# 2023 PacificSource Health Plans Prior Authorization Criteria

Last Modified: 3/22/2023
(All criteria reviewed at least once per year)

PacificSource HEALTH PLANS

## Contents

POLICY NAME:
**ABILIFY MAINTENA**
Affected Medications: ABILIFY MAINTENA (aripiprazole suspension, reconstituted)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design |
| **Required Medical Information:** | • Diagnosis of schizophrenia and on maintenance treatment OR Diagnosis of bipolar I disorder and on maintenance treatment **AND**<br>• Documentation of established tolerability to oral aripiprazole. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented failure or contraindication to Risperdal Consta<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Psychiatrist or receiving input from a psychiatry practice<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:
**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>**<u>Chronic granulomatous disease</u>**<br>• Diagnosis established by a molecular genetic test identifying a gene-related mutation associated with CGD<br><br>**<u>Severe, malignant osteopetrosis</u>**<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>**<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>Chronic Granulomatous Disease</u>**<br>• Patient is on a prophylactic regimen with an antibacterial agent and an antifungal agent<br><br>**<u>All indications</u>**<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced |

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

POLICY NAME:
**ACTIQ**
Affected Medications: FENTANYL citrate oral transmucosal lozenge

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | • Used to manage breakthrough pain due to a current cancer condition or cancer related complication |
| **Appropriate Treatment Regimen & Other Criteria:** | • A long-acting opioid is being prescribed for around-the-clock treatment of the cancer pain<br>• The patient is opioid tolerant (They have been taking at least 60mg of oral morphine per day, 25mcg/hr of transdermal fentanyl, 30mg of oral oxycodone daily, 8 mg of oral hydromorphone daily, 25mg of oral oxymorphone daily, or an equianalgesic dose of another opioid for ≥ 1 week.<br>• The patient is NOT taking a strong or moderate cytochrome P450 3A4 inhibitor, OR the patient is taking a strong or moderate 3A4 inhibitor and the patient will be carefully monitored and dosage adjustments will be made if necessary. |
| **Exclusion Criteria:** | • Use in the management of acute and/or postoperative pain including surgery/post-surgery, trauma/post-trauma, and acute medical illness (acute abdominal pain, pelvic pain, muscle spasm)<br>• Use as pre-anesthesia (preoperative anxiolysis and sedation and/or supplement to anesthesia) |
| **Age Restriction:** | • 16 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:** | • Approval: 6 months, unless otherwise specified |

POLICY NAME:
**ADCIRCA**
Affected Medications: ALYQ, tadalafil (PAH) 20 mg, TADLIQ

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design. |
| **Required Medical Information:** | • Pulmonary arterial hypertension (PAH) (WHO Group 1) confirmed by right heart catheterization<br>• Etiology of PAH (idiopathic, heritable, or associated with connective tissue disease)<br>• NYHA/WHO Functional Class II or III symptoms<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blocker) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Inadequate response or intolerance to sildenafil citrate (Revatio)<br>• Requests for oral suspension: must also be unable to swallow tablets<br>• Subsequent approvals require documentation of treatment success such as improved walking distance or improvements in functional class |
| **Exclusion Criteria:** | • Concomitant nitrate therapy on a regular or intermittent basis<br>• Concomitant use of Guanylate Cyclase Stimulators (eg. riociguat) |
| **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:** | • 12 months, unless otherwise specified |

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

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| **Covered Uses:** | • Treatment of premenopausal women with a mental health diagnosis of acquired, generalized hypoactive sexual desire disorder (HSDD), also known as female sexual interest/arousal disorder.<br>○ **Acquired** HSDD refers to HSDD that develops in a patient who previously had no problems with sexual desire.<br>○ **Generalized** HSDD refers to HSDD that occurs regardless of the type of stimulation, situation, or partner. |
| **Required Medical Information:** | • Mental health diagnosis according to Diagnostic and Statistical Manual of Mental Disorders, fifth edition (DSM-5) diagnostic criteria for female sexual interest/arousal disorder:<br>A. Lack of, or significantly reduced, sexual interest/arousal, as manifested by at least three of the following:<br>1. Absent/reduced interest in sexual activity.<br>2. Absent/reduced sexual/erotic thoughts or fantasies.<br>3. No/reduced initiation of sexual activity, and typically unreceptive to a partner's attempts to initiate.<br>4. Absent/reduced sexual excitement/pleasure during sexual activity in almost all or all (approximately 75%-100%) sexual encounters (in identified situational contexts or, if generalized, in all contexts).<br>5. Absent/reduced sexual interest/arousal in response to any internal or external sexual/erotic cues (e.g., written, verbal, visual).<br>6. Absent/reduced genital or non-genital sensations during sexual activity in almost all or all (approximately 75%-100%) sexual encounters (in identified situational contexts or, if generalized, in all contexts).<br>B. The symptoms in Criterion A have persisted for a minimum duration of approximately 6 months.<br>C. The symptoms in Criterion A cause clinically significant distress in the individual.<br>D. The sexual dysfunction is not better explained by a nonsexual mental disorder or as a consequence of severe relationship distress (e.g., partner violence) or other |

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| | significant stressors and is not attributable to the effects of a substance/medication or another medical condition. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Addyi<br>  ○ Documentation of current and previous alcohol use<br>  ○ Documentation of appropriate patient counseling regarding alcohol use<br>  ○ 100 mg once daily<br>• Vyleesi<br>  ○ Documentation that patients in heterosexual relationships are using an effective form of contraception<br>  ○ 1.75 mg as needed 45 minutes before anticipated sexual activity<br><br>**<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Diagnosis of HSDD or female sexual interest/arousal disorder without meeting requirements of DSM-5 criteria<br>• Postmenopausal females<br>• Males<br><br>**Addyi**<br>• Hepatic impairment<br>• Concomitant use with moderate/strong CYP3A4 inhibitors<br><br>**Vyleesi**<br>• Uncontrolled hypertension or known cardiovascular disease |
| **Age Restriction:** | • Premenopausal women only |
| **Prescriber/Site of Care Restrictions:** | • Risk Evaluation and Mitigation Strategy (REMS) certified for Addyi<br>• 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:** | **Quantity Limitations**<br>• Addyi<br>  ○ Limited to #1 per day<br>• Vyleesi<br>  ○ Limited to #8 per month<br>• Initial approval: 2 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ADENOSINE DEAMINASE (ADA) REPLACEMENT**
Affected Medications: ADAGEN (pegademase bovine), 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:** | • A confirmed diagnosis of adenosine deaminase severe combined immune deficiency (ADA-SCID)<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 AND<br>• For Revcovi requests, documentation that treatment with Adagen was unsuccessful<br><br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced |
| **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:** | • Immunologist or prescriber experienced in severe combined immune deficiency (SCID)<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Initial approval: 4 months<br>• Reauthorization: 6 months |

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

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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?<br>a. Treatment of patients with Erythropoetic protoporphyria (EPP) with phototoxic reactions | Yes – Go to appropriate section below | No – Criteria not met |
| **Erythropoetic protoporphyria (EPP)** | | |
| 1. Is there documentation of a diagnosis of Erythropoetic protoporphyria confirmed with mutation in the Ferrochelatase (FECH) gene OR mutation of the ALAS2 gene? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documentation of an increase in total erythrocyte protoporphyrin with at least 85% metal-free protoporphyrin? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documented symptoms of erythropoietic protoporphyria phototoxicity that causes dysfunction significantly impacting activities of daily living? | Yes – Document and go to # 4 | No – Criteria not met |
| 4. Is there documented associated neuropathic pain that has not responded to analgesics after a minimum of 12 weeks? | Yes – Document and go to # 5 | No – Criteria not met |

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| 5. Is the drug prescribed and managed by a specialist at a recognized Porphyria Center? | Yes – Approve up to 6 months | No – Criteria not met |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy (e.g. decreased severity and number of phototoxic reactions, increased duration of sun exposure, increased quality of life, etc) as assessed by the prescribing provider? | 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** | | |
| • **Scenesse**<br>  ○ Availability: 16 mg implant.<br>  ○ Dosing: 16 mg under the skin every 2 months (60 days) | | |

POLICY NAME:
**AFINITOR**
Affected Medications: AFINITOR, AFINITOR DISPERZ (everolimus), everolimus (2.5 mg, 5 mg, 7.5 mg, 10 mg), everolimus soluble tablet (2 mg, 3 mg, 5 mg)

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher<br>• Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | **<u>Oncology Indication</u>**<br>• Documentation of performance status, all prior therapies used, and prescribed treatment regimen<br><br>**<u>Tuberous Sclerosis Complex (TSC)-Associated Partial-Onset Seizures</u>**<br>• Documentation of monotherapy failure for seizure control with 2 different antiepileptic regimens AND<br>• Documentation of treatment failure with epidiolex (cannabadiol solution) adjunct therapy<br>• Documentation that therapy is being used as adjunct therapy for seizures |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of medication review and / or avoidance with strong CYP3A4 inhibitors, CYP3A4 inducers, PgP inhibitors<br>• Reauthorization requires documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Hypersensitivity to rapamycin derivatives<br><br>**<u>Oncology Indication</u>**<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: Oncologist<br>• TSC-Associated Partial-Onset Seizures: Neurologist or specialist in the treatment of TSC<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Initial approval: 3 months (2 week initial partial fill), unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**AGALSIDASE BETA**
Affected Medications: FABRAZYME (agalsidase beta)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design. |
| **Required Medical Information:** | • Diagnosis of Fabry disease<br>• Diagnosis confirmed by enzyme assay demonstrating a deficiency of alpha-galactosidase enzyme activity or by DNA testing<br>• The patient has clinical signs and symptoms of Fabry disease.<br>• The patient is male OR<br>• The patient is female and the patient has documented substantial disease manifestations (Renal dysfunction, Cardiovascular dysfunction, Cerebrovascular complications, Pulmonary complications, Neurologic/neuropathic dysfunction (pain) and diagnosis has been confirmed with genetic testing<br>• Patient weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Reauthorization will require documentation of treatment success and 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<br>• Prescribed by or in consultation with a prescriber experienced in the treatment of Fabry disease |
| **Coverage Duration:** | • Initial approval: 4 months, unless otherwise specified<br>• Subsequent approval: 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 benefit design.<br>  ○ Treatment of relapsing forms of multiple sclerosis (MS), to include relapsing-remitting disease and active secondary progressive disease in adults. |
| **Required Medical Information:** | • Diagnosis of a relapsing form of MS confirmed by MRI (Revised McDonald diagnostic criteria for multiple sclerosis) AND<br>• Documentation of inadequate response to Tysabri (natalizumab) AND one additional medication indicated for MS<br><br>**<u>Secondary-Progressive MS (SPMS)</u>**<br>• Documentation of prior history of relapsing-remitting MS (RRMS) with progressive increase in disability over at least 6 months, independent of, or in the absence of, relapses<br>• Documentation of active disease classified as the presence of clinical relapse or inflammatory activity (i.e., new or enlarging T2 lesions or gadolinium enhancing lesions on MRI) in the last 2 years<br>• Documentation of Expanded Disability Status Scale (EDSS) score of 3.0 to 6.5 |
| **Appropriate Treatment Regimen & Other Criteria:** | • Initial dose of 12 mg intravenously (IV) daily on 5 consecutive days.<br>• For second treatment course one year later, 12 mg IV daily on 3 consecutive days.<br>• Subsequent courses (12 mg IV daily on 3 consecutive days) may be administered, as needed, at least 12 months after the last dose of any prior treatment course<br>• Reauthorization requires provider attestation of treatment success |
| **Exclusion Criteria:** | • Patients infected with Human Immunodeficiency Virus (HIV)<br>• Treatment of primary progressive MS |

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| | • Treatment of clinically isolated syndrome<br>• Concurrent use of medications indicated for the treatment of relapsing-remitting MS |
| **Age Restriction:** | • Greater than or equal to 17 years of age |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or 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: For subsequent courses (3 doses for 3 days) following any previous course, unless otherwise specified |

POLICY NAME:
**ALGLUCOSIDASE ALFA**
Affected Medications: LUMIZYME

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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:<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><br>**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • 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 |

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

POLICY NAME:
**ALOSETRON**
Affected Medications: LOTRONEX (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 syndrome 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 of the following:<br>  ○ Dicyclomine<br>  ○ Hyoscyamine<br>  ○ Diphenoxylate-atropine<br>  ○ Amitriptyline or nortriptyline<br>• Reauthorization: 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 or older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care<br>• Gastroenterologist |

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| **Coverage Duration:** | • Initial approval: 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>  • Indicated for chronic augmentation and maintenance therapy in adults with clinical evidence of emphysema due to severe hereditary deficiency of Alpha1-PI (alpha1-antitrypsin deficiency) |
| **Required Medical Information:** | • Documentation of severe alpha1-antitrypsin (AAT) deficiency with emphysema or Chronic Obstructive Pulmonary Disease (COPD) that includes ALL of the following:<br>  ○ Baseline (pretreatment) alpha1-antitrypsin serum concentration less than 11 micromol/L, **OR** less than 57 mg/dL by nephelometry, **OR** less than 80 mg/dL by radial immunodiffusion<br>  ○ Forced Expiratory Volume in one second (FEV1) between 30-64% of predicted, **OR** FEV1 that is between 65-80% of predicted, but has declined by at least 100 mL per 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><br>• Coverage of Aralast NP, Glassia, or Zemaira will require a documented intolerable adverse event to Prolastin-C<br><br>• Dosing: 60 mg/kg intravenously once weekly<br><br>**<u>Reauthorization</u>** 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 lung or liver transplant |

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| **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:** | • Approval: 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 |
| **Required Medical Information:** | **<u>Lambert-Eaton myasthenic syndrome to reduce symptoms</u>**<br>• Documentation of diagnosis of Lambert-Eaton myasthenic syndrome (LEMS) confirmed by all of the following:<br>○ Records of electrodiagnostic studies, including repetitive nerve stimulation (RNS)<br>○ Anti-P/Q-type voltage-gated calcium channel (VGCC) antibody testing<br>○ Reproducible post-exercise increase in compound muscle action potential (CMAP) amplitude of at least 60 percent compared with pre-exercise baseline value or a similar increment on high-frequency repetitive nerve stimulation without exercise.<br>• Documented clinical failure to at least 12 weeks of each of the following:<br>○ Pyridostigmine<br>○ Immunosuppressive agents such as Corticosteroids (dosed at 1mg/kg/day), Azathioprine and Mycophenolate<br>○ Intravenous Immune Globulin (IVIG) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Lambert-Eaton myasthenic syndrome to reduce symptoms</u>**<br>**Adults (any weight) and pediatric patients weighing 45 kg or more:**<br>• 15 to 30 mg/day in 3 to 4 divided doses; May increase based on response and tolerability in 5 mg increments every 3 to 4 days. Maximum 80 mg/day.<br>**Pediatric patients weighing less than 45 kg:** |

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| | • 5 to 15 mg/day in 3 to 4 divided doses; May increase based on response and tolerability in 2.5 mg increments every 3 to 4 days. Maximum 40 mg/day.<br><br><u>Reauthorization requires documentation of treatment success</u> confirmed by updated electromyography records. |
| **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<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:
**ANIFROLUMAB**
Affected Medications: SAPHNELO (anifrolumab)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by benefit design.<br>  ○ Systemic Lupus Erythematosus |
| **Required Medical Information:** | • Documentation of systemic lupus erythematosus 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 standard combination therapy including hydroxychloroquine OR chloroquine with one of the following:<br>  ○ cyclosporine, azathioprine, methotrexate, or mycophenolate mofetil<br>AND<br>• Documented failure with at least 12 weeks of subcutaneous Benlysta<br><br>**<u>Dosing</u>**:<br>• 300 mg every 4 weeks<br><br>**<u>Reauthorization</u>**:<br>• Documentation of treatment success or a clinically significant improvement such as a decrease in flares or corticosteroid use |
| **Exclusion Criteria:** | • Saphnelo is not approved to be used in combination with other biologic therapies<br>• Saphnelo is not approved to be used in severe active lupus nephritis or severe active central nervous system lupus |
| **Age Restriction:** | • Must be 18 years or older |

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| **Prescriber/Site of Care Restrictions:** | • Prescribed by 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:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ANTIEMETICS**
Affected Medications: Akynzeo capsules (netupitant 300 mg and palonosetron hydrochloride 0.5 mg), Akynzeo (fosnetupitant 235 mg and palonosetron 0.25 mg), Varubi (rolapitant 0.5 mg)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• **Varubi** (rolapitant)<br>  ○ Prevention of delayed nausea and vomiting associated with initial and repeat courses of emetogenic cancer chemotherapy, including, but not limited to, highly emetogenic chemotherapy<br>• **Akynzeo for injection** (fosnetupitant and palonosetron)<br>  ○ Prevention of acute and delayed nausea and vomiting associated with initial and repeat courses of highly emetogenic cancer chemotherapy.<br>  ○ Akynzeo injection is not approved for use in anthracycline or cyclophosphamide-based chemotherapy or chemotherapy not considered highly emetogenic<br>• **Akynzeo capsules** (netupitant and palonosetron HCl)<br>  ○ Prevention of acute and delayed nausea and vomiting associated with initial and repeat courses of cancer chemotherapy, including, but not limited to, highly emetogenic chemotherapy |
| **Required Medical Information:** | • For chemotherapy induced nausea and vomiting (CINV)-documentation of planned chemotherapy regimen<br>• Highly emetogenic chemotherapy (HEC): Carboplatin, carmustine, cisplatin, cyclophosphamide, dacarbazine, doxorubicin, epirubicin, ifosfamide, mechlorethamine, streptozocin, FOLFOX regimen<br>• The following can be considered HEC in certain patients: Dactinomycin, daunorubicin, irinotecan, methotrexate (250 mg/m2 or greater), oxaliplatin, trabectedin |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Prevention of Chemotherapy induced Nausea and vomiting (CINV) in Adults</u>**<br>• **Varubi**:<br>  ○ Documentation of highly emetogenic chemotherapy (HEC); OR |

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| | ○ Moderately emetogenic chemotherapy and failure with a 5HT3-antagonist (i.e. ondansetron or granisetron) in combination with dexamethasone while receiving the current chemotherapy regimen<br>• **Akynzeo**<br>○ requires a highly emetogenic chemotherapy (HEC) regimen AND<br>○ failure with another generically available 5-HT3 receptor antagonist (e.g. ondansetron, granisetron or palonosetron) and NK1 receptor antagonist (e.g. aprepitant, fosaprepitant or rolapitant) while receiving the current chemotherapy regimen<br>• **Akynzeo** is NOT covered for: Breakthrough emesis or repeat dosing in multi-day emetogenic chemotherapy regimens<br>**<u>Prevention of Chemotherapy induced Nausea and vomiting (CINV) in Pediatric Patients (1 month to less than 17 years old)</u>**<br>• Documentation of emetogenic chemotherapy<br>• Varubi - Not being used for acute nausea and vomiting<br><br>Maximum 1 vial per 7 days for Akynzeo; 1 vial per 14 days for Varubi<br><br>**Reauthorization** requires documentation of treatment success and initial criteria to be met. |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with an oncologist (For CINV) |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization (no renewal for PONV): 6 months, unless otherwise specified |

POLICY NAME:
**ANTIHEMOPHILIC FACTORS**
Affected Medications: Advate, Adynovate, Afstyla, Alphanate, Alphanate/VWF Complex/Human, Alphanine SD, Alprolix, Benefix, Corifact, Eloctate, Esperoct, Feiba NF, Helixate FS, Hemofil M, Humate P, Idelvion, Ixinity, Jivi, Koate DVI, Kogenate FS, Kovaltry, Monoclate-P, Mononine, 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 benefit 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>• Patient 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>**<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 |

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| | hemophilia B, or severe VWD<br>○ For Vonvendi for routine prophylaxis: documentation of severe Type 3 VWD<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 |
| **Appropriate Treatment Regimen & Other Criteria:** | • Approval based on necessity and laboratory titer levels<br><br>**<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<br><br>**<u>Hemophilia B (factor IX deficiency)</u>**<br>• For **Benefix**, **Idelvion** and **Rebinyn**: documented 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**: documentation of failure or contraindication to Humate P AND Alphanate<br><br>**<u>All Indications</u>**<br>• Coverage for a non-preferred product requires documentation of one of the following: |

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| | ○ 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 |
| **Exclusion Criteria:** | • History of anaphylaxis or severe hypersensitivity to any component of the chosen agent<br>• 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 and Adynovate: 12 years and older<br>• Vonvendi: 18 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<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br>• Perioperative management: 1 month, unless otherwise specified |

POLICY NAME:
**ANTITHYMOCYTE GLOBULIN**
Affected Medications: ATGAM

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Management of allograft rejection in renal transplant patients<br>  ○ Treatment of moderate to severe aplastic anemia in patients unsuitable for bone marrow transplantation<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better<br>• Myelodysplastic Syndromes (MDS) |
| **Required Medical Information:** | • For MDS: Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing<br>  ○ Aplastic anemia: 10 to 20 mg/kg once daily for 8 to 14 days, then if needed, may administer every other day for 7 more doses for a total of 21 doses in 28 days OR 40 mg/kg daily for 4 days<br>  ○ MDS: 40 mg/kg once daily for 4 days<br>  ○ Renal transplant rejection: 10 to 15 mg/kg once daily for 14 days. Additional alternate-day therapy up to a total of 21 doses may be given. |
| **Exclusion Criteria:** | • All uses not listed in covered uses are considered experimental and are excluded from coverage<br>• Oncology: Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<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 |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care<br>• Specialist in oncology, hematology or transplant medicine |

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| **Coverage Duration:** | Approval: Maximum 4 weeks per dosing above, unless otherwise specified |

POLICY NAME:
**ANTITHROMBIN ALFA**
Affected Medications: ATRYN

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | • Diagnosed hereditary antithrombin deficiency via reduced plasma antithrombin level (not in midst of acute illness or surgery that could give falsely low antithrombin levels)<br>• Can be given for prophylaxis if negative personal/family history of thromboembolic events in high risk-settings as in surgery and pregnancy.<br>• Patient weight<br>• Documentation of intended dose based on reasonable projections and current dose utilization and product labeling. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Confirmed diagnosis of Hereditary Antithrombin deficiency<br><br>**<u>Peri-partum thromboembolic prophylaxis</u>**<br>• If positive personal/family history of VTE, ATryn recommended prior to and at the time of delivery when anticoagulation cannot be administered, and used until anticoagulation can be resumed<br>• If negative personal history of VTE, patient may need single dose of ATryn<br>• ATryn use is limited to third trimester<br>• If positive personal/family history of VTE, ATryn recommended<br>• Can be concomitantly given with LMWH or heparin<br><br>**<u>Peri-operative thromboembolic event prophylaxis</u>**<br>• Used during warfarin interruption leading up to surgical procedure (with or without heparin)<br>• Utilized until patient can resume warfarin therapy |
| **Exclusion Criteria:** | • Hypersensitivity to goats and goat milk protein<br>• Administration within first two trimesters of pregnancy<br>• Active thromboembolic event |
| **Age Restriction:** | • 18 – 65 years of age |
| **Prescriber/Site of Care Restrictions:** | • OB-GYN, MD<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Approval: 1 month, unless otherwise specified |

POLICY NAME:
**ANTI-AMYLOID MONOCLONAL ANTIBODY**
Affected Medications: ADUHELM (Aducanumab-avwa)

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| **Covered Uses:** | • Aducanumab (Aduhelm) 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:
**APOMORPHINE**
Affected Medications: KYNMOBI, APOKYN, APOMORPHINE 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 advanced Parkinson's Disease (PD)<br>• Documentation of at least one well defined acute intermittent hypomobility or "off" episode for 2 hours or more during the waking day, despite optimized therapy with other agents |
| **Appropriate Treatment Regimen & Other Criteria:** | • Concurrent therapy with levodopa/carbidopa at the maximum tolerated dose and a second agent from one of the following alternate anti-Parkinson's 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>• **Apokyn** requires documentation of failure or contraindication to Kynmobi<br><br>**<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Use as monotherapy or first line agent<br>• Concomitant use of 5-HT3 antagonists (ondansetron, granisetron, palonosetron, alosetron) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by 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:
**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. |
| **Required Medical Information:** | • Diagnosis of Mycobacterium avium complex (MAC) lung disease confirmed by a MAC-positive sputum culture<br>• Documentation of failure to obtain a negative sputum cultures 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:** | • Arikayce must be used as part of a multi-drug regimen and will not be approved for use as a single agent treatment<br>• To be used with Lamira Nebulizer system only<br>• **Reauthorization** requires documentation of negative sputum culture obtained within the last 30 days.<br>• The ATS/IDSA guidelines state that patients should continue to be treated until they have negative cultures for 1 year. Treatment beyond the first recertification approval (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:** | |
| **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 infectious disease specialist |

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

POLICY NAME:
**ARISTADA**
Affected Medications: ARISTADA (aripiprazole lauroxil), ARISTADA INITIO

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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 schizophrenia<br>• Documentation of established tolerability with oral aripiprazole for a minimum of 14 days prior to initiating treatment with Aristada.<br>• For initial authorization only: documented plan for ensuring oral adherence during first 21 days of initial Aristada is required.<br>• Documentation of anticipated dosing based on oral aripiprazole maintenance dose.<br>• Documentation of comprehensive antipsychotic treatment regimen (including dosing and frequency of all formulations).<br>• Documentation of Food and Drug Administration (Food and Drug Administration (FDA)) approved dose and frequency for the requested formulation.<br>• For Aristada Initio: Documentation of clinical rationale to avoid 21 day oral aripiprazole loading dose due to history of patient non-compliance or risk for hospitalization |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | • Repeated dosing (greater than 1 dose) of Aristada Initio<br>• Women who are pregnant, lactating, or breastfeeding.<br>• Patients with dementia-related psychosis<br>• Prior inadequate response to oral aripiprazole (unless poor adherence was a contributing factor)<br>• No current, or within the last 2 years, diagnosis of:<br>  ○ Major Depressive Disorder<br>  ○ Comorbid schizoaffective disorder<br>  ○ Amnestic or other cognitive disorder<br>  ○ Bipolar disorder<br>  ○ Dementia<br>  ○ Delirium |

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| **Age Restriction:** | • 18 years of age or 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:** | Aristada lauroxil<br>• Initial approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br><br>Aristada Initio, Approval: 1 month, unless otherwise specified |

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

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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 Philadelphia chromosome-positive chronic myeloid leukemia (CML) in chronic phase |
| **Appropriate Treatment Regimen & Other Criteria:** | • Failure or intolerance with imatinib and one additional tyrosine kinase inhibitor (TKI)<br>OR<br>• For patients with documented T315I positive mutation, documented clinical failure with ponatinib<br><br>Quantity limit in Philadelphia-positive CML with T315I mutation:<br>• 40 mg tablets #300 per 30 days.<br><br>Quantity limits in Philadelphia-positive CML previously treated with 2 or more TKIs:<br>• 40 mg tablets #60 per 30 days.<br>• 20 mg tablets #60 per 30 days.<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 approval: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

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

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| **Covered Uses:** | • All 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: |

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| | ○ 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<br>• 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 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>**<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy. |
| **Exclusion Criteria:** | • Diagnosis of infantile-onset Pompe Disease<br>• Concurrent treatment with Lumizyme |
| **Age Restriction:** | • 1 year of age or older |

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| **Prescriber/Site of Care Restrictions:** | • Metabolic specialist, endocrinologist, biochemical geneticist, or physician experienced in the management of Pompe disease. |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**AVATROMBOPAG**
Affected Medications: DOPTELET (avatrombopag maleate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Complete blood count with differential and platelet count<br>• Liver function tests<br><br>**<u>For thrombocytopenia in patients with Chronic Liver Disease undergoing medical or dental procedures</u>**<br>• Documentation of planned procedure including date |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>All indications:</u>**<br>• Documentation of all therapies tried/failed<br>• Documented inability to respond adequately to Promacta<br>• Documentation of splenectomy status<br><br>**<u>Thrombocytopenia in patients with Chronic Liver Disease undergoing medical or dental procedures</u>**<br>**<u>Dosage as either:</u>**<br>• Platelet count less than 40,000/mcl: 60 mg orally once daily with food for 5 consecutive days beginning 10 to 13 days prior to scheduled procedure **OR**<br>• Platelet count of 40,000/mcl to less than 50,000/mcl: 40 mg orally once daily with food for 5 consecutive days beginning 10 to 13 days prior to scheduled procedure<br><br>**<u>Reauthorization:</u>**<br>• Response to treatment with platelet count of at least 50,000/mcL or above without significant liver function abnormalities during procedure<br><br>**<u>Thrombocytopenia in Patients with Chronic Immune Thrombocytopenia (ITP):</u>**<br>• Documentation of platelet count less than 20,000/mcl AND<br>• Documentation of clinically significant bleeding AND<br>• Documentation of splenectomy OR failure with at least 2 therapies for ITP, including corticosteroids or immunoglobulin (defined as platelets did not increase to at least 50,000/mcl) |

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| | **<u>Reauthorization:</u>**<br>• Response to treatment with platelet count of at least 50,000/mcl (not to exceed 400,000/mcl) **OR**<br>• The platelet counts have not increased to at least 50,000/mcl and the patient has NOT been on the maximum dose for at least 4 weeks. |
| **Exclusion Criteria:** | • Platelet count above 50,000/mcL at baseline<br>• History of thrombosis<br>• Platelet transfusion or receipt of blood containing platelets within 7 days of screening for procedure<br>• Use of heparin, warfarin, NSAIDs, ASA, verapamil, or antiplatelet therapy with ticlopidine or glycoprotein IIb/IIIa antagonists, or erythropoietin stimulating agents within 7 days of screening for procedure<br>• History of hematological malignancy or myelodysplastic syndrome |
| **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><br>**<u>Thrombocytopenia in patients with Chronic Liver Disease undergoing medical or dental procedures</u>**<br>• Prescribed by or in consultation with hematologist or gastroenterology/liver specialist<br><br>**<u>Thrombocytopenia in Patients with Chronic Immune Thrombocytopenia (ITP):</u>**<br>• Prescribed by or in consultation with a hematologist |
| **Coverage Duration:** | • Thrombocytopenia with Chronic Liver Disease undergoing procedure: 1 month or for a specific procedure, unless otherwise specified<br>• Thrombocytopenia in Patients with Chronic Immune Thrombocytopenia (ITP)<br>  ○ Initial Approval: 4 months, unless otherwise specified<br>  ○ Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**AVONEX**
Affected Medications: AVONEX, AVONEX PEN

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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:** | • Reauthorization requires provider attestation of treatment success |
| **Exclusion Criteria:** | • Concurrent use of other disease-modifying medications for treatment of MS |
| **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 neurologist |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**AXICABTAGENE CILOLEUCEL**
Affected Medications: YESCARTA (axicabtagene ciloleucel)

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | • Documentation of disease staging, all prior therapies used, performance status of 0-1<br>• Patient weight<br>• Documentation of adequate organ and marrow function |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Relapsed or Refractory Large B-cell Lymphoma</u>**<br>• Diagnosed with one of the following:<br>  ○ Diffuse large B-cell lymphoma (DLBCL) not otherwise specified (including DLBCL arising from follicular lymphoma)<br>  ○ High-grade B-cell lymphoma<br>  ○ Primary mediastinal large B-cell lymphoma<br>AND<br>• Falls into one of the following categories:<br>  ○ Disease has relapsed, or has been refractory, after 2 or more lines of systemic therapy<br>  ○ Disease is refractory to first line chemoimmunotherapy OR has relapsed within 12 months of first line chemoimmunotherapy<br>  ▪ First-line therapy must have included rituximab and an anthracycline<br><br>**<u>Relapsed or Refractory Follicular Lymphoma</u>**<br>• Disease has relapsed, or has been refractory, after 2 or more lines of systemic therapy<br><br>Approved for one-time single infusion only |
| **Exclusion Criteria:** | • Central nervous system lymphoma<br>• History of allogeneic hematopoietic stem cell transplantation (HSCT)<br>• ECOG performance status of 2 or greater<br>• Absolute lymphocyte count less than 100/ul<br>• CrCl less than 60 mL/min<br>• Hepatic transaminases more than 2.5x the upper limit of normal |

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| | • Cardiac ejection fraction less than 50%<br>• Active serious infection |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Oncologist and Health care facilities must be enrolled and comply with the Risk Evaluation and Mitigation Strategies (REMS) requirement<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 1 month, unless otherwise specified (one infusion only) |

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><br>• **Dosing:** 28 days on and 28 days off<br><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 approval: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**BEDAQUILINE**
Affected Medications: SIRTURO (bedaquiline fumarate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• Pulmonary multi-drug resistant tuberculosis (MDR-TB) |
| **Required Medical Information:** | Patient has failed, is resistant, or is allergic to quad therapy of any combination of the following:<br>• Isoniazid<br>• Rifampin<br>• Ethambutol<br>• Pyrazinamide<br>• Fluoroquinolone<br>• Capreomycin (Kanamycin, Amikacin, Streptomycin)<br>• Ethionamide/Prothinamide<br>• Cycloserine/Terizidone<br>• Aminosalicylic acid (acidic salt) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of being administered by directly observed therapy (DOT)<br>• Baseline ECG<br>• BMP (including K, Ca, Mg documentation of correction if needed)<br>• LFTs |
| **Exclusion Criteria:** | • Drug-sensitive TB (DS-TB)<br>• Latent Infection due to Mycobacterium tuberculosis<br>• Extrapulmonary TB (e.g. central nervous system)<br>• QTc greater than 500 milliseconds |
| **Age Restriction:** | • 5 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with infectious disease specialist.<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 24 weeks, 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:** | **<u>Systemic Lupus Erythematosus:</u>**<br>• Documentation of systemic lupus erythematosus 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>• Documentation of patient's current weight (intravenous requests only)<br><br>**<u>Lupus Nephritis:</u>**<br>• Documentation of lupus nephritis disease stage III, IV, or V<br>• Documentation of patient's current weight (intravenous requests only) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Systemic Lupus Erythematosus:</u>**<br><br>• Failure with at least 12 weeks of standard combination therapy including hydroxychloroquine OR chloroquine with one of the following:<br>○ cyclosporine, azathioprine, methotrexate, or mycophenolate mofetil<br>• For adult patients (18 years of age and older): Intravenous (IV) formulation requires documented inability to use subcutaneous formulation.<br>• **<u>Reauthorization</u>**: Documentation of treatment success defined as a clinically significant improvement in SLE Responder Index-4 (SRI-4) or decrease in flares/corticosteroid use. |

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| | **<u>Lupus Nephritis:</u>**<br><br>• No dialysis in the past 12 months AND estimated glomerular filtration rate (eGFR) equal to or above 30 ml/min/1.73m2<br>• Failure of at least 12 weeks of standard therapy with mycophenolate mofetil AND cyclophosphamide<br>• For adult patients (18 years of age and older): Intravenous (IV) formulation requires documented inability to use subcutaneous formulation.<br>• **<u>Reauthorization</u>**: Documentation of treatment success defined as an improvement in eGFR, reduction in urinary protein: creatinine ratio, or decrease in flares/corticosteroid use<br><br>Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced |
| **Exclusion Criteria:** | • Benlysta is not approved to be used in combination with other biologic therapies<br>• Benlysta is not approved to be used in severe active central nervous system lupus |
| **Age Restriction:** | • Intravenous formulation: 5 years of age and older<br>• Subcutaneous formulation: 18 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:<br>• Systemic Lupus Erythematosus - 12 months, unless otherwise specified<br>• Lupus Nephritis<br>  ○ Initial: 6 months, unless otherwise specified<br>  ○ Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**BELINOSTAT**
Affected Medications: BELEODAQ (belinostat)

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher. |
| **Required Medical Information:** | • Documentation of staging, all prior therapies used, performance status and anticipated treatment course<br>• Complete blood count (CBC) with differential, creatinine clearance (CrCl), liver function tests<br>• Documentation of UGT1A1*28 allele status |
| **Appropriate Treatment Regimen & Other Criteria:** | • Appropriate dose reduction based on absolute neutrophil count (ANC) OR homozygous UGT1a1*28 allele<br>• Reauthorization: documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • 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:** | • Oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

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

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by benefit design<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | • Documentation of von Hippel-Lindau (VHL) disease as defined by VHL germline mutation and the presence of at least one measurable solid tumor located in the kidney, brain/spine, or pancreas<br>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course<br>• |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization:** documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• Metastatic 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:** | • 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:
**BENRALIZUMAB**
Affected Medications: FASENRA (benralizumab subcutaneous injection)

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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 for use in combination with another monoclonal antibody (Dupixent, Nucala, Xolair, Cinqair)? | Yes – Criteria not met, combination use is experimental | No – Go to #3 |
| 3. Is the request to treat a diagnosis according to one of the Food and Drug Administration (FDA)-approved indications?<br>○ Add-on maintenance treatment of patients with severe asthma aged 12 years and older with an eosinophilic phenotype | Yes – Go to appropriate section below | No – |
| **Severe Eosinophilic Asthma** | | |
| 1. Is there documentation of severe eosinophilic asthma defined by the following:<br>○ Baseline eosinophil count of at least 150 cells/µL<br>**AND**<br>○ FEV1 less than 80% at baseline or FEV1/FVC reduced by at least 5% from normal | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documented use of high-dose inhaled corticosteroid (ICS) plus a long-acting beta agonist (LABA) for at least three months with continued symptoms? | Yes – Document and go to #3 | No – Criteria not met |

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| 3. Is there a documented history of 2 or more asthma exacerbations requiring oral or systemic corticosteroid treatment in the past 12 months while on combination inhaler treatment and at least 80% adherence? | Yes – Go to #5 | No – Go to #4 |
| 4. Is there documentation that chronic daily oral corticosteroids are required? | Yes – Go to #5 | No – Criteria not met |
| 5. Is the drug prescribed by or in consultation with an Allergist, Immunologist, or Pulmonologist? | Yes – Approve up to 6 months | No – Criteria not met |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider? | Yes – Go to #2 | No – Criteria not met |
| 2. Is the request for use in combination with another monoclonal antibody (e.g., Dupixent, Nucala, Xolair, Cinqair)? | Yes – Criteria not met, combination use is experimental | No – Go to #3 |
| 3. 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** | | |
| • **Fasenra**<br>○ Availability: 30 mg/mL pre-filled syringe or auto-injector<br>○ Dosing: 30 mg every 4 weeks for the first 3 doses, then 30 mg every 8 weeks thereafter | | |

*Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced for all medical infusion drugs

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. |
| **Required Medical Information:** | • Documentation of one of the following:<br>  ○ Cystathionine beta-synthase (CBS) deficiency<br>  ○ 5,10-methylenetetrahydrofolate reductase (MTHFR) deficiency<br>  ○ Cobalamin cofactor metabolism (cbl) defect<br>  ○ Vitamin B12 and folic acid serum levels |
| **Appropriate Treatment Regimen & Other Criteria:** | • Vitamin B6, B12, and folate supplementation<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Uncorrected vitamin B12 or folic acid levels |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**BETASERON**
Affected Medications: BETASERON (interferon beta-1b)

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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 diagnosis of a relapsing form of multiple sclerosis (including clinically isolated syndrome, replapsing-remitting disease, and active secondary progressive disease) confirmed with magnetic resonance imaging (MRI) |
| **Appropriate Treatment Regimen & Other Criteria:** | • **Reauthorization**: provider attestation of treatment success |
| **Exclusion Criteria:** | • Concurrent use of medications indicated for the treatment of relapsing form of multiple sclerosis<br>• For treatment of primary progressive multiple sclerosis |
| **Age Restriction:** | • Adults (18 years of age and older) |
| **Prescriber/Site of Care Restrictions:** | • 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:
**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 able to provide a minimum number of cells |
| **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 |

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| | 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:** | • Ages 4 years 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

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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 **AND**<br>• As indicated per NCCN, documentation of performance status 0-1 **AND**<br>• If patient is at risk of thrombocytopenia: Documentation that risks (DVT, intra-abdominal thrombosis, gastrointestinal perforations, hemorrhage) have been reviewed and that benefit of therapy outweighs risks |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Non-Small Cell Lung Cancer (NSCLC)</u>**<br>• Approval will be limited to NCCN category 1 recommended therapies for first line treatment of advanced NSCL cancer<br><br>Reauthorization: documentation of disease responsiveness to therapy<br><br>**<u>Stage III or IV Epithelial Ovarian, Fallopian Tube, or Primary Peritoneal Cancer following initial surgical resection</u>**<br>• Approval will be limited for up to 22 cycles of therapy<br><br>**<u>All Indications</u>**<br>• Coverage for a non-preferred product (Avastin, Alymsys) 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 |

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| | 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 |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, on 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:
**BEXAROTENE**
Affected Medications: TARGRETIN (bexarotene), Bexarotene

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | **<u>Targretin Gel</u>**<br>• Documentation of cutaneous T-cell lymphoma (CTCL) stage IA or IB<br>• Diagnosis confirmed by biopsy (exclusion of other T cell lymphomas with cutaneous involvement)<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 |
| **Appropriate Treatment Regimen & Other Criteria:** | • Patient has been instructed on the importance and proper utilization of appropriate contraceptive methods.<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Pregnancy. |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Oncologist<br>• Dermatologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 4 months (2 week initial partial fill), unless otherwise specified<br>• Approval: 12 months, unless otherwise noted |

POLICY NAME:
**BEZLOTOXUMAB**
Affected Medications: ZINPLAVA (bezlotoxumab)

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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 diarrhea (at least 3 unformed stools in 24 hour) or radiographic evidence of ileus or toxic megacolon<br>• Stool positive for GDH antigen AND Toxin A & B OR PCR positive<br>• If GDH positive/toxin negative OR GDH negative/toxin positive, PCR MUST be positive<br>• Patient must be receiving concurrent treatment for *Clostridium difficile*: metronidazole (intravenous or oral), oral vancomycin, fidaxomicin |
| **Appropriate Treatment Regimen & Other Criteria:** | • Patients at high risk for CDI recurrence (must have at least one risk factor): age greater than 65, one or more episodes of *Clostridium Difficile* infection (CDI) in previous 6 months, immunocompromised status, clinically severe CDI (as defined by Zar score greater than or equal to 2). |
| **Exclusion Criteria:** | • Stool NEGATIVE for GDH and Toxin, or PCR negative if incongruent GDH/toxin<br>• Heart Failure |
| **Age Restriction:** | • Age 18 years or greater |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: One treatment may be given while patient is receiving antibiotic therapy for treatment of C. difficile (usually 14 days) |

POLICY NAME:
**BIMATOPROST IMPLANT**
Affected Medications: DURYSTA (bimatoprost intracameral implant)

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| 1. 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, experimental/investigational |
| **Open-Angle Glaucoma (OAG) or Ocular Hypertension (OHT)** | | |
| 1. Is there a documented diagnosis of Open-Angle Glaucoma (OAG) or Ocular Hypertension (OHT) with a baseline intraocular pressure (IOP) at least 22 mmHg? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there a documented history of positive response to prostaglandin drops (E.g., latanoprost, bimatoprost)? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documented medical justification supporting inability to manage regular glaucoma eye drop use (e.g., due to age or comorbidities including visual impairment)? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is there a Diagnosis of corneal endothelial cell dystrophy (e.g., Fuchs' Dystrophy)? | Yes – Criteria not met; contraindication | No – Go to #5 |
| 5. Is there a history of corneal transplantation or endothelial cell transplant (e.g., Descemet's | Yes – Criteria not met; contraindication | No – Go to #6 |

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| Stripping Automated Endothelial Keratoplasty (DSAEK))? | | |
| 6. Is the drug being prescribed by or in consultation with an ophthalmologist? | Yes – Go to #7 | No – Criteria not met |
| 7. Is the request for repeat implantation? | Yes – Criteria not met; repeat implantation currently considered experimental | No – Approve up to 2 months (1 implant per impacted eye) without renewal |
| **Quantity Limitations** | | |
| • **Durysta**<br>o A single intracameral bimatoprost implant per eye. Should not be readministered to an eye that received a prior Durysta | | |

POLICY NAME:
**BLINATUMOMAB**
Affected Medications: BLINCYTO

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | • Documentation of disease staging, all prior therapies used, performance status and anticipated treatment course **AND**<br>• Philadelphia chromosome status **AND**<br>• Documentation of ECOG performance status of 1 or 2 OR Karnofsky performance score greater than 50% |
| **Appropriate Treatment Regimen & Other Criteria:** | • Blincyto should permanently be discontinued for the following adverse reactions: grade 4 cytokine release syndrome, grade 4 neurological toxicity, or two Blincyto induced seizures<br>• Maximum approval: 9 cycles for Relapsed or Refractory Acute Lymphoblastic Leukemia (ALL), 4 cycles for ALL in 1st or 2nd remission with minimal residual disease (MRD) |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 30 weeks for Relapsed or Refractory ALL; 24 weeks for ALL in 1st or 2nd remission with MRD<br>• Reauthorization: 48 weeks for Relapsed or Refractory ALL (4 cycles of continued therapy x 12 weeks each, to complete a maximum of 9 cycles total); NO reauthorization for ALL in 1st or 2nd remission with MRD, unless otherwise specified |

