## Oral Ulcers Associated with Behcet's Disease

**Preferred Pharmacy Drugs:** Otezla

1. Diagnosis of oral ulcers associated with Behcet's Disease
2. Clinical treatment failure to one of the following: colchicine, prednisone, azathioprine

## Plaque Psoriasis (PP)

**Preferred Pharmacy Drugs:** Hadlima, Hyrimoz (Cordavis), adalimumab-adaz, Enbrel, Cosentyx, Otezla, Skyrizi, Tremfya, Selarsdi, Yesintek
**Preferred Medical Drugs:** Inflectra, Renflexis
**Non-preferred Medical Drugs:** Remicade, Infliximab (J1745), Avsola, Starjemza IV

1. Diagnosis of moderate-to-severe plaque psoriasis
2. Body surface area (BSA) involvement greater than or equal to 10% despite current treatment or hand, foot, or mucous membrane involvement
3. Documented clinical failure of two systemic therapies: methotrexate, cyclosporine, acitretin, phototherapy (UVB, PUVA)

## Prurigo Nodularis (PN)

**Preferred Pharmacy Drugs:** Dupixent

1. Diagnosis of prurigo nodularis confirmed by biopsy
2. Documented treatment failure with super high potency topical corticosteroid (such as clobetasol propionate 0.05%, halobetasol propionate 0.05%)

3. Documentation of treatment failure with one of the following: phototherapy, methotrexate, cyclosporine

### Psoriatic Arthritis (PsA)

**Preferred Pharmacy Drugs:** Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Otezla, Cosentyx, Xeljanz, Tremfya, Rinvoq, Skyrizi, Selarsdi, Yesintek
**Preferred Medical Drugs:** Inflectra, Renflexis, Simponi Aria
**Non-preferred Medical Drugs:** Remicade, Infliximab (J1745), Avsola, Orencia IV, Cosentyx IV, Starjemza IV

1. Diagnosis of moderate-to-severe Psoriatic Arthritis
2. Documented treatment failure with methotrexate (preferred) OR sulfasalazine, cyclosporine, or leflunomide with documentation methotrexate is contraindicated or not tolerated

### Rheumatoid Arthritis (RA)

**Preferred Pharmacy Drugs:** Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Xeljanz, Rinvoq
**Preferred Medical Drugs:** Inflectra, Renflexis, Simponi Aria, Tofidence IV, Tyenne IV
**Non-preferred Medical Drugs:** Remicade, Infliximab (J1745), Avsola, Actemra IV, Avtozma IV, Orencia IV

1. Diagnosis of moderate-to-severe Rheumatoid Arthritis
2. Documented treatment failure with methotrexate (preferred) OR sulfasalazine, hydroxychloroquine, or leflunomide with documentation methotrexate is contraindicated or not tolerated.

### Severe Asthma

**Preferred Pharmacy Drugs:** Tezspire

1. Diagnosis of severe asthma
2. Documented use of high-dose inhaled corticosteroid (ICS) plus a long-acting beta agonist (LABA) for at least three months with continued symptoms

### Ulcerative Colitis (UC)

**Preferred Pharmacy Drugs:** Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Rinvoq, Xeljanz, Skyrizi, Tremfya, Selarsdi, Yesintek
**Preferred Medical Drugs:** Inflectra, Renflexis, Skyrizi IV, Selarsdi IV, Yesintek IV, Tremfya IV
**Non-preferred Medical Drugs:** Remicade, Infliximab (J1745), Avsola, Entyvio, Omvoh, Otulfi IV, Pyzchiva IV, Steqeyma IV, Wezlana IV, Imuldosa IV, Stelara IV, Starjemza IV

1. Diagnosis of moderate-to-severe Ulcerative Colitis supported by endoscopy/colonoscopy/sigmoidoscopy or biopsy

### Uveitis

**Preferred Pharmacy Drugs:** Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz

1. Diagnosis of noninfectious uveitis (intermediate uveitis, panuveitis, or posterior uveitis)
2. Intermediate or panuveitis: documented treatment failure with one immunosuppressive agent (methotrexate, azathioprine, mycophenolate) AND one systemic calcineurin inhibitor (cyclosporine, tacrolimus)
3. Posterior uveitis: documented treatment failure with Yutiq AND Retisert

## Age Limits

As defined by FDA labeling for the requested indication.

## Prescriber Limits

Prescribed by, or in consult with, appropriate specialist for the indication.

## Quantity Limits

Infusion medications: dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced.

Adalimumab: 40mg every week or 80mg every other week for RA, PP, CD, or UC will require documentation of lost or inadequate response after a minimum of 16 weeks with standard maintenance dosing.

Entyvio: 300mg every 4 weeks will require treatment failure to Entyvio at standard dosing for at least 6 months

## Duration of Approval

Initial: 6 months, unless otherwise specified
Renewal: 24 months, unless otherwise specified

POLICY NAME:
**TARPEYO**
Affected Medications: TARPEYO (budesonide delayed release capsule 4 mg)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Reduce the loss of kidney function in adults with primary immunoglobulin A nephropathy (IgAN) who are at risk for disease progression |
| **Required Medical Information:** | • Diagnosis of primary immunoglobulin A nephropathy (IgAN) confirmed with biopsy<br>• Documentation of proteinuria greater than or equal to 0.5 g/day (with labs taken within 30 days of request)<br>• Documented estimated glomerular filtration rate (eGFR) greater than or equal to 35 mL/min/1.73m$^2$ |
| **Appropriate Treatment Regimen & Other Criteria:** | • Persistent proteinuria (greater than or equal to 0.5 g/day) despite a minimum 12-week trial with **each** of the following:<br>  ○ Maximally tolerated angiotensin-converting enzyme (ACE) inhibitor **OR** angiotensin receptor II blocker (ARB)<br>  ○ Alternative glucocorticoid therapy, such as prednisone or methylprednisolone (or adverse effect with two or more glucocorticoid therapies, which is not associated with the corticosteroid class)<br><br>**No reauthorization** – Recommended duration of therapy is 9 months followed by a 2-week dose taper prior to discontinuation |
| **Exclusion Criteria:** | • Treatment of other glomerulopathies or nephrotic syndrome |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a nephrologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 10 months, unless otherwise specified |

POLICY NAME:
**TASIMELTEON**
Affected Medications: HETLIOZ LQ SUSPENSION, TASIMELTEON

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of Non-24-Hour Sleep-Wake Disorder (Non-24)<br>  ○ Treatment of nighttime sleep disturbances in Smith-Magenis Syndrome (SMS) |
| **Required Medical Information:** | **Non-24**<br>• Documentation of being totally blind with no light perception<br>• Diagnosis of Non-24 hour sleep wake disorder meeting ALL of the following:<br>  ○ Documented history of insomnia, excessive daytime sleepiness, or both, that alternates with asymptomatic periods<br>  ○ Symptoms have been present for at least three months<br>  ○ Drift in rest-activity patterns demonstrated by at least 4 weeks of data from daily sleep logs and actigraphy<br>  ○ Documentation that other sleep disorders were treated or ruled out using a sleep study<br><br>**Smith-Magenis Syndrome (SMS)**<br>• Diagnosis of Smith-Magenis Syndrome (SMS) confirmed by both of the following:<br>  ○ Genetic test showing mutation or deletion of the retinoic acid-induced 1 (RAI1) gene<br>  ○ Documentation of significant nighttime sleep disturbances |
| **Appropriate Treatment Regimen & Other Criteria:** | **Non-24**<br>• Documentation of treatment failure with at least 12 weeks of melatonin<br>**Smith-Magenis Syndrome (SMS)**<br>• Documented trial and failure with treatment regimen that includes both melatonin taken at bedtime AND acebutolol taken during daytime for at least 12 weeks<br><br>**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Sleep disorders other than Non-24 and SMS such as insomnia, shift work disorder, jet lag disorder, irregular sleep-wake rhythm disorder, delayed sleepwake phase disorder, advanced sleep-wake rhythm disorder<br>• Sleep disturbances caused by taking sedative or stimulant central nervous system-active drugs<br>• Sleep disturbances caused by other conditions |
| **Age Restriction:** | • Non-24: 18 years of age and older<br>• SMS:<br>  ○ Capsules: 16 years of age and older<br>  ○ Suspension: 3 to 15 years of age |

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| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with a neurologist or sleep specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**TEDIZOLID**
Affected Medications: SIVEXTRO injection, SIVEXTRO tablets