POLICY NAME:
**BOTOX**
Affected Medications: BOTOX (onabotulinum toxin A)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design |
| **Required Medical Information:** | • Pertinent medical records and diagnostic testing<br>• Complete description of the site(s) of injection<br>• Strength and dosage of botulinum toxin used |
| **Appropriate Treatment Regimen & Other Criteria:** | • For use in Food and Drug Administration (FDA)-approved or compendia supported indications not otherwise excluded by benefit design that are not listed below, failure of first-line recommended and conventional therapies is required<br>• Approved first-line for: focal dystonia, hemifacial spasm, orofacial dyskinesia, blepharospasm, severe writer's cramp, laryngeal spasm or dysphonia, upper/lower limb spasticity or other conditions of central focal spasticity botulinum toxin is the preferred mode of therapy.<br><br>**<u>Idiopathic or neurogenic detrusor over-activity (Overactive Bladder (OAB)) and Urinary incontinence associated with neurologic condition</u>**<br>• Inadequate response to, or intolerance to, at least 2 incontinence anticholinergic drugs (such as oxybutynin, solifenacin, tolterodine)<br><br>**<u>Chronic migraine</u>**<br>• Documentation of chronic migraine defined as headaches on at least 15 days per month of which at least 8 days are with migraine AND documented failure with an adequate trial (at least 8 weeks) of an oral migraine preventive therapy as follows:<br>  ○ Propranolol 40 mg daily, Metoprolol 100 mg daily<br>  ○ Amitriptyline 25 mg daily<br>  ○ Topiramate 50 mg daily, Valproic acid, Divalproex sodium<br><br>**<u>Primary Axillary Hyperhidrosis</u>** |

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| | • TSH level AND inadequate response to two or more alternative therapies (topical aluminum chloride 20%, iontophoresis, oral glycopyrrolate, oral oxybutynin)<br><br>**Achalasia (Cardiospasm) -** must meet 1 of the following<br>• Failure or intolerance to peroral endoscopic myotomy (POEM) or laparoscopic Heller myotomy AND failure or intolerance to pneumatic dilation<br>• Not a candidate for POEM, surgical myotomy, or pneumatic dilation due to high risk of complications<br><br>**Anal fissure**<br>• Documented failure or intolerance to an 8 week trial of each of the following:<br>  ○ Rectiv ointment<br>  ○ Topical diltiazem or topical nifedipine<br><br>Number of treatments must not exceed the following:<br>• Idiopathic or neurogenic detrusor over-activity (OAB)/ Urinary incontinence associated with neurologic condition: 2 treatments/12 months<br>• Chronic migraine: initial treatment limited to two injections given 3 months apart, subsequent treatment approvals limited to 4 treatments per 12 months<br>• Primary axillary hyperhidrosis: 2 treatments/12 months<br>• Anal fissure: 2 treatments/12 months<br>• All other indications maximum of 4 treatments/12 months unless otherwise specified<br><br>**Reauthorization:**<br>• **Chronic migraine continuation of treatment:** Additional treatment requires that the member has achieved or maintained a 50% reduction in monthly headache frequency since starting therapy with Botox.<br>• All other indications: Documentation of treatment success and clinically significant response to therapy. |
| **Exclusion Criteria:** | • Cosmetic procedures<br>• For intradetrusor injections: documented current/recent urinary tract infection or urinary retention |

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| | • Possible medication overuse headache: headaches occurring 15 or more days each month in a patient with pre-existing headache-causing condition possibly due to<br>○ Use of ergotamines, triptans, opioids, or combination analgesics at least 10 days per month for at least three months<br>○ Use of simple analgesics (acetaminophen, aspirin, or an NSAID) at least 15 days per month for at least 3 months<br>○ Use of combination of any previously mentioned products without overuse of any one agent if no causative pattern can be established |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Blepharospasm, strabismus: ophthalmologist or neurologist<br>• Chronic migraine: treatment is administered in consultation with a neurologist or headache specialist.<br>• OAB or urinary incontinence due to neurologic condition: urologist or neurologist<br>• Anal fissure: gastroenterologist or colorectal surgeon<br>• Documentation of consultation with any of the above specialists mentioned |
| **Coverage Duration:** | Chronic migraine:<br>• Initial approval: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br><br>Idiopathic or neurogenic detrusor over-activity (OAB)/ Urinary incontinence associated with neurologic condition:<br>• Initial approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br><br>Anal Fissure:<br>• Approval: 3 months (one treatment), unless otherwise specified<br><br>All other indications<br>• Approval 12 months, unless otherwise specified |

POLICY NAME:
**BREXANOLONE**
Affected Medications: ZULRESSO (brexanolone)

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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:** | • Documentation of major depressive episode as diagnosed by DSM-5 Criteria<br>  ○ Five or more of the following symptoms present during the same two-week period and represent a change from previous function. Must include either (1) depressed mood or (2) lack of interest or pleasure<br>  ▪ Depressed mood most of the day, nearly every day, as indicated by either subjective report (eg, feels sad, empty, hopeless) or observations made by others (eg, appears tearful). (NOTE: In children and adolescents, can be irritable mood.)<br>  ▪ 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>  ▪ Significant weight loss when not dieting or weight gain (eg, a change of more than 5% of body weight in a month), or decrease or increase in appetite nearly every day. (NOTE: In children, consider failure to make expected weight gain.)<br>  ▪ Insomnia or hypersomnia nearly every day<br>  ▪ Psychomotor agitation or retardation nearly every day (observable by others, not merely subjective feelings of restlessness or being slowed down)<br>  ▪ Fatigue or loss of energy nearly every day<br>  ▪ Feelings of worthlessness or excessive or inappropriate guilt (which may be delusional) nearly every day (not merely self-reproach or guilt about being sick)<br>  ▪ Diminished ability to think or concentrate, or indecisiveness, nearly every day (either by their subjective account or as observed by others) |

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| | ▪ 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>AND<br>○ Symptoms cause clinically significant distress or impairment in social, occupational, or other important areas of functioning AND<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>• Documentation of Edinburgh Postnatal Depression Scale score (greater than 13), HAM-D score (greater than 14 points), PHQ-9 score (greater than 10 points), or MADRS score (greater than 20 points) indicating moderate to severe postpartum depression (PPD) |
| **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>• Administered as a continuous infusion over a total of 60 hours (2.5 days) as follows<br>○ 0 to 4 hours: Initiate with a dosage of 30 mcg/kg/hour<br>○ 4 to 24 hours: Increase dosage to 60 mcg/kg/hour<br>○ 24 to 52 hours: Increase dosage to 90 mcg/kg/hour (a reduction in dosage to 60 mcg/kg/hour may be considered during this time period for patients who do not tolerate 90 mcg/kg/hour)<br>○ 52 to 56 hours: Decrease dosage to 60 mcg/kg/hour<br>○ 56 to 60 hours: Decrease dosage to 30 mcg/kg/hour |
| **Exclusion Criteria:** | • Greater than 6 months postpartum |
| **Age Restriction:** | |

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| **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 psychiatrist or other licensed medical provider with specialty in psychiatry |
| **Coverage Duration:** | • One month, one time approval per pregnancy, unless otherwise specified |

POLICY NAME:
**BREXUCABTAGENE AUTOLEUCEL**
Affected Medications: TECARTUS (brexucabtagene autoleucel)

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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 |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Relapsed or Refractory Mantle Cell Lymphoma (MCL) or B-cell precursor Acute lymphoblastic leukemia (B-ALL)</u>**<br>• Patient has experienced disease progression after their last regimen or is refractory to their most recent therapy<br>• Prior treatment for MCL must include:<br>  ○ Anthracycline or bendamustine-containing chemotherapy, AND<br>  ○ Anti-CD20 monoclonal antibody (i.e. rituximab), AND<br>  ○ Bruton tyrosine kinase inhibitor (ibrutinib or acalabrutinib)<br>• Prior treatment for B-ALL must include:<br>  ○ Tyrosine kinase inhibitor AND<br>  ○ Besponsa or Blincyto<br><br>Approved for one-time single infusion only |
| **Exclusion Criteria:** | • Active hepatitis B, hepatitis C, or human immunodeficiency virus<br>• Prior allogeneic hematopoietic stem cell transplant<br>• Detectable cerebrospinal fluid malignant cells or brain metastases<br>• Platelet count of less than 75,000/uL, creatinine clearance less than 60 mL/min, cardiac ejection fraction less than 50%, or baseline oxygen saturation less than 92% on room air |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Must be prescribed by oncologist<br>• Oncologist and administering health care facility must be certified and in compliance with the Risk Evaluation and Mitigation Strategies (REMS) requirements |

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| | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 1 month, unless otherwise specified (one infusion only) |

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 benefit design.<br>  ○ The treatment of X-linked hypophosphatemia (XLH)<br>  ○ The treatment of FGF23-related hypophosphatemia in tumor induced osteomalacia (TIO) associated with phosphaturic mesenchymal tumors that cannot be curatively resected or localized |
| **Required Medical Information:** | **<u>All Indications</u>**<br>• Documentation of diagnosis by:<br>  ○ A blood test demonstrating:<br>  ○ Decreased phosphate **AND**<br>  ○ Increased FGF-23 **AND**<br>  ○ Decreased 1,25-(OH)2D **AND**<br>  ○ Normal parathyroid hormone (PTH) **AND**<br>  ○ A urine test demonstrating:<br>  • 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 AND<br>• Alternative renal phosphate-wasting disorders have been ruled out |
| **Appropriate Treatment Regimen & Other Criteria:** | For all diagnoses:<br>• Documentation of trial/failure with oral phosphate and calcitriol supplementation in combination for at least 12 months, or contraindication to therapy<br>• Dose adjustments are not made more frequently than every 4 weeks |

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| | **<u>X-Linked Hypophosphatemia</u>**<br>• Dosing<br>  ○ Pediatrics weighing less than 10 kg,<br>  ▪ Initial: 1 mg/kg, rounded to the nearest 1 mg, every 2 weeks<br>  ○ Pediatrics weighing 10 kg or greater,<br>  ▪ Initial: 0.8 mg/kg, rounded to nearest 10 mg, every 2 weeks; up to a maximum of 90 mg<br>  ○ Adults<br>  ▪ Initial: 1 mg/kg, rounded to nearest 10 mg, every 4 weeks; up to maximum of 90 mg<br><br>**<u>Tumor-Induced Osteomalacia</u>**<br>• Dosing<br>  ○ Pediatrics (2 years to less than 18 years of age),<br>  ▪ Initial: 0.4 mg/kg, rounded to the nearest 10 mg, every 2 weeks<br>  ▪ Maximum dose: 2 mg/kg (not to exceed 180 mg) every 2 weeks<br>  ○ Adults,<br>  ▪ Initial: 0.5 mg/kg, rounded to the nearest 10 mg, every 4 weeks<br>  ▪ Maximum dose: 2 mg/kg (not to exceed 180 mg) every 2 weeks<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 normalization of serum phosphate levels AND improvement in radiographic imaging of skeletal abnormalities. |
| **Exclusion Criteria:** | • Oral phosphate or active vitamin D analogs within the last week<br>• Severe renal impairment and/or end stage renal disease |
| **Age Restriction:** | • X-Linked Hypophosphatemia: Patient is at least 6 months of age<br>• Tumor-Induced Osteomalacia: Patient is at least 2 years of age |

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| **Prescriber Restrictions:** | • Must be administered by a healthcare provider.<br>• Prescribed by or in consultation with a Nephrologist or Endocrinologist, or provider experienced in managing patients with metabolic bone disease |
| **Coverage Duration:** | • Initial approval: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

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

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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 AND documentation of all of the following prior to treatment initiation:<br>  ○ Stage 3 or 4 CKD (baseline eGFR of 15 – 59 mL/min)<br>  ○ Serum total 25-hydroxyvitamin D level is less than 30 ng/mL<br>  ○ Corrected serum calcium is below 9.8 mg/dL<br>  ○ Serum phosphorus level is within normal range (less than 5.0 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: vitamin D2 (ergocalciferol), calcitriol, doxercalciferol, paricalcitol<br><br>• Dosing: 30 to 60 mcg per day (maximum of two capsules per day)<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<br>• Persistently elevated serum calcium and phosphorus levels, above the normal range |

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| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subjects to utilization of the most cost-effective site of care<br>• Prescribed by, or in consultation with, a nephrologist or endocrinologist |
| **Coverage Duration:** | • Approval: 12 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:** | **<u>All Indications</u>**<br>• Patient weight<br>• Documentation that cannabidiol 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>  ○ Rufinamide, topiramate, felbamate, or clobazam<br><br>**<u>Dravet Syndrome (DS)</u>**<br>• Documentation of at least 4 convulsive seizures in the last month while on stable antiepileptic drug therapy<br>• Documented treatment and inadequate seizure control with at least four guideline directed therapies including:<br>  ○ Valproate **and**<br>  ○ Clobazam **and**<br>  ○ Topiramate **and**<br>  ○ Clonazepam, levetiracetam, or zonisamide<br><br>**<u>Tuberous Sclerosis Complex (TSC)</u>**<br>• Documentation of monotherapy failure for seizure control with two antiepileptic regimens AND<br>• Documentation of failure with at least one adjunctive therapy for seizure control |
| **Appropriate Treatment** | Dosing: |

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| **Regimen & Other Criteria:** | • Lennox-Gastaut Syndrome or Dravet Syndrome: Not to exceed 20 mg/kg per day<br>• Tuberous Sclerosis Complex: Not to exceed 25 mg/kg per day<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 monotherapy for seizure control |
| **Age Restriction:** | • Greater than or equal to 1 year |
| **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 approval: 6 months unless otherwise specified<br>• Reauthorization: 12 months unless otherwise specified |

POLICY NAME:
**CAPLACIZUMAB**
Affected Medications: CABLIVI (caplacizumab)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design<br>○ Treatment of adult patients with acquired thrombotic thrombocytopenic purpura (aTTP), in combination with plasma exchange and immunosuppressive therapy |
| **Required Medical Information:** | Must have documentation containing all of the following:<br>• Diagnosis of acquired thrombotic thrombocytopenic purpura (aTTP)<br>• Cablivi was initiated in the inpatient setting in combination with plasma exchange therapy.<br>• Cablivi will be used in combination with immunosuppressive therapy (such as corticosteroids)<br>• Total treatment duration will be limited to 58 days beyond the last therapeutic plasma exchange |
| **Appropriate Treatment Regimen & Other Criteria:** | Dosing:<br>• First day of treatment: IV followed by SubQ: 11 mg IV at least 15 minutes prior to plasma exchange, followed by 11 mg SubQ after completion of plasma exchange on day 1.<br>• Subsequent treatment days (during daily plasma exchange): SubQ: 11 mg once daily following plasma exchange.<br>• Treatment after plasma exchange period: SubQ: 11 mg once daily, continuing for 30 days following the last daily plasma exchange; if sign(s) of persistent underlying disease remain present (eg, suppressed ADAMTS13 activity levels) after initial treatment course, treatment may be extended up to a maximum of 28 days.<br>• Discontinuation: Discontinue caplacizumab if >2 recurrences of acquired thrombotic thrombocytopenic purpura (aTTP) occur during treatment.<br><br>**Reauthorization** Request is for a new (different) episode requiring the re-initiation of plasma exchange for the treatment of aTTP. (Documentation of date of prior episode & documentation date of new episode required) |

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| **Exclusion Criteria:** | |
| **Age Restriction:** | • 18 years and older |
| **Prescriber/Site of Care Restrictions:** | • Treatment by or in consultation with a hematology specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 2 months, unless otherwise specified<br>• Reauthorization: 2 months (for new episode), 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. |
| **Required Medical Information:** | • 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 |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Acute hyperammonemia</u>**<br>• Ammonia level greater than 100 micromol/L<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><br>**<u>Chronic hyperammonemia due to N-Acetylglutamate Synthase (NAGS) deficiency</u>**<br>• Ammonia level greater than or equal to 50 micromol/L<br>• NAGS deficiency confirmed by enzymatic, biochemical, or genetic testing<br>• Prescribed in combination with a protein-restricted diet<br><br>**<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy. |
| **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 |
| **Age Restriction:** | |

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| **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:** | • Initial approval: 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. |
| **Required Medical Information:** | • Confirmed diagnosis of late infantile neuronal ceroid lipofuscinosis type 2 (CLN2) with one of the following:<br>  ○ Documented deficient tripeptidyl peptidase-1 (TPP1) activity in leukocytes<br>  ○ Pathogenic variants/mutations in each allele of TPP1/CLN2 gene **AND** baseline motor, speech and vision function documented by the physician<br>• Documentation of mild to moderate functional impairment at baseline using the CLN2 Clinical Rating Scale, defined as:<br>  ○ A combined motor and language domain score of 3 to 6 **AND**<br>  ○ A score of at least 1 in each of these two domains<br>• Planned treatment regimen including doses, frequency<br>• Planned monitoring parameters for infections and side effects |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing: 300 mg administered once every other week by intraventricular infusion<br><br>Reauthorization:<br>• Documentation of continuing to meet initial review criteria AND<br>• Documentation of clinical responsiveness to therapy with disease stability/improvement defined as a score of one or higher in the motor domain of the CLN2 Clinical Rating Scale. |
| **Exclusion Criteria:** | • Patients with acute intraventricular access device-related complications (e.g., leakage, device failure or signs of device-related infection such as swelling, erythema of the scalp, extravasation of fluid, or bulging of the scalp around or above the intraventricular access device)<br>• Other form of neuronal ceroid lipofuscinosis<br>• Patients with ventriculoperitoneal shunts |
| **Age Restriction:** | • Between 3 years to 16 years of age |

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| **Prescriber/Site of Care Restrictions:** | • Must be prescribed by a neurologist or in consultation with a neurologist with expertise in the diagnosis of CLN2<br>• Must be administered by, or under the direction of a physician knowledgeable in intraventricular administration<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 3 months, unless otherwise specified<br>• Reauthorization: 6 months, unless otherwise specified |

POLICY NAME:
**CGRP INHIBITORS**

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| **PA policy applicable to:**<br>**Preferred drugs:** Ajovy, Emgality<br>**Medical infusion drugs:** Vyepti | | |
| 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 for combined use with Botox for the treatment of chronic migraine? | Yes – Criteria not met, considered experimental | No – Go to #3 |
| 3. Is the request to treat a diagnosis according to one of the Food and Drug Administration (FDA)-approved indications (not otherwise excluded by plan design) at the approved dosing? | Yes – Go to appropriate section below | No – Criteria not met |
| **Chronic or Episodic Migraine in adults**<br>**Preferred Drugs – Ajovy, Emgality**<br>**Medical Infusion Drugs – Vyepti** | | |
| 1. Is there a diagnosis of chronic migraine defined as headaches on at least 15 days per month of which at least 8 days are with migraine at baseline? | Yes – Document and go to #3 | No – Go to #2 |
| 2. Is there a diagnosis of episodic migraine with at least 8 migraine days per month at baseline? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is the request for treatment of possible medication overuse headache? Headaches occurring 15 or more days each month in a | Yes – Criteria not met | No – Go to #4 |

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| patient with pre-existing headache-causing condition possibly due to<br>a. Use of ergotamines, triptans, opioids, or combination analgesics at least 10 days per month for at least three months<br>b. Use of simple analgesics (acetaminophen, aspirin, or an NSAID) at least 15 days per month for at least 3 months<br>c. Use of combination of any previously mentioned products without overuse of any one agent if no causative pattern can be established | | |
| 4. Is there documented treatment failure with an adequate trial (at least 8 weeks) of an oral migraine preventive therapy as follows:<br>a. Propranolol 40 mg daily, metoprolol 100 mg daily<br>b. Amitriptyline 25 mg daily<br>c. Topiramate 50 mg daily, valproic acid, divalproex sodium | Yes – Document and go to #5 | No – Criteria not met |
| 5. Is the request for treatment with Vyepti? | Yes – Go to #6 | No – Approve up to 6 months |
| 6. Is there documented treatment failure or intolerable adverse event to one of the preferred drugs (Ajovy, Emgality) AND Botox? | Yes – Approve up to 6 months | No – Criteria not met |
| **Episodic Cluster Headaches - Emgality** | | |
| 1. Is there a history of episodic cluster headaches with at least two cluster periods | Yes – Approve up to 6 months | No – Criteria not met |

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| lasting from 7 days to 1 year (when untreated) that were separated by pain-free remission periods of at least one month? | (Maximum 6 fills per year) | |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success defined as a 50% reduction in monthly headache frequency since starting treatment? | 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** | | |

- **Ajovy**
  - Availability: 225 mg/1.5 mL syringe
  - Dosing: 225 mg every 30 days or 675 mg (3x 225 mg injection) every 90 days
- **Emgality**
  - Availability: 120 mg/1 mL syringe or auto-injector; 100 mg/mL syringe (carton of 3)
  - Dosing:
    - Chronic migraine: 240 mg single loading dose then 120 mg every 30 days
    - Episodic cluster headache: 300 mg at the start of a cluster period and then 300 mg monthly until the end of the cluster period – Maximum 6 fills annually
- **Vyepti**
  - Availability: 100 mg/1 mL single-use vial
  - Dosing: 100 mg infusion every 3 months. Some patients may benefit from a dosage of 300 mg every 3 months

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 | Yes – Go to #7 | No – Criteria not met |

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| specialist? | | |
| 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 | | |
| 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 |

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| 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** | | |
| • **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:
**CHOLBAM**
Affected Medications: CHOLBAM (cholic acid) 50 & 250mg capsules

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | • Patient weight, dose and frequency<br>• Baseline liver function tests (AST, ALT, GGT, ALP, total bilirubin, INR)<br>• Diagnosis confirmed by assessment of serum or urinary bile acid levels using mass spectrometry (Fast Atom Bombardment ionization - Mass Spectrometry (FAB-MS) analysis) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose: 10 to 15 mg/kg orally once daily, or in two divided doses<br>• Dose if concomitant familial hypertriglyceridemia: 11 to 17 mg/kg orally once daily, or in two divided doses<br>• Reauthorization requires documentation of clinically significant improvement in liver function as determined by meeting TWO of the following criteria: ALT or AST values reduced to less than 50 U/L, or baseline levels reduced by 80%; total bilirubin values reduced to less than or equal to 1 mg/dL; no evidence of cholestasis on liver biopsy; body weight increased by 10% or stable at greater than the 50$^{th}$ percentile<br>• Treatment should be discontinued if liver function does not improve after 3 months of start of treatment |
| **Exclusion Criteria:** | • Treatment of extrahepatic manifestations (such as neurologic symptoms) of bile acid synthesis disorders due to single enzyme defects or peroxisomal disorders including Zellweger spectrum disorders |
| **Age Restriction:** | • 3 weeks and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with hepatologist or gastroenterologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**CINACALCET**
Affected Medications: Cinacalcet

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Secondary hyperparathyroidism in adult patients with chronic kidney disease (CKD) on dialysis<br>  ○ Hypercalcemia in adult patients with primary hyperparathyroidism<br>  ○ Hypercalcemia in adult patients with parathyroid carcinoma<br>• Persistent hyperparathyroidism post-renal transplant |
| **Required Medical Information:** | • Documentation confirming one of the following:<br>  ○ Diagnosis of secondary hyperparathyroidism with documentation of chronic kidney disease (CKD)<br>  ▪ Must be on dialysis<br>  ▪ Intact parathyroid hormone (iPTH) level greater than 300 pg/mL<br>  ○ Diagnosis of primary hyperparathyroidism with hypercalcemia<br>  ▪ Baseline serum calcium level (corrected for albumin) greater than 1.0 mg/dL above the testing laboratory's upper limit of normal<br>  ▪ Unable to undergo parathyroidectomy<br>  ○ Diagnosis of parathyroid carcinoma with hypercalcemia<br>  ▪ Disease is unresectable or no longer amenable to surgical intervention<br>  ○ Diagnosis of persistent hyperparathyroidism and hypercalcemia post renal transplant<br>  ▪ Baseline serum calcium level (corrected for albumin) greater than 1.0 mg/dL above the testing laboratory's upper limit of normal<br>  ▪ Parathyroid hormone (PTH) concentration at least 2 times above the testing laboratory's upper limit of normal |

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| **Appropriate Treatment Regimen & Other Criteria:** | Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Serum calcium is less than the lower limit of the normal range |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with an endocrinologist, nephrologist, or oncologist<br>• All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**CIALIS**
Affected Medications: CIALIS 2.5mg, 5mg, tadalafil 2.5mg, 5mg

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| **Covered Uses:** | • Treatment of symptomatic benign prostatic hyperplasia (BPH)<br>• Mental health diagnosis of sexual dysfunction |
| **Required Medical Information:** | • For mental health diagnosis, follow Diagnostic and Statistical Manual of Mental Disorders, fifth edition (DSM-5) diagnostic criteria:<br>A. At least one of the three following symptoms must be experienced on almost all or all (approximately 75%-100%) occasions of sexual activity (in identified situational contexts or, if generalized, in all contexts):<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>B. The symptoms in Criterion A have persisted for a minimum duration of approximately 6 months.<br>C. The symptoms in Criterion A cause clinically significant distress in the individual.<br>D. The sexual dysfunction is not better explained by a nonsexual mental disorder or as a consequence of severe relationship distress or other significant stressors and is not attributable to the effects of a substance/medication or another medical condition. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Benign Prostate Hyperplasia (BPH): failure of at least two generic (alfuzosin ER, doxazosin, finasteride, prazosin, tamsulosin, etc.)<br><br>**<u>Renal function impairment</u>**<br>• **BPH dose adjustment:**<br>○ CrCl 30 – 50 ml/min: 2.5 mg once daily initially; may increase to 5 mg once daily<br>○ CrCl <30ml/min: not recommended<br>• **Erectile dysfunction dose adjustment:** |

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| | ○ CrCl 30 – 50 ml/min: 5 mg once daily initially; maximum dosage is 10 mg (not to be given more frequently than every 48 hours)<br>○ CrCl <30 ml/min: 5 mg (not more frequently than every 72 hours) (maximum dosage)<br><br>**<u>Hepatic function impairment</u>**<br>• **BPH**<br>○ Child-Pugh class C: use is not recommended<br>• **Erectile dysfunction**<br>○ Child-Pugh class A or B: dose should not exceed 10 mg once daily<br>○ Child-Pugh class C: use is not recommended<br><br>Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Erectile dysfunction unrelated to mental health diagnosis of sexual dysfunction |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care<br>• Mental health diagnosis of sexual dysfunction – prescribed by, or in consultation with, a mental health provider |
| **Coverage Duration:** | • Limited to #1 per day<br>• Approval: 12 months, unless otherwise specified |

POLICY NAME:
**CILTACABTAGENE AUTOLEUCEL**
Affected Medications: CARVYKTI (ciltacabtagene autoleucel)

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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 priortherapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Relapsed or Refractory Multiple Myeloma (MM)</u>**<br>• Treatment with four or more prior lines of therapy that included all of the following:<br>  ○ An immunomodulatory agent<br>  ○ A proteasome inhibitor<br>  ○ An anti-CD38 monoclonal antibody<br>• Patient has experienced disease progression after their last regimen or is refractory to their most recent therapy<br><br>**<u>Approved for one-time single infusion only</u>** |
| **Exclusion Criteria:** | • Previously received any chimeric antigen receptor T-cell (CAR-T) therapy<br>• Previously received any B-cell maturation antigen (BCMA) targeted therapy<br>• ECOG score of 2 or greater<br>• History of active or prior significant central nervous system (CNS) disease<br>• Plasma cell leukemia<br>• Allogeneic stem cell transplant within 6 months before apheresis or ongoing treatment with immunosuppressants<br>• Creatinine clearance less than 40 mL/min<br>• Absolute lymphocyte concentration less than 300/µL<br>• Absolute neutrophil count less than 750 cells/mm3<br>• Platelet count less than 50,000/mm3<br>• Hepatic transaminases greater than 3 times the upper limit of normal<br>• Cardiac ejection fraction less than 45% |

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| | • Active serious infection |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Must be prescribed by an oncologist<br>• Oncologist and administering health care facility must be certified and in compliance with the Risk Evaluation and Mitigation Strategies (REMS) requirements<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 1 month, unless otherwise specified (one infusion only) |

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

<table>
<tr><td>Covered Uses:</td><td>All Food and Drug Administration (FDA) approved OR compendia supported indications not otherwise excluded by plan design.<br>• Treatment of relapsing forms of multiple sclerosis (MS), to include relapsing-remitting disease (RRMS) and active secondary progressive (SPMS) disease, in adults.</td></tr>
<tr><td>Required Medical Information:</td><td>• Diagnosis of relapsing or active secondary progressive forms of Multiple Sclerosis (MS) confirmed with MRI (Revised McDonald diagnostic criteria for multiple sclerosis)<br>o Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS<br>• Documentation of previous therapies tried/failed with duration of trial.<br>• Complete blood count (CBC) with differential including lymphocyte count at baseline.<br>• Transaminase within 6 months before initiation of treatment</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td>• Documented failure with at least two other disease-modifying therapies (DMTs) for multiple sclerosis (MS) for at least 3 months<br>• <u>Reauthorization (1 time only</u><u>):</u><br>• Documentation of clinical treatment success<br>• Administer second course starting at least 43 weeks after the last dose of the first course<br>• Dosing according to the approved label:
<table>
<tr><td>Weight Range</td><td colspan="2">Dose in mg (number of 10 mg tablets) per cycle</td></tr>
<tr><td>Kg</td><td>First Cycle</td><td>Second Cycle</td></tr>
<tr><td>40* to less than 50</td><td>40 mg (4 tablets)</td><td>40 mg (4 tablets)</td></tr>
<tr><td>50 to less than 60</td><td>50 mg (5 tablets)</td><td>50 mg (5 tablets)</td></tr>
<tr><td>60 to less than 70</td><td>60 mg (6 tablets)</td><td>60 mg (6 tablets)</td></tr>
<tr><td>70 to less than 80</td><td>70 mg (7 tablets)</td><td>70 mg (7 tablets)</td></tr>
<tr><td>80 to less than 90</td><td>80 mg (8 tablets)</td><td>70 mg (7 tablets)</td></tr>
<tr><td>90 to less than 100</td><td>90 mg (9 tablets)</td><td>80 mg (8 tablets)</td></tr>
<tr><td>100 to less than 110</td><td>100 mg (10 tablets)</td><td>90 mg (9 tablets)</td></tr>
<tr><td>110 and above</td><td>100 mg (10 tablets)</td><td>100 mg (10 tablets)</td></tr>
</table>
*The use of MAVENCLAD in patients weighing less than 40 kg has not been investigated</td></tr>
</table>

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| **Exclusion Criteria:** | • Patients with current malignancy<br>• Patients infected with Human Immunodeficiency Virus (HIV)<br>• Treatment naïve<br>• Treatment beyond 2 years |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with a neurologist or a Multiple Sclerosis (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 – at least 43 weeks after last dose, 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. |
| **Required Medical Information:** | • Documentation of dose based on reasonable projections and current dose utilization and product labeling, diagnosis, baseline factor level, circulating factor activity (% of normal or units/dL) and rationale for use<br>• Patient weight<br>• Documentation with one of the following diagnostic categories:<br>  ○ On-demand treatment and control of bleeding episodes<br>  ○ Perioperative management of bleeding in patients with mild and moderate hereditary Factor X deficiency<br>  ○ Routine prophylaxis to reduce the frequency of bleeding episodes<br>• Reauthorization (Routine Prophylaxis only): requires documentation of planned treatment dose, number of acute bleeds since last approval with severity and cause of bleed |
| **Appropriate Treatment Regimen & Other Criteria:** | • Food and Drug Administration (Food and Drug Administration (FDA))-approved dosing |
| **Exclusion Criteria:** | • Maintenance therapy (not Food and Drug Administration (FDA)-approved) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br>• Perioperative management: 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:** | • Approval: 3 months, unless otherwise specified |

POLICY NAME:
**CONTINUOUS GLUCOSE MONITORS**
Affected Products: Freestyle Libre, Freestyle Libre 2, Freestyle Libre 3, Dexcom G6, Dexcom G7

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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 diabetes mellitus diagnosis **AND**<br>• Currently on insulin treatment of at least 3 subcutaneous (SubQ) injections daily OR on an insulin pump, **AND**<br>• Performing at least 4 blood glucose testings per day with a home blood glucose monitoring device, **AND**<br>• Requiring frequent insulin dose adjustments based on home blood glucose monitoring readings |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | • Type 2 diabetes not on intensive insulin therapy<br>• Use of continuous glucose monitor while pregnant<br>• Use of continuous glucose monitor while on dialysis |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care<br>• In-person visit for diabetes management with requesting provider, within 6 months prior to request, documenting need for continuous glucose monitoring (CGM) |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**COPAXONE**
Affected Medications: Copaxone 20mg/ml, Copaxone 40mg/ml

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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:** | • Documentation of dose and frequency as the 20 mg/mL and 40 mg/mL formulations are not interchangeable<br><br>**Reauthorization**: provider attestation of treatment success |
| **Exclusion Criteria:** | • Concurrent use of other disease-modifying medications for the treatment of MS |
| **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:
**CORLANOR**
Affected Medications: CORLANOR (ivabradine)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Inappropriate sinus tachycardia |
| **Required Medical Information:** | <u>Chronic heart failure</u><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>• Documentation of tried or currently receiving one beta blocker (metoprolol succinate extended release, carvedilol, or carvedilol extended release) at the maximally tolerated dose for heart failure treatment OR<br>• Documentation of medical reason for avoidance of beta-blockers<br><br><u>Inappropriate sinus tachycardia</u><br>• Heart rate of at least 100 beats per minute, with average mean heart rate of at least 90 beats per minute over 24 hours not due to appropriate physiologic response or primary abnormality (hyperthyroidism or anemia)<br>• Symptomatic (palpitations, shortness of breath, dizziness, and/or decreased exercise capacity)<br>• Documentation for absence of identifiable causes of sinus tachycardia and exclusion of atrial tachycardia |
| **Appropriate Treatment Regimen & Other Criteria:** | • Effective contraception is recommended in women of child-bearing age<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy; 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>• Resting heart rate of less than 60 bpm prior to treatment<br>• Sick sinus syndrome, sinoatrial block, third-degree atrioventricular block (unless stable with functioning demand pacemaker)<br>• Severe hepatic impairment (Child-Paugh class C) |

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| | • Heart rate maintained exclusively by pacemaker<br>• Concomitant use with strong CYP3A4 inhibitors/inducers |
| **Age Restriction:** | |
| **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:** | • Initial Approval: 12 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**COVERAGE OF DESCOVY AT TIER 0 COPAY**
Affected Medications**:** DESCOVY (emtricitabine and tenofovir alafenamide)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design<br>  ○ HIV-1 infection, Pre-exposure prevention (PrEP) |
| **Required Medical Information:** | **<u>For HIV-1 PrEP:</u>**<br>• Documented treatment failure or intolerable adverse event to emtricitabine 200 mg/tenofovir disoproxil fumerate 300 mg |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | • Treatment of HIV-1 infection (not used for PrEP) |
| **Age Restriction:** | |
| **Prescriber Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 12 months |

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 benefit design.<br>  ○ To reduce the frequency of vasoocclusive crises (VOCs) in adults and pediatric patients aged 16 years and older with sickle cell disease. |
| **Required Medical Information:** | • 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:** | **Reauthorization** requires documentation of treatment success defined by a decrease in the number of sickle cell-related 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 hemoglobin oxygen affinity modulator (voxelotor) |
| **Age Restriction:** | • Greater than or equal to 16 years of age |
| **Prescriber Restrictions:** | • Prescribed by or in consultation with hematologist.<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 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: CYSTAGON (cysteamine bitartrate), 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 |
| **Required Medical Information:** | • Diagnosis of nephropathic cystinosis<br>• The diagnosis was confirmed by the presence of increased cysteine concentration in leukocytes (generally 3-23 nmol half-cysteine/mg protein) or by DNA testing (mutations in the CTNS gene) or by demonstration of cysteine corneal crystals by the slit lamp examination |
| **Appropriate Treatment Regimen & Other Criteria:** | For Procysbi request:<br>• Documented treatment failure, intolerance, or clinical rationale for avoidance of Cystagon |
| **Exclusion Criteria:** | • Documented history of hypersensitivity to cysteamine or penicillamine |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**DALFAMPRIDINE**
Affected Medications: AMPYRA (dalfampridine), dalfampridine

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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 Multiple Sclerosis with documented impairment, but able to walk with or without assistance<br>• Documentation of baseline timed 25 foot walk test |
| **Appropriate Treatment Regimen & Other Criteria:** | **Coverage for brand Ampyra**: requires documented treatment failure or intolerance to a minimum of 2 separate generic manufacturers of dalfampridine.<br><br>**Reauthorization** requires documentation of treatment success defined as a stabilization or improvement from baseline in timed walking speed (timed 25 foot walk). |
| **Exclusion Criteria:** | • History of seizures |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or after consultation with a neurologist or an MS specialist.<br>• All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**DASATINIB**
Affected Medications: SPRYCEL (dasatinib)

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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 of Philadelphia chromosome-positive mutation status |
| **Appropriate Treatment Regimen & Other Criteria:** | • For patients with Chronic Myeloid Leukemia (CML) and low risk score, documented clinical failure with Imatinib<br>• For patients with Acute Lymphoblastic Leukemia (ALL), documented clinical failure with imatinib<br><br>**Reauthorization** requires documentation of disease responsiveness to therapy (as applicable, BCR-ABL1 transcript levels, cytogenetic response) |
| **Exclusion Criteria:** | • 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:** | • All approvals are subject to utilization of the most cost-effective site of care<br>• Prescribed by, or in consultation with, an oncologist |
| **Coverage Duration:** | • Initial Authorization: 4 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. |
| **Required Medical Information:** | • Diagnosis of hepatic veno-occlusive disease (VOD), also known as sinusoidal obstruction syndrome (SOS), **AND**<br>• Renal and/or pulmonary dysfunction following hematopoietic stem cell transplantation (HSCT) **AND**<br>• Weight prior to HSCT, dose and frequency **AND**<br>• Renal function data<br>  ○ Serum creatinine (SCr) prior to admission for HSCT conditioning, during conditioning before HSCT, or Creatinine clearance (CrCl) or glomerular filtration rate (GFR) prior to admission<br>  ○ Current SCr, CrCl, or GFR<br>• Pulmonary function data<br>  ○ Oxygen saturation on room air or requirement for oxygen supplementation/ventilator dependence<br><br><u>Reauthorization Criteria</u><br>• 21 days of therapy have been completed **AND**<br>• Total bilirubin level is still above normal (normal varies by lab, ~0.1-1.2 mg/dL or 1.71-20.5 microM/L) |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | • Renal dysfunction secondary to an alternate etiology<br>• Insufficiently severe renal dysfunction defined as:<br>  ○ SCr less than 3x the value at admission for HSCT conditioning **OR**<br>  ○ SCr less than 3x the lowest value during conditioning before HSCT **OR**<br>  ○ CrCl or GFR greater than 40% of admission value **OR**<br>  ○ Not dialysis dependent after HSCT<br>• Pulmonary dysfunction secondary to an alternate etiology<br>• Insufficiently severe pulmonary dysfunction<br>  ○ Oxygen saturation greater than 90% on room air **OR** |

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| | ○ No documented requirement for oxygen supplementation/ventilator dependence<br>• Preexisting liver cirrhosis<br>• Any of the following without diagnosis of VOD or SOS with renal or pulmonary dysfunction following HSCT, hyperbilirubinemia, ascites, weight gain, and/or hepatomegaly<br>• Prior solid organ transplant<br>• Dialysis dependence at the time of HSCT<br>• Oxygen dependence during conditioning<br>• Hemodynamic instability (requirement for multiple pressors or inability to maintain mean arterial pressure with single-pressor support).<br>• Concomitant use of medications increasing hemorrhagic risk (e.g.-anticoagulants and/or fibrinolytics)<br>• Presence of active bleeding |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | |
| **Coverage Duration:** | • Authorization: 1 month, unless otherwise specified<br>• Reauthorization: 2 weeks, may only reauthorize total of two times, unless otherwise specified |

POLICY NAME:
**DEFLAZACORT**
Affected Medications: Emflaza (deflazacort)

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| **Covered Uses:** | • All 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:** | • Chart note documentation showing a trial of prednisone causing one of the following:<br>  ○ Unmanageable and clinically significant weight gain/obesity after at least 3 months of treatment or<br>  ○ Psychiatric/behavioral issues (e.g., abnormal behavior, aggression, irritability) that persists beyond the first six weeks of prednisone treatment<br>• Reauthorization requires a documented improvement from baseline or stabilization of motor function |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 2 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by a specialist with experience in the treatment of DMD<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:
**DEUTETRABENAZINE**
Affected Medications: AUSTEDO (deutetrabenazine)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | **<u>Chorea related to Huntington's Disease</u>**<br>• Diagnosis of Huntington's Disease with Chorea requiring treatment<br>• Total functional capacity score of 5 or higher on a scale of 13 (A score <5 indicates moderate to severe impairment of function, requiring a full-time caregiver- was excluded from clinical trials)<br><br>**<u>Tardive Dyskinesia</u>**<br>• Diagnosis of tardive dyskinesia requiring treatment defined as 10 or greater on AIMS.<br>• History of dopamine receptor antagonist (Antipsychotic, metoclopramide) use for 3 months if less than 60 years old.<br>• History of dopamine receptor antagonist (Antipsychotic, metoclopramide) use for 1 month if 60 years old and older. |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Chorea related to Huntington's Disease</u>**<br>• Maximum labeled dose: 48 mg/day (Dose is typically started at 6 mg/day and titrated upward to effect or tolerability)<br>• Reauthorization requires documentation of treatment success defined as a clinically significant improvement in function or decrease in Chorea<br>  ○ If disease has progressed to the point of inability to walk/need for a full-time caregiver reauthorization is not appropriate<br><br>**<u>Tardive Dyskinesia</u>**<br>• Documented inability to discontinue offending agent or persistent dyskinesia in spite of cessation<br>• Maximum labeled dose: 48 mg/day (Dose is typically started at 12 mg/day, 6mg twice daily, and titrated upward to effect or tolerability) |

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| | • Reauthorization requires documentation of treatment success defined as a clinically significant improvement with a decrease in AIMS score from baseline. |
| **Exclusion Criteria:** | • Untreated or inadequately treated depression or suicidal ideation<br>• Concomitant use of an MAOI (monoamine oxidase inhibitor) (must be >14 days post discontinuing therapy)<br>• Concomitant use of tetrabenazine (Xenazine)<br>• Severe hepatic impairment |
| **Age Restriction:** | • Safety and effectiveness in pediatric patients have not been established. |
| **Prescriber 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 approval: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

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

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| **Covered Uses:** | • All 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) and ongoing hemodialysis treatment<br>• Documentation of history of significant pruritus associated with initiation of HD<br>• Documentation of normal serum magnesium, parathyroid hormone, and phosphate<br>• Documentation of patient's current dry weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of inadequate relief after a trial lasting at least one month for each of the following first-line recommended or conventional therapies:<br>  ○ Topical therapies (such as emollients or analgesics)<br>  ○ Oral antihistamines (such as hydroxyzine or diphenhydramine)<br>  ○ Gabapentin or pregabalin<br><br>**Reauthorization** will require documented treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Peritoneal dialysis<br>• Severe hepatic impairment |
| **Age Restriction:** | • Greater than or equal to 18 years of age |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by nephrologist or a specialist with experience in the treatment of CKD-aP<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:
**DIMETHYL FUMARATE**
Affected Medications: TECFIDERA (dimethyl fumarate), dimethyl fumarate

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded from plan benefits. |
| **Required Medical Information:** | • Diagnosis of relapsing forms of multiple sclerosis confirmed with magnetic resonance imaging (MRI)<br>• Complete blood count with lymphocyte count (within 6 months) before initiating treatment, then annually and as clinically indicated |
| **Appropriate Treatment Regimen & Other Criteria:** | • Initial dose of 120mg twice daily for 7 days, then increasing to 240mg twice daily thereafter<br>• Hold therapy for four weeks if lymphocyte count is less than 500/$mm^3$ for greater than 6 months<br>• No concurrent use of medications indicated for the treatment of relapsing-remitting multiple sclerosis<br>• Not approved for primary progressive multiple sclerosis<br>• Reauthorization: provider attestation of treatment success |
| **Exclusion Criteria:** | • Pre-existing low lymphocyte counts (less than 500/$mm^3$) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or after consultation with a neurologist or an MS specialist.<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise noted. |

POLICY NAME:
**DINUTUXIMAB**
Affected Medications: UNITUXIN (dinutuximab)

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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, disease staging, all prior therapies used, and prescribed dosing regimen<br>• Documentation of Neuroblastoma, High risk, with at least a partial response to prior first-line multi-agent, multimodality therapy |
| **Appropriate Treatment Regimen & Other Criteria:** | • Maximum duration: 5 cycles<br>• Must be used in combination with granulocyte-macrophage colony-stimulating factor, interleukin-2, and 13-cis-retinoic acid<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Hold therapy if Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 5 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. |
| **Required Medical Information:** | • Confirmed diagnosis of Long Chain 3 hydroxyacyl-Coa dehydrogenase deficiency or Very long-chain acyl-CoA dehydrogenase deficiency based on trifunctional protein gene analysis or enzyme assay.<br>• Documentation of patient weight and total prescribed daily caloric intake<br>• Documentation of severe disease despite diet management as evidenced by one of the following:<br>  ○ Hypoglycemia after short periods of fasting<br>  ○ Evidence of functional cardiomyopathy<br>  ○ Frequent severe major medical episodes requiring emergency room acute care or hospitalization (3 within the past year or 5 with past 2 years)<br>  ○ Elevated creatinine kinase (chronic or episodic) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose not to exceed 35% of Daily Caloric Intake<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:** | • Endocrinologist or provider experience in 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:
**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:** | • Known hypersensitivity to dornase alfa, Chinese Hamster Ovary cell products, or any component of the product. |
| **Age Restriction:** | • 1 month or older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 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. |
| **Required Medical Information:** | • Diagnosis of neurogenic orthostatic hypotension (nOH) caused by one of the following:<br>  ○ Primary autonomic failure such as Parkinson's disease (PD), multiple system atrophy (MSA), or pure autonomic failure (PAF)<br>  ○ Dopamine beta-hydroxylase deficiency<br>  ○ Non-diabetic autonomic neuropathy<br>• Documentation of severe symptomatic orthostatic hypotension (decrease in systolic blood pressure of at least 20 mmHg or diastolic blood pressure of at least 10 mmHg within 3 minutes after standing) affecting activities of daily living<br>• Baseline dizziness score (item 1) on the Orthostatic Hypotension Symptom Assessment (OHSA) |
| **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 defined as a greater than or equal to 1 point improvement on OHSA item 1 score. |
| **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 approval: 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. |
| **Required Medical Information:** | • Diagnosis of idiopathic Parkinson's Disease (PD) based on presence of bradykinesia and at least one other cardinal PD feature (tremor, rigidity, postural instability) AND<br>• Levodopa responsive with clearly defined "On" periods AND<br>• Persistent motor complications with disabling "Off" periods for a minimum of 3 hours/day, despite optimal medical therapy with oral levodopa-carbidopa, and at least two other classes of anti-PD therapy (i.e. COMT, MAO-B inhibitor, or dopamine agonist) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Duopa is delivered as a 16-hour infusion through either a naso-jejunal tube for SHORT-term administration or through a PEG-J for LONG-term administration<br>• Reauthorization will require 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 a neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • 12 months, unless otherwise specified |

POLICY NAME:
**DUPILUMAB**
Affected Medications: DUPIXENT (dupilumab subcutaneous injection)

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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 for use in combination with another monoclonal antibody (Fasenra, Nucala, Xolair, Cinqair)? | Yes – Criteria not met; combination use is experimental | No – Go to #3 |
| 3. Is the request to treat a diagnosis according to one of the Food and Drug Administration (FDA)-approved indications?<br>○ Add-on maintenance treatment in patients with moderate-to-severe asthma aged 12 years and older with an eosinophilic phenotype or with oral corticosteroid dependent asthma<br>○ Treatment of patients aged 6 months and older with moderate-to-severe atopic dermatitis whose disease is not adequately controlled with topical prescription therapies or when those therapies are not advisable<br>○ Add-on maintenance treatment in adult patients with inadequately controlled chronic rhinosinusitis with nasal polyposis (CRSwNP)<br>○ Treatment of adult and pediatric patients aged 12 years and older, weighing at least 40 kg, with eosinophilic esophagitis (EoE) | Yes – Go to appropriate section below | No – Criteria not met |

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| ○ Treatment of adults (ages 18 years and older) with prurigo nodularis (PN) | | |
| **Moderate-to-Severe Eosinophilic Asthma** | | |
| 1. Is there documentation of severe eosinophilic asthma defined by the following:<br>○ Baseline eosinophil count at least 300 cells/µL<br>**AND**<br>○ FEV1 less than 80% at baseline or FEV1/FVC reduced by at least 5% from normal | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documented use of high-dose inhaled corticosteroid (ICS) plus a long-acting beta agonist (LABA) for at least three months with continued symptoms? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there a documented history of 2 or more asthma exacerbations requiring oral or systemic corticosteroid treatment in the past 12 months while on combination inhaled treatment and at least 80% adherence? | Yes – Go to #5 | No – Go to #4 |
| 4. Is there documentation that chronic daily oral corticosteroids are required? | Yes – Go to #5 | No – Criteria not met |
| 5. Is the drug prescribed by or in consultation with an Allergist, Immunologist, or Pulmonologist? | Yes – Approve up to 6 months | No – Criteria not met |
| **Moderate-to-severe atopic dermatitis** | | |
| 1. Is there documentation of severe inflammatory skin disease defined as functional impairment (inability to use | Yes – Go to #2 | No – Criteria not met |

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| hands or feet for activities of daily living, or significant facial involvement preventing normal social interaction)? | | |
| 2. Is there a documented body surface area (BSA) effected of at least 10% OR hand, foot or mucous membrane involvement? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documented failure with at least 6 weeks of treatment with one of the following: Tacrolimus ointment, pimecrolimus cream, Eucrisa? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is there documented treatment failure with two of the following for at least 12 weeks: Phototherapy, cyclosporine, azathioprine, methotrexate, mycophenolate? | Yes – Document and go to #5 | No – Criteria not met |
| 5. Is the drug prescribed by or in consultation with a specialist in the treatment of atopic dermatitis (Such as a dermatologist)? | Yes – Approve up to 6 months | No – Criteria not met |
| **Chronic rhinosinusitis with nasal polyps (CRSwNP)** | | |
| 1. Is there documentation of chronic sinusitis after total ethmoidectomy with a need for revision endoscopic sinus surgery due to continued symptoms of nasal congestion/obstruction from recurrent bilateral sinus obstruction due to nasal polyps? | Yes – Go to #2 | No – Criteria not met |
| 2. Is there documented failure with at least 1 intranasal corticosteroid (such as fluticasone) after ethmoidectomy? | Yes – Document and go to #3 | No – Criteria not met |

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| 3. Is there documented failure with Sinuva implant? | Yes – Go to #4 | No – Criteria not met |
| 4. Is the drug prescribed by a specialist in the treatment of nasal polyps (otolaryngologist)? | Yes – Approve up to 6 months | No – Criteria not met |
| **Eosinophilic Esophagitis (EoE)** | | |
| 1. Is there a confirmed diagnosis of EOE by endoscopic biopsy? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is the patient's age 12 years or older and body weight above or equal to 40 kg? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there a history of TWO or more dysphagia episodes per week despite current treatment? | Yes – Go to # 4 | No – Criteria not met |
| 4. Is there documented treatment failure (minimum of at least a 12-week trial) to both of the following:<br>a. High dose (twice daily dosing) Proton Pump Inhibitor (PPI)<br>b. Swallowed inhaled respiratory corticosteroid therapy (such as fluticasone or budesonide) | Yes – Go to #5 | No – Criteria not met |
| 5. Is the drug prescribed by or in consultation with a specialist in the treatment of EoE such as a gastroenterologist or allergy/immunology specialist? | Yes – Approve up to 6 months | No – Criteria not met |
| **Prurigo Nodularis (PN)** | | |

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| 1. Is there a presence of at least 20 PN lesions in total on legs, arms, and trunk. | Yes –go to #2 | No – Criteria not met |
| 2. Has PN been diagnosed by a dermatologist for at least 3 months and confirmed with skin biopsy? | Yes –go to #3 | No – Criteria not met |
| 3. Is there documented Worst-Itch Numeric Rating Scale (WI-NRS) score of at least 7 on a scale of 0 (no itch) to 10 (worst imaginable itch) in the past week? | Yes – go to # 4 | No – Criteria not met |
| 4. Is there documented treatment failure (minimum of at least a 2-week trial) to medium to super high potency topical corticosteroid AND one of the following for at least 12 weeks?<br>i. Phototherapy (NBUVB or PUVA)<br>ii. Methotrexate<br>iii. Cyclosporine | Yes – Go to #5 | No – Criteria not met |
| 5. 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 |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider? | Yes – Go to #2 | No – Criteria not met |
| 2. Is the request for use in combination with another monoclonal antibody (Fasenra, Nucala, Xolair, Cinqair)? | Yes – Criteria not met, combination use is experimental | No – Go to #3 |

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| 3. 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**

- **Dupixent**
  - Availability: 300 mg/2 mL pre-filled syringe or pre-filled pen, 200 mg/1.14 mL pre-filled syringe or pre-filled pen, 100 mg/0.67 mL pre-filled syringe
  - Dosing:
    - **Atopic Dermatitis:**
      - Children greater than or equal to 6 months and up to 5 years of age (no initial loading dose is recommended):
        - 5 to less than 15 kg: 200 mg every 4 weeks
        - 15 to less than 30 kg: 300 mg every 4 weeks
      - Children 6 to 17 years of age:
        - 15 to less than 30 kg: Initial dose of 600 mg (two 300 mg injections) followed by 300 mg every 4 weeks
        - 30 to less than 60 kg: Initial dose of 400 mg (two 200 mg injections) followed by 200 mg every other week
        - Greater than 60 kg: Initial dose of 600 mg (two 300 mg injections) followed by 300 mg every other week
      - Adults 18 years and older:
        - Initial dose of 600 mg (two 300 mg injections), followed by 300 mg given every other week

    - **Asthma**:

      Children 6 to 11 years old: NO LOADING DOSE RECOMMENDED
      - 15 kg to less than 30 kg: 100 mg every other week OR 300 mg every 4 weeks
      - 30 kg or greater: 200 mg every other week

      Adults and adolescents 12 years of age and older: Initial dose of 400 mg (two 200 mg injections) followed by 200 mg given every other week or initial dose of 600 mg (two 300 mg injections) followed by 300 mg given every other week

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| ▪ **CRSwNP**: 300 mg every other week<br><br>▪ **Eosinophilic Esophagitis:**<br>• Adults and children (12 years of age and older):<br>- 40kg or greater: 300 mg every week<br><br>▪ **Prurigo Nodularis:**<br>• Initial dose of 600 mg (two 300 mg injections) followed by 300 mg every other week<br><br>*Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced for all medical infusion drugs | |
| **Coverage Duration:** | • Initial approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ECULIZUMAB**
Affected Medications: SOLIRIS (eculizumab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design.<br>  ○ List indications<br>• Compendia-supported uses that will be covered (if applicable) |
| **Required Medical Information:** | • Documentation of complete treatment course<br>• Complete blood count (CBC), reticulocyte count, lactate dehydrogenase (LDH), packed RBC transfusion requirement<br>• Patients must be administered a meningococcal vaccine at least two weeks prior to initiation of Soliris therapy and revaccinated according to current ACIP guidelines<br><br>**<u>Paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis</u>**<br>• Platelet count greater than or equal to 30,000/mcl<br>• LDH levels greater than or equal to 1.5 times the upper limit of normal range.<br>• Flow cytometry shows GPI deficient red blood cell clone (type III cells) greater than or equal to 10%<br>• 4 or more blood transfusions required in the previous 12 months<br><br>**<u>Atypical hemolytic uremic syndrome (aHUS) to inhibit complement-medicated thrombotic microangiopathy</u>**<br>• Clinical presentation of: microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury<br>• LDH levels greater than or equal to 1.5 times the upper limit of normal range.<br>• ADAMTS13 activity level greater than 10%<br>• Patient has failed to respond to five days of plasma therapy<br>• 4 or more blood transfusions required in the previous 12 months<br><br>**<u>Generalized Myasthenia Gravis (gMG)</u>**<br>• Diagnosis of generalized Myasthenia Gravis (gMG) confirmed by:<br>  ○ A history of abnormal neuromuscular transmission test OR |

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| | ○ A positive edrophonium chloride test OR<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 anti-acetylcholine receptor (AchR) antibodies<br>• MG-Activities of Daily Living (MG-ADL) total score of greater than or equal to 6<br>• Documentation of baseline Quantitative Myasthenia Gravis (QMG) score<br>• Documentation of gMG treatment history showing the following:<br>○ Currently on a stable dose of at least one gMG therapy (acetylcholinesterase inhibitor, corticosteroid, or non-steroidal immunosuppressive therapy (NSIST))<br>○ One of the following:<br>- Documented treatment failure with an adequate trial (one year or more) of at least 2 immunosuppressive therapies (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)<br>OR<br>- Documented 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<br>• Documented treatment failure with Vyvgart and Ultomiris<br><br>**<u>Neuromyelitis Optica Spectrum Disorder (NMOSD)</u>**<br>• Diagnosis of NMOSD with AQP4-IgG requiring all of the following:<br>○ At least one core clinical characteristic:<br>▪ Optic neuritis<br>▪ Acute myelitis<br>▪ Area postrema syndrome: Episode of otherwise unexplained hiccups or nausea and vomiting<br>▪ Acute brainstem syndrome |

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| | ▪ Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMSOD-typical diencephalic MRI lesions<br>▪ Symptomatic cerebral syndrome with NMOSD-typical brain lesions<br>○ Positive test for AQP4-IgG using best available detection method<br>○ Exclusion for alternative diagnoses<br>• Documented treatment failure with 12 weeks of at least 2 of the following immunosuppressive therapies: azathioprine, mycophenolate, methotrexate<br>• Documented treatment failure with 12 weeks of at least 1 of the following: mitoxantrone (authorization required), rituximab (authorization required)<br>• Documented treatment failure with Enspryng and Uplizna (authorization required for both) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis</u>**<br>○ 600 mg weekly for the first 4 weeks, followed by<br>○ 900 mg for the fifth dose 1 week later, then<br>○ 900 mg every 2 weeks thereafter<br><br>**<u>Atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy</u>**<br>• Appropriate weight based adjustment if younger than 18 years old or less than 40kg; <u>otherwise</u>:<br>○ 900 mg weekly for the first 4 weeks, followed by<br>○ 1200 mg for the fifth dose 1 week later, then<br>○ 1200 mg every 2 weeks thereafter<br><br>**<u>Generalized Myasthenia Gravis (gMG)</u>**<br>• 900 mg weekly for the first 4 weeks, followed by<br>• 1200 mg for the fifth dose 1 week later, then<br>• 1200 mg every 2 weeks thereafter<br><br>**<u>Neuromyelitis Optica Spectrum Disorder (NMOSD)</u>**<br>• 900 mg weekly for the first 4 weeks, followed by<br>• 1200 mg for the fifth dose 1 week later, then |

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| | • 1200 mg every 2 weeks thereafter<br><br>**Dose Adjustment in Case of Plasmapheresis, Plasma Exchange, or Fresh Frozen Plasma Infusion**<br>• For adult and pediatric patients with aHUS, and adult patients with gMG or NMOSD, supplemental dosing of Soliris is required in the setting of concomitant plasmapheresis or plasma exchange, or fresh frozen plasma infusion<br><br>**<u>Reauthorization requires:</u>**<br>• gMG, NMOSD: documentation of treatment success<br>• PNH, aHUS: updated serum LDH and Hb labs, and blood transfusion history, showing treatment success and clinically significant response to therapy |
| **Exclusion Criteria:** | • Concurrent use with other monoclonal antibodies (rituximab, inebilizumab, tocilizumab, etc.) or IVIG<br>• Current meningitis infection<br>• Shiga toxin E. coli related hemolytic uremic syndrome (STEC-HUS). |
| **Age Restriction:** | • PNH, gMG and NMOSD: 18 years of age or older<br>• aHUS: 2 months of age or older |
| **Prescriber/Site of Care Restrictions:** | • PNH: hematologist<br>• aHUS: hematologist or nephrologist<br>• gMG: neurologist<br>• NMOSD: neurologist or neuro-opthalmologist<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:
**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:** | • Definite or probable Amyotrophic lateral sclerosis (ALS) based on El Escorial revised (Airlie House) 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 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>• Reauthorization: Treatment success as determined by prescriber including retaining most activities of daily living |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • 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 approval: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**EFGARTIGIMOD ALPHA**
Affected Medications: VYVGART (efgartigimod alpha)

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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 generalized Myasthenia Gravis (gMG) confirmed by:<br>  ○ A history of abnormal neuromuscular transmission test OR<br>  ○ A positive edrophonium chloride test OR<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 anti-acetylcholine receptor (AchR) antibodies<br>• MG-Activities of Daily Living (MG-ADL) total score of 5 or greater<br>• Documentation of baseline Quantitative Myasthenia Gravis (QMG) score |
| **Appropriate Treatment Regimen & Other Criteria:** | • Prior to initiating therapy for gMG, the following criteria must be met:<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<br>  ○ AND one of the following:<br>  - Documented treatment failure with an adequate trial (one year or more) of at least 2 immunosuppressive therapies (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)<br>  OR<br>  - Documented 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<br><br>**Dosing**:<br>10 mg/kg (max dose 1200 mg) IV once weekly for 4 weeks. |

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| | Administer subsequent treatment cycles based on clinical evaluation, but no sooner than 8 weeks from initiation of the previous cycle.<br><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 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 |
| **Exclusion Criteria:** | • IgG levels less than 600 mg/dL at baseline<br>• Concurrent use with other antibody fragments, monoclonal antibodies (rituximab, eculizumab, etc.), or maintenance IVIG |
| **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:
**ELAGOLIX**
Affected Medications: ORILISSA (Elagolix oral tablets), ORIAHNN (Elagolix/estradiol/norethindrone acetate)

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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 |
| **Uterine Fibroids – Oriahnn** | | |
| 1. Is there attestation of premenopausal status? | Yes –Go to #2 | No – Criteria not met |
| 2. Is there attestation that the member does not have a history of osteoporosis? | Yes – Go to #3 | No – Criteria not met |
| 3. Is there attestation from the provider that the member is not pregnant and does not have plans to become pregnant? | Yes – Go to #4 | No – Go to |
| 4. Is there documentation of a diagnosis of heavy menstrual bleeding associated with uterine leiomyomas? | Yes – Approve up to 6 months | No – Criteria not met |
| **Pain due to endometriosis – Orilissa** | | |
| 1. Is there attestation of premenopausal status? | Yes – Go to #2 | No – Criteria not met |

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| 2. Is there attestation that the member does not have a history of osteoporosis? | Yes – Go to #3 | No – Criteria not met |
| 3. Is there attestation from the provider that the member is not pregnant and does not have plans to become pregnant? | Yes – Go to #4 | No – Criteria not met |
| 4. Is there documentation of a diagnosis of moderate to severe pain associated with endometriosis? | Yes – go to #5 | No – Criteria not met |
| 5. Is there documentation of a trial and inadequate relief after at least three months of first-line therapy with nonsteroidal anti-inflammatory drugs (NSAIDs) and continuous (no placebo pills) hormonal contraceptives? | Yes – Document and approve up to 6 months | No – Criteria not met |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider? | 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 18 months for:<br>• Oriahnn<br>• Orilissa 150 mg once daily* | No – Criteria not met |
| **Quantity Limitations** | | |
| • **Oriahnn**<br>  ○ 56 tablets per 28 days<br>• **Orilissa**<br>  ○ 150 mg: 30 tablets per 30 days | | |

- 200 mg: 60 tablets per 30 days

*Maximum treatment duration for 200 mg twice daily, or 150 mg once daily with moderate hepatic impairment (Child-Pugh Class B) is 6 months. Reauthorization not allowed

POLICY NAME:
**ELAPRASE**
Affected Medications: ELAPRASE (idursulfase)

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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 Hunter syndrome (Mucopolysaccharidosis type II, MPS II)<br>• Diagnosis confirmed by enzyme assay demonstrating a deficiency of iduronate 2-sulfatase enzyme activity or by DNA testing that shows pathologic iduronate 2-sulfatase gene mutation<br>• Documentation of baseline values for 6-minute walk test (6-MWT) and/or percent predicted forced vital capacity (FVC)<br>• Must have symptoms attributable to MPS II such as: developmental delay, cognitive impairment, frequent infections, hearing loss, hepatosplenomegaly, hernias, impaired respiratory function, joint pain, skeletal deformities, sleep apnea or valvular heart disease |
| **Appropriate Treatment Regimen & Other Criteria:** | • In case of anaphylaxis or severe allergic reaction, there will be appropriate medical support readily available when Elaprase is administered<br>• QL- 0.5 mg/kg infusion once weekly<br><br>**Reauthorization**: Documentation of clinical response and toleration of agent<br>• Clinical Response: Demonstrated a response to therapy compared to pretreatment baseline: stabilization or improvement in 6-MWT and/or FVC **AND**<br>• Toleration of agent: absence of unacceptable toxicity from the drug.<br>Examples of unacceptable toxicity include the following: severe hypersensitivity including anaphylactic reactions, antibody development and serious adverse reactions, acute respiratory complications, acute cardiorespiratory failure, etc. |
| **Exclusion Criteria:** | |
| **Age** | • 5 years of age and older |

| Restriction: | |
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| **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 physician who specializes in the treatment of inherited metabolic disorders |
| **Coverage Duration:** | • Initial approval 3 months, unless otherwise specified<br>• Subsequent approval 12 months unless otherwise specified |

POLICY NAME:
**ELIGLUSTAT**
Affected Medications: CERDELGA (eliglustat)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Gaucher disease type 1 (GD1) |
| **Required Medical Information:** | • Diagnosis must be documented in the members chart notes within the past 6 months<br>• Diagnosis confirmed by enzyme assay<br>• Documentation of cytochrome P450 2D6 (CYP2D6) genotype by a FDA-approved test indicating CYP2D6 extensive metabolizers, intermediated metabolizers, or poor metabolizers<br>• Documentation of complete and current treatment course<br>• Documentation of baseline tests such as hemoglobin level, platelet count, liver function tests, renal function tests. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of failure, intolerance, or clinical rationale for the avoidance of combination therapy with imiglucerase (Cerezyme), and failure with imiglucerase (Cerezyme) monotherapy<br><br>**Extensive or Immediate Metabolizers of CYP2D6**<br>• Quantity limit - 84 mg capsules #60 per 30 days<br><br>**Poor Metabolizers of CYP2D6**<br>• Quantity limit - 84 mg capsules #30 per 30 days<br><br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy. |
| **Exclusion Criteria:** | • CYP2D6 ultrarapid metabolizers<br>• Moderate or severre hepatic impairment<br>• Pre-existing cardiac disease (congestive heart failure, myocardial infarction, bradycardia, heart block, arrhythmias, and long QT syndrome)<br>• Treatment with Class 1A (e.g., quinidine, procainaminde) and Class III (e.g., amiodarone, sotalol) antiarrhythmic medications<br>• Presence of moderate to severe renal impairment or end stage renal disease |

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| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subjects to utilization of the most cost-effective site of care<br>• Metabolic disease specialist |
| **Coverage Duration:** | • Approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

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 |

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| **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:
**ELOSULFASE ALFA**
Affected Medications: VIMIZIM (elosulfase alfa)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design. |
| **Required Medical Information:** | • Diagnosis of Mucopolysaccharidosis type IVA (MPS IVA; Morquio A syndrome) confirmed by an enzyme assay<br>• Medical history of musculoskeletal conditions such as knee deformity, kyphosis, hip dysplasia, prior spinal fusion surgery, and arthralgia<br>• Baseline six minute walk test (6-MWT) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Recommended dose is 2 mg per kg once every week<br>• Available in 5 mL vial containing 5 mg of Vimizim<br><br>Reauthorization requires documentation of treatment success defined as improved six minute walk test<br><br>Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced for all medical infusion drugs |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 5 years 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:** | • Initial approval: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ELTROMBOPAG**
Affected Medications: PROMACTA (eltrombopag), PROMACTA PACKET

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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>• Complete blood count with differential and platelet count<br>• Liver function test<br><br>**<u>Thrombocytopenia in patients with chronic Immune Thrombocytopenia (ITP)</u>**<br>• All therapies tried/failed<br>• Documentation of splenectomy status<br><br>**<u>Thrombocytopenia in patients with chronic hepatitis C</u>**<br>• Documentation of plan to initiate interferon-based therapy<br>• Child-Pugh score<br><br>**<u>Severe aplastic anemia</u>**<br>• All immunosuppressive therapies tried/failed<br>• Documentation of planned treatment regimen<br>• Baseline hemoglobin and absolute neutrophil count (ANC) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Thrombocytopenia in patients with chronic ITP</u>**<br>• Documentation of platelet count less than 20,000/mcl **AND**<br>• Documentation of clinically significant bleeding **AND**<br>• Documentation of splenectomy OR failure with at least 2 therapies for ITP, including corticosteroids or immunoglobulin (defined as platelets did not increase to at least 50,000/mcl)<br><br>**Reauthorization**<br>• Response to treatment with platelet count of at least 50,000/mcl (not to exceed 400,000/mcl) **OR**<br>• The platelet counts have not increased to a platelet count of at least 50,000/mcl 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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| | • Documentation of platelet count less than 75,000/mcl **AND**<br>• Documentation of compensated liver disease<br><br>**Reauthorization:**<br>• Response to treatment with platelet count of at least 90,000/mcl but less than 400,000/mcl and no significant liver function abnormalities<br><br>**<u>Severe aplastic anemia</u>**<br>• Documentation of platelet count less than or equal to 30,000/mcl **AND**<br>• Documentation of insufficient response to at least 1 prior immunosuppressive therapy<br><br>**Reauthorization** after initial approval requires hematologic response to treatment defined as meeting 1 or more of the following criteria:<br>○ Platelet count increases to 20,000/mcl above baseline, or stable platelet counts with transfusion independence for a minimum of 8 weeks;<br>○ Hemoglobin increase by greater than 1.5 g/dL, or a reduction in greater than or equal to 4 units RBC transfusions for 8 consecutive weeks;<br>○ ANC increase of 100% or an ANC increase greater than 500/mcl<br>• Discontinue therapy if hematologic response not achieved after 16 weeks of treatment, if platelet count greater than 400,000/mcl, or significant liver function abnormalities<br><br>Oral suspension formulation requires documented medical inability to use Promacta tablets |
| **Exclusion Criteria:** | **<u>All indications</u>**<br>• History of hematological malignancy or myelodysplastic syndrome<br><br>**<u>Thrombocytopenia in patients with chronic hepatitis C</u>**<br>• Hepatitis C treatment with direct-acting antiviral agents used without interferon<br>• Child-Pugh score greater than 6 |

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| | • History of ascites or hepatic encephalopathy |
| **Age Restriction:** | **<u>Thrombocytopenia in patients with ITP</u>**<br>• 1 year and older<br><br>**<u>Thrombocytopenia in patients with chronic hepatitis C and patients with severe aplastic anemia</u>**<br>• 18 years and older<br><br>**<u>Severe Aplastic Anemia</u>**<br>• 2 years and older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subjects to utilization of the most cost-effective site of care<br><br>**<u>Thrombocytopenia in patients with ITP and patients with severe aplastic anemia</u>**<br>• Prescribed by or consultation with hematologist<br><br>**<u>Thrombocytopenia in patients with chronic hepatitis C</u>**<br>• Prescribed by or consultation with hematologist, hepatologist, gastroenterologist, or ID specialist |
| **Coverage Duration:** | **<u>Thrombocytopenia in patients with ITP</u>**<br>• Initial approval: 3 months, unless otherwise specified<br>• Renewal with sufficient platelet increase: 12 months, unless otherwise specified<br>• Renewal with insufficient platelet increase: 3 months, unless otherwise specified<br><br>**<u>Thrombocytopenia in patients with chronic hepatitis C</u>**<br>• Initial approval: 2 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br><br>**<u>Severe aplastic anemia</u>**<br>• Initial approval: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br><br>**<u>Severe aplastic anemia in combination with cyclosporine and Atgam</u>**<br>• Approval: 6 months only |

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

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design<br>• Treatment of adult and pediatric (newborn and older) patients with primary hemophagocytic lymphohistiocytosis (HLH) with refractory, recurrent or progressive disease or intolerance with conventional HLH therapy. |
| **Required Medical Information:** | <u>Diagnosis of primary hemophagocytic lymphohistiocytosis (HLH)</u><br><br>• Medical records (e.g., chart notes, laboratory values) confirming the following:<br>  ○ Confirmation of a gene mutation known to cause primary HLH (e.g., PRF1, UNC13D); AND<br>  ○ Confirmation that 5 of the following clinical characteristics are present:<br>  • Fever 101.3°F or higher<br>  • Splenomegaly<br>  • **Two** of the following cytopenias in the peripheral blood:<br>  ○ Hemoglobin less than 9 g/dL; or<br>  ○ Platelet count less than 100 x $10^9$/L; or<br>  ○ Neutrophils less than 1 x 109/L<br>  • **One** of the following:<br>  ○ Hypertriglyceridemia defined as fasting triglycerides 3 mmol/L or higher or 265 mg/dL or higher; or<br>  ○ Hypofibrinogenemia defined as fibrinogen 1.5 g/L or lower<br>  • Hemophagocytosis in bone marrow or 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 mg/L or higher<br>  • Soluble CD25 (i.e., soluble IL-2 receptor) 2,400 U/ml or higher |

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| | AND<br>• Patient has refractory, recurrent or progressive disease or intolerance with conventional HLH therapy (i.e., etoposide + dexamethasone); **and**<br>• Emapalumab will be administered with dexamethasone; and<br>• Patient is a candidate for stem cell transplant; **and**<br>• Emapalumab is being used as part of the induction or maintenance phase of stem cell transplant, which is to be discontinued at the initiation of conditioning for stem cell transplant; **and**<br>• Dosing is in accordance with the United States Food and Drug Administration approved labeling; **and**<br>• Approval is for no more than 6 months |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | • Emapalumab for the treatment of secondary HLH |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Must be prescribed by or in consultation with a prescriber experienced in the treatment 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 (not to exceed 6 months total of treatment) |

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 |

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| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval duration: 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 |
| **Required Medical Information:** | • Diagnosis of major depressive disorder |
| **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 or tetracyclic antidepressant<br>  ○ Bupropion<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:** | • 12 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:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**ENDOTHELIN RECEPTOR ANTAGONISTS**
Affected Medications: BOSENTAN (bosentan), AMBRISENTAN (ambrisentan), OPSUMIT (macitentan)

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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) |
| **Required Medical Information:** | **PAH World Health Organization (WHO) Group 1**<br>• Documentation of PAH confirmed by right-heart catheterization<br>• Etiology of PAH (idiopathic, heritable, or associated with connective tissue disease)<br>• New York Heart Association (NYHA)/WHO Functional Class II, III, or IV symptoms.<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blocker), unless there are contraindications such as low systemic blood pressure (systolic blood pressure less than 90), low cardiac index, or 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, unless the patient has cardiopulmonary comorbidities (defined as risk factors for heart failure with preserved ejection fraction [HFpEF], such as obesity, diabetes, coronary heart disease, hypertension, and/or a low diffusing capacity for carbon monoxide [DLCO])<br><br>**For all Opsumit (macitentan) requests:**<br>• Documented failure with an adequate trial (at least 12 weeks) of BOTH ambrisentan and bosentan<br><br>**Reauthorization** requires documentation of treatment success defined as improved walking distance or improvements in functional class. |
| **Exclusion Criteria:** | • Pregnancy<br>• Idiopathic Pulmonary Fibrosis (IPF), including IPF patients with PAH (WHO Group 3) |
| **Age Restriction:** | |

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| **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:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**ENFUVIRTIDE**
Affected Medications: FUZEON (enfuvirtide)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications |
| **Required Medical Information:** | • Documentation confirming HIV-1 infection |
| **Appropriate Treatment Regimen & Other Criteria:** | • The patient has used Fuzeon for greater than or equal to 6 months AND<br>• Current viral load and CD4+ count is documented AND<br>• The patient had a positive or stable virologic response to Fuzeon<br>**OR**<br>• The patient has NOT used Fuzeon for greater than or equal to 6 months AND<br>• Baseline viral load and CD4+ count is documented AND<br>• Evidence of HIV-1 replication despite ongoing antiretroviral therapy AND<br>• Fuzeon is prescribed in combination with an optimized antiretroviral regimen |
| **Exclusion Criteria:** | • Initial therapy in patients who are antiretroviral naive |
| **Age Restriction:** | • Age 6 years of age and older (weighing at least 11 kg) |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 24 months (if the patient has already used Fuzeon for greater than or equal to 6 months), unless otherwise specified<br>• Approval: 6 months (if the patient has NOT already used Fuzeon for greater than or equal to 6 months), unless otherwise specified |

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

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications<br>  ○ Neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive |
| **Required Medical Information:** | **<u>Neuromyelitis Optica Spectrum Disorder (NMOSD)</u>**<br>• Diagnosis of NMOSD with AQP4-IgG requiring all of the following:<br>  ○ At least one core clinical characteristic:<br>  • Optic neuritis<br>  • Acute myelitis<br>  • Area postrema syndrome: episode of otherwise unexplained hiccups or nausea and vomiting<br>  • Acute brainstem syndrome<br>  • Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMSOD-typical diencephalic MRI lesions<br>  • Symptomatic cerebral syndrome with NMOSD-typical brain lesions<br>  ○ Positive test for AQP4-IgG using best available detection method<br>  ○ Exclusion for alternative diagnoses<br>• History of at least 1 attack in the past year, or at least 2 attacks in the past 2 years, requiring rescue therapy<br>• Expanded Disability Status Scale (EDSS) score of 6.5 or less<br>• Documented treatment failure with 12 weeks of at least 2 of the following immunosuppressive therapies: azathioprine, mycophenolate, methotrexate<br>• Documented treatment failure with 12 weeks of at least 1 of the following: mitoxantrone (authorization required), rituximab (authorization required)<br><br>Reauthorization requires documentation of treatment success. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing: 120 mg SQ at weeks 0, 2, and 4, followed by a maintenance dosage of 120 mg every 4 weeks |

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| **Exclusion Criteria:** | • Active Hepatitis B Virus (HBV) infection<br>• Active or untreated latent tuberculosis<br>• Concurrent use with other monoclonal antibodies (rituximab, eculizumab, tocilizumab, inebilizumab etc.) or IVIG |
| **Age Restriction:** | • 18 years or older |
| **Prescriber/Site of Care Restrictions:** | • 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:
**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. |
| **Required Medical Information:** | **<u>Pulmonary arterial hypertension (PAH) WHO Group 1</u>**<br>• Documentation of PAH confirmed by right-heart catheterization<br>• Documentation of acute vasoreactivity testing (positive result requires trial/failure to calcium channel blocker) unless there are contraindications such as low systemic blood pressure (systolic blood pressure less than 90), low cardiac index, or presence of severe symptoms (functional class IV)<br>• Patient weight, planned dose and frequency |
| **Appropriate Treatment Regimen & Other Criteria:** | • PAH: for initiation of therapy patient must have mean pulmonary artery pressure of at least 20 mm Hg, pulmonary capillary wedge pressure less than or equal to 15 mm Hg, and pulmonary vascular resistance of at least 3 Wood units **AND**<br>• Failure of the following therapy classes: PDE5 inhibitors **AND**<br>• Endothelin receptor antagonists (exception for severe disease, WHO class IV)<br>• Subsequent approval requires documentation of treatment success: exercise endurance, echocardiographic testing, hemodynamic testing, BNP, functional class |
| **Exclusion Criteria:** | • **<u>Flolan</u>**: Heart failure caused by reduced left ventricular ejection fraction<br>• **<u>Veletri</u>**: Long-term use in patients with heart failure due to severe left ventricular systolic dysfunction; long-term use patients who develop pulmonary edema during dose initiation |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with a cardiologist or pulmonologist |

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| | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 3 months unless otherwise specified<br>• Reauthorization: 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, MUSE PELLET, STAXYN, STENDRA, TADALAFIL (10 mg, 20 mg)

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| **Covered Uses:** | • Treatment for a mental health diagnosis of erectile dysfunction (ED), also known as erectile disorder. |
| **Required Medical Information:** | • Mental health diagnosis according to Diagnostic and Statistical Manual of Mental Disorders, fifth edition (DSM-5) diagnostic criteria for erectile disorder:<br>A. At least one of the three following symptoms must be experienced on almost all or all (approximately 75%-100%) occasions of sexual activity (in identified situational contexts or, if generalized, in all contexts):<br>1. Marked difficulty in obtaining an erection during sexual activity.<br>2. Marked difficulty in maintaining an erection until the completion of sexual activity.<br>3. Marked decrease in erectile rigidity<br>B. The symptoms in Criterion A have persisted for a minimum duration of approximately 6 months.<br>C. The symptoms in Criterion A cause clinically significant distress in the individual.<br>D. The sexual dysfunction is not better explained by nonsexual mental disorder or as a consequence of severe relationship distress or other significant stressors and is not attributable to the effects of a substance/medication or another medical condition. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Must have failure to formulary alternative tadalafil 2.5 mg or 5 mg tablets |
| **Exclusion Criteria:** | • Diagnosis of erectile dysfunction (ED) without meeting requirements of DSM-5 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 |

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| **Age Restriction:** | |
| **Coverage Duration:** | • Approval: 12 months |

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 migraines described as being moderate-severe<br>• Documentation of inadequate response or contraindication to all of the following:<br>  ○ Minimum of two prescription strength NSAIDs or combination analgesics (e.g. ibuprofen, naproxen, or acetaminophen plus aspirin plus caffeine)<br>  ○ Minimum of 1 oral 5-hydroxytryptamine-1 (5HT1) receptor agonists (e.g. sumatriptan, naratriptan, rizatriptan, or zolmitriptan)<br>  ○ Minimum of 1 NON-oral 5HT1 agonist (e.g. sumatriptan, zolmitriptan) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Injection doses should not exceed 3 mg in a 24 hour period, and 6 mg in one week<br>  • QL 12mL/30 days<br>• Nasal solutions should not exceed 2 mg per day, no additional benefit shown<br>  • QL 8 mL/30 days<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 |

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| **Age Restriction:** | • Patients 18 years and older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 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 FDA (Food and Drug Administration)-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 |

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| | ○ 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, Procrit, Mircera) is provided when any of the following criteria is met:<br>○ For Epogen or Procrit, a documented intolerable adverse event to the preferred product Retacrit, and the adverse event was not an expected adverse event attributed to the active ingredient<br>○ For Mircera, a documented inadequate response or intolerable adverse event to the preferred product, Retacrit<br>○ Currently receiving treatment with Mircera, excluding via samples or manufacturer's patient assistance programs |
| **Exclusion Criteria:** | • Use in combination with another erythropoiesis stimulating agent (ESA) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Must be prescribed by, or in consultation with, a specialist (hematologist, oncologist, nephrologist) |
| **Coverage Duration:** | • Approval: 6 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 |
| **Required Medical Information:** | • Diagnosis of secondary hyperparathyroidism on hemodialysis<br>• Documentation of baseline laboratory values: Calcium (corrected or free), Phosphate, Vitamin D<br>• Parathyroid hormone (PTH) levels persistently greater than 9 times the Upper Limit of Normal (ULN) for the assay used<br>• Documentation of failure or rationale for avoidance for all standard treatments for hyperparathyroidism: Calcitriol oral (capsule or solution) and injection, Paricalcitol oral and injection, Doxercalciferol oral and injection, Cinacalcet |
| **Appropriate Treatment Regimen & Other Criteria:** | • Patient does not have any Food and Drug Administration (FDA) labeled contraindications to therapy<br>• Reauthorization will require documentation of reduction of PTH to within the target range of 2-9 times the ULN |
| **Exclusion Criteria:** | • Known hypersensitivity to etelcalcetide or any of its excipients.<br>• Diagnosis of parathyroid carcinoma, primary hyperparathyroidism or with chronic kidney disease who are not on hemodialysis |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • 12 months, unless otherwise specified |

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>  ○ As an adjunct to other low-density lipoprotein-cholesterol (LDL-C) lowering therapies (LLTs) for the treatment of patients with homozygous familial hypercholesterolemia (HoFH) |
| **Required Medical Information:** | • Diagnosis of HoFH confirmed by at least 1 of the following:<br>  ○ Genetic testing showing multiple mutant alleles across the following gene loci: low-density lipoprotein receptor (LDLR), apolipoprotein B (apo B), proprotein convertase subtilisin kexin type 9 (PCSK9) or low-density lipoprotein receptor adaptor protein 1 (LDLRAP1).<br>  ○ Untreated LDL-C greater than 500 mg/dL or treated LDL-C greater than 300 mg/dL AND one of the following: (1) history of cutaneous or tendinous xanthoma prior to age 10 years or (2) documentation of untreated LDL-C greater than 190 mg/dL consistent with heterozygous familial hypercholesterolemia in both parents<br>• Documentation of baseline untreated LDL-C |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure defined as an LDL-C greater than 100mg/dL despite at least six months of adherent therapy with all of the following, unless contraindicated or not tolerated:<br>  ○ High intensity statin therapy (atorvastatin, rosuvastatin)<br>  ○ Ezetimibe<br>  ○ PCSK9 inhibitor (Praluent, Repatha), unless double-null or LDLR activity 15% or less<br><br>**<u>Reauthorization</u>**: 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<br><br>**<u>Dosing</u>**:<br>• Evkeeza: 15mg/kg IV once every 4 weeks<br>• Juxtapid |

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| | ○ Initial dose: 5mg daily<br>○ Max dose: 60mg daily |
| **Exclusion Criteria:** | • Combination therapy with Juxtapid and Evkeeza is considered experimental and is not a covered benefit |
| **Age Restriction:** | • Evkeeza: 12 years of age and older<br>• Juxtapid: 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Endocrinologist, cardiologist, or lipid 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:
**EVOLOCUMAB**
Affected Medications: REPATHA (evolocumab)

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| 1. Is the request for continuation of therapy currently approved by PacificSource? | Yes – Go to renewal criteria | No – Go to #2 |
| 2. Is the diagnosis being treated according to one of the covered indications below? | Yes – Go to appropriate section below | No – Criteria not met |
| **Primary or Familial Hyperlipidemia** | | |
| 1. Is there an untreated (no lipid-lowering therapy) LDL-cholesterol level of at least 190 mg/dL? | Yes – Document and go to #4<br>_____ | No – Go to #2 |
| 2. Is there a current LDL-cholesterol level of at least 100 mg/dL after at least three months of adherent use with maximally-tolerated statin therapy? | Yes – Document and go to #4 | No – Go to #3 |
| 3. Is there a current LDL-cholesterol level of at least 100 mg/dL and statin intolerance defined as:<br>o Intolerable statin-associated muscle symptoms lasting at least two weeks confirmed with at least two attempts of statin re-challenge (including two different statins, one of which being either atorvastatin or rosuvastatin) or<br>o Rhabdomyolysis with statin-associated elevation in creatine kinase (CK) level to at least 10 times upper limit of normal | Yes – Document LDL and go to #4 | No – Criteria not met |
| 4. 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 |
| **Clinical Atherosclerotic Cardiovascular Disease (ASCVD)** | | |
| 1. Is there a history of Clinical Atherosclerotic Cardiovascular Disease (ASCVD) or a cardiovascular event?<br>o Acute coronary syndromes, myocardial infarction (MI), stable or unstable angina, coronary or other arterial revascularization procedure (e.g., CABG, PTCA), stroke of presumed atherosclerotic origin, transient ischemic attack | Yes – Go to #2 | No – Criteria not met |

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| (TIA), peripheral arterial disease of presumed atherosclerotic origin, findings from CT angiogram or catheterization consistent with clinical ASCVD | | |
| 2. Is there a current LDL-Cholesterol of at least 70 mg/dL after at least three months of adherent use with maximally-tolerated (moderate or high-intensity) statin therapy? | Yes – Document and go to #4 | No – Go to #3 |
| 3. Is there a current LDL-Cholesterol of at least 70 mg/dL and a history of statin intolerance defined as:<br>○ Intolerable statin-associated muscle symptoms lasting at least two weeks confirmed with at least two attempts of statin re-challenge (including two different statins, one of which being either atorvastatin or rosuvastatin) or<br>○ Rhabdomyolysis with statin-associated elevation in creatine kinase (CK) level to at least 10 times upper limit of normal | Yes – Document LDL and go to #4 | No – Criteria not met |
| 4. 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** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider? | 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** | | |
| • **Repatha:** 140 mg every 2 weeks OR 420 mg once monthly<br>○ Repatha Solution Prefilled Syringe or Auto-Injector 140 mg/mL – 2 injections (2 mL) per 28 days<br>○ Repatha Pushtronex System Solution Cartridge 420 mg/3.5 mL – 1 injection (3.5 mL) per 28 days<br>• Moderate-intensity statins: Atorvastatin, fluvastatin 80 mg daily, lovastatin 40 mg, pitavastatin 2 mg or greater, pravastatin 40 mg or greater, rosuvastatin, simvastatin 20 mg or greater | | |

POLICY NAME:
**Food and Drug Administration (FDA) APPROVED DRUG – Drug or Indication Not Yet Reviewed By Plan for Formulary Placement**
Affected Medications: New Medications or Indications of Existing Drugs Not Yet Reviewed By Plan for Formulary Placement

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| **Covered Uses:** | • Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit 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:** | • Prescriber restrictions based on package insert requirements |
| **Coverage Duration:** | • Case by case based on member need |

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. |
| **Required Medical Information:** | • Documented diagnosis of Dravet syndrome (DS) or Lennox-Gastaut Syndrome (LGS)<br>• Patient Weight<br>• Documentation that therapy is being used as adjunct therapy for seizures<br><br>**<u>Dravet Syndrome</u>**<br>• Documentation of at least 6 convulsive seizures in the last 6 weeks while on stable antiepileptic drug therapy<br>• Documentation of baseline cardiac function testing<br><br>**<u>Lennox-Gastaut Syndrome (LGS)</u>**<br>• Documentation of at least 8 drop seizures per month while on stable antiepileptic drug therapy. |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Dravet Syndrome</u>**<br>• Documented treatment and inadequate control of seizures with Epidiolex AND at least four of the following therapies:<br>  ○ Valproate, clobazam, clonazepam, levetiracetam, or topiramate<br><br>**<u>Lennox-Gastaut Syndrome (LGS)</u>**<br>• Documented treatment and inadequate control of seizures with Epidolex AND at least three guideline directed therapies including:<br>  ○ Valproate, lamotrigine, rufinamide, topiramate, felbamate, or clobazam<br><br>• Dosing: not to exceed 26 mg daily<br>• Reauthorization: documentation of treatment success as determined by treating provider |
| **Exclusion Criteria:** | • Concomitant use of, or within 14 days of the administration of monoamine oxidase inhibitors. |

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| **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:
**FENTANYL (Oral-Intranasal)**
Affected Medications: ABSTRAL, FENTORA, FENTANYL CITRATE, LAZANDA, ONSOLIS, SUBSYS

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications |
| **Required Medical Information:** | • Long-Acting opioid is being prescribed for around-the clock treatment of the cancer pain.<br>• The patient is opioid tolerant (Patients are considered opioid tolerant if they have been taking at least 60 mg of oral morphine per day, 25 mcg of transdermal fentanyl/hr, 30 mg of oral oxycodone daily, 8 mg of oral hydromorphone daily, 25 mg oral oxymorphone daily or an equianalgesic dose of another opioid for a week or longer). |
| **Appropriate Treatment Regimen & Other Criteria:** | • For breakthrough pain in patients with cancer and for breakthrough chronic (non-cancer) pain<br>• Patient is unable to swallow, has dysphagia, esophagitis, mucositis, or uncontrollable nausea/vomiting **OR**<br>• Patient is unable to take **2** other short-acting narcotics (eg, oxycodone, morphine sulfate, hydromorphone, etc) secondary to allergy or severe adverse events **AND**<br>• Patient is on or will be on a long-acting narcotic (eg, Duragesic), or the patient is on intravenous, subcutaneous, or spinal (intrathecal, epidural) narcotics (eg, morphine sulfate, hydromorphone, fentanyl citrate). |
| **Exclusion Criteria:** | • Patients taking strong or moderate cytochrome P450 3A4 inhibitor(s), who will not be carefully monitored and will not have dosing adjustments made if necessary.<br>• Use in the management of acute and/or postoperative pain including surgery/post-surgery, trauma/post-trauma, acute medical illness (acute abdominal pain, pelvic pain, muscle spasm). |