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Acute bacterial skin and skin structure infections (ABSSSI) caused by susceptible isolates of the following Gram-positive microorganisms:<br>  ▪ Staphylococcus aureus (including methicillin-resistant [MRSA] and methicillin-susceptible [MSSA] isolates)<br>  ▪ Streptococcus pyogenes<br>  ▪ Streptococcus agalactiae<br>  ▪ Streptococcus anginosus Group (including Streptococcus anginosus, Streptococcus intermedius, and Streptococcus constellatus)<br>  ▪ Enterococcus faecalis |
| **Required Medical Information:** | • Documentation of confirmed or suspected diagnosis<br>• Documentation of treatment history and current treatment regimen<br>• Documentation of culture and sensitivity data<br>• Documentation of planned treatment duration |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing is in accordance with FDA labeling<br><br>Requests for the intravenous formulation will require both of the following:<br>• Documentation of treatment failure, contraindication, or intolerable adverse event with intravenous linezolid **AND**<br>• Documentation of treatment failure, contraindication, or intolerable adverse event with at least 2 of the following drugs/drug classes:<br>  ○ Vancomycin<br>  ▪ Avoidance of vancomycin due to nephrotoxicity will require documentation of multiple (at least 2 consecutive) increased serum creatinine concentrations (increase of 0.5 mg/dL [44 mcmol/L] or at least 50 percent increase from baseline, whichever is greater), without an alternative explanation<br>  ○ Daptomycin<br>  ○ Cephalosporin (cefazolin)<br><br>Requests for the oral tablet formulation will require both of the following:<br>• Documentation of treatment failure, contraindication, or intolerable adverse event with oral linezolid **AND**<br>• Documentation of treatment failure, contraindication, or intolerable adverse event with at least 2 of the following drugs/drug classes:<br>  ○ Trimethoprim-sulfamethoxazole<br>  ○ Tetracycline (doxycycline, minocycline)<br>  ○ Clindamycin |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an infectious disease specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage** | • Authorization: 1 month, unless otherwise specified |

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| **Duration:** | |

POLICY NAME:
**TEDUGLUTIDE**
Affected Medications: GATTEX (teduglutide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of Short Bowel Syndrome (SBS) |
| **Required Medical Information:** | • Documentation of confirmed SBS diagnosis<br>• Dependence on parenteral nutrition (PN) and/or intravenous (IV) fluids at least 12 consecutive months continuously<br>• Receiving three or more days per week of PN support such as fluids, electrolytes, and/or nutrients |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of inability to be weaned from PN despite use of the following conventional measures:<br>  ○ Dietary manipulations, oral rehydration solutions<br>  ○ Antidiarrheal/motility agents: loperamide or diphenoxylate<br>  ○ Antisecretory agents: H2 receptor antagonists or proton pump inhibitors<br>**OR**<br>• Developed significant complications or severe impairment in quality of life related to parenteral nutrition use (such as loss of vascular access sites, recurrent catheter-related bloodstream infections, and liver disease)<br>• Dose does not exceed 0.05 mg/kg daily<br>**<u>Reauthorization</u>** requires documentation of clinically significant benefit defined by parenteral support reduction of 1 day or greater a week |
| **Exclusion Criteria:** | • Weight of less than 10 kg<br>• Onset or worsening of gallbladder/biliary disease<br>• Onset or worsening of pancreatic disease<br>• Presence of any gastrointestinal malignancy<br>• Presence of intestinal or stomal obstruction |
| **Age Restriction:** | • 1 year of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a gastroenterologist or SBS specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 6 months, unless otherwise specified |

POLICY NAME:
**TENAPANOR**
Affected Medications: XPHOZAH (tenapanor)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of hyperphosphatemia associated with chronic kidney disease (CKD) |
| **Required Medical Information:** | • Diagnosis of hyperphosphatemia associated with CKD and currently on dialysis treatment<br>• Documentation of progressively or persistently elevated serum phosphate that is greater than 5.5 mg/dL over the past 6 months despite adherence to phosphate binders and dietary restrictions<br>• Documentation that Xphozah (tenapanor) will be used as add-on therapy to phosphate binder therapy unless contraindicated or clinically significant adverse effects were experienced |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with at least an 8-week trial, at maximally indicated doses, of at least two of the following:<br>  ○ calcium acetate<br>  ○ lanthanum carbonate<br>  ○ sevelamer<br>  ○ Velphoro (sucroferric oxyhydroxide)<br>  ○ Auryxia (ferric citrate)<br><br>**Reauthorization** requires documentation of treatment success defined as reduction in serum phosphorus from pretreatment level and maintenance of serum phosphorus level at 5.5 mg/dL or lower |
| **Exclusion Criteria:** | • Known or suspected mechanical gastrointestinal obstruction |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a nephrologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**TENOFOVIR ALAFENAMIDE**
Affected Medications**:** VEMLIDY (tenofovir alafenamide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ For the treatment of chronic hepatitis B virus (HBV) infection in adults and pediatric patients 6 years of age and older with compensated liver disease |
| **Required Medical Information:** | • Documentation confirming diagnosis of chronic hepatitis B infection<br>• Documentation of compensated liver disease (Child-Pugh A) within 12 weeks prior to anticipated start of therapy |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of one or more of the following:<br>  ○ Inadequate virologic response or intolerable adverse event to tenofovir disoproxil fumarate<br>  ○ CrCl less than or equal to 80 mL/min within 12 weeks prior to anticipated start date OR high risk for acute renal injury (i.e., nephrotoxic medications)<br>  ○ Diagnosis of osteoporosis, osteopenia, or high risk for developing osteoporosis with supporting documentation (i.e., chronic use of steroids or other drugs that worsen bone density, poor nutrition, early menopause)<br><br>**Reauthorization**: documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Decompensated hepatic impairment (Child-Pugh B or C) |
| **Age Restriction:** | • 6 years of age and older |
| **Prescriber Restrictions:** | • Prescribed by, or in consultation with, a hepatologist, gastroenterologist, or infectious disease specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**TEPLIZUMAB-MZWV**
Affected Medications: TZIELD (teplizumab-mzwv)

<table>
<tr><td>Covered Uses:</td><td>
<ul>
<li>All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
<ul>
<li>Type 1 diabetes mellitus, to delay the onset of Stage 3 type 1 diabetes in adults, and pediatric patients with Stage 2 type 1 diabetes</li>
</ul></li>
</ul>
</td></tr>
<tr><td>Required Medical Information:</td><td>
<ul>
<li>Diagnosis of Stage 2 type 1 diabetes, confirmed by both of the following:
<ul>
<li>Positive for two or more of the following pancreatic islet cell autoantibodies within the past 6 months:
<ul>
<li>Glutamic acid decarboxylase 65 (GAD) autoantibodies</li>
<li>Insulin autoantibody (IAA)</li>
<li>Insulinoma-associated antigen 2 autoantibody (IA-2A)</li>
<li>Zinc transporter 8 autoantibody (ZnT8A)</li>
<li>Islet cell autoantibody (ICA)</li>
</ul></li>
<li>Dysglycemia on oral glucose tolerance testing (OGTT) within the past 6 months, as shown by one of the following:
<ul>
<li>Fasting blood glucose between 110 mg/dL and 125 mg/dL</li>
<li>2 hour glucose greater than or equal to 140 mg/dL and less than 200 mg/dL</li>
<li>30, 60, or 90 minute value on OGTT greater than or equal to 200 mg/dL on two separate occasions</li>
</ul></li>
</ul></li>
<li>Documentation that the patient has a first-degree or second-degree relative with type 1 diabetes and one of the following:
<ul>
<li>If first-degree relative (brother, sister, parent, offspring), patient must be between 8 and 45 years of age</li>
<li>If second-degree relative (niece, nephew, aunt, uncle, grandchild, cousin), patient must be between 8 and 20 years of age</li>
</ul></li>
<li>Documentation of the patient’s current body surface area (BSA) or height and weight to calculate BSA</li>
<li>Treatment plan, including planned dose and frequency</li>
</ul>
</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td>
Approved for one-time 14-day infusion only, based on the following dosing schedule:
<table>
<tr><td>Treatment Day</td><td>Dose</td></tr>
<tr><td>Day 1</td><td>65 mcg/m$^2$</td></tr>
<tr><td>Day 2</td><td>125 mcg/m$^2$</td></tr>
<tr><td>Day 3</td><td>250 mcg/m$^2$</td></tr>
<tr><td>Day 4</td><td>500 mcg/m$^2$</td></tr>
<tr><td>Days 5- 14</td><td>1,030 mcg/m$^2$</td></tr>
</table>
<ul>
<li>Availability: 2 mg/2 mL (1 mg/mL) single-dose vials</li>
<li>Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced</li>
</ul>
</td></tr>
<tr><td>Exclusion Criteria:</td><td>
<ul>
<li>Prior treatment with Tzield</li>
<li>Diagnosis of Stage 3 type 1 diabetes (clinical type 1 diabetes)</li>
<li>Diagnosis of Type 2 diabetes</li>
</ul>
</td></tr>
</table>

| | |
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| | • Current active serious infection or chronic infection<br>• Pregnant or lactating |
| **Age Restriction:** | • 8 to 45 years of age<br>• See Required Medical Information for age requirements based on first-degree or second-degree relative |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 3 months, unless otherwise specified (one 14-day infusion only) |

POLICY NAME:
**TEPROTUMUMAB-TRBW**
Affected Medications: TEPEZZA (teprotumumab-trbw)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Thyroid Eye Disease (TED) regardless of TED activity or duration |
| **Required Medical Information:** | • Documentation that baseline disease is under control prior to starting therapy, as defined by one of the following:<br>  ○ Patient is euthyroid (thyroid function tests are within normal limits)<br>  ○ Patient has recent and mild hypo- or hyperthyroidism (thyroid function tests show free thyroxine (T4) and free triiodothyronine (T3) levels less than 50% above or below normal limits) and will undergo treatment to maintain euthyroid state<br>• TED has an appreciable impact on daily life, defined as:<br>  ○ Proptosis greater than or equal to 3 mm increase from baseline (prior to diagnosis of TED) and/or proptosis greater than or equal to 3 mm above normal for race and gender<br>  **OR**<br>  ○ Current moderate-to-severe active TED with a Clinical Activity Score (CAS) greater than or equal to 4 (on the 7-item scale) for the most severely affected eye and symptoms such as: lid retraction greater than or equal to 3 mm, moderate or severe soft tissue involvement, diplopia, and/or proptosis greater than or equal to 3 mm above normal for race and gender |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br>• Evidence of stable, well-controlled disease if comorbid inflammatory bowel disease (IBD) or diabetes<br>• Documented failure to intravenous or oral steroid at appropriate dose over 12 weeks |
| **Exclusion Criteria:** | • Use of more than one course of Tepezza treatment<br>• Prior orbital irradiation, orbital decompression, or strabismus surgery<br>• Decreasing visual acuity, new defect in visual field, color vision defect from optic nerve involvement within the previous 6 months<br>• Corneal decompensation that is unresponsive to medical management |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 7 months, maximum approval (total of 8 doses) with no reauthorization, unless otherwise specified |

POLICY NAME:
**TESTOSTERONE**
Affected Medications: TESTOPEL (testosterone pellets), JATENZO (testosterone undecanoate capsules), TLANDO (testosterone undecanoate capsules)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Testosterone replacement therapy in adult males for conditions associated with a deficiency or absence of endogenous testosterone: primary hypogonadism or hypogonadotropic hypogonadism<br>• Gender Dysphoria |
| **Required Medical Information:** | **<u>All Indications</u>**<br>• If 65 years of age and older, must provide documentation of a yearly evaluation that includes ALL the following:<br>  ○ The need for continued hormone replacement therapy<br>  ○ Education on the risks of hormone replacement therapy (heart attack, stroke)<br>  ○ Discussion about the limited efficacy and safety for hormone replacement therapy in patients experiencing an age-related decrease in testosterone levels<br><br>**<u>Hypogonadism in Adults</u>**<br>• Confirmed low testosterone level (total testosterone less than 300 ng/dl or morning free or bioavailable testosterone less than 5 ng/dL) or absence of endogenous testosterone |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Hypogonadism in Adults</u>**<br>• Requests for oral testosterone (e.g., Jatenzo, Tlando) require documented treatment failure with testosterone injections AND generic transdermal testosterone<br>• Requests for Testopel require all of the following:<br>  ○ Documented treatment failure with testosterone injections AND generic transdermal testosterone<br>  ○ Documented treatment plan, including dosage in milligrams or number of pellets to be administered and frequency<br>  ▪ Maximum dosage: 450 mg per treatment<br>  ▪ Maximum of 4 treatments in 12 months<br><br>**<u>Reauthorization</u>:**<br>• Hypogonadism in Adults: Documentation of a recent testosterone level within normal limits |
| **Exclusion Criteria:** | • Treatment of sexual dysfunction<br>• Treatment of symptoms of menopause |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | Testopel:<br>• Authorization: 12 months (maximum of 4 treatments), unless otherwise specified<br><br>All other formulations: |

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| | • Authorization: 24 months, unless otherwise specified |

POLICY NAME:
**THALIDOMIDE**
Affected Medications: THALOMID (thalidomide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved or compendia-supported indications not otherwise excluded by plan design<br>  ○ Multiple Myeloma (MM)<br>  ○ Erythema Nodosum (ENL)<br>  ○ Systemic light chain amyloidosis<br>  ○ AIDS-related aphthous stomatitis<br>  ○ Waldenström macroglobulinemia<br>  ○ Graft-versus-host disease, chronic (refractory)<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | **Multiple Myeloma**<br>• NCCN (National Comprehensive Cancer Network) regimen with evidence level of 2A or higher<br><br>**Systemic light chain amyloidosis**<br>• NCCN (National Comprehensive Cancer Network) regimen with evidence level of 2A or higher<br><br>**Waldenström Macroglobulinemia**<br>• NCCN (National Comprehensive Cancer Network) regimen with evidence level of 2A or higher<br><br>**AIDS-related or Severe recurrent aphthous stomatitis**<br>• Documented trial and failure with BOTH topical and systemic corticosteroids<br><br>**Erythema Nodosum Leprosum (ENL)**<br>• Acute treatment of the cutaneous manifestations of moderate to severe ENL (Type 2 reaction)<br>• Maintenance therapy for prevention and suppression of the cutaneous manifestations of ENL recurrence<br><br>**Reauthorization:** Documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Pregnancy<br>• Karnofsky Performance Status less than or equal to 50% or ECOG performance score greater than or equal to 3 |
| **Age Restriction:** | • 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist or infectious disease specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

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POLICY NAME:
**TOBRAMYCIN INHALATION**
Affected Medications: BETHKIS (tobramycin), KITABIS PAK (tobramycin), TOBI (tobramycin), TOBI PODHALER (tobramycin), TOBRAMYCIN NEBULIZED SOLUTION

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Management of Cystic Fibrosis (CF) patients with Pseudomonas aeruginosa |
| **Required Medical Information:** | • Diagnosis of Cystic Fibrosis (phenotyping not required).<br>• Culture and sensitivity report confirming presence of pseudomonas aeruginosa in the lungs<br>• Baseline forced expiratory volume in 1 second (FEV1)<br>  ○ Tobi Podhaler: FEV1 equal to or between 25% and 80%<br>  ○ Bethkis: FEV1 equal to or between 40% and 80%<br>  ○ Kitabis Pak: FEV1 equal to or between 25% and 75% |
| **Appropriate Treatment Regimen & Other Criteria:** | • For Tobi Podhaler, Kitabis Pak, Bethkis, and Tobi: Documentation of failure with nebulized tobramycin or clinical rationale for avoidance<br>• Use is limited to a 28 days on and 28 days off regimen<br><br>**Reauthorization** requires documentation of improved respiratory symptoms and need for long-term use |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a pulmonologist, or provider who specializes in CF<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**TOFERSEN**
Affected Medications: QALSODY (tofersen)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Amyotrophic lateral sclerosis (ALS) associated with a mutation in the superoxide dismutase 1 (*SOD1*) gene (SOD1-ALS) |
| **Required Medical Information:** | • Documentation of "definite" or "probable" ALS diagnosis based on revised El Escorial (Airlie House) or Awaji criteria<br>• Documentation of a confirmed SOD1 genetic mutation<br>• Forced vital capacity (FVC) greater than or equal to 50% as adjusted for age, sex, and height (from a sitting position)<br>• Baseline plasma neurofilament light chain (NfL) value<br>• Patient currently retains most activities of daily living defined as at least 2 points on all 12 items of the ALS functional rating scale-revised (ALSFRS-R) |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires documentation of treatment success and a clinically significant response to therapy, defined as both of the following:<br>• Reduction in plasma NfL from baseline<br>• The patient's baseline functional status has been maintained at or above baseline level or not declined more than expected given the natural disease progression<br>• Patient is not dependent on invasive mechanical ventilation (e.g., intubation, tracheostomy) |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist, neuromuscular specialist, or specialist with experience in the treatment of ALS<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**TOLVAPTAN**
Affected Medications: JYNARQUE, TOLVAPTAN (15 mg, 30 mg)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ **Tolvaptan:** treatment of clinically significant hypervolemic and euvolemic hyponatremia (serum sodium less than 125 mEq/L OR less marked hyponatremia that is symptomatic and has resisted correction with fluid restriction), including patients with heart failure and Syndrome of Inappropriate Antidiuretic Hormone (SIADH)<br>  ○ **Jynarque:** to slow kidney function decline in adults at risk of rapidly progressing autosomal dominant polycystic kidney disease (ADPKD) |
| **Required Medical Information:** | **<u>Hyponatremia</u>**<br>• Serum sodium less than 125 mEq/L at baseline<br>**OR**<br>• Serum sodium less than 135 mEq/L at baseline and symptomatic (nausea, vomiting, headache, lethargy, confusion)<br><br>**<u>ADPKD</u>**<br>• Diagnosis of typical ADPKD confirmed by family history, imaging, and if applicable, genetic testing<br>• Estimated glomerular filtration rate (eGFR) greater than or equal to 25 mL/min/1.73m$^2$<br>• High risk for rapid progression determined by Mayo imaging class 1C, 1D, or 1E |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Hyponatremia</u>**<br>• Treatment is initiated or re-initiated in a hospital setting prior to discharge<br><br>**<u>ADPKD</u>**<br>• Documentation of intensive blood pressure control with an angiotensin-converting enzyme (ACE) inhibitor or angiotensin receptor blocker (ARB), unless contraindicated<br><br>**<u>Reauthorization (for ADPKD)</u>** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Patients requiring intervention to raise serum sodium urgently to prevent or treat serious neurological symptoms<br>• Patients who are unable to sense or respond to thirst<br>• Hypovolemic hyponatremia<br>• Anuria<br>• Uncorrected urinary outflow obstruction |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a nephrologist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | **<u>Hyponatremia</u>**<br>• Authorization: 1 month (no reauthorization), unless otherwise specified<br><br>**<u>ADPKD</u>**<br>• Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**TOPICAL AGENTS FOR CUTANEOUS T-CELL LYMPHOMA (including Mycosis fungoides and Sézary syndrome)**
Affected Medications: VALCHLOR (mechlorethamine topical gel), TARGRETIN (bexarotene gel)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• Documentation of cutaneous T-cell lymphoma (CTCL), stage and type confirmed by biopsy.<br>• Extent of skin involvement (limited/localized or generalized) |
| **Appropriate Treatment Regimen & Other Criteria:** | **Limited/localized skin involvement (topical bexarotene and mechlorethamine)**<br>• Documented clinical failure to ALL of the following:<br>  ○ Topical corticosteroids (high or super-high potency) such as clobetasol, betamethasone, fluocinonide, halobetasol<br>  ○ Topical imiquimod<br>  ○ Phototherapy<br><br>**Generalized skin involvement (topical mechlorethamine only)**<br>• Documentation of failure or contraindication to at least 1 skin-directed therapy<br><br>**Reauthorization:** documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• Pregnancy |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

# Topical Skin Disease Agents

Included Products: Vtama cream, Zoryve cream/foam, Opzelura

## SCOPE & EXCLUSIONS

Included Indications: All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. Drug compendia supported indications may be covered.