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| | • Use as pre-anesthesia (preoperative anxiolysis and sedation and/or supplement to anesthesia). |
| **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:** | • Approval: 12 months, unless otherwise specified. |

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

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | • Susceptibility cultures matching flucytosine activity |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing: maximum 150 mg/kg/day |
| **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:** | • Approval: 6 weeks, or lesser requested duration, 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 from by plan design. |
| **Required Medical Information:** | **Iluvien**<br>• Diagnosis of clinically significant diabetic macular edema **AND**<br>• Documentation of past treatment with corticosteroids without a clinically significant rise in intraocular pressure **AND**<br>• Documentation of insufficient response to initial therapy with intravitreal bevacizumab (or another anti-VEGF therapy) **AND**<br>• Documentation of insufficient response to laser photocoagulation<br><br>**Retisert and Yutiq**<br>• Diagnosis of recurrent non-infectious uveitis with documentation of slit-lamp examination and dilated fundus examination<br>• Authorization for Retisert requires documented clinical failure with Yutiq |
| **Appropriate Treatment Regimen & Other Criteria:** | **Iluvien**<br>• One intravitreal implant per 36 months as monotherapy<br>• If the physician determines that adjunctive therapy with anti-VEGF is necessary (e.g. worsening visual acuity, retinal volume, or fluorescein leakage with Iluvien monotherapy), the request will be reviewed and determination will be made based on medical necessity. Adjunctive therapy with Avastin (bevacizumab) will be the preferred option.<br><br>**Retisert and Yutiq**<br>• One intravitreal implant per 30 months (Retisert) or 36 months (Yutiq)<br>• Documented failure with<br>  ○ A 12-week trial with a systemic corticosteroid (such as prednisone) AND<br>  ○ At least one immunosuppressive agent: methotrexate, azathioprine, mycophenolate, AND<br>  ○ At least one calcinuerin inhibitor (cyclosporine, tacrolimus) AND |

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| | ○ At least two of the following ocular steroids: Ozurdex, Triesence, Trivaris<br>AND<br>• Authorization for Retisert requires documented clinical failure with Yutiq |
| **Exclusion Criteria:** | • Active or suspected ocular or periocular infections<br>• Glaucoma or documentation of past treatment with corticosteroids with a clinically significant rise in intraocular pressure<br>• Concurrent use of intravitreal implants and injections: Ozurdex (dexamethasone), Triesence (triamcinolone), Trivaris (triamcinolone) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • **Iluvien**: 36 months, unless otherwise specified<br>• **Retisert**: 30 months, unless otherwise specified<br>• **Yutiq**: 36 months, unless otherwise specified |

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 |
| **Required Medical Information:** | • Complete blood count with differential and platelet count<br>• Liver function test<br><br>**<u>Thrombocytopenia in patients with Chronic Immune thrombocytopenia (ITP)</u>**<br>• All therapies tried/failed<br>• Documentation of splenectomy status |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Thrombocytopenia in patients with Chronic ITP</u>**<br>• Documentation of platelet count less than 20,000/mcl and clinical bleeding<br>• Must fail at least 2 therapies for ITP – a thrombopoietin receptor agonist and another including corticosteroids, immunoglobulins, immunosuppression, or splenectomy<br>• **Continuation of therapy** requires response to treatment with platelet count of at least 50,000/mcl without significant liver function abnormalities<br>• Discontinue therapy after 12 weeks if platelet count does not increase to a level sufficient to avoid clinically important bleeding |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber Restrictions:** | • Prescribed by or consultation with hematologist |
| **Coverage Duration:** | • Initial approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 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:
**GALAFOLD**
Affected Medications: GALAFOLD (migalastat)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications |
| **Required Medical Information:** | • Diagnosis of Fabry disease confirmed by:<br>  ○ Enzyme assay demonstrating a deficiency of alpha-galactosidase enzyme activity<br>  **AND**<br>  ○ Genetic testing confirming the presence of at least one amenable galactosidase alpha (GLA) variant.<br>• The patient has clinical signs and symptoms of Fabry disease. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Concurrent use with Fabrazyme |
| **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 prescriber experienced in the treatment of Fabry disease |
| **Coverage Duration:** | • Initial approval: 4 months, unless otherwise specified<br>• Subsequent approval: 12 months, unless otherwise specified |

POLICY NAME:
**GALSULFASE**
Affected Medications: NAGLAZYME (galsulfase)

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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 the Food and Drug Administration (FDA)-approved indication of mucopolysaccharidosis VI (MPS VI or Maroteaux-Lamy syndrome)? | Yes – Go to section below | No – Criteria not met |
| **Indication: Mucopolysaccharidosis VI (MPS VI or Maroteaux-Lamy syndrome)** | | |
| 1. Is there documentation of a diagnosis of mucopolysaccharidosis VI (MPS VI or Maroteaux-Lamy syndrome) | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documentation of a confirmed diagnosis by an enzyme assay demonstrating a deficiency in Nacetylgalactosamine 4-sulfatase (arylsulfatase B) enzyme activity or by DNA testing? | Yes – Document and go to 3 | No – Criteria not met |
| 3. Is there documentation of a current body weight for dosing calculations? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is the requested dose within the Food and Drug Administration (FDA)-approved label and PacificSource quantity limitations? | Yes – Approve up to 3 months, unless otherwise specified | No – Criteria not met |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider? | Yes – Go to #2 | No – Criteria not met |

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| 2. Is the requested dose within the Food and Drug Administration (FDA)-approved label and PacificSource quantity limitations? | Yes – Approve up to 12 months, unless otherwise specified | No – Criteria not met |
| **Quantity Limitations** | | |
| • **Naglazyme**<br>o Availability: 5 mg/5 mL single-use vial<br>o Dose: 1 mg/kg of body weight* administered once weekly as an intravenous infusion.**<br><br>*Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced.<br>**All approvals are subject to utilization of the most cost-effective site of care | | |

POLICY NAME:
**GANAXOLONE**
Affected Medications: ZTALMY

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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 generic testing<br>• Documentation of inadequately controlled seizures despite current treatment<br>• Documented treatment failure with at least two therapies for seizure management |
| **Appropriate Treatment Regimen & Other Criteria:** | **Dosing**:<br>• Dosage for patients weighing 28 kg or less:<br>  ○ Starting dose of 6 mg/kg three times daily (18 mg/kg/day)<br>  ○ Maximum dose of 21 mg/kg three times daily (63 mg/kg/day).<br>• Dosage for patients weighing over 28 kg:<br>  ○ Starting dose of 150 mg three times daily (450 mg daily)<br>  ○ Maximum dose of 600 mg three times daily (1800 mg daily).<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 or 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 |

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

POLICY NAME:
**GILENYA**
Affected Medications: GILENYA (fingolimod), Fingolimod

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| **Covered Uses:** | All Food and Drug Administration (FDA)-approved indications not otherwise excluded from plan benefits. |
| **Required Medical Information:** | • Diagnosis of relapsing forms of multiple sclerosis confirmed with magnetic resonance imaging (MRI)<br>• Recent documentations of complete blood count, liver function tests, and an electrocardiogram |
| **Appropriate Treatment Regimen & Other Criteria:** | • No concurrent use of any medications indicated for the treatment of relapsing-remitting multiple sclerosis<br>• Not approved for primary progressive multiple sclerosis<br>• Maximum dose: 0.5 mg once daily<br>• Documentation of varicella serology and varicella zoster virus vaccination if antibody negative for those without a history of chicken pox or prior vaccination<br>• Reauthorization: provider attestation of treatment success |
| **Exclusion Criteria:** | • Varicella or Zostavax/Shingrex vaccination within the last month<br>• Myocardial infarction, unstable angina, stroke, transient ischemic attack, decompensated heart failure requiring hospitalization or Class III/IV heart failure in the last 6 months<br>• History or presence of Mobitz Type II second-degree or third-degree AV block or sick sinus syndrome, unless patient has a functioning pacemaker<br>• Baseline QTc interval is equal to or greater than 500 msec<br>• Current use of Class Ia or Class III anti-arrhythmic drugs |
| **Age Restriction:** | • At least or greater than 10 years old (per Food and Drug Administration (FDA) labeling) |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by a Neurologist or an MS specialist.<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:
**GIVOSIRAN**
Affected Medications: GIVLAARI (givosiran)

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| **Covered Uses:** | All Food and Drug Administration (FDA)-approved indications not otherwise excluded from plan benefits. |
| **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 including medications, smoking, drinking, medications, and infections<br>• Documentation of baseline liver function tests |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of active acute disease defined as at least 2 documented porphyria attacks within the last six months requiring Hemin administration that are not attributable to a specific exacerbating factor<br>• Documented 12-week trial and failure of prophylactic hemin administration<br>• For women:<br>  ○ Documented 12-week trial and failure of gonadotropin releasing hormone analogue (ex. Leuprolide) OR<br>  ○ Documentation that attacks are not related to the luteal phase of the menstrual cycle<br><br>**Reauthorization** will require documentation of greater than 50% reduction in baseline acute attack frequency |
| **Exclusion Criteria:** | • Active HIV, Hepatitis C, or Hepatitis B infection(s)<br>• History of Pancreatitis<br>• Concomitant use with prophylactic hemin |
| **Age Restriction:** | • Greater than or equal to 12 years of age |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with physicians that specialize 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, VICTOZA, OZEMPIC, RYBELSUS

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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 |
| **Required Medical Information:** | • Lab test confirmation of diagnosis (one of the following at baseline):<br>  ○ A1c equal to or greater than 6.5%<br>  ○ Fasting plasma glucose equal to or greater than 126 mg/dl<br>  ○ Oral glucose tolerance test 2 hour blood sugar level equal to or greater than 200 mg/dl<br>AND<br>• Documented treatment failure with metformin or metformin extended release |
| **Appropriate Treatment Regimen & Other Criteria:** | Reauthorization: documentation of disease responsiveness to therapy |
| **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). |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 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. |
| **Required Medical Information:** | |
| **Appropriate Treatment Regimen & Other Criteria:** | • Hypogonadotropic hypogonadism secondary to a pituitary deficiency in males<br>• Perioperative use in male infants/toddlers with hypospadias and chordee OR total epispadias and bladder exstrophy<br>• Prepubertal cryptorchidism not caused by anatomic obstruction<br><br>• **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:** | • Approval: 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>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better |
| **Required Medical Information:** | **<u>Prostate/Breast Cancer</u>**<br>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | • For endometriosis: documentation of a trial and inadequate relief after at least three months of first-line therapy with nonsteroidal anti-inflammatory drugs (NSAIDs) and continuous (no placebo pills) hormonal contraceptives<br>• Reauthorization for oncologic uses requires documentation of disease responsiveness to therapy<br>**Dosing**<br>• Breast Cancer: 3.6 mg every 28 days<br>• Endometrial thinning: 3.6 mg for 1 or 2 doses with each depot is given 28 days apart. (When 1 depot is given, endometrial ablation surgery should be performed at 4 weeks. If 2 depots are given, surgery should be performed within 2-4 weeks following the second depot dosage)<br>• Endometriosis: 3.6 mg every 28 days for 6 months<br>• Prostate Cancer: 3.6 mg depot 8 weeks before radiotherapy, followed in 28 days by the Zoladex 10.8 mg depot, can be administered. Alternatively, four injections of 3.6 mg depot can be administered at 28-day intervals, two depots preceding and two during radiotherapy. |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• For gynecologic uses, prior use of Zoladex for a 6-month period |
| **Age Restriction:** | • 18 years and up for endometriosis and endometrial thinning |
| **Prescriber/Site of Care Restrictions:** | • Oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | Oncologic uses<br>• Initial approval: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified<br><br>Endometriosis<br>• Approval: 6 months with no reauthorization, unless otherwise specified |

POLICY NAME:
**GROWTH HORMONE (Somatropin) Injectables**
Affected Medications: GENOTROPIN MINIQUICK, HUMATROPE, HUMATROPE COMBOPACK, NORDITROPIN FLEXPRO, NORDITROPIN, NORDIFLEX, NUTROPIN AQ, NUTROPOIN AQ NUSPIN 10, NUTROPIN AQ NUSPIN 5, NUTROPIN AQ PEN, NUTROPIN, OMNITROPE, PROTROPIN, SAIZEN, SAIZEN CLICK EASY, SKYTROFA, ZOMACTON, SKYTROFA

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | **All indications:**<br>• Documentation of baseline height, height velocity, bone age, and patient weight<br>• Patient must try Norditropin prior to use of any other growth hormone agent<br>• Documentation of clinical failure with an adequate trial (at least 12 weeks each) of Norditropin AND one additional growth hormone agent prior to Skytrofa approval<br><br>**Growth hormone deficiency or Pituitary dwarfism**<br>• For initial approval, documentation of the following is required:<br>  ○ Diagnosis of growth hormone deficiency or pituitary dwarfism AND<br>  ○ Low serum values for GH stimulation test, IGF-I, and IGFBP-3 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>**Turner's syndrome**<br>• For initial approval, documentation of the following is required:<br>  ○ 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 WHO growth curves at equivalent age, AND<br>  • No secondary factor present that would explain observed growth delays |

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| | ▪ 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>○ 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>○ 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<br><br>**<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 age 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 |

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| | • Height standard deviation score (HSDS) at start of growth hormone treatment of -2.5<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>• Dose for children less than 4 years with baseline HSDS between -2 to -3 must not exceed starting dose 0.033mg/kg/day<br>• Max dose of 0.067mg/kg/day for all other ages<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-I outside reference range for patients 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:** Documented IGF-I within normal reference range for age and sex, clinical improvement |
| **Appropriate Treatment Regimen & Other Criteria:** | • Height velocity impairment |
| **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 |

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| | deviation score (SDS) <-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:** | • Pediatric endocrinologist<br>• Endocrinologist for adult indication<br>• All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 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 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 with genotyping<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:** | • Prescribed by, or in consultation with, a hepatologist, gastroenterologist, liver transplant physician, or infectious disease specialist |

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| | • 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 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 | SOF/VEL x 12 weeks<br>Mavyret x 8 weeks |
| | 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 + | Non-cirrhotic or compensated cirrhosis | Mavyret + SOF + RBV x 16-24 weeks<br>Vosevi + RBV x 24 weeks |

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| sofosbuvir | | |

Abbreviations: DAA = direct-acting antiviral; PEG = pegylated interferon; RBV = ribavirin; SOF/VEL = sofosbuvir/velpatasvir

*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 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 |

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| 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), VANTAS (histrelin acetate implant)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>• Gender dysphoria |
| **Required Medical Information:** | **<u>Central Precocious Puberty</u>**<br>• Documentation of central precocious puberty (CPP) confirmed by basal luteinizing hormone (LH), follicle-stimulating hormone (FSH), and either estradiol or testosterone concentrations<br><br>**<u>Gender Dysphoria</u>**<br>• Documentation of current Tanner stage 2 or greater OR Documentation of baseline and current estradiol and testosterone levels to confirm onset of puberty<br>• Documentation from a licensed mental health professional (LMHP) confirming diagnosis and addressing the patient's general identifying characteristics:<br>  ○ The initial and evolving gender and any associated mental health concerns, and other psychiatric diagnoses<br>  ○ The duration of the referring licensed mental health professional's relationship with the client, including the type of evaluation and psychotherapy to date<br>  ○ The clinical rationale for supporting the client's request for hormone therapy and statement that the client meets eligibility criteria; and<br>  ○ Permission to contact the licensed mental health professional for coordination of care<br>• Comprehensive mental health evaluation should be provided in accordance with most current version of the World Professional Association for Transgender Health (WPATH) Standards of Care |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>All Indications</u>**<br>• Approval requires documented treatment failure with leuprolide<br><br>• QL: 50 mg implant every 12 months |

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| | • Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • Equal or greater than 2 years old |
| **Prescriber/Site of Care Restrictions:** | • Central Precocious Puberty: Prescribed by or in consultation with endocrinologist<br>• Gender dysphoria: Diagnosis made and prescribed by, or in consultation with, a specialist in the treatment of gender dysphoria<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:
**HEREDITARY ANGIOEDEMA (HAE)**
Affected Medications: Berinert, Icatibant Acetate, Sajazir, Firazyr, Ruconest, Kalbitor, Cinryze, Haegarda, Takhzyro, Orladeyo

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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 for acute treatment to be used in combination with another HAE drug used for acute treatment such as Berinert, Ruconest or Icatibant Acetate? | Yes- Criteria not met | No – go to #3 |
| 3. Is the request for prophylactic treatment to be used in combination with another HAE drug used for prophylactic treatment such as Haegarda, Takhzyro, Cinryze? | Yes- Criteria not met | No – go to #4 |
| 4. Is the request for Orladeyo in the setting of End-Stage Renal Disease or those requiring hemodialysis? | Yes- Criteria not met | No – go to #5 |
| 5. Is the official diagnosis of hereditary angioedema (HAE) documented in the member's chart and documentation of requested number of units or doses and current weight? | Yes – Go to #6 | No – Criteria not met |
| 6. Have all other causes of acquired angioedema (e.g., medications, auto-immune diseases) been excluded? | Yes – Go to #7 | No – Criteria not met |
| 7. Is there a laboratory confirmed diagnosis for HAE type I or II?<br>a. Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing test) AND ONE of the following:<br>i. C1-inhibitor functional level less than 50% of the lower limit of | Yes – Go to #9 | No – Go to #8 |

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| normal as defined by the laboratory performing test<br>ii. C1-inhibitor antigenic level less than 50% of the lower limit of normal as defined by the laboratory performing test | | |
| 8. Is there a family history of angioedema and the angioedema was refractory to a trial of antihistamine (e.g., cetirizine) for at least one month or confirmed factor 12 (FXII) mutation? | Yes – Go to #9 | No – Criteria not met |
| 9. Is the request for one of the following:<br>a. Acute treatment to treat 3 or less attacks per month?<br>b. Acute treatment to treat more than 3 attacks per month?<br>c. Prophylactic treatment? | Yes – Go to appropriate section | No – Criteria not met |
| **Acute treatment of HAE with 3 or less attacks per month**<br>**Drugs: Berinert, Icatibant Acetate, Sajazir, Firazyr, Ruconest, Kalbitor** | | |
| 1. Is there documentation of requested number of units or doses and current weight? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documentation of the number of acute attacks requiring treatment in the past year? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is the request for Berinert? | Yes – Go to #4 | No – Go to #5 |
| 4. Is there a documented treatment failure (or documented intolerable adverse event) to Ruconest or one of the following:<br>a. Member is less than 13 years of age?<br>b. Request is to treat laryngeal attacks?<br>c. Currently receiving treatment with | Yes – Go to #5 | No – Criteria not met; Berinert requires failure with Ruconest |

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| Berinert, excluding via samples or manufacturer's patient assistance programs? | | |
| 5. Is the drug prescribed by, or in consultation with, an allergist/immunologist or physician that specializes in HAE or related disorders? | Yes – Go to #6 | No – Criteria not met |
| 6. Is the requested dose within the Food and Drug Administration (FDA)-approved label and age restrictions?<br><br>Authorization for therapy for acute treatment will provide a sufficient quantity to cover the number of attacks experienced in the last year plus 1 additional dose. Limited to having medication on hand to treat average number of acute attacks per month plus 1 additional dose. | Yes – Approve up to 3 months | No – Criteria not met |
| **Acute Treatment of HAE with more than 3 attacks per month**<br>**Drugs: Berinert, Icatibant Acetate, Sajazir, Firazyr, Ruconest, Kalbitor** | | |
| 1. Is there documentation of requested number of units or doses and current weight? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documentation of current treatment, or failure, intolerance, or clinical rationale for avoidance of prophylactic therapies such as Haegarda, Takhzyro, Cinryze? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documentation of the number of acute attacks requiring treatment in the past year? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is the request for Berinert? | Yes – Go to #5 | No – Go to #6 |

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| 5. Is there a documented treatment failure (or documented intolerable adverse event) to Ruconest or one of the following:<br>a. Member is less than 13 years of age?<br>b. Request is to treat laryngeal attacks?<br>c. Currently receiving treatment with Berinert, excluding via samples or manufacturer's patient assistance programs? | Yes – Go to #6 | No – Criteria not met; Berinert requires failure with Ruconest |
| 6. Is the drug prescribed by, or in consultation with, an allergist/immunologist or physician that specializes in HAE or related disorders? | Yes – Go to #7 | No – Criteria not met |
| 7. Is the requested dose within the Food and Drug Administration (FDA)-approved label and age restrictions?<br><br>Authorization for therapy for acute treatment will provide a sufficient quantity to cover the number of attacks experienced in the last year plus 1 additional dose. Limited to having medication on hand to treat average number of acute attacks per month plus 1 additional dose. | Yes – Approve up to 3 months | No – Criteria not met |
| **Prophylactic treatment of HAE**<br>**Drugs – Cinryze, Haegarda, Takhzyro, Orladeyo** | | |
| 1. Did treatment with acute therapy (i.e. Kalbitor, Firazyr, Berinert or Ruconest) not result in meaningful outcomes such as decreased severity of attacks, avoidance of hospitalization? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documentation of number of acute attacks requiring treatment in the past year? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is at least ONE of the following present:<br>a. Disabling symptoms for at least 5 days | Yes – Go to #4 | No – Criteria not met |

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| per month<br>b. Laryngeal edema or history of laryngeal edema<br>c. A history of self-limiting, non-inflammatory subcutaneous angioedema, without uticaria, which is recurrent and lasts greater than 12 hours<br>d. Self-limiting, recurrent abdominal pain without a clear organic cause lasting greater than 6 hours | | |
| 4. Is there a history or TWO or more severe attack(s) per month on average for the past 3 months (defined as an attack that significantly interrupts daily activities despite short-term treatment)? | Yes – Document and go to #5 | No – Criteria not met |
| 5. Is the request for Cinryze or Orladeyo? | Yes – Go to #6 | No – Go to #7 |
| 6. Is there a documented treatment failure (or documented intolerable adverse event) to both Haegarda AND Takhzyro or the following:<br>a. Currently receiving treatment with requested drug for prophylaxis, excluding via samples or manufacturer's patient assistance programs and have had a greater than or equal to 50% reduction of frequency and severity of HAE attacks requiring acute therapy from baseline? | Yes – Go to #7 | No – Criteria not met |
| 7. Is the drug prescribed by, or in consultation with, an allergist/immunologist or physician that specializes in HAE or related disorders? | Yes – Go to #8 | No – Criteria not met |
| 8. Is the requested dose within the Food and Drug Administration (FDA)-approved label and age restrictions? | Yes – Approve up to 3 months | No – Criteria not met |

| Renewal Criteria | | |
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| 1. Is there documentation of number of acute attacks treated in the past year AND documentation of treatment success defined as reduction of frequency and severity of HAE attack episodes by greater than or equal to 50% from baseline? | 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**

- **Berinert:** Approved for acute treatment of HAE attacks in adult and pediatric patients.
  - Treatment of acute attacks dosed at 20 units/kg IV.
- **Icatibant, Sajazir, Firazyr**: Approved for acute treatment of HAE attacks in patients 18 and older.
  - Treatment of acute attacks dosed at 30mg SQ. Additional doses may be administered at 6 hour intervals if response is inadequate or symptoms recur. Maximum 3 doses in 24 hours.
- **Ruconest**: Approved for acute treatment of HAE attacks (non-laryngeal) in patients 13 and older.
  - Treatment of acute attacks dosed at 50 units/kg IV, not to exceed 4200 units per dose. If attack symptoms persist, a second dose may be administered. Not to exceed 2 doses in 24 hours. (Effectiveness not demonstrated in patients with laryngeal attacks)
- **Kalbitor**: Approved for acute treatment of HAE attacks in patients 12 years and older.
  - Treatment of acute attacks dosed at 30mg SQ. If attack persists, an additional dose of 30mg may be given within 24 hours.
- **Cinryze**: Approved for routine prophylaxis of HAE attacks in patients 6 years and older.
  - Cinryze Prophylaxis: 1000 units IV twice a week.
    - Doses up to 2,500 units (not exceeding 100 units/kg) may be appropriate if inadequate response with 1000 units.
- **Haegarda**: Approved for routine prophylaxis of HAE attacks in patients 6 years and older.
  - Haegarda Prophylaxis: 60 units/kg SC twice a week.
- **Takhzyro**: Approved for routine prophylaxis of HAE attacks in patients 12 years and older
  - Takhzyro Prophylaxis: 300mg SC every 2 weeks.
    - If patient is dosing every 2 weeks and has been attack free for 6 months, dosing will

be reduced to every 4 weeks.

- **Orladeyo**: Approved for routine prophylaxis of HAE attacks in patients 12 years and older.
  - Orladeyo Prophylaxis: 150 mg once daily.

*Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced for all medical infusion drugs.

POLICY NAME:

**HEREDITARY TYROSINEMIA (HT-1) AGENTS**
Affected Medications: NITYR, ORFADIN

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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 hereditary tyrosinemia type 1 confirmed by biochemical testing (e.g. detection of succinylacetone in urine) and appropriate clinical picture of the patient or by DNA testing<br>• Current patient weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Use as an adjunct to dietary restriction of tyrosine and phenylalanine<br>• Dosing: Initial- 0.5 mg/kg twice daily<br>Maximum: 2 mg/kg/day<br>• Orfadin requires documented failure with or contraindication to Nityr<br>• Reauthorization: documentation of treatment success confirmed by urine or plasma succinylacetone reduction since starting therapy and documented adherence to medical/nutritional therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with physicians that specialize 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 approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**Hormone Supplementation under 18 years of age**
Affected Medications: Depo-Estradiol oil, Estradiol twice weekly patch, Estradiol weekly patch, Estradiol tablets, Estradiol gel, Menest, Divigel transdermal, Elestrin gel, Estrogel, Estropipate, Evamist, Premarin tablets, Testosterone Cypionate solution, Testosterone enanthate, testosterone transdermal, Androxy tablets, Testred capsule, Methitest tablets, Alora Patches, Climara patches, Delestrogen oil, Estrace tablets, Estradiol valerate oil, Lyllana Patch, Menostar Patch, Minivelle Patch, Premarin solution, Vivelle-dot patches

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>• Gender dysphoria<br>  ○ Applies to patients under the age of 18 |
| **Required Medical Information:** | **<u>Gender dysphoria</u>**<br>• Documentation of current Tanner stage 2 or greater OR Documentation of baseline and current estradiol and testosterone levels to confirm onset of puberty<br>• Documentation from a licensed mental health professional (LMHP) confirming diagnosis and addressing the patient's general identifying characteristics;<br>  ○ The initial and evolving gender and any associated mental health concerns, and other psychiatric diagnoses;<br>  ○ The duration of the referring licensed mental health professional's relationship with the client, including the type of evaluation and psychotherapy to date;<br>  ○ The clinical rationale for supporting the client's request for cross-hormone therapy and statement that the client meets eligibility criteria;<br>  ○ Informed consent required from both patient and guardian documented by prescribing provider<br>  ○ Permission to contact the licensed mental health professional for coordination of care<br>• Comprehensive mental health evaluation should be provided in accordance with most current version of the World Professional Association for Transgender Health (WPATH) Standards of Care<br><br>• **Note:** For requests following pubertal suppression therapy, an updated or new comprehensive mental health evaluation |

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| | must be provided prior to initiation of hormone supplementation |
| **Appropriate Treatment Regimen & Other Criteria:** | Reauthorization requires documentation of treatment success |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care<br>• Gender Dysphoria: Diagnosis made and prescribed by, or in consultation with a specialist in the treatment of gender dysphoria |
| **Coverage Duration:** | • Authorization: 12 months, unless otherwise specified |

POLICY NAME:
**HYDROCORTISONE ORAL GRANULES**
Affected Medications: ALKINDI SPRINKLE

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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>• Current glucocorticoid replacement therapy regimen, if applicable |
| **Appropriate Treatment Regimen & Other Criteria:** | • Total daily dose of replacement therapy regimen must be the equivalent of 10 mg or less of hydrocortisone<br>  ○ For doses of greater than 10 mg daily, coverage will not be granted<br>• Documented treatment failure with 6 months of compounded hydrocortisone oral capsules or oral solution<br>• Starting dose: 8-10 mg/m2/day in 3 divided doses<br>  ○ Exception: infants with Congenital Adrenal Hyperplasia may start at a dose of 10-15mg/m2/day in 3 divided doses<br>• When switching patients from other oral hydrocortisone replacement therapy regimens, total daily dose should be equal<br>• Response to therapy should be evaluated monthly in the first three months after starting, every three months in older infants, every six months thereafter while still growing and yearly in those who have achieved adult height<br>• Dose should be reduced if there is evidence of excessive glucocorticoid replacement (excessive weight gain with decreased height velocity, facial plethora, or other symptoms or signs of Cushing syndrome)<br><br>**Reauthorization**:<br>• All initial criteria must be met |

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| | • 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) in adults and pediatric patients 6 years of age and older |
| **Required Medical Information:** | • Documented diagnosis of TSC.<br>  • Presence of facial angiofibromas (at least 2 mm in diameter with redness in each) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with laser therapy and/or surgery, unless contraindicated<br>• Reauthorization requires documentation of a positive clinical response to therapy (decrease in size and/or redness of FAs) |
| **Exclusion Criteria:** | • Those on systemic mammalian target of rapamycin inhibitors<br>• Non-facial angiofibroma |
| **Age Restriction:** | • 6 years of age and older |
| **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:
**IDECABTAGENE VICLEUCEL**
Affected Medications: Abecma (idecabtagene vicleucel)

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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 |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Relapsed or Refractory Multiple Myeloma (MM)</u>**<br>• Treatment with four or more prior lines of therapy, including:<br>○ Immunomodulatory agent<br>○ Proteasome inhibitor AND<br>○ Anti-CD38 monoclonal antibody.<br>• Patient has experienced disease progression after their last regimen or is refractory to their most recent therapy<br><br>Approved for one-time single infusion only |
| **Exclusion Criteria:** | • ECOG score of 2 or greater<br>• Creatinine clearance of less than or equal to 45 mL/minute<br>• Alanine aminotransferase greater than 2.5 times upper limit of normal<br>• Left ventricular ejection fraction less than 45%<br>• Absolute neutrophil count less than 1000 cells/mm^3<br>• Platelet count less than 50,000/mm^3 |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Must be prescribed by an oncologist<br>• Oncologist and administering health care facility must be certified and in compliance with the Risk Evaluation and Mitigation Strategies (REMS) requirements<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Approval: 1 month, unless otherwise specified (one infusion only) |

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), Familial Mediterranean Fever (FMF), Adult-Onset Still's Disease (AOSD), Systemic Juvenile Idiopathic Arthritis (SJIA), Cryopyrin-Associated Periodic Syndromes (CAPS). |
| **Required Medical Information:** | • Patient weight<br><br><u>Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS)</u><br>• Confirmed diagnosis of TRAPS with frequent and/or severe recurrent disease AND documented genetic defect of TNFRSF1A gene<br><br><u>Hyperimmunoglobulin D syndrome (HIDS)</u><br>• Confirmed diagnosis including presence of heterozygous or homozygous mutation in the mevalonate kinase (MVK) gene<br>• Documented frequent and severe attacks with substantive quality-of-life detriment<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 aged 2 years and older<br>• Documentation of active joint count<br><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) |
| **Appropriate Treatment** | <u>Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS)</u> |

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| **Regimen & Other Criteria:** | • Documented clinical failure to episodic treatment with nonsteroidal anti-inflammatory drugs (NSAIDs), glucocorticoids (prednisone or prednisolone), and a minimum 12 week trial with Enbrel<br><br><u>Hyperimmunoglobulin D syndrome (HIDS)</u><br>• Documented treatment failure to episodic treatment with nonsteroidal anti-inflammatory drugs (NSAIDs), glucocorticoids, and anakinra<br><br><u>Familial Mediterranean Fever (FMF)</u><br>• Documented treatment failure with maximal tolerable dose of colchicine (3 mg daily in adults and 2 mg daily in children)<br>AND<br>• Documentation of frequent and/or severe recurrence disease despite adequate treatment with at least 12 weeks of Anakinra<br><br><u>Still's Disease</u><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><u>Cryopyrin-Associated Periodic Syndromes (CAPS)</u><br>• Documentation of treatment failure with a minimum 12 week trial with anakinra<br><br>After up to 8 weeks of therapy if the patient has had a response to therapy as determined by prescribing physician an additional 6 months authorization is allowed.<br><br>**Reauthorization:** Documentation of treatment success. |
| **Exclusion Criteria:** | • Treatment of neonatal onset multisystem inflammatory disorder (NOMID) or chronic infantile neurological cutaneous and articular syndrome (CINCA), gout, rheumatoid arthritis, chronic obstructive pulmonary disease (COPD), type 2 diabetes mellitus |

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| | • Use in combination with tumor necrosis factor (TNF) blocking agents (e.g. Enbrel, Humira, Cimzia, Remicade, Simponi), Kineret, or Arcalyst |
| **Age Restriction:** | • FMF, HIDS, juvenile idiopathic arthritis, TRAPS: 2 years of age and older<br>• CAPS: 4 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, an allergist, immunologist, or rheumatologist |
| **Coverage Duration:** | • Initial approval: 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 from benefit design. |
| **Required documentation:** | **<u>Pulmonary arterial hypertension (PAH) WHO Group 1</u>**<br>• Documentation of PAH confirmed by right-heart catheterization<br>• NYHA/WHO Functional Class III or IV symptoms<br>• Etiology of PAH: idiopathic PAH, hereditary PAH, OR 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>  ○ Shistosomiasis<br>  ○ Portal hypertension<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blocker) unless contraindications exist such as low systemic blood pressure (systolic blood pressure less than 90), low cardiac index, or presence of severe symptoms (functional class IV) |
| **Appropriate Treatment Regimen:** | • For initiation of therapy patient must have mean pulmonary artery pressure at least 25 mmHg at rest or at least 30 mmHg with exertion **AND**<br>• The pulmonary hypertension has progressed despite maximal medical and/or surgical treatment of the identified condition<br><br>**<u>Reauthorization</u>** requires documentation of treatment success including improvements in one of the following: exercise endurance, echocardiographic testing, hemodynamic testing, brain natriuretic peptide (BNP), functional class |
| **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.) |

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| **Age Restriction:** | • 18 years or older |
| **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:** | • 12 months, unless otherwise specified |

POLICY NAME:
**IMIGLUCERASE**
Affected Medications: CEREZYME (imiglucerase) (J1786) IV Infusion

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by benefit design.<br>  ○ Gaucher disease, Type 1 |
| **Required Medical Information:** | • Diagnosis of Type 1 Gaucher disease confirmed by enzyme assay.<br>• Must include current symptoms characteristic of bone involvement such as:<br>  ○ Low platelet count<br>  ○ Low hemoglobin and hematocrit levels<br>  ○ Radiologic bone disease, T-score less than -2.5 or bone pain<br>  ○ Delayed growth in children<br>• Documented patient weight, dose and frequency |
| **Appropriate Treatment Regimen & Other Criteria:** | • <u>Documented adult patients with symptomatic disease:</u> platelet count less than 60,000/microL, liver greater than 2.5 times normal size, spleen greater 15 times normal size, radiologic evidence of skeletal disease<br>• <u>Documented symptomatic children:</u> includes those with malnutrition, growth retardation, impaired psychomotor development, and/or fatigue (early presentation is associated with more severe disease)<br><br><u>Reauthorization criteria:</u><br>  • Documentation of treatment efficacy based on improved labs or patient symptoms |
| **Exclusion Criteria:** | • Gaucher disease (Type 2 or Type 3)<br>• Combination treatment with more than one targeted therapy for Gaucher disease<br>• Dose increases due to osteonecrosis and fibrosis of liver, spleen, or lung |
| **Age Restriction:** | • Greater than or equal to 2 years old |

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| **Prescriber/Site of Care Restrictions:** | • Provider experienced in the treatment of Gaucher disease<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 3 months, unless otherwise specified<br>• Reauthorization (treatment effective): 12 months, unless otherwise specified |

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

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| **Covered Uses:** | • 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>○ HIV infected children: 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<br>○ Toxic Shock Syndrome<br>○ Pediatric Acute-Onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS) |
| **Initial Approval Criteria:** | **Primary immunodeficiency (PID)/Wiskott - Aldrich syndrome**<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 |

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| | 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:<br>  ○ Titers that were drawn before challenging with vaccination AND<br>  ○ Titers that were drawn between 4 and 8 weeks after vaccination<br><br>**Idiopathic thrombocytopenia purpura (ITP)**<br>For Acute disease state:<br>• Documented use to manage acute bleeding due to severe thrombocytopenia (platelet counts less than 30); OR<br>• To increase platelet counts prior to invasive surgical procedures, such as splenectomy. (Platelets less than 100); OR<br>• Documented severe thrombocytopenia (platelet counts less than 20) and is considered to be at risk for intracerebral hemorrhage;<br>• Authorization is valid for 1 month only<br>Chronic Immune Thrombocytopenia (CIT):<br>  ○ Documentation of increased risk for bleeding as indicated by a platelet count less than 30; AND<br>  ○ History of failure, contraindication, or intolerance with corticosteroids; AND<br>  ○ Duration of illness more than 6 months; AND |

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| | ○ 10 years of age or older<br><br>**Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)**<br>• Documented baseline in strength/weakness has been documented 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; AND<br>• An abnormal or absent deep tendon reflexes in upper or lower limbs; AND<br>• Electrodiagnostic testing indicating demyelination:<br>○ Partial motor conduction block in at least two motor nerves or in 1 nerve plus one other demyelination criterion listed here in at least 1 other nerve; OR<br>○ Distal CMAP duration increase in at least 1 nerve plus one other demyelination criterion listed here in at least 1 other nerve; OR<br>○ Abnormal temporal dispersion conduction must be present in at least 2 motor nerves OR<br>○ Reduced conduction velocity in at least 2 motor nerves; OR<br>○ Prolonged distal motor latency in at least 2 motor nerves; OR<br>○ Absent F wave in at least two motor nerves plus one other demyelination criterion listed here in at least 1 other nerve; OR<br>○ Prolonged F wave latency in at least 2 motor nerves ; AND<br>• Cerebrospinal fluid analysis indicates the following:<br>○ CSF white cell count of less than 10 cells/mm3; AND<br>○ CSF protein is elevated; AND<br>• Refractory to or intolerant of corticosteroids (prednisolone, prednisone) given in therapeutic doses over at least three months<br>• Initial approval will be valid for 3 months. Subsequent authorizations will be approved for up to 1 year<br><br>**Guillain-Barre Syndrome (Acute inflammatory polyneuropathy)**<br>• Documentation that the disease is severe (aid required to walk); AND |

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| | • Onset of symptoms are recent (less than 1 month); AND<br>• Approval will be granted for a maximum of 2 rounds of therapy within 6 weeks of onset; 2 months maximum<br><br>**Multifocal Motor Neuropathy**<br>• Documented multi-focal weakness; AND<br>• Partial conduction block or abnormal temporal dispersion conduction must be present in at least 2 nerves; AND<br>• Baseline in strength/weakness has been documented using objective clinical measuring tool (e.g. INCAT, Medical Research Council (MRC) muscle strength, 6 Minute walk test, Rankin, Modified Rankin); AND<br>• Initial authorization length is 1 course (1 month) to assess viability of treatment.<br><br>**HIV infected children: Bacterial control or prevention**<br>• Approved for those 13 years of age and younger<br><br>**Myasthenia Gravis**<br>• Documented myasthenic crisis (impending respiratory or bulbar compromise); AND<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>• Approval for one course (1 month)<br><br>**Dermatomyositis/Polymyositis**<br>• Documented severe active disease state on physical exam; AND<br>• Proximal weakness in all upper and/or lower limbs; AND<br>• CPK greater than 1,000 (with documentation of previously normal CPK); AND<br>• Documented failure with a trial of corticosteroids (such as prednisone); AND<br>• Documented failure with a trial of immunosuppressants (Methotrexate, azathioprine)<br>• Initial approval will be valid for 3 months;<br>• Renewals will require current CPK lab and physical exam |

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| | **Complications of transplanted solid organ (kidney, liver, lung, heart, pancreas) and bone marrow transplant**<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>• Documented anti-GAD antibodies; AND<br>• Documented failure with at least 2 of the following treatments: benzodiazepines, baclofen, phenytoin, clonidine and/or tizanidine<br><br>**Allogeneic Bone Marrow or Stem Cell Transplant**<br>• Approved in use for prevention of acute Graft- Versus- Host Disease(GVHD) or infection (such as cytomegalovirus)<br>• Documentation that the BMT was allogeneic; AND<br>• Transplant was less than 100 days ago<br>• Authorization is valid for 3 months<br><br>**Kawasaki's Disease (Pediatric)**<br>• Approved for age 13 years or under for 1 course of treatment (1 month)<br><br>**Fetal alloimmune thrombocytopenia (FAIT)**<br>• Documentation of one or more of the following:<br>o Previous FAIT pregnancy<br>o Family history of the disease<br>o Screening reveals platelet alloantibodies<br>• Authorization is valid until delivery date only<br><br>**Hemolytic disease of the newborn**<br>• Approved for 1 course of treatment (1 month)<br><br>**Auto-immune Mucocutaneous Blistering Diseases**<br>• Diagnosis confirmed by biopsy of one of the following:<br>o Pemphigus vulgaris |

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| | ○ Pemphigus foliaceus<br>○ Bullous Pemphigoid<br>○ Mucous Membrane Pemphigoid (also known as Cicatricial Pemphigoid)<br>○ Epidermolysis bullosa aquisita<br>○ Pemphigus gestationis (Herpes gestationis)<br>○ Linear IgA dermatosis; AND<br>• Documented severe disease that is extensive and debilitating; AND<br>• Disease is progressive; AND<br>• Refractory to a trial of conventional combination therapy with corticosteroids and immunosuppressive treatment (azathioprine, cyclophosphamide, mycophenolate mofetil)<br><br>**Chronic lymphocytic leukemia with associated hypogammaglobulinemia**<br>• Documentation of an IgG level less than 500<br>AND<br>• A documented history of recurrent or chronic infections that have required intravenous antibiotics or hospitalization<br><br>**Toxic Shock Syndrome**<br>• Approved for a single course of therapy (1 month)<br><br>**Pediatric Acute-Onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated With Streptococcal Infections (PANDAS)**<br>• Documentation of active autoimmune process (neuro-inflammation or post-infectious autoimmunity) confirmed by appropriate indicators such as:<br>○ Elevated erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP)<br>○ Exacerbation of autoimmune disease (eg, thyroiditis, spondyloarthritis, rheumatoid arthritis, etc.)<br>• Abrupt and severe onset of the following symptoms between 3 years of age and the onset of puberty:<br>○ Obsessive-compulsive disorder (OCD) or severely restricted food intake AND |

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| | ○ Acute onset of at least two concurrent severe neuropsychiatric symptoms (eg, anxiety, depression, emotional lability, etc)<br>○ Documentation that symptoms cause significant interference with daily activities and overall functioning<br>• Documentation of comprehensive psychiatric evaluation<br>• Documentation of lab work and other studies excluding alternate diagnose<br>• Trial and failure of all of the following treatments in combination for at least 6 weeks:<br>○ Behavioral pharmacologic therapy (eg. Fluoxetine, fluvoxamine, sertraline) AND behavior therapies for neuropsychiatric symptoms<br>○ NSAIDs (eg. Naproxen, Diclofenac, Ibuprofen)<br>○ Oral and IV corticosteroids (eg. Prednisone, methylprednisolone)<br><br>• Approved for a single course of therapy (1 month) |
| **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**<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**<br>• Renewals will require 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**<br>• Renewals will require 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>**HIV infected children: Bacterial control or prevention**<br>• Age 13 years or less<br>**Dermatomyositis/Polymyositis** |

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<tr><td></td><td>
• Renewal will require documentation that CPK (Creatine phosphokinase) levels are lower upon renewal request; AND<br>
• Documentation of clinically significant improvement above baseline per physical exam<br>
• Approved for up to 6 months<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:<br>
• Renewal requires a documented clinically significant improvement over baseline per physical exam<br>
• Renewals will be approved for up to 6 months<br>
Chronic lymphocytic leukemia (CLL) with associated hypogammaglobulinemia<br>
• Renewal requires disease response as evidenced by a decrease in the frequency and/or severity of infections<br>
• Renewals will be approved for up to 6 months<br>
Pediatric Acute-Onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated With Streptococcal Infections (PANDAS)<br>
• Renewal requires documentation of symptomatic improvement within 4 weeks after initial dose with evident recurrence of symptoms after initial course
</td></tr>
<tr><td>Dosing:</td><td>
Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced
<table>
<tr><th>Indication</th><th>Dose</th></tr>
<tr><td>PID</td><td>Up to 800 mg/kg every 21 days</td></tr>
<tr><td>CIDP</td><td>2 g/kg divided over 2-4 days X 1,</td></tr>
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| | | then 1 g/kg every 21 days |
| | ITP | 2 g/kg divided over 5 days in a 28 day cycle |
| | FAIT | 1 g/kg/week until delivery |
| | Kawasaki's Disease (pediatric patients) | 2 g/kg x 1 single dose |
| | MMN | 2 g/kg divided over 5 days in a 28 day cycle |
| | CLL | 400 mg/kg every 3 weeks |
| | Pediatric HIV | 400 mg/kg every 28 days |
| | Guillain-Barre | 2 g/kg divided over 5 days x 1 cycle |
| | Myasthenia Gravis | 1 g/kg x 1 dose (acute attacks) |
| | Auto-immune blistering diseases | 2 g/kg divided over 5 days in a 28 day cycle |
| | Dermatomyositis/Polymyositis | 2 g/kg divided over 5 days in a 28 day cycle |
| | Bone Marrow or Stem Cell Transplant | 500 mg/kg/week x 90 days, then 500 mg/kg/month 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 |
| | Stiff Person | 2 g/kg divided over 5 days in a 28 day cycle |
| | Toxic shock syndrome | 2 g/kg divided over 5 days x 1 cycle |
| | Hemolytic disease of the newborn | 1 g/kg x 1 dose, may be repeated once if needed |
| | PANS/PANDAS | Initial dose: 1.5-2 g/kg divided over 2-5 days<br>Subsequent: monthly doses (up to 6 total doses): 1-2 g/kg divided over 2-5 days |
| **Prescriber/Site of Care Restrictions:** | • Must be prescribed by a specialist for the condition being treated (neurologist, rheumatologist, immunologist, hematologist)<br>• All approvals are subject to utilization of the most cost-effective site of care | |