Exclusions: Use in cosmetic conditions such as vitiligo. The requested products will not be given concurrently with biologics or other JAK inhibitors (such as Rinvoq, Dupixent, Adbry, Nemluvio)

## AUTHORIZATION CRITERIA

### Required Medical Information:

1. Documentation of affected body surface area (BSA) and areas of involvement
2. For Atopic Dermatitis/Plaque Psoriasis, one of the following:
    a. At least 10% BSA involvement despite current treatment
    b. Hand, foot, face, mucous membrane involvement
3. For Seborrheic Dermatitis:
    a. Moderate to severe seborrheic dermatitis with persistent itching, scaling, and erythema despite current therapy

### Atopic Dermatitis

**Group 1 Drugs:** Zoryve cream
**Group 2 Drugs:** Vtama cream
**Group 3 Drugs:** Opzelura cream

1. Treatment failure with a 6-week trial of a topical calcineurin inhibitor
2. Group 2 drugs require treatment failure with one Group 1 drug
3. Group 3 drugs require treatment failure with one Group 1 drug, one Group 2 drug, and a minimum 12-week trial of two of the following: phototherapy, cyclosporine, azathioprine, methotrexate, mycophenolate

### Plaque Psoriasis

**Group 1 Drugs:** Zoryve cream/foam
**Group 2 Drugs:** Vtama cream

1. Treatment failure with a 4-week trial of a topical vitamin D analog or tazarotene
2. Group 2 drugs require treatment failure with a Group 1 drug

### Seborrheic Dermatitis

**Group 1 Drugs:** Zoryve foam

1. Treatment failure with a 6-week trial of one calcineurin inhibitor

## RENEWAL CRITERIA

1. Documentation of treatment success.

2. Opzelura requires resolution of symptoms within the first 8 weeks of treatment and that Opzelura will be discontinued each time signs and symptoms resolve.

## DURATION OF APPROVAL

Opzelura: 12 weeks, unless otherwise specified
All others: 12 months, unless otherwise specified

## QUANTITY LIMITS

1. Quantities greater than 1 tube of Opzelura per month require documented trial and failure of a biologic indicated for the requested diagnosis.

POLICY NAME:
**TRIENTINE**
Affected Medications: TRIENTINE HYDROCHLORIDE, CUVRIOR (trientine tetrahydrochloride)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ Wilson's disease |
| **Required Medical Information:** | • Diagnosis of Wilson's disease confirmed by **ONE** of the following:<br>  ○ Genetic testing results confirming biallelic pathogenic *ATP7B* mutations (in either symptomatic or asymptomatic individuals)<br>  ○ Liver biopsy findings consistent with Wilson's disease<br>  ○ Presence of Kayser-Fleischer (KF) rings **AND** serum ceruloplasmin level less than 20 mg/dL **AND** 24-hour urinary copper excretion greater than 40 mcg<br>  ○ Presence of Kayser-Fleischer (KF) rings **AND** 24-hour urinary copper excretion greater than 100 mcg<br>  ○ Absence of KF rings with serum ceruloplasmin level less than 10 mg/dL **AND** 24-hour urinary copper excretion greater than 100 mcg |
| **Appropriate Treatment Regimen & Other Criteria:** | • For Cuvrior, must meet **BOTH** of the following:<br>  ○ Documented treatment failure with a minimum 6-month trial of penicillamine that was not due to tolerability<br>  ○ Documented intolerable adverse event to a maximally tolerated dosage of generic trientine hydrochloride and the adverse event was not an expected adverse event attributed to the active ingredient<br><br>**Reauthorization**: Documentation of treatment success and a clinically significant response to therapy as shown by normalization of free serum copper (non-ceruloplasmin bound copper) to less than 15 mcg/dL and 24-hour urinary copper in the range of 200 to 500 mcg |
| **Exclusion Criteria:** | For trientine hydrochloride:<br>• Treatment of rheumatoid arthritis<br>• Treatment of cystinuria<br>• Treatment of biliary cirrhosis |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a hepatologist, gastroenterologist, or liver transplant provider<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization:12 months, unless otherwise specified |

POLICY NAME:
**TRIPTORELIN**
Affected Medications: TRELSTAR, TRIPTODUR (triptorelin)

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher<br>• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Prostate Cancer (Trelstar)<br>  ○ Central Precocious Puberty (Triptodur)<br>• Gender Dysphoria |
| **Required Medical Information:** | **Central Precocious Puberty (CPP)**<br>• Documentation of CPP confirmed by one of the following labs:<br>  ○ Elevated basal luteinizing hormone (LH) level greater than 0.2 - 0.3 mIU/L<br>  ○ Elevated leuprolide-stimulated LH level greater than 3.3 - 5 IU/L (dependent on type of assay used)<br>• Bone age greater than 2 standard deviations (SD) beyond chronological age |
| **Appropriate Treatment Regimen & Other Criteria:** | For all Triptodur requests:<br>• Documentation of treatment failure with leuprolide<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Use as neoadjuvant androgen deprivation therapy (ADT) for radical prostatectomy |
| **Age Restriction:** | • CPP: 2 years of age through 11 years for females, 2 years of age through 12 years for males |
| **Prescriber/Site of Care Restrictions:** | • Oncology: prescribed by, or in consultation with, an oncologist<br>• CPP: prescribed by, or in consultation with, a pediatric endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Oncology Initial Authorization: 4 months, unless otherwise specified<br>• CPP Approval/Oncology Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**TROFINETIDE**
Affected Medications: DAYBUE (trofinetide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of Rett syndrome (RTT) |
| **Required Medical Information:** | • Documented diagnosis of typical RTT (per the revised diagnostic criteria for Rett Syndrome) AND a period of regression followed by recovery or stabilization<br>• Documented presence of mutation in the *MECP2* gene<br>• Documentation of all the following:<br>  ○ Partial or complete loss of acquired purposeful hand skills<br>  ○ Partial or complete loss of acquired spoken language<br>  ○ Gait abnormalities: Impaired (dyspraxic) or absence of ability<br>  ○ Stereotypic hand movements such as hand wringing/squeezing, clapping/tapping, mouthing, and washing/rubbing automatisms<br>• Current weight (within past 30 days)<br>  ○ Must weigh minimum of 9 kilograms |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires documentation of treatment success determined by treating provider |
| **Exclusion Criteria:** | • Brain injury secondary to trauma or severe infection<br>• Grossly abnormal psychomotor development in first 6 months of life |
| **Age Restriction:** | • 2 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or provider experienced in the management of Rett syndrome<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**TROGARZO**
Affected Medications: TROGARZO (ibalizumab-uiyk/IV infusion)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of human immunodeficiency virus type 1 (HIV-1) infection, in combination with other antiretrovirals, in heavily treatment-experienced adults with multidrug resistant HIV-1 infection failing their current antiretroviral regimen |
| **Required Medical Information:** | • Documentation of all prior therapies used<br>• Documentation of active antiretroviral therapy for at least 6 months<br>• Documented resistance to at least one antiretroviral agent from three different classes:<br>  ○ Nucleoside reverse-transcriptase inhibitors (NRTIs)<br>  ○ Non-nucleoside reverse-transcriptase inhibitors (NNRTIs)<br>  ○ Integrase strand transfer inhibitors (INSTIs)<br>  ○ Protease inhibitors (PIs)<br>• Documentation of current (within the past 30 days) HIV-1 RNA viral load of at least 200 copies/mL |
| **Appropriate Treatment Regimen & Other Criteria:** | • Prescribed in combination with an optimized background antiretroviral regimen<br><br>**Reauthorization** requires all of the following:<br>• Treatment plan includes continued use of optimized background antiretroviral regimen<br>• Documentation of treatment success as evidenced by one of the following:<br>  ○ Reduction in viral load from baseline or maintenance of undetectable viral load<br>  ○ Absence of postbaseline emergence of ibalizumab resistance-associated mutations confirmed by resistance testing |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an infectious disease or HIV specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization 12 months, unless otherwise specified |

POLICY NAME:
**TUCATINIB**
Affected Medications: TUKYSA (tucatinib)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• Documentation of RAS wild-type, human epidermal growth factor receptor-2 (HER2) positive, unresectable or metastatic colorectal cancer that has progressed following treatment with fluoropyrimidine, oxaliplatin, and irinotecan-based chemotherapy<br>**OR**<br>• Advanced, unresectable or metastatic, HER2-positive breast cancer with prior treatment of 1 or more anti-HER2-based regimens in the metastatic setting |
| **Appropriate Treatment Regimen & Other Criteria:** | **Colorectal cancer**<br>• Documented intolerable adverse event to Lapatinib<br><br>**Reauthorization:** documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• Colorectal cancer ONLY: previous treatment with a HER2 inhibitor |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**TYVASO**
Affected Medications: TYVASO (treprostinil inhalation)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1<br>  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 3 |
| **Required Medical Information:** | **<u>Pulmonary Arterial Hypertension (PAH) WHO Group 1</u>**<br>• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:<br>  ○ Mean pulmonary artery pressure of at least 20 mm Hg<br>  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg<br>  ○ Pulmonary vascular resistance of at least 2.0 Wood units<br>• Etiology of PAH: idiopathic PAH, hereditary PAH, OR<br>• PAH secondary to one of the following conditions:<br>  ○ Connective tissue disease<br>  ○ Human immunodeficiency virus (HIV) infection<br>  ○ Drugs<br>  ○ Congenital left to right shunts<br>  ○ Schistosomiasis<br>  ○ Portal hypertension<br>• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class III or higher symptoms<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blockers) unless there are contraindications:<br>  ○ Low systemic blood pressure (systolic blood pressure less than 90)<br>  ○ Low cardiac index<br>  OR<br>  ○ Presence of severe symptoms (functional class IV)<br><br>**<u>Pulmonary Hypertension Associated with Interstitial Lung Disease WHO Group 3</u>**<br>• Documentation of diagnosis of idiopathic pulmonary fibrosis confirmed by presence of usual interstitial pneumonia (UIP) on high resolution computed tomography (HRCT), and/or surgical lung biopsy<br>OR<br>• Pulmonary fibrosis and emphysema<br>OR<br>• Connective tissue disorder |
| **Appropriate Treatment Regimen & Other Criteria:** | • The pulmonary hypertension has progressed despite maximal medical and/or surgical treatment of the identified condition<br>• Documentation that treprostinil is used as a single route of administration (Remodulin, Tyvaso, Orenitram should not be used in combination)<br><br>**<u>WHO Group 1 only:</u>**<br>• Treatment with oral calcium channel blocking agents has been tried and failed, or has been considered and ruled out |

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| | • Treatment with combination of endothelin receptor antagonist (ERA) and phosphodiesterase 5 inhibitor (PDE5I) has been tried and failed for WHO functional class II and III<br>  ○ Ambrisentan and tadalafil<br>  ○ Bosentan and riociguat<br>  ○ Macitentan and sildenafil<br><br>**Reauthorization** requires documentation of treatment success defined as one or more of the following:<br>• Improvement in walking distance<br>• Improvement in exercise ability<br>• Improvement in pulmonary function<br>• Improvement or stability in WHO functional class |
| **Exclusion Criteria:** | • PAH secondary to pulmonary venous hypertension such as left sided atrial or ventricular disease, left sided valvular heart disease, or disorders of the respiratory system such as chronic obstructive pulmonary disease, obstructive sleep apnea or other sleep disordered breathing, alveolar hypoventilation disorders, etc. |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months unless otherwise specified<br>• Reauthorization: 12 months unless otherwise specified |

POLICY NAME:
**UBLITUXIMAB-XIIY**
Affected Medications: BRIUMVI (ublituximab-xiiy)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:<br>  ▪ Clinically isolated syndrome (CIS)<br>  ▪ Relapsing-remitting multiple sclerosis (RRMS)<br>  ▪ Active secondary progressive multiple sclerosis (SPMS) |
| **Required Medical Information:** | **<u>Relapsing Forms of MS</u>**<br>• Diagnosis confirmed with magnetic resonance imaging (MRI) per revised McDonald diagnostic criteria for MS<br>  ○ Clinical evidence alone will suffice; additional evidence desirable, but must be consistent with MS |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br>• Documentation of one of the following:<br>  ○ Documented disease progression or intolerance to rituximab (preferred products: Riabni and Ruxience)<br>  ○ Currently receiving treatment with Briumvi, excluding via samples or manufacturer's patient assistance program<br><br>**<u>Reauthorization</u>** requires documentation of treatment success |
| **Exclusion Criteria:** | • Active hepatitis B infection<br>• Concurrent use of disease-modifying medications indicated for the treatment of multiple sclerosis |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or a multiple sclerosis specialist<br>• All approved are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**UPNEEQ**
Affected Medications: UPNEEQ (oxymetazoline opthalmic solution)

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| **Covered Uses:** | • Upneeq (oxymetazoline opthalmic solution) is not considered medically necessary due to insufficient evidence of therapeutic value. |
| **Required Medical Information:** | |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | |
| **Coverage Duration:** | |

POLICY NAME:
**VAGINAL PROGESTERONE**
Affected Medications: FIRST-PROGESTERONE VGS 100 MG, FIRST-PROGESTERONE VGS 200 MG

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| **Covered Uses:** | • Prevention of preterm birth in pregnancy |
| **Required Medical Information:** | • Documentation of a current pregnancy with one or more risk factor(s) for preterm birth, including but not limited to:<br>  ○ Ethnicity (e.g., African American, American Indian/Alaska Native)<br>  ○ Lifestyle factors (e.g., smoking, drinking alcohol, using illegal drugs)<br>  ○ Being underweight or obese before pregnancy<br>  ○ Prior preterm delivery<br>  ○ Having multiple gestations (e.g., twins, triplets)<br>  ○ Short time period between pregnancies (less than 6 months between a birth and the beginning of the next pregnancy)<br>• Documentation of a short cervix (defined as cervical length less than or equal to 25 mm) confirmed by ultrasound<br>• Current week of gestation and estimated delivery date |
| **Appropriate Treatment Regimen & Other Criteria:** | • May continue until completion of 36 weeks gestation |
| **Exclusion Criteria:** | • Treatment of infertility |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a gynecologist or obstetrician<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: up to 6 months, unless otherwise specified |

POLICY NAME:
**VALOCTOCOGENE ROXAPARVOVEC-RVOX**
Affected Medications: ROCTAVIAN (valoctocogene roxaparvovec-rvox) - Available on Medical Benefit only

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Hemophilia A (Factor VIII deficiency) |
| **Required Medical Information:** | • Documentation of diagnosis of Hemophilia A<br>• Documentation of current testing with negative results for active factor VIII inhibitors on 2 consecutive occasions (at least one week apart within the past 12 months) and is not receiving a bypassing agent (e.g., Feiba)<br>• Documentation of baseline circulating level of factor with Factor VIII activity level equal to or less than 1 IU/dL or 1% endogenous factor VIII<br>• Evidence of any bleeding disorder NOT related to hemophilia A has been ruled out<br>• No detectable antibodies to AAV5 as determined by an FDA-approved/CLIA-compliant test<br>• Has received stable dosing of prophylactic Factor VIII replacement therapy on a regular basis for at least 1 year<br>• Baseline lab values (must be less than 2 times upper limit of normal):<br>  ○ ALT<br>  ○ AST<br>  ○ Total bilirubin<br>  ○ Alkaline phosphatase (ALP) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Dosing</u>**<br>• $6 \times 10^{13}$ vector genomes/kg (which is 3 mL/kg) as a single one-time dose |
| **Exclusion Criteria:** | • History of or current presence of Factor VIII inhibitors<br>• Prior gene therapy administration<br>• Active Hepatitis B or C infection or other active acute or uncontrolled chronic infection<br>• Cirrhosis<br>• Female gender at birth<br>• Allergy to mannitol |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation, with a hematologist or specialist with experience in treatment of hemophilia<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 2 months (one time infusion), unless otherwise specified |