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| **Coverage Duration:** | • Initial approval: Up to 3 months, unless otherwise specified<br>• Reauthorization: Up to 12 months, unless otherwise specified |

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>  ○ Adjunct to diet and maximally tolerated statin therapy for the treatment of adults with heterozygous familial hypercholesterolemia (HeFH) or clinical atherosclerotic cardiovascular disease (ASCVD), who require additional lowering of low-density lipoprotein cholesterol (LDL-C) |
| **Required Medical Information:** | **<u>Heterozygous Familial Hypercholesterolemia (HeFH):</u>**<br>• Diagnosis of HeFH confirmed by:<br>  ○ Genetic testing<br>  OR<br>  ○ Documented history of untreated LDL-C of greater than 190 mg/dL **AND** a first degree relative with: confirmed HeFH, LDL-C of greater than 190 mg/dL, or with known premature coronary heart disease (less than 55 years for men; less than 60 years for women).<br><br>**<u>Clinical Atherosclerotic Cardiovascular Disease (ASCVD):</u>**<br>• Diagnosis of Clinical ASCVD or a cardiovascular event, defined as:<br>  ○ Acute coronary syndromes, myocardial infarction (MI), stable or unstable angina, coronary or other arterial revascularization procedure (e.g., CABG, PTCA), stroke of presumed atherosclerotic origin, transient ischemic attack (TIA), peripheral arterial disease of presumed atherosclerotic origin, findings from CT angiogram or catheterization consistent with clinical ASCVD |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>HeFH:</u>**<br>• Documented treatment failure with statin therapy defined as:<br>  ○ Current LDL-C level of at least 100 mg/dL after a least three months of adherent use with maximally-tolerated statin therapy |

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| | OR<br>○ Current LDL-C level of at least 100 mg/dL and statin intolerance defined as:<br>- Intolerable statin-associated muscle symptoms lasting at least two weeks confirmed with at least two attempts of statin re-challenge (including 2 different statins, one of which being either atorvastatin or rosuvastatin) OR<br>- Rhabdomyolysis with statin-associated elevation in creatinine kinase (CK) level to at least 10 times the upper limit of normal<br>AND<br>• Documented treatment failure (or intolerable adverse event) to a minimum 12-week trial of Repatha<br><br>**<u>Clinical Atherosclerotic Cardiovascular Disease (ASCVD):</u>**<br>• Documented treatment failure with statin therapy defined as:<br>○ Current LDL-C level of at least 70 mg/dL after a least three months of adherent use with maximally-tolerated statin therapy<br>OR<br>○ Current LDL-C level of at least 70 mg/dL and statin intolerance defined as:<br>- Intolerable statin-associated muscle symptoms lasting at least two weeks confirmed with at least two attempts of statin re-challenge (including 2 different statins, one of which being either atorvastatin or rosuvastatin) OR<br>- Rhabdomyolysis with statin-associated elevation in creatinine kinase (CK) level to at least 10 times the upper limit of normal<br>AND<br>• Documented treatment failure (or intolerable adverse event) to a minimum 12-week trial of Repatha<br><br>**<u>Reauthorization:</u>** requires documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider |

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| | **<u>Dosing:</u>**<br>284 mg as a single injection at 0 and 3 months, then every 6 months thereafter |
| **Exclusion Criteria:** | • Concurrent use with other PCSK9 inhibitors |
| **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:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**INTRAVITREAL ANTI-VEGF THERAPY**
Affected Medications: LUCENTIS (ranibizumab injection), EYLEA (aflibercept), BEOVU (brolucizumab), SUSVIMO (ranibizumab implant), VABYSMO (faricimab), BYOOVIZ (ranibizumab-nuna), CIMERLI (ranibizumab-eqrn)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>○ Neovascular (Wet) Age-Related Macular Degeneration (AMD)<br>• Eylea, Lucentis, Susvimo, Beovu, Vabysmo, Byooviz, Cimerli<br>○ Macular Edema Following Retinal Vein Occlusion (RVO)<br>• Eylea, Lucentis, Byooviz, Cimerli<br>○ Diabetic Macular Edema (DME)<br>• Eylea, Lucentis, Vabysmo, Beovu, Cimerli<br>○ Diabetic Retinopathy (DR) in patients with Diabetes Mellitus<br>• Eylea, Lucentis, Cimerli<br>○ Myopic Choroidal Neovascularization (mCNV)<br>• Lucentis, Byooviz, Cimerli |
| **Required Medical Information:** | ○ Anticipated treatment course with dose and frequency clearly stated in chart notes. |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Eylea Dosing</u>**<br>○ **Coverage for the non-preferred product Eylea is provided when either of the following criteria is met:**<br>○ Currently receiving treatment with Eylea, 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 all of the preferred products (Avastin, Byooviz, and Cimerli)<br>• **AMD -** 2mg (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 |

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| | • **DME and DR -** 2mg (0.05 mL) every 4 weeks for the first 5 injections followed by 2 mg (0.05 mL) every 8 weeks<br><br>**<u>Lucentis Dosing</u>**<br>• **Coverage for the non-preferred product Lucentis is provided when either of the following criteria is met:**<br>  ○ Currently receiving treatment with Lucentis, 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 all of the preferred products (Avastin, Byooviz, and Cimerli)<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><br>**<u>Byooviz Dosing</u>**<br>• **AMD and RVO -** maximum 0.5 mg every 4 weeks<br>• **mCNV -** 0.5 mg monthly for up to 3 months<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 all of the preferred products (Avastin, Byooviz, and Cimerli)<br>• **AMD** – 6 mg every month for the first three doses followed by 6 mg every 8-12 weeks<br>• **DME** – 6 mg every six weeks for the first five doses followed by 6 mg every 8-12 weeks<br><br>**<u>Susvimo Dosing</u>**<br>• **Coverage for the non-preferred product Susvimo is provided when either of the following criteria is met:**<br>  ○ Currently receiving treatment with Susvimo, excluding |

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| | when the product is obtained as samples or via manufacturer's patient assistance programs.<br>○ A documented inadequate response or intolerable adverse event with all of the preferred products (Avastin, Byooviz, and Cimerli)<br>• Must be established on ranibizumab (Lucentis) injections with response to treatment for a minimum of 6 months at standard dosing (0.5 mg every 4 weeks)<br>• **AMD** – 2 mg administered continuously via ocular implant with refills every 24 weeks.<br><br>**<u>Vabysmo Dosing</u>**<br>• **Coverage for the non-preferred product Vabysmo is provided when either of the following criteria is met**:<br>○ Currently receiving treatment with Vabysmo, 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 all of the preferred products (Avastin, Byooviz, and Cimerli)<br>• **AMD** – 6 mg every 4 weeks for the first 4 injections followed by 6 mg every 8 to 16 weeks<br>○ Some patients may require continued every 4 week injections following the initial doses<br>• **DME**<br>○ Fixed interval regimen: 6 mg every 4 weeks for the first 6 injections followed by 6 mg every 8 weeks<br>○ Variable interval regimen: 6 mg once every 4 weeks for at least the first 4 injections followed by 6 mg every 4 to 16 weeks (based on visual assessments)<br>○ Some patients may require continued every 4 week injections following the initial doses<br><br>**<u>Cimerli Dosing</u>**<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 |

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| | **<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 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 approval: 6 months, unless otherwise specified<br>• Reauthorization: 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 benefit 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 |

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

POLICY NAME:
**INVEGA INJECTABLES**
Affected Medications: INVEGA TRINZA (Paliperidone Palmitate Extended-Release Injectable Suspension), INVEGA HAFYERA (Paliperidone Palmitate Extended-Release Injectable Suspension)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ Schizophrenia |
| **Required Medical Information:** | • A documented history of non-compliance, refusal to utilize oral medication, or cannot be stabilized on oral medications |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Invega Trinza</u>**<br>• Adequate treatment has been established with Invega Sustenna for at least 4 months<br>AND<br>• Documented anticipated dose and dosing schedule based on maintenance Invega Sustenna maintenance dose<br>• Once every 3 months dosing<br><br>**<u>Invega Hafyera</u>**<br>• Adequate treatment has been established with Invega Sustenna for at least 4 months or with Invega Trinza for at least one three-month injection cycle<br>AND<br>• Documented anticipated dose and dosing schedule based on maintenance Invega Sustenna or Invega Trinza maintenance dose<br>• Once every 6 months dosing<br><br>**<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy. |
| **Exclusion Criteria:** | • Diagnosis of dementia-related psychosis.<br>• Prior hypersensitivity (anaphylactic reactions and/or angioedema) to paliperidone or risperidone |
| **Age Restriction:** | |

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| **Prescriber/Site of Care Restrictions:** | • Psychiatrist or in consultation with a psychiatric practice.<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:
**IOBENGUANE I-131**
Affected Medications: Azedra (iobenguane I-131)

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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:** | • Documented diagnosis of metstatic or unresectable pheochromocytoma or paraganglioma<br>**AND**<br>• Positive adrenal/abdominal MRI or CT scan<br>**AND**<br>• Prior positive meta-iodobenzylguanidine (MIBG) scan with dosimetry<br><br>**Reauthorization**: Will require documentation of disease responsiveness to therapy |
| **Appropriate Treatment Regimen & Other Criteria:** | Dosimetric Dose<br>• Patients weighing greater than 50 kg: 185 to 222 MBq (5 or 6 mCi) intravenous<br>• Patients weighing 50 kg or less: 3.7 MBq/kg (0.1 mCi/kg) intravenous<br><br>Therapeutic Dosage: administer 2 therapeutic doses intravenously a minimum of 90 days apart<br>1. Patients weighing greater than 62.5 kg: 18,500 MBq (500 mCi)<br>• Patients weighing 62.5 kg or less: 296 MBq/kg (8 mCi/kg) |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • Must be at least 12 years old |
| **Prescriber/Site of Care Restrictions:** | • Oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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

POLICY NAME:
**IPILIMUMAB**
Affected Medications: YERVOY (ipilimumab)

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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 of use with NCCN 2A or higher level of evidence regimen |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Non-Small Cell Lung Cancer (NSCLC)</u>**<br>• Documentation of use only as first line systemic therapy for advanced or metastatic disease<br>• Documentation of use in combination with nivolumab (Opdivo)<br>• Documented current programmed death-ligand 1 (PD-L1) level<br>○ For PD-L1 less than 1%: Yervoy and Opdivo must include two cycles of chemotherapy with a platinum agent and pemetrexed (Alimta)<br><br>**<u>For all other conditions:</u>**<br>• Documentation of use with NCCN 2A or higher level of evidence regimen<br><br>**<u>Reauthorization</u>**: documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Documented prior immunotherapy treatment failure<br>• Karnofsky Performance Status 50% or less or Eastern Cooperative Oncology Group (ECOG) performance score 3 or greater |
| **Age Restriction:** | • 12 years or older for unresectable or metastatic melanoma, colorectal cancer, melanoma<br>• 18 years or older for NSCLC |
| **Prescriber/Site of Care Restrictions:** | • 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:
**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>  ○ Invasive aspergillosis<br>  ○ Invasive mucormycosis |
| **Required Medical Information:** | **Aspergillosis:**<br>• Documented clinical failure, contraindication, or intolerable adverse event to at least 6 weeks of both of the following:<br>  ○ Voriconazole<br>  ○ Posaconazole<br><br>**Mucormycosis:**<br>• Documented clinical failure, contraindication, or intolerable adverse event to at least 6 weeks of 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) |
| **Appropriate Treatment Regimen & Other Criteria:** | All Indications:<br>• Susceptibility cultures matching isavuconazonium activity<br>• Exceptions made for empiric therapy as long as treatment is adjusted when susceptibility cultures are available.<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 |

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| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 3 months, unless otherwise specified |

POLICY NAME:
**KALYDECO**
Affected Medications: KALYDECO (ivacaftor)

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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 cystic fibrosis (CF) diagnosis.<br>• Documentation confirming Food and Drug Administration (FDA) approved mutation 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>• Liver Function Testing prior to Kalydeco initiation, every 3 months during first year of treatment, and annually thereafter. |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Homozygous F508del mutation |
| **Age Restriction:** | • Ivacaftor oral granules are approved in patients 4 months of age and older.<br>• Ivacaftor oral tablets are approved in patients 6 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 approval: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

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

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by benefit design. |
| **Required Medical Information:** | • Documentation of anticipated treatment course, including target phenylalanine (Phe) level set by specialist<br>• Documentation of failure to Phe restricted diet as monotherapy<br>• Current patient weight<br>• Baseline (pre-treatment) blood Phe levels<br><br>Baseline Phe concentration must be consistent with the following:<br>• Age less than or equal to 12 years: Phe level must be greater than 6mg/dL (360 microM)<br>• Age greater than or equal to 12 years: Phe level must be greater than 10mg/dL (600 microM)<br>• During pregnancy: Phe level must be greater than 6mg/dL (360 microM)<br><br>Reauthorization after initial approval requires documentation of updated Phe labs decreased by 30% or greater from baseline<br>• Treatment with Kuvan should be discontinued in patients whose blood Phe has not decreased by at least 30 percent from baseline<br><br>Reauthorization for continued long-term approval (12 months) requires updated Phe labs meeting one of the following criteria:<br>• Phe level less than 30 percent of baseline OR<br>• Phe level lower than baseline and meets specialist's target level |
| **Appropriate Treatment Regimen & Other Criteria:** | If patient has failed monotherapy with Phe restricted diet and treatment with Kuvan is warranted, treatment must be consistent with the following:<br>○ Phe restricted diet must be maintained during Kuvan treatment **AND**<br>○ Initial dose must be 10mg/kg/day x 1 month<br>○ If blood Phe does not decrease from baseline after 1 month, dose can be increased to 20mg/kg/day x 1 month |

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| **Exclusion Criteria:** | • Prior intolerance or allergic reaction to requested medication<br>• Doses greater than 20mg/kg/day |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Specialist in metabolic disorders or endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 2 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**LARONIDASE**
Affected Medications: ALDURAZYME

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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 one the following type I mucopolysaccharidosis:<br>  ○ Hurler Mucopolysacchardiosis I (MPS I H)<br>  ○ Herler-Scheie Mucopolysaccharidosis I (MPS I H/S)<br>  ○ Scheie form of Mucopolysacchardiosis (MPS I S) with moderate to severe symptoms<br>• Diagnosis confirmed by an enzyme assay showing deficiency of alpha-L-iduronidase enzyme activity or by DNA testing<br>• Patient weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Appropriate medical support readily available when Aldurazyme is administered in case of anaphylaxis or severe allergic reaction<br>• Pretreatment with antipyretics and/or antihistamines prior to infusion<br>• QL: 0.58 mg/kg intravenous once weekly<br>• Reauthorization: documentation of treatment success defined as improvement in percent predicted forced vital capacity (FVC), six-minute walk test, sleep apnea, shoulder flexion, and activities of daily living |
| **Exclusion Criteria:** | • Treatment of central nervous system manifestation of the disorder |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 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 NTRK gene-fusion, as determined by an FDA approved test. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Requires previous treatment with Rozlytrek<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:** | • 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:
**LEUPROLIDE**
Affected Medications: LUPRON DEPOT 3.75 MG and 11.25 MG AND LUPRON DEPOT-PED 11.25 MG; LUPRON DEPOT 7.5 MG, 22.5 MG, 30 MG, and 45 MG AND LUPRON DEPOT-PED 15 MG AND ELIGARD; LEUPROLIDE ACETATE OR INJECTION SOLUTION, LUPANETA KITS, FENSOLVI, CAMCEVI 45 MG.

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design<br>• NCCN (National Comprehensive Cancer Network) indications level 2A or higher<br>• Gender dysphoria |
| **Required Medical Information:** | **<u>Endometriosis</u>**<br>• Documentation of moderate to severe pain due to endometriosis<br>• Documentation of a trial and inadequate relief (or contraindication) after at least three months of first-line therapy with nonsteroidal anti-inflammatory drugs (NSAIDs) and continuous (no placebo pills) hormonal contraceptives<br><br>**<u>Preoperative anemia due to uterine leiomyomata</u>**<br>• Documentation of leiomyoma-related surgery in 6 or less months<br>• Documentation of planned use in combination with iron supplements<br><br>**<u>Gender dysphoria</u>**<br>• Documentation of current Tanner stage 2 or greater OR documentation of baseline and current estradiol and testosterone levels to confirm onset of puberty<br>• Documentation from a licensed mental health professional (LMHP) confirming diagnosis and addressing the patient's general identifying characteristics;<br>o The initial and evolving gender and any associated mental health concerns, and other psychiatric diagnoses;<br>o The duration of the referring licensed mental health professional's relationship with the client, |

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| | including the type of evaluation and psychotherapy to date;<br>○ The clinical rationale for supporting the client's request for hormone therapy and statement that the client meets eligibility criteria; and<br>○ Permission to contact the licensed mental health professional for coordination of care<br>• Comprehensive mental health evaluation should be provided in accordance with most current version of the World Professional Association for Transgender Health (WPATH) Standards of Care<br><br>**<u>Central precocious puberty</u>**<br>• Documentation of central precocious puberty (CPP) confirmed by basal luteinizing hormone (LH), follicle-stimulating hormone (FSH), and either estradiol or testosterone concentrations |
| **Appropriate Treatment Regimen & Other Criteria:** | • Women of childbirth age should have pregnancy ruled out and a plan to use a non-hormonal based contraceptive during therapy<br><br>**<u>Endometriosis</u>**<br>• Lupron Depot 3.75 and 11.25mg<br><br>**<u>Preoperative anemia due to uterine leiomyomata</u>**<br>• Lupron Depot 3.75 and 11.25mg<br>• Planned treatment of 6 months or less<br>• Must be given in conjunction with iron supplementation<br><br>**<u>Central precocious puberty</u>**<br>• Leuprolide Acetate, Lupron Depot-Ped 11.25 and 15mg, Fensolvi 45mg<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) |

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| **Age Restriction:** | • Endometriosis and preoperative uterine leiomyomata: age 18 years or older<br>• Central precocious puberty (CPP): age 11 or younger (females), age 12 or younger (males) |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with oncologist, endocrinologist, or gynecologist for endometriosis<br>• Gender Dysphoria: Diagnosis made and prescribed by, or consultation with a specialist in the treatment of gender dysphoria<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>o Corticotropin (ACTH)-producing pituitary tumor (Cushing's disease)<br>o Ectopic ACTH secretion by a non-pituitary tumor<br>o Cortisol secretion by an adrenal adenoma<br>AND<br>• Documentation that surgery is not an option or has not been curative<br>AND<br>• A mean of at least three 24-hour Urine Free Cortisol (mUFC) levels greater than 1.5 times the upper limit of normal (ULN) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented clinical failure to a minimum 8 week trial of the maximally tolerated dose of ketoconazole<br>OR<br>• Intolerable adverse event to ketoconazole, and the adverse event was not an expected adverse event attributed to the active ingredient<br><br><u>Reauthorization:</u> documentation of treatment success as determined by mUFC less than or equal to the ULN based on central laboratory results |
| **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 |

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

POLICY NAME:
**LISOCABTAGENE MARALEUCEL**
Affected Medications: BREYANZI (lisocabtagene maraleucel)

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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 |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Relapsed or Refractory B-cell Lymphoma</u>**<br>• Diagnosed with one of the following:<br>  ○ Diffuse large B-cell lymphoma (DLBCL) not otherwise specified (including DLBCL arising from indolent lymphoma)<br>  ○ High-grade B-cell lymphoma<br>  ○ Primary mediastinal large B-cell lymphoma<br>  ○ Follicular lymphoma grade 3B<br>AND<br>• Disease is refractory to first line chemoimmunotherapy or relapse within 12 months of first-line chemotherapy<br>OR<br>• Disease has relapsed, or has been refractory, after 2 or more lines of systemic therapy<br><br>Approved for one-time single infusion only |
| **Exclusion Criteria:** | • ECOG status greater than 2<br>• Creatinine clearance less than 30 mL/min<br>• Alanine aminotransferase greater than 5 times the upper limit of normal<br>• Left ventricular ejection fraction less than 40%<br>• Primary CNS lymphoma<br>• Prior CAR-T therapy |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Must be prescribed by an Oncologist<br>• Must be administered at a Risk Evaluation and Mitigation Strategies (REMS)-certified healthcare facility |

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| | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 1 month, unless otherwise specified (one infusion only) |

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

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| **Covered Uses:** | • All 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 |

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| | ○ Round all total daily doses to the nearest 25 mg increment<br><br><u>Reauthorization</u>:<br>• Documentation of treatment success and initial criteria to be met. |
| **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:
**LONG ACTING INJECTABLE RISPERIDONE**
Affected Medications: PERSERIS (risperidone subcutaneous injection), RISPERDAL CONSTA (risperidone intramuscular injection)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>○ Schizophrenia<br>○ Bipolar I disorder maintenance treatment as monotherapy or as adjunctive therapy to lithium and valproate (Risperdal Consta only) |
| **Required Medical Information:** | **<u>Treatment Initiation</u>**<br>• A documented history of non-compliance, refusal to utilize oral medication, or cannot be stabilized on oral medications<br>• Documentation of established tolerability to oral risperidone (if risperidone-naïve)<br>• Requests for Perseris require documentation of treatment failure or clinical rationale for avoidance of Risperdal Consta<br><br>**<u>Continuation of Therapy</u>**<br>• Documentation showing that member is stable on current treatment with Perseris or Risperdal Consta |
| **Appropriate Treatment Regimen & Other Criteria:** | • Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Diagnosis of dementia-related psychosis<br>• Prior hypersensitivity reaction (anaphylactic reactions and/or angioedema) to paliperidone or risperidone |
| **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 subjects to utilization of the most cost-effective site of care |

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

POLICY NAME:
**LUMASIRAN**
Affected Medications: OXLUMO (lumasiran)

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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:** | <u>Requirements for Initial Authorization:</u><br>• Must have genetic testing confirming diagnosis of PH1 via presence of AGXT mutation<br><br>AND<br><br>• ONE of the following:<br>  ○ Elevated urine oxalate (Ox) excretion as measured by BSA-normalized daily UOx output greater than upper limit of normal<br>  ○ Elevated UOx excretion as measured by UOx: creatinine ratio above age-specific upper limit of normal.<br>  ○ Elevated plasma oxalate (POx) concentration (POx concentration greater than upper limit of normal)<br><br>Reference Value for Urinary Oxalate (UOx) Excretion in 24-hour urine samples:<br>  ○ All ages: less than 0.5 mmol/1.73 m2/day |
| **Appropriate Treatment Regimen & Other Criteria:** | • Oxlumo is supplied in 0.5 mL single-use vials containing 94.5 mg<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced.<br><br><u>Oxlumo Weight-Based Dosing</u><br>• Body weight less than 10 kg<br>  ○ Loading Dose: 6 mg/kg once monthly for 3 doses<br>  ○ Maintenance Dose: Start 1 month after last loading dose; 3 mg/kg once monthly<br><br>• Body weight between 10 kg to less than 20 kg<br>  ○ Loading Dose: 6 mg/kg once monthly for 3 doses |

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| | ○ Maintenance Dose: Start 1 month after last loading dose; 6 mg/kg once every 3 months<br><br>• Body weight 20 kg or greater<br>○ Loading Dose: 3 mg/kg once monthly for 3 doses<br>○ Maintenance Dose: Start 1 month after last loading dose; 3 mg/kg once every 3 months<br><br><u>Requirements for Reauthorization:</u><br>• Liver or kidney transplant has not occurred since previous authorization<br><br>AND<br><br>• ONE of the following criteria related to treatment success:<br>○ Must show reduction from baseline urine or plasma oxalate levels at 6 months.<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:** | • History of liver or kidney transplant.<br>• Genetic tests positive for other form of primary hyperoxaluria including type 2 and type 3 primary hyperoxaluria.<br>• Secondary hyperoxaluria |
| **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 nephrologist, urologist, geneticist, or physician specialized in the treatment of PH1 |
| **Coverage Duration:** | • Initial Authorization: 6 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>  ○ For the treatment of thrombocytopenia in adult patients with chronic liver disease who are scheduled to undergo a procedure |
| **Required Medical Information:** | • Complete blood count with differential and platelet count<br>• Liver function tests |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of planned procedure and baseline platelet count<br>• Dosing:<br>  ○ 3 mg orally once daily for 7 days beginning 8 to 14 days prior to scheduled procedure.<br>  ○ Patients should undergo their procedure 2-8 days after the last dose.<br>• Documented inability to respond adequately to Promacta<br><br>**Reauthorization** requires documented response to treatment with platelet count of at least 50,000/mcL without significant liver function abnormalities during procedure |
| **Exclusion Criteria:** | • Platelet count above 50,000/mcL at baseline<br>• A history of splenectomy, partial splenic embolization, or thrombosis and those with Child-Pugh class C liver disease, absence of hepatopetal blood flow, or a prothrombotic condition other than chronic liver disease |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with hematologist or gastroenterology/liver specialist |

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| **Coverage Duration:** | • Approval: 1 month (7 days of treatment), based on planned procedure date, unless otherwise specified |

POLICY NAME:
**MACRILEN**
Affected Medications: Macrilen (macimorelin acetate for oral solution 60mg)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | • Clinical context making growth hormone deficiency (GHD) likely<br>• Recent insulin-like growth factor-1 (IGF-1) level that is low for age/gender |
| **Appropriate Treatment Regimen & Other Criteria:** | • A documented history of seizure disorder or cardiovascular disease preventing the use of Insulin Tolerance Test (ITT) **AND**<br>• Inability to complete glucagon stimulation testing as a means of diagnosis<br>• Dosing: single oral dose of 0.5 mg/kg |
| **Exclusion Criteria:** | • Body Mass Index greater than 40 kg/m2 |
| **Age Restriction:** | • Adults at least 18 years of age |
| **Prescriber/Site of Care Restrictions:** | • Endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 1 month, unless otherwise specified |

POLICY NAME:
**MAKENA**
Affected Medications: MAKENA and Hydroxyprogesterone Caproate

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design.<br>• NCCN indications with evidence level of 2A or higher |
| **Required Medical Information:** | **<u>Oncology Indications</u>**<br>• Documentation of performance status, all prior therapies used, and prescribed treatment regimen. Consider holding therapy if Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater.<br>• Documentation of trial and failure prescription progesterone products (medroxyprogesterone, progestin-based therapies)<br>**<u>Preterm Labor Prevention</u>**<br>• Singleton pregnant patient<br>• History of singleton spontaneous preterm birth (less than 37 weeks)<br>• Expected date of delivery |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | • Current or history of any of the following:<br>o Multiple gestations or other risk factors for preterm birth<br>o Thrombosis or thromboembolic disorders<br>o Known or suspected breast cancer or other hormone-sensitive cancer, or history of these conditions<br>o Undiagnosed abnormal vaginal bleeding unrelated to pregnancy<br>o Cholestatic jaundice of pregnancy<br>o Liver tumors, benign or malignant, or active liver disease<br>o Uncontrolled hypertension |
| **Age Restriction:** | • 16 years of age or older |

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| **Prescriber/Site of Care Restrictions:** | • Oncology use: Oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Oncology: Initial, 4 Months. Reauthorization, 12 months.<br>• Preterm Labor Prevention: Approval: 21 weeks, unless otherwise specified |

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

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| 1. Is the request for add on maintenance therapy for Cystic Fibrosis? | Yes – Go to #2 | No – Criteria not met |
| 2. Is the diagnosis of Cystic Fibrosis (CF) confirmed by appropriate diagnostic or genetic testing?<br>a. Additional testing should include evaluation of overall clinical lung status and respiratory function (eg pulmonary function tests, lung imaging, etc.) | Yes – Go to #3 | No – Criteria not met |
| 3. Is there documentation that the Bronchitol Tolerance Test has been passed? | Yes – Go to #4 | No – Criteria not met |
| 4. Is the request for continuation of therapy currently approved through insurance? | Yes – Go to renewal criteria | No – Go to appropriate section below |
| **Indication: Add on maintenance therapy for Cystic Fibrosis** | | |
| 1. Is there documented failure of 6 months with twice daily hypertonic saline defined as one of the following despite at least 80% adherence with hypertonic saline:<br>a. Increase in pulmonary exacerbations from baseline?<br>b. Decrease in FEV1? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Will Bronchitol be used in conjunction with standard therapies for Cystic Fibrosis? | Yes – Approve up to 12 months | No – Criteria not met |
| **Renewal Criteria** | | |

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| 1. Is there documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider? | 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 |

POLICY NAME:
**MARALIXIBAT**
Affected Medications: LIVMARLI (Maralixibat)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design<br>• Cholestatic pruritus in patients with Alagille syndrome (ALGS) |
| **Required Medical Information:** | • Documentation of Alagille syndrome confirmed by:<br>  ○ Genetic test detecting a JAG1 or NOTCH2 mutation, or<br>  ○ Liver biopsy<br>• Documentation of patient's current weight<br>• Documentation of history of significant pruritus |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented failure with an adequate trial (at least 30 days) of all of the following: rifampin, ursodiol, AND cholestyramine<br><br>**Reauthorization**:<br>• Documented treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Decompensated cirrhosis<br>• History or presence of other concomitant liver disease (such as biliary atresia, liver cancer, non-PFIC related cholestasis)<br>• Prior liver transplant |
| **Age Restriction:** | • 1 year and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by a gastroenterologist or a specialist with experience in the treatment of 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<br>• Reauthorization: 12 months, unless otherwise specified |

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

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| **Covered Uses:** | • All 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 post-transplant CMV infection<br>• Documentation of patient's current weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented clinical failure (not due to drug intolerance) with an adequate trial (at least 14 days) of at least one of the following: ganciclovir, valganciclovir, cidofovir or foscarnet<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. |
| **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: 4 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 |
| **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:** | • **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:** | • 18 years or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by 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: 3 months<br>• Reauthorization: 12 months |

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><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:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**MECHLORETHAMINE**
Affected Medications: VALCHLOR (mechlorethamine hydrochloride)

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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:** | • Diagnosis of Stage IA or Stage IB mycosis fungoides-type cutaneous T-cell lymphoma<br>• Extent of skin involvement (limited/localized or generalized) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of all prior therapies used for the given indication<br>• Documentation of counseling on applicable special handling procedure<br><br>**<u>Limited/localized skin involvement</u>**<br>• Documentation of failure or contraindication of at least 1 topical retinoid (such as tretinoin 0.05%) AND topical corticosteroid<br><br>**<u>Generalized skin involvement</u>**<br>• Documentation of failure or contraindication to at least ≥1 skin-directed therapy (topical corticosteroids, topical retinoids, phototherapy, topical chemotherapy [e.g. carmustine], topical imiquimod, local radiation)<br><br>**Reauthorization:**<br>• Documentation of monitoring for non-melanoma skin cancer<br>• Documentation of improvement with treatment based either on CAILS score or decrease in severity of scaling, plaque elevation or surface area |
| **Exclusion Criteria:** | • Use in the management of onychomycosis,<br>• Treatment or prevention of vaginal or vulvovaginal candidiasis, |

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| | tinea cruris, tinea manuum, tinea pedis, tinea faciei, tinea capitis, tinea barbae, tinea corporis, tinea versicolor (pityriasis versicolor), or other superficial fungal infections.<br>• Coverage is not recommended for circumstances not listed in the Covered Uses. |
| **Age Restriction:** | • Age 18 years and older. |
| **Prescriber/Site of Care Restrictions:** | • Oncologist or Dermatologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**MEDICAL NECESSITY**
Affected Medications: Abilify MyCite, Abiraterone 500 mg tablet, Absorica, Absorica LD, Acanya, Aciphex, Actemra SQ, Acthar HP, Acuvail, Acyclovix, Aczone, Adapalene pads, Adcirca, Adlarity, Adlyxin, Admelog, Advicor, Adzenys ER, Adzenys XR, Aerospan, Afrezza, Aimovig, AirDuo, AirDuo Digihaler, Aklief, Allopurinol 200 mg tablet, Allzital, Alprazolam Dispersible, Alprazolam Intensol, Altoprev, Alvesco, Ameluz, Amitiza, Amphetamine ER suspension, Amrix, Amturnide, Amzeeq, Ancobon, Androgel, Androxy, Apadaz, APAP/Caff/Dihydrocodeine, Apidra, Aplenzin, Arazlo, Aripiprazole Dispersible, Armonair Digihaler, Armonair Respiclick, Arymo ER, Asacol HD (Mesalamine), Asmanex, Asmanex HFA, Aspruzyo, Astepro Solution, Auvelity, Auvi-Q, Aveed, Azathioprine tablet (75 mg, 100 mg), Azelex, Azesco, Azstarys, Basaglar, Baxdela, Beconase, Belbuca, Beser kit/lotion, Bevespi Aerophere, BiDil, Biifenac, Brexafemme, Breztri, Bridion, Briviact, Bryhali, Budesonide 9 mg ER tablet, Bunavail, Bupap, Bupropion XL 450 mg, Butisol, Butrans Patch, Bydureon, Bydureon BCise, Byetta, Bynfezia, Byvalson, Calcipotriene/Betamethasone Dipropionate Suspension, Cambia, Capex Shampoo, Capital/Codeine, Carac, Carbinoxamine 6 mg Tab, Carisoprodol/ASA, Carisoprodol/ASA/Codeine, CaroSpir, Carticel Implant, Cataflam, Cephalexin 750 mg capsule, Cephalexin tablet, Cequa, Chlorpheniramine/Codeine, Chlorzoxazone 250 mg tablet, Cibinqo, Cimzia, Ciprodex OTIC, Cipro HC Otic, Clemastine Syrup, Clindamycin Phosphate/Benzoyl Peroxide Gel 1.2-2.5%, Clindavix, Clobetex, Codar AR, Colazal, Conjupri, Consensi, Conzip, Coreg CR, Cosopt PF, Cotempla XR-ODT, Crinone, Cuprimine, Cuvposa, Cyclobenzaprine ER, Cyclosporine in Klarity, Dapsone 7.5% Gel, Dartisla ODT, Debacterol, Degludec, Delzicol, Demser, Denavir, Denavir Cream, DermacinRx Lexitral cream pack, Dermalid, Desonate Gel, Desonide Gel, Desonide Lotion, DesRx Gel, Dexilant, Dexlansoprazole DR, Dhivy, Diclofenac 1.3% Patch, Diclofenac Potassium Capsule, Diclofenac Potassium 25 MG Tablet, Diclofenac Sod Soln 1.5% & Capsaicin Cream 0.025% Ther Pack, Diclofex DC Cream, Diclopak, Diclosaicin Cream, Diclotral pack, Diclotrex, Diclovix DM Pak, 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, Econasil, Edarbi, Edarbyclor, Egaten, Egrifta, Elepsia XR, Elidel, Elyxyb, Emend, Enablex, Enalapril Oral Solution, Enstilar Foam, Entadfi, Epaned, Epanova, Epclusa, Eprontia, Equetro, Esbriet, Eskata, Evzio, Exjade, Exservan, Extavia, Extina foam 2%, Fabior foam, Fenofibrate 120 mg, Fenoprofen, Fenortho, First-lansoprazole, First-omeprazole, Flector Patch, Fleqsuvy, Flolipid, Flowtuss, Fluocinonide, Fluopar Kit, Fluorouracil 0.5% cream, Flurandrenolide, Forfivo XL, Fortamet, Fortesta GEL, Fosamax Plus D, Fulyzaq, Furoscix, Gabacaine Pak, Gabapal, Giazo, Gimoti, Glatiramer, Glatopa, Gleevec, Gloperba, Glumetza, Glycate, Glycopyrrolate 1.5 mg

tablet, Gocovri, Gonitro, GPL Pak, Halog, Halcinonide Cream, Harvoni, Harvoni Pak, Helidac, Hemady, Humalog, Humalog Junior KwikPen, Hemangeol, Humatin, Humulin, Humulin 70/30 KwikPen, Humulin N, Humulin R-100, Hycofenix, Ibsrela, Ibuprofen/Famotidine, Igalmi, Ilumya, Imbruvica 70 mg capsule, Imbruvica 140 mg & 280 mg tablet, Imiquimod 3.75%, Impeklo, Impoyz, Imvexxy, Inbrija, Indocin suppository, Indomethacin 20 mg capsule, Inflatherm Kit, Inflatherm Pak, Infugem, Ingrezza, Innolet Insulin, Insulin Aspart, Insulin Degludec, Insulin Glargine, Insulin Glargine-yfgn, Insulin Lispro, Intrarosa, Invokamet, Invokamet XR, Invokana, Isordil Titradose, Isotretinoin 25 mg and 35 mg capsule, Ivermectin tablet, Jadenu, Jadenu Sprinkle Packet, Jentadueto, Jentadueto XR, Jublia, Karbinal ER, Katerzia, Kazano, K-bicarb, Kenalog Aerosol, Kenalog Susp, Keragel, KeragelT, Kerendia, Kerydin, Kesimpta, Ketek, Ketorolac nasal spray, Keveyis, Kevzara, Kineret, Kisqali, Kisqali-Femara Co-Pak, Klisyri, Kombiglyze XR, Lampit, Lescol XL, Letairis, Levamlodipine, Levorphanol tartrate, Lexette, Lexuss, Lialda, Licart, Lido GB 300 Kit, Lidostream, Lidotin Pak, Lifems, Lipritin Pak, Liptruzet, Lithostat, LMR Plus Lidocaine, Lofena, Lonhala Magnair, Loreev XR, Lubiprostone, Lucemyra, Luzu, Lybalvi, Lyrica, Lyrica CR Tablet, Lyumjev, Lyumjev Kwikpen, Lyvispah, Meclofen, Meloxicam Capsule, Memantine ER capsule, Mentax Cream 1%, Metaclopramide, Metaxall, Metaxall CP, Metformin ER (mod), Metformin ER (OSM), Metformin Solution, Methadone Intensol, Methadose, Methamphetamine 5 mg Tablet, MethylTESTOSTERone Capsule, Metyrosine, Migraine Pack, Minocycline ER, Minolira, Mitigare, Monocycline ER, MorphaBond, MorphaBond ER, Motegrity, Motofen, Mounjaro, Mycapssa, Myfembree, Mytesi, Nalocet, Namenda XR, Namzaric, Naprelan, Naproxen-Esomeprazole, Nascobal, Natesto GEL, Neo-Synalar cream, Nesina, Nexletol, Nexlizet, Nitisinone, Nocdurna, Noctiva, Nolix, Nopioid TC Kit, Norgesic Forte, Noritate, Norliqva, Noroxin, Northera, Nourianz, Novolin 70/30 Relion, Novolin N Relion, Novolin R Relion, Novolog Relion, NuDiclo Solupak, Nurtec ODT, Nuvakaan Kit, Nuvakaan II Kit, Nuvigil, Nuzyra, Olumiant, Olysio, Omeprazole-Sodium Bicarb, Omnaris, Ondansetron 24 mg tablet, Onexton, Onfi, Onglyza, Onmel, Onzetra Xsail, Oracea, Oralair, Orencia SQ, Orphenadrine-Aspirin-Caffeine tablet, Orphengesic Forte, Ortikos, Oseni, Otrexup, Oxaydo, Oxycodone-Acetaminophen (2.5 mg-300 mg, 5 mg-300 mg, 7.5 mg-300 mg, 10 mg-300 mg), Ozobax, Pamelor, Panlor, Panretin gel, Paromomycin, Pazeo, Pedizolpak, Penicillamine Capsule 250 mg, Pennsaid Solution, Pentican Pak, Percocet, Pertzye, Pheburane, Picato, Pioglitazone-Glimepiride, Pirfenidone 534 mg tablet, Pradaxa, Praluent, Prevacid SoluTab, Prialt, Prilo Patch, Prilopentin, Primlev, Primsol, Pristiq, ProAir Digihaler, Prolate, Prudoxin, Purified Cortrophin Gel, Purixan, Qbrelis, Qbrexza, Qdolo, Qelbree, QilliChew ER, Qmiiz, QNASL, Qtern, Qudexy XR, Quillivant XR, Quinixil, Quinosone, Qulipta, Qwo, Ranexa, Rasuvo, Rayos, Recarbrio, Reditrex, Relexxii, Relion Insulins, Reltone, Retin-A Micro Pump Gel (0.06 %, 0.08 %), Reyvow, Rhofade, Rhopressa, Ribasphere, Ridaura, Riomet, Riomet ER, Rocklatan, Ryaltris, Ryvent, Ryzodeg 70/30, Sabril, Samsca, Sarafem, Savaysa, Seconal, Seebri Neohaler, Seglentis,

Segluromet, Semglee, Sensipar, Sernivo, Seysara, Siklos, Sila III Pak, Siliq Subcutaneous Injection, Simponi, Simvastatin Suspension, Skelaxin, Skelid, Soaanz, Soliqua, Solodyn, Solosec, Sorilux, Sotyktu, Sovaldi, Sovaldi Pak, Sporanox Solution, Spritam, Sprix, Steglatro, Steglujan, Striant, Striant BUCCAL, Stromectol, Sublocade, Suboxone, Sumatriptan-Naproxen, Sure Result DSS premium pack, Symbyax, Sympazan, Symproic, Synalar, Syndros, Syprine, Taclonex Suspension, Talicia, Taltz, Tanzeum, Targadox, Tasoprol, Tavaborole, Tazarotene Foam, Technivie, Thalitone, Thiola, Thiola EC, Thyquidity, Ticlopidine, Tiglutik, Tioptonin, Tivorbex, Tizanidine Capsule, Tolak, Tolsura, Tosymra, Tovet Kit, Tracleer, Tradjenta, Treximet, Tri-Luma, Trixylitral kit, Trokendi XR, Trudhesa, Trulance, Tudorza Pressair, Twyneo, Tyrvaya, Tyzeka, Tyzine, Ultravate, Ultresa, Uptravi, Utibron Neohaler, Valsartan oral solution, Vanatol LQ, Vanos, Varophen, Vasotec, Vecamyl, Vectical, Veltassa, Venlafaxine Besylate ER, Venlafaxine ER tablets, Veramyst, Veregen, Verkazia, Vesicare LS, Vexasyn, Vexasyn gel, Vfend oral suspension, V-Go, Viberzi, Vibramycin, Victrelis, Viekira, Vimovo, Viokace, Vivlodex, Vogelxo, Voquezna Dual Pak, Voriconazole oral suspension, Vtol LQ solution, Vyzulta, Wakix, Winlevi, Wynorza, Xaciato, Xadago, Xartemis XR, Xatmep, Xcopri, Xelitral Pack, Xelstrym, Xenleta, Xerese, Xermelo, Xhance, Ximino, Xtampza ER, Xultophy, Xyosted, Yosprala, Yupelri, Zanaflex capsule, Zcort, Zebutal, Zecuity, Zelnorm, Zembrace, Zenevix, Zepatier, Zetonna, Zileuton ER, Zinbryta, Zipsor, Zolpak, Zolpimist, Zonalon, Zonisade, Zorvolex, ZTLido, Z-Tuss, Zubsolv, Zyclara, Zypitamag, Zyprexa Relprevv, 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 compendia supported dosing. |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |

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| **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:
**MELPHALAN**
Affected Medications: EVOMELA (melphalan)

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | • Indication: palliative treatment for multiple myeloma: Not covered, use melphalan IV/oral<br>• Indication: high-dose conditioning prior to hematopoietic stem cell transplant (HSCT) for multiple myeloma<br>• Currently approved for HSCT and provide the tentative date of the stem cell transplant<br>• Weight: if patient weights more than 130% of ideal body weight, use ideal body weight for body surface area calculation<br>• Body surface area to determine dose (if patient weights more than 130% of ideal body weight, use ideal body weight) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Food and Drug Administration (FDA)-approved dosing by body surface area (100mg/m2) daily for 2 days on day -3 and day -2 prior to autologous stem cell transplantation on day 0 |
| **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:** | • Approval duration: 1 month (for 2 days treatment), unless otherwise specified |

POLICY NAME:
**MEPOLIZUMAB**
Affected Medications: NUCALA (mepolizumab)

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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?<br>○ Add-on maintenance treatment of patients with severe asthma aged 6 years and older with an eosinophilic phenotype<br>○ Treatment of adult patients with eosinophilic granulomatosis with polyangiitis (EGPA)<br>○ Treatment of patients aged 12 years and older with hypereosinophilic syndrome (HES)<br>○ Add-on maintenance treatment of chronic rhinosinusitis with nasal polyps (CRSwNP) in adult patients 18 years of age and older with inadequate response to nasal corticosteroids (NCS). | Yes – Go to appropriate section below | No – Criteria not met |
| **Severe Eosinophilic Asthma** | | |
| 1. Is there documentation of severe eosinophilic asthma defined by the following:<br>○ Baseline eosinophil count at least 150 cells/μL<br>**AND** | Yes – Document and go to #2 | No – Criteria not met |