POLICY NAME:
**VAMOROLONE**
Affected Medications: AGAMREE (vamorolone)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Duchenne muscular dystrophy (DMD) in patients 2 years of age and older |
| **Required Medical Information:** | • Confirmation of Duchenne muscular dystrophy (DMD) diagnosis by genetic testing or biopsy showing lack of muscle dystrophin<br>• Documentation of being ambulatory without needing an assistive device such as a wheelchair, walker, or cane<br>• Baseline motor function assessment from one of the following:<br>  ○ Time to Stand Test (TTSTAND)<br>  ○ 6-minute walk test<br>  ○ North Star Ambulatory Assessment (NSAA)<br>  ○ Motor Function Measure (MFM)<br>  ○ Hammersmith Functional Motor Scale (HFMS)<br>• Patient weight and planned treatment regimen |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with a 6-month trial of prednisone, or intolerable adverse event causing one of the following:<br>  ○ Clinically significant weight gain defined as greater than or equal to 10% of body weight gain over a 6-month period<br>  ○ Psychiatric/behavioral issues (e.g., abnormal behavior, aggression, irritability) that persists beyond the first six weeks of prednisone treatment<br><br>**Reauthorization** requires a documented improvement from baseline or stabilization of motor function demonstrated by a motor function assessment tool |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 2 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**VARIZIG**
Affected Medications: VARIZIG (varicella zoster immune globulin (human) IM injection)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ For post exposure prophylaxis of varicella in high-risk individuals |
| **Required Medical Information:** | Documentation of immunocompromised patient, defined as:<br>• Newborns of mothers with signs and symptoms of varicella shortly before or after delivery (five days before to two days after delivery)<br>• Hospitalized premature infants born at least 28 weeks of gestation who are exposed during their hospitalization and whose mothers do not have evidence of immunity<br>• Hospitalized premature infants less than 28 weeks of gestation or who weigh 1000 grams or less at birth and were exposed to varicella during hospitalization, regardless of mother's immunity status to varicella<br>• Immunocompromised children and adults who lack evidence of immunity to varicella<br>• Pregnant women who lack evidence of immunity to varicella<br>  ○ Lack evidence of immunity to varicella is defined as: those who are seronegative for varicella zoster antibodies OR those with unknown history of varicella |
| **Appropriate Treatment Regimen & Other Criteria:** | • If repeat dose is necessary due to re-exposure, use more than 3 weeks after initial administration |
| **Exclusion Criteria:** | • Coagulation disorders |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 6 months, unless otherwise specified |

POLICY NAME:
**VELMANASE ALFA-TYCV**
Affected Medications: LAMZEDE (velmanase alfa-tycv)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ The treatment of non-central nervous system manifestations of alpha-mannosidosis |
| **Required Medical Information:** | • Diagnosis of alpha-mannosidosis (AM) confirmed by enzyme assay demonstrating alpha-mannosidase activity less than 10% of normal activity<br>• Documentation of symptoms consistent with AM such as hearing impairment, difficulty walking, skeletal abnormalities, or intellectual disabilities |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** will require documentation of treatment success such as improvement in motor function, forced vital capacity (FVC), or reduction in frequency of infections |
| **Exclusion Criteria:** | • AM with only central nervous system manifestations and no other symptoms |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care<br>• Prescribed by, or in consultation with, a specialist familiar with the treatment of lysosomal storage disorders |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**VERTEPORFIN**
Affected Medications: VISUDYNE (verteporfin)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of predominantly classic subfoveal choroidal neovascularization (CNV) due to one of the following:<br>  ▪ Age-related macular degeneration (AMD)<br>  ▪ Pathologic myopia<br>  ▪ Presumed ocular histoplasmosis |
| **Required Medical Information:** | • Documented diagnosis of subfoveal CNV due to one of the following:<br>  ○ Neovascular AMD<br>  ○ Pathologic myopia<br>  ○ Presumed ocular histoplasmosis<br>• Documentation of current body surface area (BSA) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Neovascular AMD and Pathologic Myopia</u>**<br>• Documentation of one of the following:<br>  ○ Currently receiving treatment with Visudyne, excluding via samples or manufacturer's patient assistance program<br>  ○ Documented treatment failure or intolerance following a minimum 3-month trial with both of the following: Avastin and ranibizumab (preferred product: Byooviz)<br><br>**Dosing**<br>• 6 mg/$m^2$ BSA<br>  ○ Every 3 month dosing is permitted with evidence of choroidal neovascular leakage (see reauthorization criteria)<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br><br>**Reauthorization** requires documentation of the following:<br>• Positive response to therapy (e.g., improved or stable visual acuity, reduced central macular thickness)<br>• Evidence of recurrent or persistent leakage on fluorescein angiogram or optical coherence tomography (OCT), performed at least 3 months after the last treatment |
| **Exclusion Criteria:** | • Concurrent therapy with vascular endothelial growth factor (VEGF) inhibitors<br>• Treatment of non-neovascular (dry) AMD |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**VIGABATRIN**
Affected Medications: VIGABATRIN, VIGADRONE (vigabatrin)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Refractory complex partial seizures (focal seizures with impaired awareness)<br>  ○ Infantile spasms |
| **Required Medical Information:** | **<u>Infantile Spasms</u>**<br>• Used as monotherapy for pediatric patients (1 month to 2 years of age)<br><br>**<u>Refractory Complex Partial Seizures (focal seizures with impaired awareness)</u>**<br>• Used as adjunctive therapy only |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Refractory complex partial seizures (focal seizures with impaired awareness)</u>**<br>• Documentation of treatment failure with at least 2 alternative therapies: carbamazepine, phenytoin, levetiracetam, topiramate, oxcarbazepine, or lamotrigine<br><br>**<u>Reauthorization</u>** will require documentation of treatment success and a reduction in seizure severity, frequency, and/or duration |
| **Exclusion Criteria:** | • Use as a first line agent for complex partial seizures (focal seizures with impaired awareness) |
| **Age Restriction:** | • Infantile Spasms: 1 month to 2 years of age<br>• Refractory complex partial seizures (focal seizures with impaired awareness): greater than 2 years of age |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **<u>Infantile Spasms</u>**<br>• Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months (or up to 2 years of age), unless otherwise specified<br><br>**<u>Refractory Complex Partial Seizures (focal seizures with impaired awareness)</u>**<br>• Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**VIJOICE**
Affected Medications: VIJOICE (alpelisib)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of severe manifestations of PIK3CA-related overgrowth spectrum (PROS) in patients who require systemic therapy |
| **Required Medical Information:** | • Documented diagnosis of PROS, to include any of the following:<br>  ○ CLAPOS syndrome<br>  ○ CLOVES syndrome<br>  ○ Diffuse capillary malformation with overgrowth (DCMO)<br>  ○ Dysplastic megalencephaly (DMEG)<br>  ○ Facial infiltrating lipomatosis (FIL)<br>  ○ Fibroadipose hyperplasia (FAH)/fibroadipose overgrowth (FAO)/hemihyperplasia multiple lipomatosis (HHML) syndrome<br>  ○ Fibroadipose vascular anomaly (FAVA)<br>  ○ Hemimegalencephaly (HMEG)<br>  ○ Klippel-Trenaunay syndrome (KTS)<br>  ○ Lipomatosis of nerve (LON)<br>  ○ Megalencephaly-capillary malformation (MCAP) syndrome<br>  ○ Muscular hemihyperplasia (HH)<br>• Documentation of PIK3CA gene mutation<br>• Documentation of clinical manifestations that were assessed by the treating provider as severe or life-threatening and necessitating systemic treatment<br>• Documentation that clinical manifestations are a direct result of a lesion that is both of the following:<br>  ○ Inoperable, as defined by the treating provider<br>  ○ Causing functional impairment<br>• Documentation of one or more target lesion(s) identified on imaging within 6 months prior to request, including location(s) and volume of lesion(s) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Treatment failure (or intolerable adverse event) with sirolimus for at least 6 months at a dose of at least 2 mg daily in patients with lymphatic, venous, or combined manifestations of disease<br><br>**Reauthorization** will require documentation of both of the following:<br>  ○ Radiological response, defined as greater than or equal to a 20% reduction from baseline in the sum of measurable target lesion volume, confirmed by at least one subsequent imaging assessment<br>  ○ Absence of greater than or equal to a 20% increase from baseline in any target lesion, progression of non-target lesions, or appearance of a new lesion |
| **Exclusion Criteria:** | • Treatment of PIK3CA-mutated conditions other than PROS |
| **Age Restriction:** | • 2 years of age and older |

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| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist with experience in the treatment of PROS<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**VISTOGARD**
Affected Medications: VISTOGARD (uridine triacetate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ For the emergency treatment of adult and pediatric patients:<br>  ▪ Following a fluorouracil or capecitabine overdose regardless of the presence of symptoms, **OR**<br>  ▪ Who exhibit early-onset, severe, or life-threatening toxicity affecting the cardiac or central nervous system, and/or early-onset, unusually severe adverse reactions (e.g., gastrointestinal toxicity and/or neutropenia) within 96 hours following the end of fluorouracil or capecitabine administration |
| **Required Medical Information:** | • Documentation of fluorouracil or capecitabine administration<br>• Documentation of overdose **OR** early-onset, severe adverse reaction, or life-threatening toxicity |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing is in accordance with FDA labeling |
| **Exclusion Criteria:** | • Non-emergent treatment of adverse events associated with fluorouracil or capecitabine<br>• Use more than 96 hours following the end of fluorouracil or capecitabine administration |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 7 days, unless otherwise specified |