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| ○ FEV1 less than 80% at baseline or FEV1/FVC reduced by at least 5% from normal | | |
| 2. Is there documented use of high-dose inhaled corticosteroid (ICS) plus a long-acting beta agonist (LABA) for at least three months with continued symptoms? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there a documented history of 2 or more asthma exacerbations requiring oral or systemic corticosteroid treatment in the past 12 months while on inhaled combination treatment and at least 80% adherence? | Yes – Go to #5 | No – Go to #4 |
| 4. Is there documentation that chronic daily oral corticosteroids are required? | Yes – Go to #5 | No – Criteria not met |
| 5. Is the drug prescribed by or in consultation with an Allergist, Immunologist, or Pulmonologist? | Yes – Approve up to 6 months | No – Criteria not met |
| **Eosinophilic granulomatosis with polyangiitis (EGPA)** | | |
| 1. Is there a confirmed diagnosis of relapsing or refractory eosinophilic granulomatosis with polyangiitis (EGPA) with the following:<br>○ Chronic rhinosinusitis<br>○ Asthma<br>○ Blood eosinophilia (at least 1,500 cells/microL and/or 10% eosinophils on differential) at baseline<br>○ Diagnosis must be confirmed by a | Yes – Document and go to #2 | No – Criteria not met |

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| second clinical opinion | | |
| 2. Is there documented relapsing disease while on the highest tolerated oral corticosteroid dose? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documentation that the disease has been refractory to at least two oral immunosuppressive drugs for at least 12 weeks each (Azathioprine, Methotrexate, Leflunomide)? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is the drug prescribed by a specialist in the treatment of eosinophilic granulomatosis with polyangiitis (EGPA) (immunologist or rheumatologist)? | Yes – Approve up to 6 months | No – Criteria not met |
| **Hypereosinophilic Syndrome** | | |
| 1. Is there documentation of hypereosinophilic syndrome (HES) with all of the following:<br>• Blood eosinophil count greater than 1000 cells/mcL<br>• Disease duration greater than 6 months<br>• At least 2 flares within the past 12 months<br>• Lab work showing Fip1-like1-platelet-derived growth factor receptor alpha (FIP1L1-PDGFRα) mutation negative disease<br>• Non-hematologic secondary HES has been ruled out (drug hypersensitivity, parasitic helminth infection, HIV | Yes – Document and go to #2 | No – Criteria not met |

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| infection, non-hematologic malignancy) | | |
| 2. Is the HES currently controlled using the highest tolerated glucocorticoid dose (defined as an improvement in clinical symptoms and a decrease in eosinophil count by at least 50% from baseline)? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documentation showing that the patient has a lymphocytic variant of HES (L-HES)? | Yes – Document and go to #5 | No – Go to #4 |
| 4. Is there documentation of treatment failure to at least 12 weeks of hydroxyurea? | Yes – Document and go to #5 | No – Criteria not met |
| 5. Is there documentation of treatment failure with interferon-alfa? | Yes – Document and go to #6 | No – Criteria not met |
| 6. Is the drug prescribed by a specialist for the treatment of HES (e.g., immunologist or hematologist)? | Yes – Approve up to 6 months | No – Criteria not met |
| **Chronic Rhinosinusitis with Nasal Polyps (CRSwNP)** | | |
| 1. Is there documentation of chronic sinusitis after total ethmoidectomy with a need for revision endoscopic sinus surgery due to continued symptoms of nasal congestion/obstruction from recurrent bilateral sinus obstruction due to nasal polyps? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documented failure with at least 1 intranasal corticosteroid (such as fluticasone) after ethmoidectomy? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documented failure with Sinuva implant? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is the drug prescribed by a specialist in the treatment of nasal polyps | Yes – Approve up to 6 months | No – Criteria not met |

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| (otolaryngologist)? | | |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider? | Yes – Go to #2 | No – Criteria not met |
| 2. Is the request for use in combination with another monoclonal antibody (Fasenra, Dupixent, Xolair, Cinqair)? | Yes – Criteria not met, combination use is experimental | No – Go to #3 |
| 3. 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** | | |

- **Nucala**
  - Availability: 100 mg/mL single-use vial or prefilled syringe or auto-injector
  - Dosing:
    - Severe asthma: 100 mg every 4 weeks for age 12 and up, 40 mg every 4 weeks for age 6 to 11
    - EGPA: 300 mg every 4 weeks
    - HES: 300 mg every 4 weeks
    - CRSwNP: 100 mg every 4 weeks

*Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced for all medical infusion drugs

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. |
| **Required Medical Information:** | • Documentation of treatment of opioid-induced constipation (OIC) in a patient with:<br>○ Advanced illness who is receiving palliative care **OR**<br>○ Chronic non-cancer pain who has taken opioids for at least 4 weeks<br>**AND**<br>• Trial and inadequate treatment response or contraindication to Movantik and Linzess for patients with OIC in non-cancer pain |
| **Appropriate Treatment Regimen & Other Criteria:** | • Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Known or suspected mechanical gastrointestinal 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 approval: 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 |
| **Required Medical Information:** | • Weight<br>• Baseline serum leptin levels, HbA1c, fasting glucose, fasting triglycerides, fasting serum insulin<br>• Prior Myalept use will require test of anti-metrepeptin antibodies |
| **Appropriate Treatment Regimen & Other Criteria:** | • Serum leptin < 6.0 ng/mL females and < 3.0 ng/mL males, obtained on at least 2 occasions<br>• If treating acquired generalized lipodystrophy with concurrent hypertriglyceridemia defined as triglycerides ≥ 500 mg/dL despite optimizing with statin and/or fibrate<br>• If treating acquired generalized lipodystrophy with concurrent diabetes, baseline HbA1c ≥ 7% despite optimal treatment with metformin, TZD, sulfonylurea, GLP-1 agonist or DPP-4 inhibitor, SGLT-2, and insulin<br>• Treatment success defined by improvement in HbA1c, fasting glucose, and fasting triglycerides<br>• Worsening metabolic control and/or severe infection: indicators of possible anti-metreleptin antibodies<br>• Maximum daily dose for individuals <40 kg: 0.13 mg/kg<br>• Maximum daily dose for individuals >40 kg: 10 mg/day<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **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 evidence of generalized lipodystrophy |
| **Age Restriction:** | • Age ≥ 1 year |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an Endocrinologist<br>• Myalept is available only through the MYALEPT REMS Program |

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| | • All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial: 4 months, unless otherwise specified<br>• Subsequent: 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<br>**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) |

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| | <u>Reauthorization criteria – 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 or older - for Paget's disease of bone only |
| **Prescriber/Site of Care Restrictions:** | |
| **Coverage Duration:** | • Approval: 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 |
| **Required Medical Information:** | • Current weight<br>• Documentation of Visceral leishmaniasis **OR** Cutaneous leishmaniasis **OR** Mucosal leishmaniasis |
| **Appropriate Treatment Regimen & Other Criteria:** | • Food and Drug Administration (FDA)-approved dosing of 30 to 44 kg: one 50 mg capsule twice daily for 28 consecutive days OR 45 kg or greater: one 50 mg capsule three times daily for 28 consecutive days<br>• Weight equal to or greater than 30kg (66lbs) |
| **Exclusion Criteria:** | • Pregnancy (category D)<br>• Sjögren-Larsson-Syndrome |
| **Age Restriction:** | • Age less than 12 years of age<br>• Weight less than 30 kg (66 lbs) |
| **Prescriber/Site of Care Restrictions:** | • Infectious Disease Specialist<br>• All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 1 month unless otherwise specified |

POLICY NAME:

**MITAPIVAT**

Affected Medications: PYRUKYND (mitapivat tablet)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design<br>  ○ Hemolytic anemia |
| **Required Medical Information:** | • Diagnosis of pyruvate kinase deficiency (PKD), defined as documented presence of at least 2 mutant alleles in the pyruvate kinase liver and red blood cell (PKLR) gene, of which at least 1 is a missense mutation<br><br>**AND**<br>• If receiving regular transfusions, documentation of both of the following:<br>  ○ A minimum of 6 transfusion episodes in the 12-month period prior to treatment<br>  ○ Baseline transfusion amount, including date of transfusion and number of red blood cell (RBC) units transfused<br><br>**OR**<br>• If not receiving regular transfusions, documentation of both of the following:<br>  ○ No more than 4 transfusions in the 12-month period prior to treatment and no transfusions in the 3-month period prior to treatment<br>  ○ Baseline hemoglobin (Hb) AND must be less than or equal to 10 g/dL |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization:** documentation of treatment success and a clinically significant response to therapy, defined as:<br>• For patients receiving regular transfusions at baseline: must document greater than or equal to a 33% reduction in RBC units transfused compared to baseline<br>• For patients not receiving regular transfusions at baseline: must document greater than or equal to a 1.5 g/dL increase in Hb from baseline sustained at 2 or more scheduled visits AND no transfusions were needed<br>• Discontinue therapy after 6 months if no benefit in transfusion |

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<td>requirement or Hb has been observed<br><br>• Dose: Approve 5 mg, 20 mg, and 50 mg tablets (QL of 56 per 28 days) per dosing schedule below<br><br>Table 1: Dose Titration Schedule<br><table><tr><th>Duration</th><th>Dosage</th></tr><tr><td>Week 1 through Week 4</td><td>5 mg twice daily</td></tr><tr><td>Week 5 through Week 8</td><td>If Hb is below normal range or patient has required a transfusion within the last 8 weeks:<br>• Increase to 20 mg twice daily and maintain for 4 weeks.<br>If Hb is within normal range and patient has not required a transfusion within the last 8 weeks:<br>• Maintain 5 mg twice daily.</td></tr><tr><td>Week 9 through Week 12</td><td>If Hb is below normal range or patient has required a transfusion within the last 8 weeks:<br>• Increase to 50 mg twice daily and maintain thereafter.<br>If Hb is within normal range and patient has not required a transfusion within the last 8 weeks:<br>• Maintain current dose (5 mg twice daily or 20 mg twice daily).</td></tr><tr><td>Maintenance</td><td>If Hb decreases, consider up-titration to the maximum of 50 mg twice daily as per the above schedule.</td></tr></table></td>
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<td>Exclusion Criteria:</td>
<td>• Homozygous for the c.1436G>A (p.R479H) variant or have 2 non-missense variants (without the presence of another missense variant) in the PKLR gene<br>• Splenectomy scheduled during treatment or have undergone within the 12-month period prior to starting treatment<br>• Previous bone marrow or stem cell transplant<br>• Receiving hematopoietic stimulating agents or anabolic steroids (including testosterone preparations) within 28 days prior to treatment</td>
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| **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:
**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. |
| **Required Medical Information:** | • Documentation of chronic sinusitis status post total ethmoidectomy indicated for revision endoscopic sinus surgery due to continued symptoms of nasal congestion/obstruction from recurrent bilateral sinus obstruction due to sinonasal polyposis |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of failure with at least 1 intranasal corticosteroid after ethmoidectomy |
| **Exclusion Criteria:** | • History of previous Sinuva implant use<br>• Known history of resistant or poor response to oral steroids<br>• Acute bacterial or invasive fungal sinusitis<br>• Immune deficiency (including cystic fibrosis) |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Otolaryngologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 1 month, unless otherwise specified<br>• Reauthorization: Not eligible, There are no studies evaluating repeat implantation of the SINUVA Sinus Implant |

POLICY NAME:
**MONOMETHYL FUMARATE**
Affected Medications: BAFIERTAM (monomethyl 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), to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease, in adults |
| **Required Medical Information:** | • Diagnosis of relapsing or active secondary progressive forms of Multiple Sclerosis (MS) confirmed with MRI (Revised McDonald diagnostic criteria for multiple sclerosis)<br>  ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS<br>• Complete blood count with lymphocyte count, and liver function tests (within 6 months) before initiating treatment, then CBC annually and as clinically indicated |
| **Appropriate Treatment Regimen & Other Criteria:** | • Initial dose of 95 mg twice daily for 7 days, then increasing to 190 mg twice daily thereafter<br>• Hold therapy for four weeks if lymphocyte count is less than 500/mm3 for greater than 6 months<br>• No concurrent use of disease-modifying therapy for the treatment of multiple sclerosis<br>• Not approved for primary progressive multiple sclerosis<br>• Reauthorization: provider attestation of treatment success |
| **Exclusion Criteria:** | • Pre-existing low lymphocyte counts (less than 500/mm3) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or after consultation with a neurologist or MS specialist |
| **Coverage Duration:** | • Approval: 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), and 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, FULPHILA, 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)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br><br>**Neupogen, Nivestym, Releuko & Zarxio**<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 Acute Myeloid Leukemia Receiving Induction or Consolidation Chemotherapy</u>**<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>**<u>Patients with Cancer Receiving Bone Marrow Transplant</u>**<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>**<u>Patients Undergoing Autologous Peripheral Blood Progenitor Cell Collection and Therapy</u>**<br>• Indicated for the mobilization of autologous hematopoietic progenitor cells into the peripheral blood for collection by leukapheresis. |

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| | **<u>Patients With Severe Chronic Neutropenia</u>**<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>**Leukine**<br><br>**<u>Use Following Induction Chemotherapy in Acute Myelogenous Leukemia</u>**<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.<br>**<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. Mobilization allows for the collection of increased numbers of progenitor cells capable of engraftment as compared with collection without mobilization. After myeloablative chemotherapy, the transplantation of an increased number of progenitor cells can lead to more rapid engraftment, which may result in a decreased need for supportive care. Myeloid reconstitution is further accelerated by administration of Leukine following peripheral blood progenitor cell transplantation.<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>**<u>Use in Myeloid Reconstitution After Allogeneic Bone Marrow Transplantation</u>**<br>• Indicated for acceleration of myeloid recovery in patients undergoing allogeneic BMT from HLA-matched related donors.<br>**<u>Use in Bone Marrow Transplantation Failure or Engraftment Delay</u>** |

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| | • Indicated in patients who have undergone allogeneic or autologous BMT in whom engraftment is delayed or has failed. Fulphila & Udenyca<br><br>**Neulasta, Fulphila, Udenyca, Ziextenzo, Nyvepria, Fylnetra, 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)</u>**<br>• Indicated to increase survival in patients acutely exposed to myelosuppressive doses of radiation<br>• Not indicated for the mobilization of peripheral blood progenitor cells for hematopoietic stem cell transplantation<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 (Neupogen/Granix/Zarxio/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 |

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| | ○ Neutropenia related to HIV/AIDS<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<br>• Documentation of therapy intention (curative, palliative) for prophylaxis of febrile neutropenia<br>• Documentation of risk factors both medication therapy regimen and patient specific<br>• Documentation of planned treatment course<br>• Documentation of current patient weight |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Filgrastim products:</u>**<br><br>**When requested via the MEDICAL benefit:**<br>Coverage for the non-preferred products, Neupogen, 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, 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:</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</u>**<br><br>Coverage for the non-preferred products, Neulasta, Fylnetra, and |

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| | Nyvepria is provided when the member meets the following criteria:<br>• Documented treatment failure or intolerable adverse event to Ziextenzo, Fulphila and Udenyca<br><br>**<u>Eflapegrastim product</u>**<br><br>Coverage for the non-preferred product, Rolvedon, is provided when the member meets the following criteria:<br>• Documented treatment failure or intolerable adverse event to the preferred Pegfilgrastim products (Ziextenzo, Fulphila, and Udenyca)<br><br>For prophylaxis of febrile neutropenia (FN) or other dose-limiting neutropenic events for patients receiving myelosuppressive anticancer drugs:<br>• Meets one of the following:<br>○ **Curative Therapy**: High risk (greater than 20% risk) OR intermediate risk (10-20% risk) for febrile neutropenia based on chemotherapy regimen with documentation of significant risk factors for serious medical consequences OR has experienced a dose-limiting neutropenic event on a previous cycle of current chemotherapy to be continued<br>○ **Palliative Therapy**: Myeloid growth factors will not be approved upfront for prophylaxis of febrile neutropenia in the palliative setting. Per the NCCN, 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>For Treatment of Severe Chronic Neutropenia,<br>• Must meet **<u>ALL of</u>** the following:<br>○ Congenital neutropenia, cyclic neutropenia, OR idiopathic neutropenia<br>○ Current documentation of ANC less than 500 cells/microL<br>○ Neutropenia symptoms (fever, infections, oropharyngeal ulcers) |

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| | ○ CBC with differential and platelet counts, bone marrow morphology, and karyotype |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by oncologist, hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 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 in children of both sexes<br>  ○ Management of endometriosis |
| **Required Medical Information:** | **<u>Central Precocious Puberty</u>**<br>• Documentation of central precocious puberty (CPP) confirmed by basal luteinizing hormone (LH), follicle-stimulating hormone (FSH), and either estradiol or testosterone concentrations<br><br>**<u>Endometriosis</u>**<br>• Documentation of moderate to severe pain due to endometriosis |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Endometriosis</u>**<br>• Documentation of a trial and inadequate relief (or contraindication) after at least three months of first-line therapy with nonsteroidal anti-inflammatory drugs (NSAIDs) and continuous (no placebo pills) hormonal contraceptives<br>• Maximum treatment duration 6 months total<br>  ○ Retreatment is not recommended |
| **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): age 11 or younger (females), age 12 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:** | Authorization:<br>• Endometriosis (no reauthorization): 6 months, 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. |
| **Required Medical Information:** | • Documentation of treatment of opioid-induced constipation in a patient with chronic non-cancer pain who have taken opioids for at least 4 weeks.<br>**AND**<br>• Trial and inadequate treatment response or contraindication to polyethylene glycol 3350 (PEG 3350) and at least one other laxative. |
| **Appropriate Treatment Regimen & Other Criteria:** | **Dosing:**<br>• Discontinue if opioid pain medication is also discontinued<br><br>Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Known or suspected mechanical gastrointestinal obstruction.<br>• Concomitant use of strong CYP3A4 inhibitors (e.g. clarithromycin, ketoconazole) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 6 months, unless otherwise specified |

POLICY NAME:
**NATALIZUMAB**
Affected Medications: TYSABRI (natalizumab)

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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), to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease, in adults.<br>  ○ Crohn's Disease (CD) |
| **Required Medical Information:** | • Documentation of prior treatments<br>• Screening for seropositivity for anti-JC virus antibodies prior to Tysabri therapy<br><br>**<u>Adults with Multiple Sclerosis (MS)</u>**<br>• Documentation of diagnosis of relapsing forms of multiple sclerosis confirmed with magnetic resonance imaging (Revised McDonald diagnostic criteria for MS)<br>  ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS<br>• **<u>Clinically Isolated Syndrome (CIS)</u>**<br>  ○ Documentation of CIS as shown by patients who do not fulfill McDonald criteria for a diagnosis of MS but have an abnormal brain MRI with one or more hyperintense T2 lesions that are characteristic of MS in at least two of four MS-typical regions at presentation or within three to six months of the event<br>• **<u>Secondary-Progressive MS (SPMS)</u>**<br>  ○ Documentation of prior history of relapsing-remitting MS (RRMS) with progressive increase in disability over at least 6 months, independent of, or in the absence of, relapses<br>  ○ Documentation of active disease classified as the presence of clinical relapse or inflammatory activity (i.e. new or enlarging T2 lesions or gadolinium enhancing lesions on MRI) in the last 2 years |

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| | ○ Documentation of Expanded Disability Status Scale (EDSS) score of 3.0 to 6.5<br><br>**<u>Adults with Crohn's disease (CD)</u>**<br>• Patient has moderately to severely active disease despite current treatment |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>All Uses</u>**<br>• Dosing: 300 mg IV every four weeks.<br>• **Reauthorization for patients with baseline positive JCV:** documentation of response to therapy and periodic MRI to monitor for PML occurrence<br><br>**<u>Adults with 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>Adults with CD</u>**<br>• Patient has moderately to severely active CD with evidence of inflammation (e.g., elevated C-reactive protein)<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 **AND**<br>• Documented clinical failure with at least 12 weeks of infliximab (Inflectra, Renflexis)<br>• Discontinue in patients that have not experienced therapeutic benefit by 12 weeks of induction therapy, and in patients that cannot discontinue chronic concomitant steroids within 6 months of starting therapy. |
| **Exclusion Criteria:** | • Concurrent use of medications indicated for the treatment of RRMS<br>• A diagnosis of Primary Progressive Multiple Sclerosis (PPMS) |

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| | • Concurrent or combined treatment with multiple targeted immune modulators (such as Humira, Stelara, infliximab or Entyvio)<br>• History of Progressive Multifocal Leukoencephalopathy (PML) |
| **Age Restriction:** | • Adutls aged 18 and older |
| **Prescriber/Site of Care Restrictions:** | • MS: prescribed by or in consultation with a neurologist<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 |
| **Coverage Duration:** | **<u>Adults with MS:</u>**<br>• Approval: 12 months, unless otherwise specified.<br><br>**<u>Adults with CD:</u>**<br>• Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**NAXITAMAB**
Affected Medications: DANYELZA (naxitamab)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>○ Treatment of relapsed or refractory high-risk neuroblastoma in the bone or bone marrow in patients who have demonstrated a partial response, minor response, or stable disease to prior 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 prescribed dosing regimen.<br>• Diagnosis of neuroblastoma as defined per the International Neuroblastoma Response Criteria (INRC):<br>○ An unequivocal histologic diagnosis from tumor tissue by light microscopy [with or without immunohistochemistry, electron microscopy, or increased urine (or serum) catecholamines or their metabolites]<br>OR<br>○ Evidence of metastases to bone marrow on an aspirate or trephine biopsy with concomitant elevation of urinary or serum catecholamines or their metabolites<br>• Evidence of high-risk neuroblastoma, including:<br>○ Stage 2/3/4/4S disease with amplified MYCN (any age)<br>○ Stage 4 disease in patients greater than 18 months of age<br>• Disease is evaluable in the bone and/or bone marrow, as documented by histology and/or appropriate imaging [e.g. metaiodobenzylguanidine (MIBG) scan]<br>• Documented history of previous treatment with at least one systemic therapy to treat disease outside of the bone or bone marrow |
| **Appropriate Treatment Regimen & Other Criteria:** | • Must be used in combination with granulocyte-macrophage colony-stimulating factor (GM-CSF).<br><br>**<u>Dosing</u>**:<br>• Availability: 40 mg/10 mL single-dose vial |

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| | • 3 mg/kg/day (maximum: 150 mg/day) on days 1, 3, and 5 of each treatment cycle (in combination with GM-CSF). One treatment cycle is 4 or 8 weeks.<br><br>Reauthorization will require documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• Patients with progressive disease |
| **Age Restriction:** | • 1 year of age or older |
| **Prescriber/Site of Care Restrictions:** | • Must be prescribed by or in consultation with a hematologist/oncologist with expertise in neuroblastoma<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:
**NILOTINIB**
Affected Medications: TASIGNA (nilotinib)

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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-positive mutation, BCR-ABL1 positive mutation |
| **Appropriate Treatment Regimen & Other Criteria:** | • For patients with low-risk score, documented clinical failure with Imatinib<br><br>**Reauthorization** requires documentation of treatment success (as applicable, BCR-ABL1 transcript levels, cytogenetic response) |
| **Exclusion Criteria:** | • Karnofsky Performance Status ≤50% or ECOG performance score ≥3<br>• Hypokalemia, hypomagnesemia, or long QT syndrome |
| **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:
**NIRAPARIB**
Affected Medications: ZEJULA (niraparib)

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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, disease staging, all prior therapies used, and anticipated treatment course<br>• Confirmed dignosis of ovarian, fallopian tube, or primary peritoneal cancer |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Maintenance therapy post primary treatment</u>**<br>• Documentation of platinum-sensitive disease prior to surgical resection<br>• Documentation of imaging results<br>• Documentation of BRCA mutation status<br>  ○ If mutation is present or suspected, documented intolerable adverse event to Lynparza<br>  ○ If mutation not present, niraparib must be preferred agent<br><br>**<u>Maintenance therapy for recurrent disease</u>**<br>• Documentation of platinum-sensitive disease<br>• Documented intolerable adverse event to the preferred product Lynparza<br><br>**<u>Treatment for disease progression</u>**<br>• Documentation of a deleterious or suspected deleterious BRCA mutation<br>  ○ If mutation is present or suspected, documented intolerable adverse event to Lynparza<br>OR<br><br>• Documentation of homologous recombination deficiency (HRD) positive status defined by:<br>  ○ Genomic instability and who have progressed more than six months after response to the last platinum-based chemotherapy, AND<br>  ○ No deleterious or suspected deleterious BRCA mutation |

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| | Reauthorization: documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• Clinical failure or progression on a previous PARP inhibitor |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • 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:
**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), TRIVISC (Sodium hyaluronate intra-articular injection), 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 | | |
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| 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
- Trivisc: 3 injections per course
- Visco-3: 3 injections per course

POLICY NAME:
**NOXAFIL**
Affected Medications: NOXAFIL (posaconazole), posaconazole

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Susceptibility cultures matching posaconazole activity<br>• Current body weight (for pediatric patients) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Treatment of invasive aspergillosis</u>**<br>• Documentation of resistance (or intolerable adverse event) to voriconazole<br><br>**<u>Prophylaxis of invasive Aspergillus and Candida infections</u>**<br>• Documentation of severely immunocompromised state, such as hematopoietic stem cell transplant (HSCT) recipients with graft versus-host disease (GVHD) or those with hematologic malignancies with prolonged neutropenia from chemotherapy<br>• Documentation of resistance (or intolerable adverse event) to one other compendia-supported systemic agent (e.g. fluconazole, itraconazole, voriconazole)<br><br>**<u>Treatment of oropharyngeal candidiasis (OPC):</u>**<br>• Documented failure (or intolerable adverse event) to 10 days or more of treatment with all of the following:<br>o Fluconazole<br>o Itraconazole |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • Posaconazole delayed release tablets – 2 years of age and older, who weigh greater than 40kg<br>• Noxafil oral suspension –13 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:** | • Approval: 6 months, unless otherwise specified |

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: amyotrophic lateral sclerosis (ALS), extrapyramidal and cerebellar disorders (Parkinson's disease, multiple system atrophy, progressive supranuclear palsy), multiple sclerosis (MS), traumatic brain injury, Alzheimer's disease and other dementias, or 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:** | • Using for non-FDA approved or compendia supported indications |
| **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 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. |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Presumptive diagnosis of MoCD Type A can be based on any one of the following</u>**:<br>• Family history<br>  ○ Affected siblings 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 (usually appear within the first 28 days after birth but can also present later):<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>**<u>Genetic confirmation using a panel which includes MOCS1 to confirm MoCD Type A</u>**:<br>• In patients with a presumptive diagnosis of MoCD Type A, the diagnosis must be confirmed immediately using a genetic test<br><br>**<u>Dosing</u>**:<br>• Available as: 9.5 mg single-dose vial for reconstitution.<br>• Administered via intravenous (IV) infusion. |

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| | • One year of age or older: 0.9 mg/kg (based on actual body weight) once daily.<br>• Less than one year of age (by gestational age): dosing is based on actual body weight and should be titrated to the target dose of 0.9 mg/kg/day over a period of 3 months Please refer to label instructions for titration schedule.<br><br>**<u>Reauthorization</u>**:<br>• Documentation of clinically significant response to therapy as determined by prescribing physician<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:** | • All approvals are subject to utilization of the most cost-effective site of care<br>• Prescribed by or in consultation with one of the following: neonatologist, pediatrician, pediatric neurologist, neonatal neurologist, or geneticist. |
| **Coverage Duration:** | • Initial Authorization: 1 month, unless otherwise specified<br>• Reauthorization: 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 |
| **Required Medical Information:** | |
| **Appropriate Treatment Regimen & Other Criteria:** | • Diagnosis of Parkinson's disease (PD) **AND**<br>• Presence of psychotic symptoms: hallucinations and/or delusions described as severe and frequent that started after the PD diagnosis **AND**<br>• Failure or contraindication to 30-day trial with quetiapine fumarate tablets<br>• Reauthorization requires documentation of treatment success and 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:** | • 12 months, unless otherwise specified |

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

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| **Covered Uses:** | • All FDA approved indications not otherwise excluded by benefit design<br>  ○ Spinal Muscular Atrophy |
| **Required Medical Information:** | • Patient must have documentation of a confirmed diagnosis of spinal muscular atrophy (SMA) type 1, 2, or 3 defined as ONE of the following genetic tests of 5q13 demonstrating:<br>  ○ Homozygous SMN1 gene deletion OR<br>  ○ Homozygous SMN1 gene mutation OR<br>  ○ Compound heterozygous SMN1 gene mutation<br>• Patient has at least 2 or more copies of the SMN2 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 ventilator use status<br>  ○ Is the patient ventilator dependent (using it 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 |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with or intolerable adverse event on Evrysdi<br>• Loading dose: 12 mg once every 14 days for 3 doses; then 12 mg once 30 days after the third dose<br>• Maintenance dose: 12 mg once every 4 months<br>**Reauthorization**: documentation of clinically significant improvement from baseline motor function demonstrated by:<br>  ○ Improvement from baseline motor function score documented within one month of renewal request AND<br>  ○ More areas of motor function improved than worsened<br>  ○ HINE-2:<br>  ▪ at least a 2-point increases in ability to kick OR |

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| | ▪ at least a 1-point increase in the motor milestones of head control, rolling, sitting, crawling, standing or walking using Section 2 of the Hammersmith Infant Neurologic Exam (HINE) AND<br>▪ More areas of motor function improved than worsened<br>○ Hammersmith Functional Motor Scale (HFSME)<br>▪ At least 3 points increase in score from pretreatment baseline AND<br>▪ More areas of motor function improved than worsened<br>○ Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)<br>▪ At least a 4 point increase in score from the pretreatment baseline AND<br>▪ More areas of motor function improved than worsened<br>○ Upper Limb Module (ULM)<br>▪ At least a 3 point increase from pretreatment baseline<br>○ 6-Minute Walk Test (6MWT)<br>▪ At least a 30 meter increase from pretreatment baseline |
| **Exclusion Criteria:** | • SMA type 4<br>• Ventilator dependent (using at least 16 hours per day on at least 21 of the last 30 days)<br>○ Does not apply to patients who require non-invasive ventilator assistance<br>• Prior treatment with Zolgensma (AVXS-101) |
| **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 pediatric neurologist or provider who is experienced in treatment of spinal muscular atrophy |
| **Coverage Duration:** | • Initial approval: 5 doses to be administered in a 6 month period, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**OBETICHOLIC ACID**
Affected Medications: OCALIVA (obeticholic acid)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Liver function tests (including alkaline phosphatase and bilirubin)<br>• Child-Pugh score<br>• Lipid profile |
| **Appropriate Treatment Regimen & Other Criteria:** | • The patient has an alkaline phosphatase level (ALP) at least 1.67 times the upper limit of normal (ULN) and/or bilirubin above the upper limit of normal while on ursodiol (with demonstrated adherence) after at least 12 months of therapy or has demonstrated a clinical inability to tolerate ursodiol<br>  ○ ULN ALP defined as 118 U/L for females or 124 U/L for males<br>  ○ ULN of total bilirubin defined as 1.1 mg/dL for females or 1.5 mg/dL for males<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>• Compensated cirrhosis with evidence of portal hypertension (e.g., ascites, gastroesophageal varices, persistent thrombocytopenia) |
| **Age Restriction:** | • 18 years and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by hepatologist<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:
**OCRELIZUMAB**
Affected Medications: OCREVUS (ocrelizumab)

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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 primary progressive multiple sclerosis (MS) in adults, relapsing forms of MS, to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease, in adults<br>• Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS |
| **Appropriate Treatment Regimen & Other Criteria:** | • Relapsing forms of MS: Documented disease progression or intolerable adverse event with rituximab (Authorization required, biosimilar products preferred)<br>• Primary Progressive MS (PPMS): Documentation of at least one year of disease progression and Baseline Expanded Disability Status Scale (EDSS) of 3-6.5<br>• Initial dose: 300 mg infusion followed two weeks later by a second 300 mg infusion<br>• Subsequent doses: Single 600 mg infusion every 6 months<br>• Reauthorization:<br>  ○ Relapsing forms of MS - Documentation of treatment success<br>  ○ PPMS- Documentation of treatment success as determined by treating provider (based on clinical and MRI findings)<br>  ○ Lack of disability progression (progression defined as when the EDSS score increased by 1 point or more from the baseline EDSS if the baseline EDSS was 5.5 points or less, or by 0.5 points or more if the baseline EDSS was more than 5.5 points) |
| **Exclusion Criteria:** | • Use outside of the Food and Drug Administration (Food and Drug Administration (FDA))-approved indications of relapsing or primary progressive forms of Multiple Sclerosis (MS)<br>• Active HBV infection |

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| | • Use with any other disease-modifying therapy for Multiple Sclerosis |
| **Age Restriction:** | • Safety and effectiveness of Ocrevus in pediatric patients have not been established<br>• Clinical studies of Ocrevus did not include sufficient numbers of subjects aged 65 and over to determine whether they respond differently from younger subjects |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or after consultation with a neurologist or an MS specialist.<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 6 months (2 initial infusions as noted above), unless otherwise specified<br>• Reauthorization: 12 months (2 infusions as noted above), unless otherwise specified |

POLICY NAME:
**ODEVIXIBAT**
Affected Medications: BYLVAY (odevixibat)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design<br>  ○ Pruritus due to progressive familial intrahepatic cholestasis (PFIC) |
| **Required Medical Information:** | • 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>• Documentation of patient's current weight<br>• Documentation of history of significant pruritus |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented failure with an adequate trial (at least 30 days) of all of the following: rifampin, ursodiol, AND cholestyramine<br><br>**<u>Reauthorization</u>**:<br>• Documented treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Prior hepatic decompensation events<br>• Concomitant liver disease (e.g., biliary atresia, liver cancer, non-PFIC related cholestasis)<br>• INR greater than 1.4<br>• ALT or total bilirubin greater than 10-times the upper limit of normal (ULN)<br>• Prior liver transplant |
| **Age Restriction:** | • 3 months and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by a hepatologist or a specialist with experience in the treatment of PFIC<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:
**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 |
| **Required Medical Information:** | • Documentation of nicotine use.<br>• If active nicotine user, documentation risks have been reviewed<br>• including decreased efficacy of therapy<br>• Documentation of a pregnancy test in females of reproductive potential prior to initiating treatment with nintedanib<br>• Documentation of baseline liver function tests in all patients, at regular<br>• intervals during the first three months, then periodically<br>• thereafter or as clinically indicated<br><br>AND<br>• Documentation of diagnosis of idiopathic pulmonary fibrosis<br>• Presence of usual interstitial pneumonia (UIP) on high resolution<br>• computed tomography (HRCT), and/or surgical lung biopsy AND<br>• Documentation of baseline forced vital capacity (FVC) greater<br>• than or equal to 50% of the predicted value AND<br>• Documentation of predicted diffuse capacity for carbon<br>• monoxide (DLCO) greater than or equal to 30%<br><br>OR<br>• Documentation of diagnosis of Systemic Sclerosis-Associated Interstitial Lung Disease from the American College of Rheumatology / European League Against Rheumatism classification criteria<br>• Documentation of onset of disease (first non-Raynaud symptom) of less than 7 years AND<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 forced vital capacity (FVC) greater than or equal to 40% of predicted<br>• Documentation of predicted diffuse capacity for carbon monoxide (DLCO)30-89% of predicted]<br>OR |

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| | • Documentation of a diagnosis of chronic fibrosing interstitial lung diseases with a progressive phenotype<br>• Documentation of relevant fibrosis (greater than 10% fibrotic features) on chest high resolution computed tomography (HRCT) scan with clinical signs of progression (defined as Forced Vital capacity (FVC) decline at least 10%, Forced Vital capacity (FVC) decline at least 5% with worsening symptoms and/or imaging in the previous 24 months and<br>• Forced Vital capacity (FVC) greater than or equal to 45% of predicted and<br>• Diffuse capacity for carbon dioxide (DLCO) 30% to less than 80% of predicted |
| **Appropriate Treatment Regimen & Other Criteria:** | • Pregnancy should be avoided while on Ofev and for at least 3 months after the last dose.<br>• Treatment of patients with moderate (Child Pugh B) and severe (Child Pugh C) hepatic impairment with OFEV is not recommended.<br>• The safety, efficacy, and pharmacokinetics of nintedanib have not been studied in patients with severe renal impairment (less than 30 mL/min CrCl) and end-stage renal disease.<br>• Reauthorization requires documentation of treatment success |
| **Exclusion Criteria:** | • Documentation of airway obstruction (such as pre-bronchodilator FEV/FVC less than 0.7)<br>• Concomitant administration of moderate or strong CYP3A4 and P-gp inhibitors / inducers should be avoided while taking Ofev<br>• Transaminases more than 5 times the upper limit of normal or elevated transaminases accompanied by symptoms (jaundice, hyperbilirubinemia).<br>• Ofev is not approved for use in combination with Esbriet |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with a pulmonologist |
| **Coverage Duration:** | • Initial approval: 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 acid sphingomyelinase deficiency (ASMD) in adult and pediatric patients |
| **Required Medical Information:** | • Documentation of acid sphingomyelinase deficiency as evidenced by<br>  ○ Enzyme assay showing diminished (less than 10% of controls) or absent acid sphingomyelinase activity (ASM)<br>  ○ Gene sequencing showing biallelic pathogenic SMPD1 mutation<br>  ○ Documentation of clinical presentation (e.g., hepatosplenomegaly, interstitial lung disease, liver fibrosis, growth restriction of childhood) outside the central nervous system<br>• Documentation of baseline measures of affected systems: (examples below)<br>  ○ Lungs: Diffusion capacity of lungs (Dlco) and pulmonary function tests (PFT)<br>  ○ Liver and spleen: volume, liver function tests, imaging<br>  ○ Bones: platelet counts, z-score (pediatric) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Dosing:</u>** 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 |

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| | **<u>Reauthorization</u>**: Documentation of improvement in patient specific disease presentation such as:<br>• Improvement in PFT or Dlco<br>• Improvement in 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 |
| **Coverage Duration:** | • Initial Authorization: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**OMALIZUMAB**
Affected Medications: XOLAIR (omalizumab)

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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 for use in combination with another monoclonal antibody (Fasenra, Nucala, Xolair, Cinqair)? | Yes – Criteria not met, combination use is experimental | No – Go to #3 |
| 3. Is the request to treat a diagnosis according to one of the Food and Drug Administration (FDA)-approved indications?<br>○ Indicated for patients 6 years of age and older with moderate to severe persistent asthma who have a positive skin test or in vitro reactivity to a perennial aeroallergen and whose symptoms are inadequately controlled with inhaled corticosteroids<br>○ Treatment of adults and adolescents 12 years of age and older with chronic idiopathic urticaria who remain symptomatic despite H1 antihistamine treatment<br>○ Treatment in adults 18 years of age and older as add on treatment of nasal polyps who have had inadequate response to nasal corticosteroids | Yes – Go to appropriate section below | No – Criteria not met |
| **Severe Allergic Asthma** | | |

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| 1. Is there documentation of severe allergic asthma defined by all of the following:<br>○ A positive skin test or in vitro reactivity to a perennial aeroallergen<br>○ A serum total IgE level at baseline of<br>▪ At least 30 IU/mL and less than 700 IU/mL in patients age 12 years or older; OR<br>▪ At least 30 IU/mL and less than 1,300 IU/mL in patients age 6 to 11 years<br>○ FEV1 less than 80% at baseline or FEV1/FVC reduced by at least 5% from normal | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documented use of high-dose inhaled corticosteroid (ICS) plus a long-acting beta agonist (LABA) for at least three months with continued symptoms? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there a documented history of 2 or more asthma exacerbations requiring oral or systemic corticosteroid treatment in the past 12 months while on combination inhaled treatment and at least 80% adherence? | Yes – Go to #5 | No – Go to #4 |
| 4. Is there documentation that chronic daily oral corticosteroids are required? | Yes – Go to #5 | No – Criteria not met |
| 5. Is the drug prescribed by or in consultation with an allergist, immunologist, or pulmonologist? | Yes – Approve up to 6 months | No – Criteria not met |
| **Chronic Idiopathic Urticaria** | | |

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| 1. Is there documentation of active chronic idiopathic urticaria and the underlying cause is not considered to be any other allergic condition or other form of urticaria? | Yes – Go to #2 | No – Criteria not met |
| 2. Is there documented avoidance of triggers (such as NSAIDs)? | Yes – Go to #3 | No – Criteria not met |
| 3. Is there documented baseline score from an objective clinical evaluation tool, such as:<br>• Urticaria Activity Score (UAS7), OR<br>• Angioedema Activity Score (AAS), OR<br>• Dermatology Life Quality Index (DLQI), OR<br>• Angioedema Quality of Life (AE-QoL), OR<br>• Chronic Urticaria Quality of Life Questionnaire (CU-QoL)? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is there documented failure of at least one month trial with scheduled dosing of up to 4-fold standard dosing of one of the following second generation H1-antihistamine products: cetirizine, fexofenadine, loratadine, desloratadine, or levocetirizine? | Yes – Document and go to #5 | No – Criteria not met |
| 5. Is there documented failure to one or more month trial on previous therapy with scheduled dosing of at least one of the following:<br>• Add-on therapy with a leukotriene antagonist (montelukast or | Yes – Document and go to #6 | No – Criteria not met |

| | | |
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| zafirlukast)<br>• Add-on therapy with a H2-antagonist (famotidine or cimetidine)<br>• Add-on therapy with cyclosporine A | | |
| 6. Is the drug prescribed by an allergist or immunologist? | Yes – Approve up to 6 months | No – Criteria not met |
| **Nasal Polyps** | | |
| 1. Is there documentation of chronic sinusitis after total ethmoidectomy with a need for revision endoscopic sinus surgery due to continued symptoms of nasal congestion/obstruction from recurrent bilateral sinus obstruction due to nasal polyps? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documented failure with at least 1 intranasal corticosteroid (such as fluticasone) after ethmoidectomy? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documented failure with Sinuva implant? | Yes – Go to #4 | No – Criteria not met |
| 4. Is the drug prescribed by a specialist in the treatment of nasal polyps (otolaryngologist)? | Yes – Approve up to 6 months | No – Criteria not met |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider? | Yes – Go to #2 | No – Criteria not met |

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| 2. Is the request for use in combination with another monoclonal antibody (Fasenra, Dupixent, Nucala, Cinqair)? | Yes – Criteria not met, combination use is experimental | No – Go to #3 |
| 3. 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**

- **Xolair**
  - Availability: 75 mg/0.5 mL & 150 mg/mL prefilled syringe; 150 mg/mL single-dose vial
  - Dosing:
    - CIU: 150 mg or 300 mg every 4 weeks
    - Asthma and Nasal Polyps: dose based on pre-treatment serum IgE (IU/mL), weight (kg), and age per FDA-approved drug label. If weight and IgE levels are outside of recommended dosing schedule, use of Xolair is considered experimental and is not covered.

*Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced for all medical infusion drugs

POLICY NAME:
**ONASEMNOGENE ABEPARVOVEC XIOI**
Affected Medications: ZOLGENSMA (onasemnogene abeparvovec xioi + IV)

<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>• Documentation of previous treatment history AND<br>• Diagnosis of spinal muscular atrophy (SMA) by genetic test showing:<br>○ Fewer than 3 copies of SMN2<br>AND<br>• Documentation of anti-adeno-associated virus (AAV) serotype 9 antibody titer less than or equal 1:50 AND<br>• Documentation of ventilator use status</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td>• Dosed 1.1 x $10^{-14}$ vectors per kilogram of body weight with prophylactic prednisolone 1 mg/kg/day prior to and following administration for a total of 30 days
<table>
<tr><th>Patient Weight Range (kg)</th><th>Dose volume (mL)</th></tr>
<tr><td>2.6-3.0</td><td>16.5</td></tr>
<tr><td>3.1-3.5</td><td>19.3</td></tr>
<tr><td>3.6-4.0</td><td>22.0</td></tr>
<tr><td>4.1-4.5</td><td>24.8</td></tr>
<tr><td>4.6-5.0</td><td>27.5</td></tr>
<tr><td>5.1-5.5</td><td>30.3</td></tr>
<tr><td>5.6-6.0</td><td>33.0</td></tr>
<tr><td>6.1-6.5</td><td>35.8</td></tr>
<tr><td>6.6-7.0</td><td>38.5</td></tr>
<tr><td>7.1-7.5</td><td>41.3</td></tr>
<tr><td>7.6-8.0</td><td>44.0</td></tr>
<tr><td>8.1-8.5</td><td>46.8</td></tr>
<tr><td>8.6-9.0</td><td>49.5</td></tr>
<tr><td>9.1-9.5</td><td>52.3</td></tr>
<tr><td>9.6-10.0</td><td>55.0</td></tr>
<tr><td>10.1-10.5</td><td>57.8</td></tr>
<tr><td>10.6-11.0</td><td>60.5</td></tr>
<tr><td>11.1-11.5</td><td>63.3</td></tr>
<tr><td>11.6-12.0</td><td>66.0</td></tr>
<tr><td>12.1-12.5</td><td>68.8</td></tr>
<tr><td>12.6-13</td><td>71.5</td></tr>
<tr><td>13.1-13.5</td><td>74.3</td></tr>
</table></td></tr>
</table>

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| **Exclusion Criteria:** | • Concurrent treatment with Spinraza<br>• Previous treatment with Zolgensma (AVXS-101) in their lifetime<br>• Advanced SMA at baseline (complete paralysis of limbs)<br>• Breathing assistance: tracheostomy, permanent ventilator dependence<br>• Pre-existing hepatic insufficiency |
| **Age Restriction:** | • Children less than 2 years old |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with a pediatric 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:** | • Approved for one dose only per lifetime, unless otherwise specified |

POLICY NAME:
**ONCOLOGY AGENTS**
Affected Medications: ABIRATERONE ACETATE, ABRAXANE, ADCETRIS, ALECENSA, ALIMTA, ALIQOPA, ALKERAN, ALUNBRIG 180mg ORAL TABLET, ARZERRA, ASPARLAS, AYVAKIT, BALVERSA, BAVENCIO, BELRAPZO, BELUMOSUDIL, BENDAMUSTINE, BENDEKA, BESPONSA, BESREMI, BLENREP, BORTEZOMIB, BOSULIF, BRAFTOVI, BRUKINSA, CABOMETYX, CALQUENCE, CAPRELSA, CLOFARABINE, CLOLAR, COMETRIQ, COPIKTRA, COSELA, COTELLIC, CYRAMZA, DACOGEN, DARZALEX, DARZALEX FASPRO, DAURISMO, DOXIL, DOXORUBICIN LIPOSOMAL, ELAHERE, EMPLICITI, ENHERTU, ERBITUX, ERIVEDGE, ERLEADA, ERLOTINIB, ERWINAZE, EXKIVITY, FARYDAK, FOTIVDA, GAZYVA, GAVRETO, GILOTRIF, HYCAMTIN, IBRANCE, IBRUTINIB, ICLUSIG, IDHIFA, IMATINIB, IMBRUVICA IMFINZI, IMJUDO, IMLYGIC IRESSA, INLYTA, INQOVI, INREBIC, ISTODAX, IXEMPRA, JAKAFI, JELMYTO, JEMPERLI, JEVTANA, KADCYLA, KEYTRUDA, KIMMTRAK, KYPROLIS, LAPATINIB, LARTRUVO, LENALIDOMIDE, LENVIMA, LIBTAYO, LONSURF, LORBRENA, LUMAKRAS, LUMOXITI, LUTATHERA, LYNPARZA, LYTGOBI, MARGENZA, MARQIBO, MATULANE, MEKINIST, MEKTOVI, MONJUVI, MYLOTARG, NAB-PACLITAXEL, NEXAVAR, NERLYNX, NILANDRON, NINLARO, NIVOLUMAB, NUBEQA, ODOMZO, ONCASPAR, ONIVYDE, ONUREG, OPDIVO, OPDUALAG, PADCEV, PEMAZYRE, PEMETREXED, PEMFEXY, PEPAXTO, PERJETA, PHOTOFRIN, PLUVICTO, POLIVY, POMALYST, PORTRAZZA, POTELIGEO, PROLEUKIN, PROVENGE, QINLOCK, RETEVMO, REVLIMID, REZUROCK, ROMIDEPSIN, ROZLYTREK, RUBRACA, RYBREVANT, RYDAPT, RYLAZE, SARCLISA, SORAFENIB, STIVARGA, SUNITINIB, SUTENT, SYNRIBO, TABRECTA, TAFINLAR, TAGRISSO, TALZENNA, TARCEVA, TAZVERIK, TECENTRIQ, TECVAYLI, TEMODAR, TEMOZOLOMIDE, TEPADINA, TEPMETKO, TIBSOVO, TORISEL, TREANDA, TRODELVY, TRUSELTIQ, TUKYSA, TYKERB, UKONIQ, VECTIBIX, VELCADE, VENCLEXTA, VERZENIO, VIDAZA, VIZIMPRO, VONJO, VOTRIENT, VYXEOS, XALKORI, XELODA, XOFIGO, XOSPATA, XPOVIO, XTANDI, YONDELIS, ZALTRAP, ZELBORAF, ZEPZELCA, ZOLINZA, ZYDELIG, ZYKADIA, ZYNLONTA

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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, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment** | • Reauthorization: documentation of disease responsiveness to therapy |

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| **Regimen & Other Criteria:** | |
| **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 approval: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**ONPATTRO**
Affected Medications: ONPATTRO (patisiran sodium)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>  ○ Treatment of the polyneuropathy of hereditary transthyretin-mediated amyloidosis in adults. |
| **Required Medical Information:** | • Documented pathogenic mutation in transthyretin (TTR; e.g. V30M mutation)<br>• Diagnosis of hereditary transthyretin (hATTR) amyloidosis with polyneuropathy<br>• Documentation of baseline polyneuropathy disability (PND) score less than or equal to IIIb **OR** baseline familial amyloid polyneuropathy (FAP) stage I or II<br>• Presence of clinical signs and symptoms of disease (e.g., peripheral/autonomic neuropathy, motor disability, cardiovascular dysfunction, renal dysfunction)<br>• Documented failure with diflunisal<br><br><u>Reauthorization:</u><br>• Documentation of either continued PND score less than or equal to IIIb **OR** patient continues to have FAP stage I or II<br>**AND**<br>• Documentation of the patient experiencing positive clinical response to patisiran (e.g., improved neurologic impairment, motor function, cardiac function, quality of life assessment, serum TTR levels, etc.) |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Hereditary transthyretin-mediated (hATTR) amyloidosis</u>**<br>Dosing:<br>• For patients weighing less than 100 kg, the recommended dosage is 0.3 mg/kg once every 3 weeks.<br>• For patients weighing 100 kg or more, the recommended dosage is 30 mg once every 3 weeks.<br><br>Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced |

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| **Exclusion Criteria:** | • Previous liver transplantation<br>• NYHA class III or IV<br>• Concomitant antisense oligonucleotide (e.g., inotersen) or tafamidis (Vyndaqel, Vyndamax) |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Physicians experienced in the management of amyloidosis<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:
**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>• Exceptions require that combined opioid use greater than 90 MME is not chronic and is being used for short term exceptional circumstances</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></td></tr>
<tr><td>Up to 20mg per day</td><td>4</td></tr>
<tr><td>21 to 40mg per day</td><td>8</td></tr>
<tr><td>41 to 60mg per day</td><td>10</td></tr>
<tr><td>Greater than 60mg per day</td><td>12</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>4</td></tr>
<tr><td>Morphine</td><td>1</td></tr>
<tr><td>Oxycodone</td><td>1.5</td></tr>
<tr><td>Oxymorphone</td><td>3</td></tr>
<tr><td>Tramadol</td><td>0.1</td></tr>
<tr><td>Buprenorphine patch</td><td>**</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 per hour over a 24-hour day.</td></tr>
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| | **Example**:<br>5 ug/hr buprenorphine patch X 24 hrs = 120 ug/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.<br><br>• 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 µg/hr buprenorphine patches dispensed for use over 28 days would work out as follows:<br><br>**Example**:<br>5 ug/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:** | • Based on exceptional circumstanse, not to exceed 3 months, unless otherwise specified |

**OPZELURA**
Affected Medications: OPZELURA CREAM (1.5%)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design.<br>  ○ Atopic dermatitis |
| **Required Medical Information:** | **<u>Severe Atopic Dermatitis</u>**<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:** | • Documented 12-week trial and clinical failure with all of the following alternatives: tacrolimus ointment, pimecrolimus cream, Eucrisa, phototherapy, cyclosporine, azathioprine, methotrexate, mycophenolate, Dupixent, AND Adbry (prior authorization required for Dupixent and Adbry).<br><br><u>Reauthorization</u><br>• No reauthorization permitted: treatment beyond 8 weeks has not been studied or found to be safe and effective. |
| **Exclusion Criteria:** | • Combination use with a monoclonal antibody (such as Dupixent)<br>• Previous 8-week treatment course<br>• Cosmetic indications, such as vitiligo |
| **Age Restriction:** | • 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist (i.e., dermatologist, allergist, or immunologist)<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Maximum of 8 weeks, unless otherwise specified. |

POLICY NAME:
**ORAL TESTOSTERONE**
Affected Medications: JATENZO, TLANDO, KYZATREX

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise exclude by plan design. |
| **Required Medical Information:** | • 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<br><br>**For member 65 years and above:**<br>• Yearly evaluation of need is completed discussing need for hormone replacement therapy.<br>• Yearly documentation that provider has educated patient on risks of hormone replacement (heart attack, stroke)<br>• Yearly documentation that provider has discussed limited efficacy and safety for hormone replacement in patients experiencing age related decrease in testosterone levels<br><br>**Gender Dysphoria hormone supplementation under 18 years of age:**<br>• Documentation of current Tanner stage 2 or greater OR Documentation of baseline and current estradiol and testosterone levels to confirm onset of puberty<br>• Documentation from a licensed mental health professional (LMHP) confirming diagnosis and addressing the patient's general identifying characteristics;<br>  ○ The initial and evolving gender and any associated mental health concerns, and other psychiatric diagnoses;<br>  ○ The duration of the referring licensed mental health professional's relationship with the client, including the type of evaluation and psychotherapy to date;<br>  ○ The clinical rationale for supporting the client's request for cross-hormone therapy and statement that the client meets eligibility criteria;<br>  ○ Informed consent required from both patient and guardian documented by prescribing provider<br>  ○ Permission to contact the licensed mental health professional for coordination of care |

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| | ○ Comprehensive mental health evaluation should be provided in accordance with most current version of the World Professional Association for Transgender Health (WPATH) Standards of Care<br><br>○ **Note:** For requests following pubertal suppression therapy, an updated or new comprehensive mental health evaluation must be provided prior to initiation of hormone supplementation |
| **Appropriate Treatment Regimen & Other Criteria:** | Kyzatrex:<br>• Documented failure with transdermal testosterone<br><br>Jatenzo, Tlando:<br>• Documented failure with transdermal testosterone<br>AND<br>• Documented failure with Kyzatrex<br><br>**Reauthorization**: documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Women (unless covered benefit for treatment of gender dysphoria) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Gender Dysphoria: Diagnosis made and prescribed by, or in consultation with, a specialist in the treatment of gender dysphoria<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Gender dysphoria: 12 months, unless otherwise specified<br>• All other indications: 24 months, unless otherwise specified |

POLICY NAME:
**ORENITRAM**
Affected Medications: ORENITRAM (treprostinil)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design. |
| **Required Medical Information:** | **<u>Pulmonary arterial hypertension (PAH) WHO Group 1</u>**<br>• Documentation of PAH confirmed by right-heart catheterization<br>• Etiology of PAH: (idiopathic, heritable, or associated with connective tissue disease)<br>• NYHA/WHO Functional Class II to III symptoms<br>• Documentation of acute vasoreactivity testing (positive result requires trial/failure to calcium channel blocker), unless there are contraindications such as low systemic blood pressure (systolic blood pressure less than 90), low cardiac index, or presence of severe symptoms (functional class IV) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of failure with Remodulin<br>• For initiation of therapy patient must have mean pulmonary artery pressure of at least 20 mmHg at rest, an elevated pulmonary vascular resistance (PVR) of at least 2.0 Wood units, and a mean pulmonary capillary wedge pressure less than 15<br>**AND**<br>• The pulmonary hypertension has progressed despite maximal medical and/or surgical treatment of the identified condition<br>• Treatment with oral calcium channel blocking agents has been tried and failed, or has been considered and ruled out<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>• Subsequent approvals require documentation of treatment success such as improved walking distance or improvements in 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:** | • 12 months, unless otherwise specified. |

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

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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 benefit design. (For non-cancer use only) |
| **Required Medical Information:** | Prostate Cancer<br>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | Prostate Cancer<br>• Documented treatment failure or intolerable adverse event with leuprolide or degarelix<br>• Dosing: 360 mg on Day 1, followed by 120 mg daily starting on Day 2<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:** | • 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:
**ORKAMBI**
Affected Medications: ORKAMBI (lumacaftor/ivacaftor)

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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 cystic fibrosis (CF) diagnosis.<br>• Documentation of Homozygous for the F508 del mutation by Food and Drug Administration (FDA)-cleared CF mutation test on both alleles of the CFTR gene<br>• Baseline forced expiratory volume in 1 second (FEV1)<br>• Documentation of baseline liver function tests |
| **Appropriate Treatment Regimen & Other Criteria:** | • 1 through 2 years and weighing between 7 kg to 9 kg: Take one packet of lumacaftor 75 mg/ivacaftor 94 mg granules every 12 hours<br>• 1 through 2 years and weighing between 9 kg to 14 kg: Take one packet of lumacaftor 100 mg/ivacaftor 125 mg granules every 12 hours<br>• 1 through 2 years and weighing 14 kg or greater: Take one packet of lumacaftor 150 mg/ivacaftor 188 mg granules every 12 hours<br>• 2 through 5 years and weighing less than 14 kg: Take one lumacaftor 100 mg/ivacaftor 125 mg packet of granules every 12 hours<br>• 2 through 5 years and weighing 14 kg or greater: Take one lumacaftor 150 mg/ivacaftor 188 mg packet of granules every 12<br>• 6 through 11 years: Take two lumacaftor 100 mg/ivacaftor 125 mg tablets every 12 hours<br>• 12 years and older: Take two lumacaftor 200 mg/ivacaftor 125 mg tablets every 12 hours<br><br>**Reauthorization**: Documentation of improvement in FEV1 from baseline, documentation of follow up liver function tests; blood pressure monitoring |
| **Exclusion Criteria:** | • Concurrent use of strong CYP3A inducers: rifampin, rifabutin, phenobarbital, carbamazepine, phenytoin, and St. John's wort |
| **Age Restriction:** | • 1 year of age and older |

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| **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 approval: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

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

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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 |
| **Persistent or recurrent Cushing's disease or patients with de novo Cushing's disease for whom pituitary surgery is not an option or has not been curative.** | | |
| 1. Is there documentation that the patient has persistent or recurrent Cushing's disease for whom surgery has not been curative OR a new diagnosis of Cushing's disease in which surgery is not an option | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documentation that the patient has a 24-hour mean Urine Free Cortisol (UFC) greater than 1.5 times the upper limit of normal (above 67 µg/24 hours). | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documentation that the patient has risk factors for Torsades de Pointe or taking other medications that may prolong the QTc interval? | No – Document and go to #4 | Yes – Criteria not met |
| 4. Is there documentation that the treatment is in consult with an | Yes – Approve up to 6 months | No – Criteria not met |

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| endocrinologist, neurologist or adrenal surgeon with confirmation of a titration schedule including urine free cortisol monitoring every 1-2 weeks until adequate clinical response is maintained? | | |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success as determined by the mean urine free cortisol levels less than or equal to the upper limit of normal based on laboratory results? | 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** | | |
| • **Isturisa 1 mg tablets**<br>  ○ 180/30<br>• **Isturisa 5 mg tablets**<br>  ○ 180/30<br>• **Isturisa 10 mg tablets**<br>  ○ 180/30 | | |

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 four 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 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 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>• Documented provider attestation that the patient has been informed of the risk of pregnancy given their fertility status (such as tubal ligation failure, misdiagnosed/temporary menopause, etc.), the severe fetal harm that can occur with pregnancy that follows the administration of Vivjoa, AND documentation that the patient acknowledges and understands these risks and agrees to be vigilant in avoiding pregnancy during Vivjoa therapy and for a minimum of 2 years following their last dose of Vivjoa.<br><br><u>Dosing:</u><br>• **Vivjoa-ONLY regimen:**<br>Day 1 – 600mg as one dose<br>Day 2 – 450 mg as one dose<br>Starting Day 14 – 150 mg every 7 days for 11 weeks (weeks 2 through 12)<br>• **Fluconazole-Vivjoa regimen:**<br>Day 1, Day 4, Day 7 – fluconazole 150 mg<br>Day 14 through Day 20 – Vivjoa 150 mg once daily<br>Starting day 28 – Vivjoa 150 mg every 7 days for 11 weeks (weeks 4 through 14)<br>• **Not to exceed one treatment course per year** |

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| | • **<u>Reauthorization</u>** 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 |
| **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:
**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. |
| **Required Medical Information:** | • Documentation of decreased corneal sensitivity (≤ 4 cm using the Cochet-Bonnet aesthesiometer) within the area of the persistent epithelial defect or corneal ulcer and outside of the area of the defect in at least one corneal quadrant<br>• Documentation of stage 2 or stage 3 neurotrophic keratitis<br>  ○ Stage 2 neurotrophic keratitis<br>  • Persistent corneal epithelial defect OR<br>  • Descemet's membrane folds and stromal swelling OR<br>  • Anterior chamber inflammatory reaction<br>  ○ Stage 3 neurotrophic keratitis<br>  • Corneal ulcer OR<br>  • Corneal perforation OR<br>  • Corneal stromal melting |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented progression in disease severity with all of the following treatments:<br>  ○ Preservative-free artificial tears, gel, or ointments<br>  ○ Therapeutic corneal or scleral contact lenses<br>  ○ Amniotic membrane transplantation and conjunctival flap surgery OR tarsorrhaphy OR cyanoacrylate glue OR soft-bandage contact lens<br>• Dose may not exceed more than 1 vial per eye per day<br>• Dosing does not exceed 8 weeks for first treatment<br>• Reauthorization will require documentation of improvement in corneal sensitivity and grade of severity determined by corneal fluorescein staining using the modified Oxford scale |
| **Exclusion Criteria:** | • Active or suspected ocular or periocular infections |
| **Age Restriction:** | |

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| **Prescriber/Site of Care Restrictions:** | • Ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Authorization: 8 weeks, unless otherwise specified<br>• Reauthorization: 8 weeks, maximum approval (total of 16 weeks), unless otherwise specified |

POLICY NAME:
**OXYBATES**
Affected Medications: XYREM (sodium oxybate), XYWAV (oxybate salts)

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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 narcolepsy and experiences episodes of cataplexy<br>**OR**<br>• Diagnosis of narcolepsy and experiences excessive daytime sleepiness (EDS) confirmed by all of the following:<br>  ○ Polysomnography and multiple sleep latency test results<br>  ○ Current evaluation of symptoms and Epworth Sleepiness Scale (ESS) score of at least 15 at baseline<br>**OR**<br>• Diagnosis of idiopathic hypersomnia (IH) and experiences EDS confirmed by all of the following (Xywav only):<br>  ○ Polysomnography and multiple sleep latency test results<br>  ○ Current evaluation of symptoms and ESS score of at least 15 at baseline |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Narcolepsy with cataplexy:</u>**<br>• Documented treatment failure with each of the following for at least 1 month, unless contraindicated:<br>  ○ Venlafaxine, fluoxetine, and a tricyclic antidepressant.<br><br>**<u>Narcolepsy or IH, with EDS:</u>**<br>• Symptoms limit ability to perform normal daily activities<br>• Current ESS score of at least 13 despite current therapy<br>• Documented treatment failure with at least 3 of the following (1 in each category required) for at least 1 month, unless contraindicated:<br>  ○ Modafinil or armodafinil<br>  ○ Methylphenidate, or dextroamphetamine, or lisdexamfetamine<br>  ○ Sunosi (required for EDS due to narcolepsy only)<br><br>**<u>Reauthorization:</u>**<br>• Narcolepsy with cataplexy: clinically significant reduction in cataplexy episodes |

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| | • Narcolepsy or IH, with EDS: clinically significant improvement in activities of daily living and in Epworth Sleepiness Scale (ESS) score |
| **Exclusion Criteria:** | • Current alcohol use disorder<br>• Concurrent use of sedative/hypnotic drugs or other central nervous system depressants |
| **Age Restriction:** | • 7 years of age or older for cataplexy or EDS due to narcolepsy<br>• 18 years of age or older for EDS due to IH |
| **Prescriber/Site of Care Restrictions:** | • Sleep specialist enrolled in Xyrem REMS program<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:
**OZANIMOD**
Affected Medications: ZEPOSIA (Ozanimod)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design:<br>  ○ Treatment of relapsing forms of multiple sclerosis (MS), to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease, in adults.<br>  ○ Ulcerative Colitis |
| **Required Medical Information:** | **<u>Relapsing Remitting MS (RRMS)</u>**<br>• Documentation of diagnosis of relapsing forms of Multiple Sclerosis (MS) confirmed with MRI (Revised McDonald diagnostic criteria for MS<br>  ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS<br>**<u>Clinically Isolated Syndrome (CIS)</u>**<br>• Documentation of CIS as shown by patients who do not fulfill McDonald criteria for a diagnosis of MS but have an abnormal brain MRI with one or more hyperintense T2 lesions that are characteristic of MS in at least two of four MS-typical regions at presentation or within three to six months of the event<br>**<u>Secondary-Progressive MS (SPMS)</u>**<br>• Documentation of prior history of relapsing-remitting MS (RRMS) with progressive increase in disability over at least 6 months independent of, or in the absence of, relapses<br>• Documentation of active disease classified as the presence of clinical relapse or inflammatory activity (i.e. new or enlarging T2 lesions or gadolinium enhancing lesions on MRI) in the last 2 years.<br>• Documentation of Expanded Disability Status Scale (EDSS) score of 3.0 to 6.5<br>**<u>Ulcerative Colitis</u>**<br>• Diagnosis supported by endoscopy/colonoscopy/sigmoidoscopy or biopsy with moderate to severely active disease despite current treatment |
| **Appropriate Treatment Regimen &** | **<u>Ulcerative Colitis</u>**<br>• Documented failure with at least two oral treatments for a minimum of 12 weeks each: corticosteroids, sulfasalazine, |

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| **Other Criteria:** | azathioprine, mesalamine, balsalazide, cyclosporine, 6-mercaptopurine<br>**AND**<br>• Documented treatment failure with (or intolerable adverse event) with all preferred pharmacy drugs (Humira, Xeljanz, Stelara)<br><br><u>Dosing:</u><br>• After treatment titration, the recommended maintenance dosage of Zeposia is 0.92 mg once daily after Day 7.<br><br>Reauthorization requires provider attestation of treatment success |
| **Exclusion Criteria:** | • Patients with PPMS<br>• Resting heart rate less than 55 beats per minute at baseline<br>• Recent myocardial infarction, stroke, prolonged Fridericia-corrected QT<br>• Active infections |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with a neurologist, multiple sclerosis specialist, or gastroenterologist appropriate for diagnosis.<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Authorization: 6 months (Ulcerative Colitis only), all other indications: 12 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**OZURDEX**
Affected Medications: OZURDEX (dexamethasone intravitreal implant)

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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 non-infectious uveitis **OR**<br>• Diagnosis of macular edema following branch retinal vein occlusion (BRVO) or central vein occlusion (CRVO) **OR**<br>• Diagnosis of clinically significant diabetic macular edema defined as:<br>  ○ Thickening of the retina less than or equal to 500 micrometers from the center of the macula **OR**<br>  ○ Hard exudates and adjacent retinal thickening less than or equal to 500 micrometers from macula center **OR**<br>  ○ Zone of retinal thickening at least 1 disc area in size (1500 micrometers) located less than or equal to 1 disc diameter from the center of the macula<br>• Past treatment with corticosteroids without a clinically significant rise in intraocular pressure **AND**<br>• Past treatment with laser photocoagulation |
| **Appropriate Treatment Regimen & Other Criteria:** | • One intravitreal implant per 6 months<br>• Must not be used concurrently with other intraocular treatments such as: Avastin, Lucentis or Eylea |
| **Exclusion Criteria:** | • Ocular or Periocular infections<br>• Glaucoma<br>• Torn or ruptured posterior lens capsule |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Ophthalmologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • 6 months, unless otherwise specified |

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

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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 |
| **Mitigation of allergic reactions due to accidental exposure to peanut** | | |
| 2. Is the request age-appropriate, as defined below?<br>○ Initial Dose Escalation and Up-Dosing: 4 to 17 years of age.<br>○ Maintenance: 4 to 17 years of age, OR 18 years of age, or greater, for those who began Palforzia maintenance before becoming 18 years of age. | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there a documented history of allergic reactions to peanut that meet the criteria below?<br>○ Signs and symptoms of a significant systemic allergic reaction to peanut, such as: hives, swelling, wheezing, hypotension, and 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 medication. | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is there documentation of a positive skin prick test (SPT) response to peanut with a wheal diameter at least 3 mm larger than | Yes – Document and go to #5 | No – Criteria not met |

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| control? | | |
| 5. Is there documentation indicating a significant impact on quality of life due to peanut allergies? | Yes – Document and go to #6 | |
| 6. Are there known contraindications to treatment with Palforzia, as defined below?<br>○ Currently uncontrolled asthma.<br>○ A history of cardiovascular disease, including uncontrolled or inadequately controlled hypertension.<br>○ A history of eosinophilic esophagitis or other eosinophilic gastrointestinal diseases.<br>○ A history of a mast cell disorders, including mastocytosis, urticarial pigmentosa, and hereditary or idiopathic angioedema. | Yes – Criteria not met | No – Document and go to #7 |
| 7. Is Palforzia being prescribed by or in consultation with an allergist or immunologist? | Yes – Approve up to 6 months | No – Criteria not met |
| **Renewal Criteria** | | |
| 1. Is this a renewal request following the completion of the Up-Dosing phase? | Yes – Document and go to #2 | No – Go to #3 |
| 2. Is there documentation of treatment success and a clinically significant response to therapy defined by a successful completion of all Up-Dosing levels for the required lengths of time? | Yes – Document and go to #4 | No – Criteria not met |

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| 3. Is there documentation of treatment success and a clinically significant response to therapy, as defined below?<br>○ An improvement in quality of life (for those in the Maintenance phase).<br>○ A decrease in SPT wheal diameter of at least 0.5mm from baseline. | Yes – Document and go to #4 | No – Criteria not met |
| 4. 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**

| Dosing Phase and Dosage Form | Quantity Limit |
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| Palforzia cap escalation | 1 kit/14 days |
| Palforzia cap level 1 | 1 kit/14 days |
| Palforzia cap level 2 | 1 kit/14 days |
| Palforzia cap level 3 | 1 kit/14 days |
| Palforzia cap level 4 | 1 kit/14 days |
| Palforzia cap level 5 | 1 kit/14 days |
| Palforzia cap level 6 | 1 kit/14 days |
| Palforzia cap level 7 | 1 kit/14 days |
| Palforzia cap level 8 | 1 kit/14 days |
| Palforzia cap level 9 | 1 kit/14 days |
| Palforzia cap level 10 | 1 kit/14 days |
| Palforzia pow level 11 (#15 for Up-Dosing) | 1 kit/14 days |
| Palforzia pow level 11 (#30 for maintenance) | 30/30 days |

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 anticipated treatment course, including target phenylalanine (Phe) level set by specialist<br>• Documentation of failure to Phe restricted diet as monotherapy<br>• Documentation of failure to Kuvan AND phenylalanine-restricted diet as dual-therapy<br>• Current patient weight<br>• Baseline (pre-treatment) blood Phe levels<br><br>Baseline Phe concentration must be consistent with the following:<br>• Phe level must be greater than 10mg/dL (600 microM)<br><br>Reauthorization after initial approval requires documentation of updated Phe labs decreased by 20% or greater from baseline<br>• Treatment with Palynziq should be discontinued in patients whose blood Phe has not decreased by at least 20% from baseline **or** a blood phenylalanine concentration ≤600 microM/L after 16 weeks with max dose of 40 mg/day<br><br>Reauthorization for continued long-term approval (12 months) requires updated Phe labs meeting one of the following criteria:<br>• Phe level less than 20 percent of baseline OR<br>• Phe level lower than baseline and meets specialist's target level |
| **Appropriate Treatment Regimen & Other Criteria:** | If patient has failed dual-therapy with Kuvan and Phe restricted diet, and Palynziq is warranted, treatment must be consistent with the following:<br>• Initial dose must be 2.5mg once weekly x 4 weeks<br>• Titration (after 4-week induction): 2.5 mg twice weekly for 1 week, then 10 mg once weekly for 1 week, then 10 mg twice weekly for 1 week, then 10 mg 4 times/week for 1 week, then 10 mg once daily for 2 week. |

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| | Maintenance (after induction and titration): 20 mg once daily for at least 24 weeks. May increase to 40 mg once daily if a response (20% reduction from baseline in blood phenylalanine **or** blood phenylalanine concentration 600 micromol/L or less) has not been achieved after administering 20 mg once daily for 24 weeks. |
| **Exclusion Criteria:** | • Prior intolerance or allergic reaction to requested medication<br>• Doses greater than 40mg/day |
| **Age Restriction:** | • 18 years and older |
| **Prescriber/Site of Care Restrictions:** | • Specialist in metabolic disorders or endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 2 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PARATHYROID HORMONE**
Affected Medications: NATPARA (parathyroid hormone)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Adjunct to calcium and vitamin D to control hypocalcemia in hypoparathyroidism |
| **Required Medical Information:** | • Documentation of the following lab values:<br>  ○ 25-hydroxyvitamin D levels within normal limits (approximately 30-74 ng/mL) while on standard of care (such as calcitriol)<br>  ○ Total serum calcium (albumin-corrected) greater than 7.5 mg/dL |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented failure with at least 8 weeks of a consistent supplementation regimen as follows:<br>  ○ Calcium 2000 mg daily<br>  ○ Vitamin D (metabolite or analog)<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 approval: 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. |
| **Required Medical Information:** | • Documentation of DEXA score within 2 years T Score less than or equal to -2.5, OR FRAX score indicating major fracture risk 20% or greater or hip fracture 3% or greater, OR non-traumatic hip or vertebral fracture. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of adequate calcium intake and vitamin D level and/or treatment<br>• Documentation of clinically significant worsening osteoporosis or five years of continuous treatment on therapeutic doses of bisphosphonates (e.g., alendronate, risedronate, ibandronate, zoledronic acid) or contraindication to intravenous bisphosphonate therapy<br>**OR**<br>• T-score -3.5 or lower, or -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>Maximum duration of therapy should not exceed 2 years</u>** |
| **Exclusion Criteria:** | • Paget's Disease<br>• Open epiphyses (i.e., pediatric or young adult patient)<br>• Bone metastases or skeletal malignancies<br>• Hereditary disorders predisposing to osteosarcoma<br>• Prior external beam or implant radiation therapy involving the skeleton<br>• Concurrent therapy with bisphosphonates, Prolia, Xgeva, or another parathyroid hormone analog<br>• Pre-existing hypercalcemia<br>• Pregnancy |
| **Age Restriction:** | • 18 years of age and older with fully fused epiphyses |

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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:** | • Approval: 24 months (no reauthorization), 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:** | **Documentation of one of the following conditions:**<br>1. Congenital heart disease (CHD):<br>a. With cardiac transplantation, cardiac bypass, or extra-corporeal membrane oxygenation<br>b. That is hemodynamically significant (e.g. acyanotic heart disease, congestive heart failure, or moderate to severe pulmonary hypertension)<br>2. Chronic lung disease (CLD) of prematurity:<br>a. In the first year of life, born less than 32 weeks gestation and requiring greater that 21% oxygen for at least the first 28 days of life<br>b. In the second year of life necessitating continued medical support within the 6 month period prior to RSV season (e.g. corticosteroids, diuretics, supplemental oxygen)<br>3. Cystic Fibrosis **and**:<br>a. Clinical evidence of CLD and/or nutritional compromise<br>b. Severe lung disease (e.g. previous hospitalization for pulmonary exacerbation in the first year of life or abnormalities on chest radiography or computed tomography that persist when stable)<br>c. A weight for length less than the 10$^{th}$ percentile<br>4. Congenital airway abnormality or neuromuscular condition (not cystic fibrosis) that impairs the ability to clear airway secretions<br>5. Premature infants without above conditions |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Prevention of serious lower respiratory tract disease caused by respiratory syncytial virus (RSV)</u>**<br>• The first dose of Synagis should be administered prior to commencement of the RSV season<br>• Remaining doses should be administered monthly throughout the RSV season (Exception: dose administration should occur immediately post cardiopulmonary bypass surgery, even if dose is administered earlier than a month from previous dose, then dosing schedule should resume monthly) |

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| | • No more than 5 monthly doses During the RSV season, November 1 through March 31<br>• Discontinue prophylaxis therapy if hospitalized for RSV |
| **Exclusion Criteria:** | • For use in the treatment of RSV disease |
| **Age Restriction:** | **Refer to numbered conditions above in "Required Medical Information":**<br>• 1a. Less than 2 years of age<br>• 1b. Less than 1 year of age<br>• 2a. Less than 1 year of age; Gestational Age less than 32 weeks<br>• 2b. Less than 2 years of age; Gestational Age less than 32 weeks<br>• 3a. Less than 1 year of age<br>• 3b. Less than 2 years of age<br>• 3c. Less than 2 years of age<br>• 4. Less than 1 year of age<br>• 5. Less than 1 year of age; Gestational Age less than 29 weeks |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | Approval:<br>• 5 months (November 1 through March 31) [5 monthly doses] , unless otherwise specified<br>• 1 month for off-season when RSV activity greater than or equal to 10% for the region according to the CDC [1 monthly dose] , 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 the following:<br>  ○ Treatment plan is a cisplatin-based regimen treating a localized, non-metastatic solid tumor |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | • Metastatic disease |
| **Age Restriction:** | • Pediatric patients greater than or equal to 1 month old and less than 18 years of age |
| **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

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved and compendia-supported indications not otherwise excluded by plan design. |
| **Required Medical Information:** | **Chronic Hepatitis C (CHC):**<br>• Documentation chronic hepatitis C virus (HCV) genotype by liver biopsy or by Food and Drug Administration (FDA)-approved serum test<br>• Baseline HCV RNA level<br>• Documentation of anti-hepatitis C virus regimen to be used with AND anticipated dose and duration of therapy<br><br>**Chronic Hepatitis B (CHB):**<br>• Documentation of HBeAg-positive or HBeAg-negative chronic hepatitis B virus (HBV) infection<br>• Baseline HBV DNA level<br>• Documentation of anti-hepatitis B virus regimen to be used with AND anticipated dose and duration of therapy<br><br>**Chronic Hepatitis C and B:**<br>• Baseline HIV-1 RNA level<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>• Current estimated creatinine clearance OR serum creatinine, height, and weight to calculate by Cockcroft-Gault within 12 weeks prior to anticipated start of therapy<br>• Current complete blood count AND liver function tests within 12 weeks prior to anticipated start of therapy<br>• Documentation if HIV/HCV/HBV coinfection<br>• Documentation of abstinence from alcohol and any illegal drug use for at least 6 months |
| **Appropriate Treatment** | **Chronic Hepatitis C:** |

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| **Regimen & Other Criteria:** | • Approve if used in combination with Food and Drug Administration (FDA)- and/or AASLD/IDSA- recommended regimen and if not otherwise excluded from PacificSource policies of other medications in the regimen<br>• Preferred regimen should include concomitant ribavirin<br><br>**Chronic Hepatitis B (one of the following 4 scenarios must be met):**<br>• HBeAg-positive **AND** baseline serum HBV DNA greater than 20,000 copies/mL **AND** baseline serum aminotransferase (ALT) two times greater than the upper limit of normal range<br>• HBeAg-positive **AND** baseline serum HBV DNA greater than 20,000 copies/mL **AND** baseline serum aminotransferase (ALT) one to two times greater than the upper limit of normal range **AND** moderate-severe inflammation/fibrosis<br>• HBeAg-negative **AND** baseline serum HBV DNA greater than 2,000 copies/mL **AND** baseline serum aminotransferase (ALT) two times greater than the upper limit of normal range<br>• HBeAg-negative **AND** baseline serum HBV DNA greater than 2,000 copies/mL **AND** baseline serum aminotransferase (ALT) one to two times greater than the upper limit of normal range **AND** moderate-severe inflammation/fibrosis<br><br>**Chronic Hepatitis C and B:**<br>• Creatinine clearance less than 50 ml/min, adjust dose: 135 mcg subcutaneously once weekly<br>• Baseline platelet count greater than or equal to 90,000 cells/mm3<br>• Baseline absolute neutrophil count 1,500 cells/mm3 or more |
| **Exclusion Criteria:** | • Treatment of patients with CHC who have had solid organ transplantation<br>• Autoimmune hepatitis<br>• Hepatic decompensation (Child-Pugh score greater than 6) |
| **Age Restriction:** | • CHC: 5 years of age or older<br>• CHB: 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with a gastroenterologist, hepatologist, or infectious disease specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • CHC: 12 weeks, unless otherwise specified (depends on regimen and diagnosis)<br>• CHB: 12 months, unless otherwise specified |

POLICY NAME:
**PEGCETACOPLAN**
Affected Medications: EMPAVELI (pegcetacoplan)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>  ○ Treatment of paroxysmal nocturnal hemoglobinuria (PNH) |
| **Required Medical Information:** | • PNH diagnosis confirmed by high-sensitivity flow cytometry evaluation<br>• Complete blood count (CBC), reticulocyte count, lactate dehydrogenase (LDH), packed RBC transfusion requirement<br>• Patients must be administered a meningococcal vaccine at least 2 weeks prior to initiation of Empaveli therapy if have not received one in the past 3 years, and revaccinated according to current ACIP guidelines<br>• Platelet count of at least 50,000<br>• At least 4 blood transfusions required in the previous 12 months for those not currently on eculizumab |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with eculizumab, defined as ongoing need for transfusions despite regular treatment for at least 6 months<br>• If switching from eculizumab, Empaveli may be initiated while continuing eculizumab at its current dose for 4 weeks. After 4 weeks, eculizumab must be discontinued.<br><br>**Reauthorization** requires documentation of treatment success, as shown by improvement in serum LDH and hemoglobin labs, and a decrease in blood transfusion requirement |
| **Exclusion Criteria:** | • Current meningitis infection<br>• History of bone marrow transplantation<br>• Use in combination with other complement-inhibitor therapy |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Must be 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: 12 months, unless otherwise specified |

POLICY NAME:
**PEGINTRON**
Affected Medications: PEGINTRON REDIPEN, PEGINTRON

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved and compendia-supported indications not otherwise excluded by plan design. |
| **Required Medical Information:** | • Documentation chronic hepatitis C virus (HCV) genotype by liver biopsy or by Food and Drug Administration (FDA)-approved serum test<br>• Baseline HCV RNA level<br>• Documentation of anti-hepatitis C virus regimen to be used with **AND** anticipated dose and duration of therapy<br>• Patient weight<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>• Current estimated creatinine clearance **OR** serum creatinine, height, and weight to calculate by Cockcroft-Gault within 12 weeks prior to anticipated start of therapy<br>• Current complete blood count **AND** liver function tests within 12 weeks prior to anticipated start of therapy<br>• Documentation if HIV/HCV/HBV coinfection<br>  • Documentation of abstinence from alcohol and any illegal drug use for at least 6 months |
| **Appropriate Treatment Regimen & Other Criteria:** | • Approve if used in combination with Food and Drug Administration (FDA)- and/or AASLD/IDSA- recommended regimen and if not otherwise excluded from PacificSource policies of other medications in the regimen<br>• Preferred regimen should include concomitant ribavirin<br>• In patients with moderate renal dysfunction (creatinine clearance 30-50 mL/min), the PegIntron dose should be reduced by 25%<br>• Patients with severe renal dysfunction (creatinine clearance 10-29 mL/min), including those on hemodialysis, should have the PegIntron dose reduced by 50% |

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| **Exclusion Criteria:** | • Autoimmune hepatitis<br>• Hepatic decompensation (Child-Pugh score greater than 6) |
| **Age Restriction:** | • 3 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with a gastroenterologist, hepatologist, or infectious disease specialist<br>• All approvals are subject to utilization of the most cost-effective site of care<br>• |
| **Coverage Duration:** | • 12 weeks, unless otherwise specified (depends on regimen and diagnosis) |

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

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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? (With a preferred drug, if applicable to this policy) | Yes – Go to appropriate section below | No – Criteria not met |
| **Chronic Gout** | | |
| 1. Is there documentation of at least 3 gout flares in the past 18 months that were uncontrolled by colchicine and/or nonsteroidal anti-inflammatory drugs (NSAIDS) or oral or injectable corticosteroids? | Yes – Document and go to #3 | No – Go to #2 |
| 2. Is there documentation of at least 1 gout tophus or chronic gouty arthritis? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documentation of baseline serum uric acid level greater than 8 mg/dL | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is there a documented contraindication, intolerance, or clinical failure (inability to reduce serum uric acid to less than 6 mg/dL) with a minimum 3 month trial of each of the following: | Yes – Document treatment and go to #5 | No – Criteria not met |

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| • Highest tolerated dose of allopurinol<br>• Highest tolerated dose of febuxostat | | |
| 5. Is there documentation of negative testing for glucose-6-phosphate dehydrogenase (G6PD) deficiency or documented lower risk making testing unnecessary? | Yes – Document and go to #6 | No – Criteria not met |
| 6. Is the drug prescribed by, or in consultation with a rheumatologist or nephrologist? | Yes – Approve up to 6 months | No – Criteria not met |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success such as reduction of symptoms or tophi AND documentation of serum uric acid level less than 6 mg/dL prior to scheduled infusion? | Yes – Document and 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** | | |
| • **Kyrstexxa (pegloticase injection)**<br>o 8 mg given as an intravenous infusion every two weeks (8 mg/mL single use vial)<br>o Limited to two vials per 28 days | | |

POLICY NAME:
**PENICILLAMINE**
Affected Medications: DEPEN (penicillamine)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | • Due to risk of fatalities due to aplastic anemia, agranulocytosis, thrombocytopenia, myasthenia gravis, and Goodpasture's Syndrome: Documented treatment plan including routine urinalysis, WBCs, hemoglobin, platelet count, liver function tests, renal function tests. |
| **Appropriate Treatment Regimen & Other Criteria:** | • For Reauthorization: Documentation of disease responsiveness 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:** | • Approval: 3 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:** | • Diagnosis of pheochromocytoma and one of the following:<br>  ○ Documentation of preoperative preparation for surgical resection.<br>  ○ Documentation of chronic treatment for metastatic pheochromocytoma. |
| **Appropriate Treatment Regimen & Other Criteria:** | • If use is projected to be greater than 14 days:<br>  ○ Documentation of failure or contraindication to a selective alpha-1 adrenergic receptor blocker (e.g., doxazosin, terazosin, prazosin)<br><br>• Initial dosing: 10 mg twice daily, increase by 10 mg every other day until optimal blood pressure response is achieved; usual range: 20-40 mg 2-3 times/day. Doses up to 240 mg/day have been reported<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 a specialist 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:
**PHESGO**
Affected Medications: PHESGO (pertuzumab-trastuzumab-hyaluronidase-zzxf)

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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 prescribed dosing regimen<br>• Baseline evaluation of left ventricular function<br>• Documentation of HER2 positivity based on 3+ IHC testing or positive FISH test |
| **Appropriate Treatment Regimen & Other Criteria:** | • Regular assessment of LVEF for all indications<br><br>**<u>All Indications</u>**<br>• Coverage for Phesgo requires documentation of one of the following:<br>  ○ A documented intolerable adverse event to all of the preferred products (Perjeta in combination with Kanjinti, and Perjeta in combination with Ogivri) and the adverse event was not an expected adverse event attributed to the active ingredients<br>  ○ Currently receiving treatment with Phesgo, excluding via samples or manufacturer's patient assistance programs<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<br>• Consider withholding therapy for at least 3 weeks for either a drop in LVEF to <40% OR LVEF 40-45% with a 10% reduction in LVEF from pre-treatment values<br>• Stage IV Breast Cancer: Previous failure/progression while on Perjeta (pertuzumab) |
| **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 |

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| **Coverage Duration:** | • For new starts to adjuvant breast cancer therapy – approve 12 months with no reauthorization<br><u>For all other clinical scenarios:</u><br>• Initial approval: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PIQRAY**
Affected Medications: PIQRAY (alpelisib)

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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 |
| **Appropriate Treatment Regimen & Other Criteria:** | • Reauthorization: documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater<br>• **Previous use of fulvestrant** |
| **Age Restriction:** | • 18 years of age or 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 approval: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**PIRFENIDONE**
Affected Medications: Pirfenidone 267 mg tablet, Pirfenidone 801 mg tablet

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise by plan design<br>  ○ Idiopathic Pulmonary Fibrosis |
| **Required Medical Information:** | • Documentation of ALL of the following:<br>  ○ Presence of usual interstitial pneumonia (UIP) on high resolution computed tomography (HRCT), and/or surgical lung biopsy<br>  ○ Baseline forced vital capacity (FVC) greater than or equal to 50 percent of the predicted value<br>  ○ Predicted diffuse capacity for carbon monoxide (DLCO) greater than or equal to 30 percent |
| **Appropriate Treatment Regimen & Other Criteria:** | • Pirfenidone is not approved for use in combination with Ofev<br>• Reauthorization requires documentation of treatment success |
| **Exclusion Criteria:** | • Transaminases more than 5 times the upper limit of normal or elevated transaminases accompanied by symptoms (jaundice, hyperbilirubinemia) |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consulation 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:
**PLEGRIDY**
Affected Medications: PLEGRIDY (peglyated interferon beta-1a)

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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 diagnosis of relapsing forms of multiple sclerosis confirmed with magnetic resonance imaging (MRI) |
| **Appropriate Treatment Regimen & Other Criteria:** | • No concurrent use of medications indicated for the treatment of relapsing-remitting multiple sclerosis<br>• Not approved for primary progressive multiple sclerosis<br>• Multiple sclerosis<br>• Reauthorization: provider attestation of treatment success |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or after a consultation with a Neurologist or a MS specialist<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:
**PONVORY**
Affected Medications: Ponvory (ponesimod)

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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), to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease, in adults |
| **Required Medical Information:** | **<u>Relapsing forms of Multiple Sclerosis (MS)</u>**<br>• Diagnosis confirmed by Magnetic Resonance Imaging (MRI) - per Revised McDonald diagnostic criteria for MS<br><br>**<u>Secondary-Progressive MS (SPMS)</u>**<br>• Documentation of prior history of relapsing-remitting MS (RRMS) with progressive increase in disability over at least 6 months, independent of, or in the absence of, relapses<br>• Documentation of active disease classified as the presence of clinical relapse or inflammatory activity (i.e., new or enlarging T2 lesions or gadolinium enhancing lesions on MRI) in the last 2 years<br>• Documentation of Expanded Disability Status Scale (EDSS) score of 3.0 to 6.5 |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of treatment failure or contraindication with at least 12 weeks of TWO of the following: Gilenya, Aubagio, Mayzent<br>• Discontinuation of treatment if liver enzymes exceed three times the upper limit of normal (ULN) with signs of liver dysfunction (unexplained nausea, vomiting, abdominal pain, fatigue, anorexia, rash with eosinophilia, or jaundice and/or dark urine)<br>• Reauthorization: provider attestation of treatment success |
| **Exclusion Criteria:** | • Recent (in the past 6 months) myocardial infarction (MI), unstable angina, stroke, transient ischemic attack (TIA), decompensated heart failure requiring hospitalization or class III or IV heart failure,<br>• Mobitz type II second- or third-degree Atrioventricular block (AV block) or sick sinus syndrome (unless patient has functioning pacemaker) |
| **Age** | • Adults over 18 |