POLICY NAME:
**VMAT2 INHIBITORS**
Affected Medications: TETRABENAZINE, AUSTEDO (deutetrabenazine), AUSTEDO XR (deutetrabenazine)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved and compendia supported indications not otherwise excluded by plan design<br>  ○ Chorea associated with Huntington's disease<br>  ○ Tardive dyskinesia |
| **Required Medical Information:** | **Chorea related to Huntington's Disease**<br>• Diagnosis of Huntington's Disease with Chorea requiring treatment<br><br>**Tardive Dyskinesia**<br>• Diagnosis of moderate to severe tardive dyskinesia including all of the following:<br>  ○ A history of at least one month of ongoing or previous dopamine receptor-blocking agent exposure<br>  ○ Presence of dyskinetic or dystonic involuntary movements that developed either while exposed to a dopamine receptor-blocking agent, or within 4 weeks of discontinuation from an oral agent (8 weeks from a depot formulation)<br>  ○ Other causes of abnormal movements have been excluded<br>• Baseline evaluation of the condition using one of the following:<br>  ○ Abnormal Involuntary Movement Scale (AIMS)<br>  ○ Extrapyramidal Symptom Rating Scale (ESRS) |
| **Appropriate Treatment Regimen & Other Criteria:** | **Tardive Dyskinesia**<br>• Persistent dyskinesia despite dose reduction or discontinuation of the offending agent<br>**OR**<br>• Documented clinical inability to reduce dose or discontinue the offending agent<br><br>**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy<br>• Tardive Dyskinesia: must include an improvement in AIMS or ESRS score from baseline |
| **Exclusion Criteria:** | • Use for Huntington's comorbid with untreated or inadequately treated depression or actively suicidal<br>• Concomitant use with another VMAT2 inhibitor or reserpine<br>• Hepatic impairment |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or psychiatrist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**VOCLOSPORIN**
Affected Medications: LUPKYNIS CAPSULE 7.9 MG ORAL

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| 1. Is the request for continuation of therapy currently approved through insurance? | Yes – Go to renewal criteria | No – Go to #2 |
| 2. Is the request to treat a diagnosis according to the Food and Drug Administration (FDA)-approved indication?<br>a. For use in combination with a background immunosuppressive therapy regimen for the treatment of adult patients with active lupus nephritis | Yes – Go to appropriate section below | No – Criteria not met |
| **Lupus Nephritis (LN)** | | |
| 1. Is there documented International Society of Nephrology/Renal Pathology Society (ISN/RPS) biopsy-proven active class III, IV and/or V disease? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Are there documented current baseline values (within the last 3 months) for all of the following?<br>a. Estimated glomerular filtration rate (eGFR)<br>b. Urine protein to creatinine ratio (uPCR)<br>c. Blood pressure | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documented treatment failure with at least 12 weeks of standard therapy with both mycophenolate mofetil (MMF) AND cyclophosphamide? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is there documented treatment failure with at least 12 weeks of subcutaneous Benlysta? | Yes – Document and go to #5 | No – Criteria not met |
| 5. Will Lupkynis be used in combination with MMF and corticosteroids or other background immunosuppressive therapy, other than cyclophosphamide? | Yes – Document and go to #6 | No – Criteria not met |
| 6. Is the drug prescribed by, or in consultation with, a rheumatologist, immunologist, nephrologist or kidney specialist? | Yes – Go to #7 | No – Criteria not met |
| 7. Is the requested dose within the Food and Drug Administration (FDA)-approved label and PacificSource quantity limitations? | Yes – Approve up to 12 months | No – Criteria not met |
| **Renewal Criteria** | | |

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| 1. Is there documentation of treatment success defined as an increase in eGFR, decrease in uPCR, or decrease in flares and corticosteroid use? | Yes – Go to #2 | No – Criteria not met |
| 2. Is the requested dose within the Food and Drug Administration (FDA)-approved label and PacificSource quantity limitations? | Yes – Approve up to 12 months | No – Criteria not met |
| **Quantity Limitations** | | |
| • **Lupkynis**<br>  ○ Starting dose: 23.7 mg twice daily (BID)<br>  ○ Starting dose must be reduced in the below situations as follows:<br>  ▪ eGFR 45 mL/min/1.73 $m^2$ or less at initiation: 15.8 mg BID<br>  ▪ Mild-to-moderate hepatic impairment (Child-Pugh A or B): 15.8 mg BID<br>  ▪ Concomitant use with moderate CYP3A4 inhibitors: 15.8 mg in morning and 7.9 mg in afternoon. | | |

POLICY NAME:
**VORETIGENE NEPARVOVEC**
Affected Medications: LUXTURNA (Voretigene neparvovec-rzyl intraocular suspension for subretinal injection)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ Inherited Retinal Dystrophies (IRD) caused by mutations in the retinal pigment epithelium-specific protein 65kDa (RPE65) gene |
| **Required Medical Information:** | • Diagnosis of a confirmed biallelic RPE65 mutation-associated retinal dystrophy (e.g., Leber's congenital amaurosis [LCA], Retinitis pigmentosa [RP], Early Onset Severe Retinal Dystrophy [EOSRD], etc.)<br>• Genetic testing documenting biallelic mutations of the RPE65 gene<br>• Visual acuity of at least 20/800 OR have remaining light perception in the eye(s) receiving treatment<br>• Visual acuity of less than 20/60 OR a visual field of less than 20 degrees<br>• Presence of neural retina and a retinal thickness greater than 100 microns within the posterior pole as assessed by optical coherence tomography with AND have sufficient viable retinal cells as assessed by the treating physician |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | • Patient has been previously enrolled in clinical trials of gene therapy for retinal dystrophy RPE65 mutations or has been previously treated with gene therapy for retinal dystrophy in the eye(s) receiving treatment<br>• Patient has other pre-existing eye conditions or complicating systemic diseases that would eventually lead to irreversible vision loss and prevent the patient from receiving full benefit from treatment (e.g., severe diabetic retinopathy) |
| **Age Restriction:** | • 12 months of age and older |
| **Prescriber/Site of Care Restrictions:** | • Ophthalmologist or retinal surgeon with experience providing sub-retinal injections |
| **Coverage Duration:** | • Authorization: 1 month - 1 injection per eye per lifetime, unless otherwise specified |

POLICY NAME:
**VOSORITIDE**
Affected Medications: VOXZOGO (vosoritide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ To increase linear growth in pediatric patients with achondroplasia with open epiphyses |
| **Required Medical Information:** | • Diagnosis of achondroplasia confirmed by molecular genetic testing showing a mutation in the fibroblast growth factor receptor type 3 (FGFR3) gene<br>• Baseline height, growth velocity, and patient weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of all the following:<br>  ○ Evaluation of epiphyses (growth plates) documenting they are open<br>  ○ Growth velocity greater than or equal to 1.5 cm/yr<br><br>**<u>Reauthorization:</u>**<br>• Evaluation of epiphyses (growth plates) documenting they remain open<br>• Growth velocity greater than or equal to 1.5 cm/yr |
| **Exclusion Criteria:** | • Hypochondroplasia<br>• Other short stature condition other than achondroplasia<br>• Evidence of growth plate closure |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a pediatric orthopedist, endocrinologist, or a provider with experience in treating skeletal dysplasia<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 12 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**XGEVA**
Affected Medications: XGEVA (denosumab), WYOST (denosumab-bbdz), OSENVELT (denosumab-bmwo), BOMNYTRA (denosumab-bnht), BILPREVDA (denosumab-nxxp)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ One of these diagnoses:<br>  ▪ Giant cell tumor<br>  ▪ Bone metastases from solid tumors<br>  ▪ Hypercalcemia of malignancy<br>  ▪ Multiple myeloma<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | • **Giant cell tumor**<br>  ○ Unresectable disease or surgical resection would likely result in severe morbidity.<br>• **Bone metastases from solid tumors**<br>• **Hypercalcemia of malignancy**<br>  ○ Refractory to bisphosphonate therapy or contraindication<br>• **Multiple myeloma**<br>  ○ Requires failure of zoledronic acid or pamidronate OR creatinine clearance less than 30 mL/min |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • **Giant cell tumor**: Adults and adolescents at least 12 years of age and skeletally mature weighing at least 45 kg<br>• **All other indications**: 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**XIAFLEX**
Affected Medications: XIAFLEX (collagenase clostridium histolyticum)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Dupuytren's contracture with a palpable cord<br>  ○ Peyronie's disease |
| **Required Medical Information:** | **Peyronie's disease:**<br><br>• Documented diagnosis of Peyronie's disease with a palpable plaque<br>• Curvature deformity is at least 30 degrees at the start of therapy<br>• Documentation of stable disease defined as symptoms that have remained unchanged for at least 3 months |
| **Appropriate Treatment Regimen & Other Criteria:** | **Dupuytren's:**<br><br>• Authorization will be limited per joint as follows: One injection per month for a maximum of three injections per cord<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy<br><br>**Peyronie's disease:**<br><br>• One treatment cycle consists of two Xiaflex injection procedures<br><br>**Reauthorization** for additional treatment cycles may be given if the curvature deformity is more than 15 degrees after the first, second or third treatment cycle, or if the prescribing healthcare provider determines that further treatment is clinically indicated<br>• Maximum of 4 treatment cycles per plaque, administered at 6-week intervals |
| **Exclusion Criteria:** | • Peyronie's plaques that involve the penile urethra |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • **Peyronie's**: prescribed by, or in consultation with, a urologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • **Dupuytren's**: 12 weeks, unless otherwise specified<br>• **Peyronie's**: 6 weeks, unless otherwise specified |

POLICY NAME:
**XIFAXAN**
Affected Medications: XIFAXAN (rifaximin)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Prevention of hepatic encephalopathy (HE)<br>  ○ Treatment of Travelers' Diarrhea caused by noninvasive strains of *Escherichia coli (E. coli)*<br>  ○ Treatment of Irritable Bowel Syndrome with Diarrhea (IBS-D)<br>• Compendia-supported uses that will be covered (if applicable)<br>  ○ Treatment of HE<br>  ○ Treatment of recurrent Clostridium difficile (C. diff)-associated diarrhea<br>  ○ Treatment of Small Intestinal Bacterial Overgrowth (SIBO) |
| **Required Medical Information:** | • Documentation of complete & current treatment course required<br>• Documentation of E-coli bacterial cultures for travelers' diarrhea<br>• Previous antibiotic history and documented allergies/hypersensitivity |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Recurrent C. diff</u>**<br>• Documentation confirming a current diagnosis of recurrent C. diff infection (CDI) with **ALL** of the following:<br>  ○ CDI symptoms resolved on prior appropriate therapy and have reappeared within 8 weeks of completing prior therapy<br>  ○ Presence of at least 3 unformed stools in 24 hours<br>  ○ Positive stool test for toxigenic Clostridium difficile<br>• Documented treatment failure with oral vancomycin<br><br>**<u>HE</u>**<br>• Documented treatment failure with at least 1 month of lactulose therapy defined as continued altered mental status or elevated ammonium levels despite adequate upward titration<br><br>**<u>Travelers' Diarrhea</u>**<br>• Documentation of **ALL** of the following:<br>  ○ Travelers' diarrhea is caused by noninvasive strains of E. coli<br>  ○ Systemic signs of infection (fever or blood in stool) are not present<br>  ○ Member is returning from an area of high fluoroquinolone resistance<br>• Documented treatment failure with a fluoroquinolone (e.g., ciprofloxacin, levofloxacin) and azithromycin<br><br>**<u>SIBO</u>**<br>• Documented diagnosis confirmed by a carbohydrate breath test<br>• Documented treatment failure with trial of at least one of the following antibiotics: amoxicillin/clavulanic acid, ciprofloxacin, metronidazole<br><br>**<u>IBS-D</u>**<br>• Documentation confirming a Rome IV diagnosis with recurrent abdominal pain, on average, at least one day per week in the last 3 months, associated with two or more of the following:<br>  ○ Related to defecation |

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| | ○ Associated with a change in stool frequency<br>○ Associated with a change in stool form (appearance)<br>• Symptom onset at least six months prior to diagnosis<br>• Documented treatment failure with **ALL** of the following:<br>○ Loperamide<br>○ Dicyclomine or hyoscyamine<br>○ Tricyclic antidepressant (e.g., amitriptyline, nortriptyline)<br><br>• **Retreatment criteria for IBS-D**: Patient must have responded to the initial treatment for at least 4 weeks with either greater than or equal to 30% improvement from baseline in the weekly average abdominal pain score OR at least a 50% reduction in number of days in a week with a daily stool consistency of Bristol Stool Scale type 6 or 7 compared with baseline (6: fluffy pieces with ragged edges, a mushy stool; 7: watery stool, no solid pieces; entirely liquid). Retreatment can be approved when recurrence of symptoms (abdominal pain or mushy/watery stool consistency) occur for 3 weeks of a rolling 4-week period. Retreatment can be approved twice per lifetime.<br><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | **Recurrent C. diff**<br>• Xifaxan exceeding 400 mg three times per day for 20 days<br><br>**HE**<br>• Xifaxan exceeding the recommended dose of 550 mg twice daily or 400 mg 3 times daily for the treatment or prevention of hepatic encephalopathy<br><br>**Travelers' Diarrhea**<br>• Xifaxan exceeding 200 mg three times per day for total of 3 days<br>• Diarrhea complicated by fever or bloody stool, or caused by bacteria other than noninvasive strains of E. coli<br><br>**SIBO**<br>• Xifaxan exceeding 550 mg three times per day for 14 days<br><br>**IBS-D**<br>• Mild cases of irritable bowel syndrome or diagnosis of irritable bowel syndrome with constipation<br>• Xifaxan exceeding 550 mg three times per day for 14 days |
| **Age Restriction:** | • 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | **Recurrent C. diff**<br>• Authorization: 20 days, unless otherwise specified<br>**HE**<br>• Authorization: 12 months, unless otherwise specified<br>**Travelers' Diarrhea**<br>• Authorization: 7 days, unless otherwise specified<br>**SIBO**<br>• Authorization: 14 days, unless otherwise specified (one treatment per lifetime) |

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| | **IBS-D**<br>• Authorization: 14 days, unless otherwise specified (maximum of 3 treatment courses per lifetime) |

POLICY NAME:
**XURIDEN**
Affected Medications: XURIDEN (uridine triacetate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Hereditary orotic aciduria |
| **Required Medical Information:** | • Diagnosis of hereditary orotic aciduria confirmed by **ONE** of the following:<br>  ○ Molecular genetic testing confirming biallelic pathogenic mutation in the UMPS gene<br>  ○ Urinary orotic acid level above the normal reference range<br>  ○ Clinical manifestations consistent with disease such as:<br>  ▪ Megaloblastic anemia<br>  ▪ Leukopenia<br>  ▪ Developmental delays<br>  ▪ Failure to thrive |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** requires documentation of treatment success based on **ONE** of the following:<br>• Improvement of hematologic abnormalities such as megaloblastic anemia and leukopenia<br>• Reduction of urinary orotic acid levels |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a metabolic specialist or geneticist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**YONSA**
Affected Medications: YONSA (abiraterone)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented inadequate response or intolerable adverse event with the preferred product abiraterone acetate<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Child-Pugh Class C<br>• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ZANIDATAMAB**
Affected Medications: ZIIHERA (zanidatamab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• Documentation that Ziihera will be administered as monotherapy.<br>• Documentation of previously treated unresectable or metastatic human epidermal growth factor receptor 2 (HER2)-positive biliary tract cancer (BTC) that has progressed following at least 1 prior systemic therapy<br>• Documentation of HER2 positivity with a score of 3+ on immunohistochemistry (IHC) testing |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure or intolerable adverse event with Enhertu (fam-trastuzumab deruxtecan)<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist.<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ZILUCOPLAN**
Affected Medications: ZILBRYSQ (zilucoplan)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive |
| **Required Medical Information:** | • Diagnosis of generalized myasthenia gravis (gMG) confirmed by one of the following:<br>  ○ A history of abnormal neuromuscular transmission test<br>  ○ A positive edrophonium chloride test<br>  ○ Improvement in gMG signs or symptoms with an acetylcholinesterase inhibitor<br>• Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV<br>• Positive serologic test for AChR antibodies<br>• MG-Activities of Daily Living (MG-ADL) total score of 6 or greater **OR** Quantitative Myasthenia Gravis (QMG) total score of 12 or greater |
| **Appropriate Treatment Regimen & Other Criteria:** | • Currently on a stable dose of at least one gMG therapy (acetylcholinesterase inhibitor, corticosteroid, or non-steroidal immunosuppressive therapy (NSIST)) that will be continued during initial treatment with Zilbrysq<br>• Documentation of one of the following:<br>  ○ Treatment failure with an adequate trial (one year or more) of at least two immunosuppressive therapies (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)<br>  ○ Has required three or more courses of rescue therapy (plasmapheresis/plasma exchange and/or intravenous immunoglobulin), while on at least one immunosuppressive therapy, over the last 12 months<br><br>**<u>Reauthorization</u>:**<br>• Documentation of treatment success and clinically significant response to therapy defined as:<br>  ○ A minimum 2-point reduction in MG-ADL score from baseline AND<br>  ○ Absent or reduced need for rescue therapy compared to baseline<br>• That the patient requires continuous treatment, after an initial beneficial response, due to new or worsening disease activity |
| **Exclusion Criteria:** | • Current or recent systemic infection within 2 weeks<br>• Concurrent use with other biologics (rituximab, eculizumab, IVIG, etc) |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ZOPAPOGENE IMADENOVEC-DRBA**
Affected Medications: PAPZIMEOS (zopapogene imadenovec-drba)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ The treatment of adults with recurrent respiratory papillomatosis (RRP) |
| **Required Medical Information:** | • Histologically confirmed diagnosis from a CLIA-certified (or comparable) laboratory report<br>• More than or equal to 3 surgeries in the previous 12 months to remove papillomas<br>• Documentation of laryngotracheal papillomas<br>• Documentation of human papillomavirus (HPV) vaccination |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with bevacizumab or cidofovir. |
| **Exclusion Criteria:** | • Prior use of Papzimeos (zopapogene imadenovec-drba) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 weeks with no reauthorization, unless otherwise specified |

POLICY NAME:
**ZUSDURI**
Affected Medications: ZUSDURI (mitomycin thermal hydrogel)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• Recurrent low-grade intermediate-risk non–muscle-invasive bladder cancer (LG-IR-NMIBC) confirmed by cystoscopy and pathology. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented recurrence after prior transurethral resection of bladder tumor (TURBT) AND<br>• Clinical justification for TURBT ineligibility (e.g. high anesthesia risk, complex anatomy or prior complications with resection) |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• Use in low-risk or high-grade non–muscle-invasive bladder cancer (NMBIC) |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist or urologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 4 months (no reauthorization), unless otherwise specified |