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| **Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with a neurologist or multiple sclerosis specialist<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:
**PRETOMANID**
Affected Medications: PRETOMANID (pretomanid)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design<br>  ○ Extensively drug resistant tuberculosis (XDR-TB)<br>  ○ Treatment-intolerant multidrug-resistant tuberculosis (TI MDR-TB)<br>  ○ Nonresponsive multidrug-resistant tuberculosis (NR MDR-TB) |
| **Required Medical Information:** | • Patient has failed, is resistant, or is allergic to quad therapy of any combination of the following: Isoniazid, Rifampin, Ethambutol, Pyrazinamide, Fluoroquinolone, Capreomycin (Kanamycin, Amikacin, Streptomycin), Ethionamide/Prothinamide, Cycloserine/Terizidone, Aminosalicylic acid (acidic salt) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of being administered by directly observed therapy (DOT) |
| **Exclusion Criteria:** | • Drug-sensitive TB (DS-TB)<br>• Latent Infection due to Mycobacterium tuberculosis<br>• Extrapulmonary TB (e.g. central nervous system) |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or after a consultation with a Neurologist or a MS specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 26 weeks, unless otherwise specified |

POLICY NAME:
**PROLIA**
Affected Medications: PROLIA (denosumab)

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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:** | • Dosage is 60 mg once every 6 months |
| **Coverage Duration:** | • Approval: 24 months, unless otherwise specified<br>• Reauthorization: 24 months, unless otherwise specified |

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

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| **Covered Uses:** | • All 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:
**QUTENZA**
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 |
| **Required Medical Information:** | • Diagnosis of neuropathic pain associated with one of the following<br>  ○ Post-herpetic neuralgia<br>  ○ Diabetic peripheral neuropathy of the feet<br><br>• Documented treatment failure with at least 12 weeks of ALL of the following:<br>  ○ gabapentin<br>  ○ pregabalin<br>  ○ carbamazepine or oxcarbazepine or valproic acid/divalproex sodium<br>  ○ amitriptyline or nortriptyline<br>  ○ topical lidocaine |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose limited to single treatment (up to 4 patches) once every 90 days.<br>• For renewal, your doctor must send in notes showing that this drug has worked well for you. |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of most cost-effective site of care<br>• Pain management specialist |
| **Coverage Duration:** | • Initial approval: 3 months (single treatment), unless otherwise specified<br>• Reauthorization: 12 months (up to 4 treatments), 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:** | • The prescribed medication will be used in combination with dietary protein restriction<br>• Documented intolerance to sodium phenylbutyrate or documented comorbid condition that prohibits a trial of sodium phenylbutyrate due to its sodium content (e.g., heart failure, renal impairment, hypertension, or edema)<br><br>**<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • Age greater than or equal to 2 months |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Approval: 3 months, unless otherwise specified<br>• Reapproval: 12 months, unless otherwise specified |

POLICY NAME:
**RAVULIZUMAB**
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. |
| **Required Medical Information:** | • Documentation of complete treatment course<br>• Complete blood count (CBC), reticulocyte count, lactate dehydrogenase (LDH), packed RBC transfusion requirement<br><br>**<u>Paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis</u>**<br>• LDH levels greater than or equal to 1.5 times the upper limit of normal range if not currently treated with complement-inhibitor therapy (eculizimab, ravulizumab-cwvz, pegcetacoplan)<br>• PNH diagnosis confirmed by high-sensitivity flow cytometry evaluation of red blood cells and white blood cells with granulocyte or monocyte clone size of greater than or equal to 5%<br>• Platelet count of at least 30,000<br>• 4 or more blood transfusions required in the past 12 months if not currently treated with complement-inhibitor therapy (eculizimab, ravulizumab-cwvz, pegcetacoplan)<br><br>**<u>Atypical hemolytic uremic syndrome (aHUS) to inhibit complement-medicated thrombotic microangiopathy</u>**<br>• Clinical presentation of: microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury<br>• LDH levels greater than or equal to 1.5 times the upper limit of normal range.<br>• ADAMTS13 activity level greater than 10%<br>• Patient has failed to respond to five days of plasma therapy<br>• 4 or more blood transfusions required in the past 12 months if not currently treated with complement-inhibitor therapy (eculizimab, ravulizumab-cwvz, pegcetacoplan) |

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| | **<u>Generalized Myasthenia Gravis (gMG)</u>**<br>• Diagnosis of generalized Myasthenia Gravis (gMG) confirmed by:<br>  ○ A history of abnormal neuromuscular transmission test OR<br>  ○ A positive edrophonium chloride test OR<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 anti-acetylcholine receptor (AchR) antibodies<br>• MG-Activities of Daily Living (MG-ADL) total score of greater than or equal to 6<br>• Documentation of baseline Quantitative Myasthenia Gravis (QMG) score<br>• Documentation of gMG treatment history showing the following:<br>  ○ Currently on a stable dose of at least one gMG therapy (acetylcholinesterase inhibitor, corticosteroid, or non-steroidal immunosuppressive therapy (NSIST))<br>  ○ One of the following:<br>  ▪ Documented treatment failure with an adequate trial (one year or more) of at least 2 immunosuppressive therapies (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate) OR<br>  ▪ Documented 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<br>• Documented treatment failure with Vyvgart |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>PNH and aHUS weight-based dosing:</u>**<br>• (5 to less than 10 kg) Loading, 600 mg IV infusion; maintenance, 300 mg 2 weeks after loading dose then every 4 weeks<br>• (10 to less than 20 kg) Loading, 600 mg IV infusion; maintenance, 600 mg 2 weeks after loading dose then every 4 weeks |

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| | • (20 to less than 30 kg) Loading, 900 mg IV infusion; maintenance, 2100 mg 2 weeks after loading dose then every 8 weeks<br>• (30 to less than 40 kg) Loading, 1200 mg IV infusion; maintenance, 2700 mg 2 weeks after loading dose then every 8 weeks<br>• (40 to less than 60 kg) Loading, 2400 mg IV infusion; maintenance, 3000 mg 2 weeks after loading dose then every 8 weeks<br>• (60 to less than 100 kg) Loading, 2700 mg IV infusion; maintenance, 3300 mg 2 weeks after loading dose then every 8 weeks<br>• (100 kg or greater) Loading, 3000 mg IV infusion; maintenance, 3600 mg 2 weeks after loading dose then every 8 weeks<br><br>**<u>gMG weight-based dosing:</u>**<br>• (40 to less than 60 kg) Loading, 2400 mg IV infusion; maintenance, 3000 mg 2 weeks after loading dose then every 8 weeks<br>• (60 to less than 100 kg) Loading, 2700 mg IV infusion; maintenance, 3300 mg 2 weeks after loading dose then every 8 weeks<br>• (100 kg or greater) Loading, 3000 mg IV infusion; maintenance, 3600 mg 2 weeks after loading dose then every 8 weeks<br><br>Switching from Soliris (eculizumab), administer loading dose 2 weeks after last eculizumab infusion, then administer maintenance doses once every 8 weeks, starting 2 weeks after the loading dose<br><br><u>Reauthorization requires documentation of treatment success</u><br>• PNH, aHUS: updated serum LDH and Hb labs, and blood transfusion history, showing treatment success and clinically significant response to therapy |
| **Exclusion Criteria:** | • Current meningitis infection<br>• History of bone marrow transplantation<br>• Use in combination with other complement-inhibitor therapy (eculizumab) |

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| **Age Restriction:** | • PNH: 1 month of age and older<br>• aHUS: 1 month of age and older<br>• gMG: 18 years and older |
| **Prescriber/Site of Care Restrictions:** | • PNH: Hematologist<br>• aHUS: Hematologist or Nephrologist<br>• gMG: Neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**REBLOZYL**
Affected Medications: REBLOZYL INJ 25MG, REBLOZYL INJ 50MG

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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 anemia in adult patients with beta thalassemia who require regular red blood cell (RBC) transfusions OR<br>• Diagnosis of anemia failing an erythropoiesis stimulating agent and requiring 2 or more red blood cell units over 8 weeks in adult patients with very low- to intermediate-risk myelodysplastic syndromes with ring sideroblasts (MDS-RS) or with myelodysplastic/myeloproliferative neoplasm with ring sideroblasts and thrombocytosis (MDS/MPN-RS-T).<br>• Baseline complete blood count (CBC) within 2 months and then prior to each administration, or more frequently as indicated<br>• Documentation of current RBC transfusion regimen<br>• Negative pregnancy test for female patients of reproductive potential |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing:<br>○ Starting dose of 1mg/kg every 3 weeks<br>○ Not to exceed 1.25mg/kg every 3 weeks (beta thalassemia)<br>○ Not to exceed 1.75mg/kg every 3 weeks (MDS-RS or MDS/MPN-RS-T)<br><br>**Reauthorization** requires documentation of a 20% reduction in red blood cell (RBC) transfusion burden 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:** | • Hematologist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**REBIF**
Affected Medications: REBIF, REBIF TITRATION PACK

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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:** | • **Reauthorization**: provider attestation of treatment success |
| **Exclusion Criteria:** | • Concurrent use of other disease-modifying medications for the treatment of MS |
| **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:
**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 (SCV) 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>• **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:** | |
| **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:
**REMODULIN**
Affected Medications: REMODULIN INJECTION (treprostinil), TREPROSTINIL INJECTION

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design. |
| **Required Medical Information:** | **<u>Pulmonary arterial hypertension (PAH) WHO Group 1</u>**<br>• Documentation of PAH confirmed by right-heart catheterization<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 to IV symptoms<br>• Documentation of acute vasoreactivity testing (positive result requires trial/failure to calcium channel blocker) unless contraindications such as low systemic blood pressure (systolic blood pressure less than 90), low cardiac index, or presense of severe symptoms (functional class IV) |
| **Appropriate Treatment Regimen & Other Criteria:** | • For initiation of therapy patient must have a mean pulmonary artery pressure of at least 20 mmHg at rest, an elevated pulmonary vascular resistance (PVR) of at least 3.0 Wood units, and a mean pulmonary capillary wedge pressure less than 15 mmHg<br>**AND**<br>• The pulmonary hypertension has progressed despite maximal medical and/or surgical treatment of the identified condition<br>• Treatment with oral calcium channel blocking agents dependent on vasoreactivity testing results has been tried and failed, or has been considered and ruled out<br>• Documentation that treprostinil is used as a single route of administration (Remodulin, Tyvaso, Orenitram should not be used in combination) |

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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 symptoms<br>  ○ Ambrisentan and tadalafil<br>  ○ Bosentan and riociguat<br>  ○ Macitentan and sildenafil<br>• Subsequent approvals require documentation of treatment success such as improved walking distance or improvements in functional class |
| **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:** | • Cardiologist or pulmonologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Approval: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**RESLIZUMAB**
Affected Medications: CINQAIR (reslizumab)

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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 for use in combination with another monoclonal antibody (Fasenra, Nucala, Xolair, Dupixent)? | Yes – Criteria not met, combination use is experimental | No – Go to #3 |
| 3. Is the request to treat a diagnosis according to one of the Food and Drug Administration (FDA)-approved indications?<br>a. Add-on maintenance treatment of patients with severe asthma aged 18 years and older with an eosinophilic phenotype | Yes – Go to appropriate section below | No – Criteria not met |
| **Severe Eosinophilic Asthma** | | |
| 1. Is there documentation of severe eosinophilic asthma defined by the following:<br>a. Baseline eosinophil count at least 400 cells/µL<br>**AND**<br>b. FEV1 less than 80% at baseline or FEV1/FVC reduced by at least 5% from normal | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documented use of high-dose inhaled corticosteroid (ICS) plus a long-acting beta agonist (LABA) for at least three months with continued symptoms? | Yes – Document and go to #3 | No – Criteria not met |

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| 3. Is there a documented history of 2 or more asthma exacerbations requiring oral or systemic corticosteroid treatment in the past 12 months while on combination inhaled treatment and at least 80% adherence? | Yes – Go to #5 | No – Go to #4 |
| 4. Is there documentation that chronic daily oral corticosteroids are required? | Yes – Go to #5 | No – Criteria not met |
| 5. Is there a documented trial and failure or intolerable adverse event with all of the preferred products – Dupixent, Fasenra, Nucala, Xolair? | Yes – Go to #6 | No – Criteria not met |
| 6. Is the drug prescribed by or in consultation with an allergist, Immunologist, or Pulmonologist? | Yes – Approve up to 6 months | No – Criteria not met |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider? | Yes – Go to #2 | No – Criteria not met |
| 2. Is the request for use in combination with another monoclonal antibody (Fasenra, Nucala, Xolair, Dupixent)? | Yes – Criteria not met, combination use is experimental | No – Go to #3 |
| 3. 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** | | |

- **Cinqair**
  - Availability: 100 mg/10 mL single-use vial
  - Dosing: 3 mg/kg infusion once every 4 weeks

*Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced for all medical infusion drugs

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

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Presence of one of the following syndromic disorders confirmed by genetic testing: complete DiGeorge Syndrome, FOXN1 deficiency, 22q11.2 deletion, CHARGE (coloboma, heart defect, choanal atresia, growth and development retardation, genital hypoplasia, ear defects including deafness) syndrome, 10p13 hemizygosity, CHD7 mutation.<br>• Congenital athymia confirmed by flow cytometry:<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 |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | • Diagnosis of Severe Combined Immunodeficiency<br>• Heart surgery planned within 4 weeks of administration of cultured thymus tissue (CTT) or 3 months after administration<br>• Prior thymus transplant<br>• Human Immunodeficiency virus (HIV) infection |
| **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. |
| **Coverage Duration:** | • Initial Authorization: 1 month (1 treatment only), unless otherwise specified |

POLICY NAME:
**REVATIO**
Affected Medications: Revatio 20mg tablet, sildenafil 20mg tablet, sildenafil 10mg/mL SUSP

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design. |
| **Required Medical Information:** | • Pulmonary Arterial Hypertension (PAH) (WHO Group 1) confirmed by right heart catheterization<br>• Etiology of PAH (idiopathic or associated with connective tissue disease)<br>• NYHA/WHO Functional Class II or III symptoms<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blocker) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Subsequent approvals require documentation of treatment success such as improved walking distance or improvements in functional class |
| **Exclusion Criteria:** | • Concomitant nitrate therapy on a regular or intermittent basis<br>• Concomitant use of riociguat, a guanylate cyclase stimulator |
| **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:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**RIBAVIRIN**
Affected Medications: RIBASPHERE 200mg, RIBATAB, RIBAPAK, REBETOL (PDL only Copegus)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | • Baseline hemoglobin level greater than 10 g/dL<br>• Baseline creatinine clearance (serum creatinine, height, weight to calculate)<br>• Baseline weight<br>• Documentation chronic hepatitis C virus genotype by liver biopsy or by Food and Drug Administration (FDA)-approved serum test<br>• Documentation of anti-hepatitis C virus regimen to be used with and anticipated duration of therapy |
| **Appropriate Treatment Regimen & Other Criteria:** | • Approve if used in combination with Food and Drug Administration (FDA)- and/or AASLD/IDSA- recommended regimen and if not otherwise excluded from PacificSource policies of other medications in the regimen |
| **Exclusion Criteria:** | • Women who are pregnant<br>• Men whose female partners are pregnant<br>• Patients with autoimmune hepatitis<br>• Patients with hemoglobinopathies (e.g., thalassemia major, sickle-cell anemia)<br>• Patients with creatinine clearance less than 50 mL/min<br>• Coadministration with didanosine<br>• Hemoglobin level  less than 8.5 g/dL |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with gastroenterologist or hepatologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 weeks, unless otherwise specified (depends on regimen) |

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 |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>All Indications:</u>**<br>• Documented treatment failure or intolerable adverse event with trial of Kineret (anakinra)<br><br>**<u>Recurrent Pericarditis:</u>**<br>• Documented treatment failure or intolerable adverse event to triple therapy with colchicine AND aspirin AND a glucocorticoid<br><br>**<u>Dosing for CAPS or Recurrent Pericarditis:</u>** |

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| | ○ Adults: loading dose of 320 mg followed by 160 mg once weekly<br>○ Pediatric patients (age 12 to 17): loading dose of 4.4 mg/kg (maximum 320 mg) followed by 2.2 mg/kg once weekly (maximum 160 mg)<br>**Dosing for DIRA:**<br>○ Adults: 320 mg once weekly<br>○ Pediatric patients (weighing 10 kg or more): 4.4 mg/kg (maximum 320 mg) once weekly<br><br>**Reauthorization** will require:<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, TNF inhibitors, or other biologics |
| **Age Restriction:** | • CAPS or Recurrent Pericarditis: 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist (such as a rheumatologist, immunologist, cardiologist, or dermatologist)<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 weeks, unless otherwise specified (depends on regimen) |

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 benefit design. |
| **Required Medical Information:** | **<u>Chronic thromboembolic pulmonary hypertension (CTEPH)</u>**<br>• WHO Group 4 with documented thromboembolic occlusion of proximal or distal pulmonary vasculature and mean pulmonary arterial pressure of at least 25 mmHg at rest in the absence of elevated pulmonary capillary wedge pressure (i.e. PCWP not more than 15 mmHg)<br><br>**<u>Pulmonary arterial hypertension (PAH)</u>**<br>• WHO Group 1 confirmed by right heart catheterization<br>• Etiology of PAH (idiopathic, heritable, or associated with connective tissue disease)<br>• NYHA/WHO Functional Class II to III symptoms<br>• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blocker) unless contraindications such as low systemic blood pressure (systolic blood pressure less than 90), low cardiac index, or presence of severe symptoms (functional class IV)<br>• Liver function tests, creatinine clearance, and baseline exercise testing (6MWD) |
| **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>• Failure/Contraindication to the following therapy classes: Phosphodiesterase type 5 (PDE5) inhibitors **AND** endothelin receptor antagonists<br>• Safety and efficacy have not been demonstrated in patients with creatinine clearance less than or equal to 15 ml/min or on dialysis |

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| | • Safety and efficacy have not been demonstrated in patients with severe (Child-Pugh class C) hepatic impairment<br><br><u>Reauthorization:</u> Subsequent approvals require documentation of treatment success such as improved walking distance or improvements in functional class |
| **Exclusion Criteria:** | • Pregnancy<br>• 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)<br>• Use in patients with symptomatic pulmonary hypertension associated with an idiopathic interstitial pneumonias (PH-IIP) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Cardiologist or a pulmonologist<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:
**RISDIPLAM**
Affected Medications: EVRYSDI (risdiplam)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design<br>  ○ Spinal Muscular atrophy type 1, 2 or 3 |
| **Required Medical Information:** | • Documentation of spinal muscular atrophy diagnosis confirmed by genetic tests demonstrating 5q-autosomal recessive disease<br>  ○ Documentation of four or fewer copies of SMN2<br>• For symptomatic patients, 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) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of Food and Drug Administration approved dosing and treatment plan<br><br>**<u>Reauthorization:</u>** documentation of clinically significant improvement from baseline motor function demonstrated by:<br>  ○ Improvement from baseline motor function score documented within one month of renewal request AND<br>  ○ More areas of motor function improved than worsened<br>  ○ HINE-2:<br>  ▪ at least a 2-point increase in ability to kick OR<br>  ▪ at least a 1-point increase in the motor milestones of head control, rolling, sitting, crawling, standing or walking using Section 2 of the Hammersmith Infant Neurologic Exam (HINE) AND<br>  ▪ More areas of motor function improved than worsened<br>  ○ Hammersmith Functional Motor Scale (HFSME)<br>  ▪ At least 3 points increase in score from pretreatment baseline AND<br>  ▪ More areas of motor function improved than worsened |

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| | ○ Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)<br>▪ At least a 4 point increase in score from the pretreatment baseline AND<br>▪ More areas of motor function improved than worsened<br>○ Upper Limb Module (ULM)<br>▪ At least a 3 point increase from pretreatment baseline<br>○ 6-Minute Walk Test (6MWT)<br>▪ At least a 30 meter increase from pretreatment baseline |
| **Exclusion Criteria:** | • SMA type 4<br>• Prior treatment with Zolgensma (AVXS-101)<br>• Concurrent therapy with Spinraza (nursinersen) |
| **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:
**RITUXIMAB**
Affected Medications: RITUXAN (rituximab), RITUXAN HYCELA, TRUXIMA (rituximab-abbs), RUXIENCE (rituximab-pvvr), RIABNI (rituximab-arrx)

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by benefit design.<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher<br>• Relapsing Remitting Multiple Sclerosis |
| **Required Medical Information:** | • Indication must be documented in the member's chart notes within the most recent 6 months<br>• Documentation of disease staging, all prior therapies used, and anticipated treatment course<br><br>**<u>Rheumatoid Arthritis</u>**<br>• Documentation of complete and current treatment course laboratory test confirming diagnosis of RA rheumatoid arthritis (anti-CCP, RF)<br>• Documentation of moderate to severe disease despite current treatment<br>• Documented current level of disease activity with one of the following (or equivalent objective scale):<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 RAPID3 of at least 2.3<br><br>**<u>Non-Hodgkin's Lymphoma (NHL)</u>**<br>• Documentation of CD20-positve B-Cell NHL<br><br>**<u>Chronic Lymphocytic Leukemia (CLL)</u>**<br>• Documentation of advanced or active CLL<br>• Binet Stage A or B with active disease<br>• Binet Stage C<br>• Modified Rai Stage 0, I, or II with symptoms<br>• Modified Rai Stage III or IV |

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| | **<u>Microscopic Polyangiitis (MPA) or Granulomatosis with Polyangiitis (GPA)</u>**<br>• Documentation of active GPA or MPA<br><br>**<u>Relapsing Remitting Multiple Sclerosis</u>**<br>• Diagnosis of relapsing form of Multiple Sclerosis (MS) confirmed with MRI (Revised McDonald diagnostic criteria for multiple sclerosis)<br>• Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS<br><br>**<u>Moderate to severe Pemphigus Vulgaris</u>**<br>• Confirmed diagnosis of pemphigus vulgaris:<br>  ○ Multiple non-healing oral ulcers persisting for at least 1 month, multiple flaccid blisters on normal skin and positive Nikolsky sign.<br>  ○ Direct immunofluorescence (DIF) showing intercellular localization of immunoglobulin on perilesional skin or mucosal biopsy<br>• Patient has failed a minimum of 12 weeks of therapy with corticosteroids **AND**<br>• Patient has failed a minimum of 12 weeks of therapy with immunosuppressants (e.g., azathioprine, mycophenolate, methotrexate, etc.)<br><br>**<u>Thrombycytopenia in patients with ITP</u>**<br>• Documentation of splenectomy status |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>All Uses</u>**<br>• Coverage of Truxima, Rituxan or Rituxan Hycela requires documentation of one of the following:<br>  ○ A documented intolerable adverse event to the preferred products, Riabni and Ruxience, and the adverse event was not an expected adverse event attributed to the active ingredient<br>  ○ Currently receiving treatment with Rituxan or Truxima, excluding via samples or manufacturer's patient assistance programs. |

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| | **<u>Oncology Uses</u>**<br>• Documentation of ECOG performance status of 1 or 2 OR Karnofsky performance score greater than 50%<br><br>**<u>Rheumatoid Arthritis (RA)</u>**<br>• Initial Course: Documented failure with two of the preferred pharmacy drugs (Humira, Enbrel, Xeljanz, Rinvoq)<br>  ○ Dose is approved for up to 2 doses of 1,000 mg given 2 weeks apart<br>• Repeat Course: Approve if 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.<br><br>**<u>Microscopic Polyangiitis and Granulomatosis with Polyangiitis</u>**<br>• For initial immunosuppression: in combination with a glucocorticoid<br>• Dose is approved for up to two doses of 1,000 mg annually<br>  ○ Higher doses (e.g., 1,000 mg x 2 every 6 months) will require documentation to support<br><br>**<u>Pemphigus Vulgaris</u>**<br>• Administered in combination with systemic glucocorticoid<br><br>**<u>Thrombocytopenia in patients with ITP</u>**<br>• Platelet count less than 20,000/mcl<br>**AND**<br>• Documented steroid-dependence to maintain platelets/prevent bleeding for at least 3 months **OR**<br>• Lack of clinically meaningful response to corticosteroids (defined as platelets did not increase to at least 50,000/mcl)<br><br>**<u>Reauthorization:</u>** documentation of disease responsiveness to therapy |

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| **Exclusion Criteria:** | • Concurrent use of: abatacept (Orencia), tocilizumab (Actemra), adalimumab (Humira), entanercept (Enbrel), infliximab (Remicade), certolizumab (Cimzia), golimumab (Simponi)<br>• Positive hepatitis B test/history of hepatitis B or positive tuberculosis test |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • For RA,GPA,MPA – Prescribed by a rheumatologist or in consultation with a rheumatologist<br>• For CLL, NHL– Prescribed by an oncologist<br>• For MS- Prescribed by or in consultation with a neurologist<br>• All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • For RA – Initial approval: 6 months, unless otherwise specified<br>• For Oncology – Initial approval: 4 months, unless otherwise specified<br>• For MPA/GPA – Initial approval: 3 months, unless otherwise specified<br>• For MS- Initial approval: 6 months (up to two doses of 1,000 mg), unless otherwise specified<br>• For PV – Initial approval: 3 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:** | • Complete blood count (CBC) with differential and platelet count<br>• Patient Weight<br><br>**Thrombocytopenia in patients with immune thrombocytopenia pupura (ITP)**<br>• All previously trialed therapies<br>• Documentation of splenectomy status<br><br>**Hematopoietic syndrome of acute radiation syndrome**<br>• Suspected or confirmed exposure to radiation levels greater than 2 gray (Gy); do not delay romiplostin if CBC is not readily available |
| **Appropriate Treatment Regimen & Other Criteria:** | **Thrombocytopenia in patients with ITP**<br>• Documentation of platelet count less than 20 x $10^9$/L **AND**<br>• Documentation of clinically significant bleeding **AND**<br>• One of the following:<br>  ○ Must fail at least 2 therapies for ITP, including corticosteroids or immunoglobulin (defined as platelets did not increase to at least 50 x $10^9$/L) **OR**<br>  ○ Documentation of splenectomy<br><br>**Hematopoietic syndrome of acute radiation syndrome**<br>• Approved for one-time single infusion of 10 mcg/kg |

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| | **Reauthorization (ITP only)**<br>• Response to treatment with platelet count of at least 50 x $10^9$/L (not to exceed 400 x $10^9$/L) OR<br>• The platelet counts have not increased to a level of at least 50 x $10^9$/L and the patient has NOT been on the maximum dose for at least 4 weeks<br><br>Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced |
| **Exclusion Criteria:** | • Treatment of thrombocytopenia due to myelodysplastic syndrome (MDS)<br>• Use in combination with another thrombopoietin receptor agonist (Promacta or Doptelet) or similar treatments. |
| **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 hematologist |
| **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>• 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 for fracture, defined as one of the following:<br>  • History of osteoporotic fracture<br>  • Multiple risk factors for fracture<br>  • 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<br>• Dosage: 210 mg once monthly, 12-month lifetime maximum |
| **Exclusion Criteria:** | • Heart attack or stroke event within the preceding year<br>• Concurrent use of bisphosphonates, parathyroid hormone analogs, or RANK ligand inhibitors |

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| | • Preexisting hypocalcemia |
| **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:** | • Approval: 12 months, unless otherwise specified |

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

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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 Lennox-Gastaut Syndrome |
| **Appropriate Treatment Regimen & Other Criteria:** | • QL: 3200 mg daily<br>• Reauthorization: documentation of treatment success |
| **Exclusion Criteria:** | • Familial Short QT syndrome |
| **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:** | • Approval: 12 months, unless otherwise specified |

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

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design<br>  ○ Plasminogen Deficiency Type 1 |
| **Required Medical Information:** | **<u>Plasminogen Deficiency type 1 (must meet all of the following):</u>**<br>• Diagnosis of symptomatic congenital plasminogen deficiency (C-PLGD) as evidenced by documentation of all of the following:<br>  ○ Genetic testing showing presence of biallelic pathogenic variants in plasminogen (PLG)<br>  ○ Baseline plasminogen activity level less than or equal to 45% of laboratory standard<br>  ○ Presence of (ligneous) pseudomembranous lesions with documented size, location and number of total lesions<br>• Abnormal plasminogen antigen plasma level less than 9 mg/dL as confirmed by an enzyme-linked immunosorbent assay<br>• Presence of clinical signs and symptoms of the disease (such as ligneous conjunctivitis, gingivitis, tonsillitis, abnormal wound healing) |
| **Appropriate Treatment Regimen & Other Criteria:** | Initial dosing: 6.6 mg/kg every three days<br><br>Obtain a trough plasminogen activity level approximately 72 hours following the initial dose and prior to the second dose (same time of day as initial dosing)<br>• If plasminogen activity is less than 10% above baseline level then increase to every 2 day dosing<br>• If between 10-20% of baseline then maintain every 3 day dosing<br>• If above 20% of baseline then change dosing to every 4 days.<br><br>**<u>Maintain dosing frequency as determined above for 12 weeks while treating active lesions</u>**<br>• If lesions do not resolve by 12 weeks, or there are new or recurrent lesions, increase dosing frequency in one-day |

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| | increments every 4-8 weeks up to Q2D dosing while reassessing clinical improvement until lesion resolution or until the lesions stabilize without further worsening.<br>• If desired clinical change does not occur by 12 weeks, check trough plasminogen activity level.<br>  ○ If plasminogen activity is greater than 10% above baseline level then consider other treatment options, such as surgical removal of the lesion in addition to plasminogen treatment.<br>  ○ If plasminogen activity is less than 10% above baseline level then obtain a second trough plasminogen activity level to confirm. If low plasminogen activity level is confirmed in combination with no clinical efficacy, consider discontinuing plasminogen treatment due to the possibility of neutralizing antibodies<br><br>***If lesions resolve by 12 weeks, continue at same dosing frequency and monitor for new or recurrent lesions every 12 weeks.<br><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>**<u>Reauthorization (must meet all of the following):</u>**<br>• Trough plasminogen activity level (taken 72 hours after dose) greater than 10% above baseline level<br>• Documented improvement (reduction) in lesion size and number<br>• Dosing may not exceed 6.6 mg/kg every 2 days. |
| **Exclusion Criteria:** | • Prior treatment failure with Ryplazim<br>• Treatment of idiopathic pulmonary fibrosis |
| **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 hematologist in coordination with Hemophilia Treatment Center (HTC) or other |

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| | specialized center of excellence |
| **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. |
| **Required Medical Information:** | • Documentation of confirmed congenital sucrose-isomaltase deficiency, diagnosed by duodenal or jejunal mucosal biopsy assayed for lactase, sucrose, isomaltase, and maltase.<br>• Reauthorization: requires documentation of treatment success (fewer stools, lower number of symptoms) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Symptoms of congenital sucrose-isomaltase deficiency include:<br>o Diarrhea<br>o Abdominal pain or cramping<br>o Bloating<br>o Gas<br>o Loose Stools<br>o Abdominal pain or cramping<br>o Bloating<br>o Nausea<br>o Vomiting |
| **Exclusion Criteria:** | • Known hypersensitivity to years, yeast products, glycerin (glycerol), or papain |
| **Age Restriction:** | • 5 months or older |
| **Prescriber/Site of Care Restrictions:** | |
| **Coverage Duration:** | • Initial approval: 1 month, 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. |
| **Required Medical Information:** | • Diagnosis of lysosomal acid lipase (LAL) deficiency or Rapidly Progressive LAL deficiency within the first 6 months of life via enzyme assay that measures the level and activity of LAL or a genetic sequencing analysis test<br>• Documentation of patient weight<br>• Documentation of prescribed treatment regimen (dose and frequency)<br>• Baseline fasting lipid panel prior to initiating therapy (not required for Rapidly Progressive LAL deficiency) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Reauthorization for Rapidly Progressive LAL deficiency requires documentation of improvement in weight for-age Z-score.<br>• Reauthorization for lysosomal acid lipase (LAL) deficiency requires documentation of improvement in fasting lipid panel<br>• If documentation of anti-drug antibodies to Kanuma, documentation of continued response to therapy required |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 1 month or older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial Approval: 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:** | |
| **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:
**SELUMETINIB**
Affected Medications: KOSELUGO (selumetinib)

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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 2 years 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<br><br>**<u>Neurofibromatosis type 1 (NF1) with inoperable plexiform neurofibromas</u>**<br>• Documentation of diagnosis of 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 (1 or more if patient has a parent who is diagnosed with NF1) as evaluated by a multidisciplinary specialist care team:<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 |

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| | ○ A heterozygous pathogenic NF1 variant with a variant allele fraction of 50% in apparently normal tissue such as white blood cells<br><br>**NCCN Indications**<br>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | **Reauthorization**: documentation of disease responsiveness to therapy<br>• For NF1: defined as lack of plexiform neurofibroma growth |
| **Exclusion Criteria:** | **Neurofibromatosis type 1 (NF1) with inoperable plexiform neurofibromas**<br>• Patient has experienced any of the following adverse effects while taking Kaselugo:<br>○ Symptomatic decreased LVEF<br>○ Grade 3 or 4 decreased LVEF<br>○ Retinal vein occlusion<br>○ Grade 4 diarrhea<br>○ Grade 3 or 4 colitis<br>○ Rhabdomyolysis<br>• Patient is unable to tolerate Kaselugo after 2 dose reductions<br><br>**NCCN Indications**<br>• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | **Neurofibromatosis type 1 (NF1) with inoperable plexiform neurofibromas**<br>• 2 to 18 years of age |
| **Prescriber/Site of Care Restrictions:** | **Neurofibromatosis type 1 (NF1) with inoperable plexiform neurofibromas**<br>• Prescribed by, or in consultation with, a pediatric oncologist or specialist with experience in the treatment of neurofibromatosis<br><br>**NCCN Indications**<br>• Prescribed by, or in consultation with, an oncologist |

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

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

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications<br>  ○ HIV (human immunodeficiency virus)-associated wasting, cachexia |
| **Required Medical Information:** | • Documentation of body mass index (BMI), weight, and ideal body weight (IBW)<br><br>For initial approval members must meet all the following criteria:<br><br>• Diagnosis of cachexia or wasting syndrome associated with HIV infection<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>• Patient has unintentionally lost more than 10% of body weight over last 12 months or more than 5% over last 6 months **OR**;<br>• Member weighs less than 90% of ideal body weight **OR**;<br>• Patient has a body mass index (BMI) less than 20 $kg/m^2$<br><br>For continuation of therapy members must meet the following criteria:<br><br>• Patients treated with Serostim for 12 or more weeks have demonstrated a response to therapy (i.e., body mass index has improved or stabilized)<br>• Currently on antiretroviral therapy |

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| **Appropriate Treatment Regimen & Other Criteria:** | • 0.1 mg/kg once daily at bedtime (maximum: 6 mg/day) OR<br>• Based on the following body weights:<br>○ Less than 35 kg, 0.1 mg/kg SUBQ at bedtime<br>○ 35 to 45 kg, 4 mg SUBQ at bedtime<br>○ 45 to 55 kg, 5 mg SUBQ at bedtime<br>○ Over 55 kg, 6 mg SUBQ at bedtime<br><br>Patients at risk for adverse effects (e.g., glucose intolerance) may be started at 0.1 mg/kg every other day. |
| **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<br>• Hypersensitivity to Serostim |
| **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<br>• Reauthorization: 8 months (maximum duration of therapy 48 weeks total) |

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. |
| **Required Medical Information:** | • Diagnosis of Cushing's Disease<br>• The patient had surgery that was not curative or is not a candidate for surgery |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>If the patient is currently receiving Signifor therapy</u>**:<br>• The patient has shown a clinically meaningful reduction in 24-hour urinary free cortisol levels and/or improvement in signs or symptoms of the disease.<br>• Electrocardiogram (ECG) obtained prior to dose adjustment<br><br>**<u>If the patient is not currently receiving Signifor:</u>**<br>• Baseline fasting plasma glucose and/or hemoglobin A1c (HgA1c) levels were obtained<br>• The patient has controlled blood glucose levels OR the patient is receiving optimized antidiabetic therapy<br>• ECG obtained<br>• Liver function tests evaluated prior to initiation |
| **Exclusion Criteria:** | • Poorly controlled diabetes mellitus (HbA1c >8%)<br>• Severe hepatic impairment (Child Pugh C) |
| **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:** | • Approval: 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. |
| **Required Medical Information:** | **<u>Acromegaly</u>**<br>Patient meets the following criteria for initiation of therapy:<br>• Clinical evidence of acromegaly<br>• Pre-treatment high insulin-like growth factor-1 (IGF-1) level for age/gender<br>• Documented inadequate response or intolerable adverse event to Somatuline Depot (lanreotide) or Somavert (pegvisomant)<br>• Patient has had an inadequate or partial response to surgery and/or radiotherapy **OR** there is a clinical reason for why the patient has not had surgery or radiotherapy (e.g., medically unstable conditions, patient is at high risk for complications of anesthesia because of airway difficulties, lack of an available skilled surgeon, patient refuses surgery or prefers the medical option over surgery, major systemic manifestations of acromegaly including cardiomyopathy, severe hypertension and uncontrolled diabetes).<br>• Members receiving treatment with Signifor LAR, excluding via samples or manufacturer's patient assistance programs, may be allowed to continue<br><br><u>Reauthorization:</u> IGF-1 level decreased or normalized.<br><br>**<u>Cushing's Disease</u>**<br>Patient meets the following criteria for initiation of therapy:<br>• Clinical evidence of Cushing's disease in whom pituitary surgery is not an option or has not been curative<br>• Mean urinary free cortisol level (mUFC) between 1.5 and 5 times the upper limit of normal<br>• Documented inadequate response, intolerable adverse event, or contraindication to ALL of the following: ketoconazole, cabergoline, mifepristone |

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| | • Initial starting dose is 10mg every 4 weeks, may be increased after 4 months to a maximum of 40mg every 4 weeks if 24-hour Urinary Free Cortisol has not normalized<br><br>Reauthorization: mUFC equal to or less than the upper limit of normal |
| **Appropriate Treatment Regimen & Other Criteria:** | • Prior to initiation of therapy hypokalemia or hypomagnesemia must be corrected.<br>• Prior to initiation of therapy baseline hemoglobin A1c (HbA1c), liver function tests, and electrocardiogram (ECG) should be obtained<br>• Blood glucose monitoring should be done weekly for the first 3 months after initiation and the first 4 to 6 weeks after dose increases<br>• New assessment of liver function should be obtained 3 weeks after initiation and then monthly for 3 months<br>• Quantity limit 1 injection (maximum 60 mg) every 28 days |
| **Exclusion Criteria:** | • Poorly controlled diabetes mellitus (HbA1c greater than 8%)<br>• Severe hepatic impairment (Child Pugh C) |
| **Age Restriction:** | • Must be 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 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:** | • Consider delaying first dose if absolute neutrophil count (ANC) is less than 1.0 x $10^9$/L, platelets are less than 75 x $10^9$/L, or hemoglobin is less than or equal to 17 g/dL<br>• Subsequent doses may be delayed if ANC is less than 1.0 x $10^9$/L, platelets are less than 50 x $10^9$/L, or hemoglobin less than or equal to 17 g/dL<br><br>**Dosing**: 11 mg/kg intravenous (IV) infusion once every 3 weeks until treatment failure<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:** | • Cytokine release syndrome: 1 month, unless otherwise specified |

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| | • All other indications:<br>  ○ Initial Authorization: 4 months, unless otherwise specified<br>  ○ Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**SIPONIMOD**
Affected Medications: MAYZENT (siponimod)

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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), to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease, in adults |
| **Required Medical Information:** | • Documentation of diagnosis of relapsing forms of Multiple Sclerosis (MS) confirmed with magnetic resonance imaging (MRI) (Revised McDonald diagnostic criteria for MS)<br>• Documentation of electrocardiogram (ECG), complete blood count (CBC), liver function tests, ophthalmic evaluation, and CYP2C9 genetic testing<br>• Documentation of antibodies to varicella zoster virus (VZV) or vaccination of antibody-negative patients prior to treatment initiation<br><br><u>Secondary-Progressive MS (SPMS)</u><br>• Documentation of prior history of relapsing-remitting MS (RRMS) with progressive increase in disability over at least 6 months, independent of, or in the absence of, relapses<br>• Documentation of active disease classified as the presence of clinical relapse or inflammatory activity (i.e., new or enlarging T2 lesions or gadolinium enhancing lesions on MRI) in the last 2 years<br>• Documentation of Expanded Disability Status Scale (EDSS) score of 3.0 to 6.5 |
| **Appropriate Treatment Regimen & Other Criteria:** | • After treatment titration, the recommended maintenance dosage of Mayzent is 2 mg taken orally once daily starting on Day 6. Dosage adjustment is required in patients with a CYP2C9*1/*3 or *2/*3 genotype<br>  ○ If one titration dose is missed for more than 24 hours, treatment needs to be reinitiated with Day 1 of the titration regimen |

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| | ○ In patients with a CYP2C9*1/*3 or *2/*3 genotype, after treatment titration, the recommended maintenance dosage of Mayzent is 1 mg taken orally once daily starting on Day 5<br>• Discontinuation of treatment if liver enzymes exceed three times the upper limit of normal (ULN) with signs of liver dysfunction (unexplained nausea, vomiting, abdominal pain, fatigue, anorexia, rash with eosinophilia, or jaundice and/or dark urine)<br><br>**Reauthorization:** provider attestation of treatment success |
| **Exclusion Criteria:** | • CYP2C9*3/*3 genotype<br>• Recent (in the past 6 months) myocardial infarction (MI), unstable angina, stroke, transient ischemic attack (TIA), decompensated heart failure requiring hospitalization, or class III or IV heart failure<br>• Mobitz type II second or third degree atrioventricular (AV) block or sick sinus syndrome (unless patient has functioning pacemaker) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or multiple sclerosis specialist<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:
**SODIUM PHENYLBUTYRATE**
Affected Medications: Buphenyl, sodium phenylbutyrate

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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 Urea Cycle Disorder (UCD)<br>• Diagnosis confirmed by blood, enzymatic, biochemical, or genetic testing<br>• The prescribed medication will be used for chronic management of UCD<br>• The patient has UCD that cannot be managed by dietary protein restriction and/or amino acid supplementation alone |
| **Appropriate Treatment Regimen & Other Criteria:** | • The prescribed medication will be used in combination with dietary protein restriction<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Should not be used in the treatment of acute hyperammonemia |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise specified |

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

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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>• Confirmed by Sleep Lab Evaluation<br>• An Epworth Sleepiness Scale score of at least 17 at baseline<br><br>**Obstructive Sleep Apnea**<br>• Confirmed by polysomnography<br>• Documentation of current CPAP utilization<br>• An Epworth Sleepiness Scale score of at least 15 at baseline |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented trial and failure, or contraindication, to all the following:<br>  ○ Modafinil<br>  ○ Armodafinil<br>  ○ Methylphenidate or dextroamphetamine or lisdexamfetamine<br>• Reauthorization:<br>  ○ Clinically significant improvement in activities of daily living and in Epworth Sleepiness Scale score |
| **Exclusion Criteria:** | • Work related conditions |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a sleep 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 |

POLICY NAME:
**SOLARAZE**
Affected Medications: SOLARAZE (diclofenac sodium 3% topical gel)

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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 treating diagnosis, including number and distribution of actinic keratosis lesions<br>• Documentation of all therapies tried/failed. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Approval requires documentation of inadequate response or intolerance to at least 2 alternative therapies used in the management of actinic keratosis such as 5-fluorouracil, imiquimod, ingenol mebutate, or photodynamic therapy<br>• Documentation of use for the shortest duration of time, consistent with patient treatment goals<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • Age greater than or equal to 18 years |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Maximum of 3 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>**<u>Octreotide, Sandostatin LAR:</u>**<br>▪ Acromegaly<br>▪ Symptomatic treatment of metastatic carcinoid tumors (carcinoid syndrome)<br>▪ Symptomatic treatment of vasoactive intestinal peptide tumors (VIPomas)<br>**<u>Lanreotide, Somatuline Depot:</u>**<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><br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | **<u>Acromegaly</u>**<br>• Initiation of therapy, patient meets the following:<br>○ Clinical evidence of acromegaly<br>○ Pre-treatment high insulin-like growth factor (IGF-1) level for age/gender<br>○ Patient had an inadequate or partial response to surgery and/or radiotherapy OR there is a clinical reason for avoidance of surgery or radiotherapy<br>○ Clinical reasons for avoidance of surgery or radiotherapy include:<br>▪ Medically unstable conditions<br>▪ Patient is at high risk for complications of anesthesia because of airway difficulties<br>▪ Lack of an available skilled surgeon |

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| | ▪ Patient refuses surgery or prefers the medical option over surgery<br>▪ Major systemic manifestations of acromegaly including cardiomyopathy<br>▪ Severe hypertension<br>▪ Uncontrolled diabetes<br><br>**<u>All other indications</u>**<br>Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>All indications</u>**<br>• May use the long-acting IM depot formulation as initial therapy OR may consider 1 or 2 doses of subcutaneous (SQ) octreotide to assess tolerability prior to starting the long-acting IM depot<br>• For patients experiencing breakthrough symptoms while taking the long-acting depot, supplementary doses of SQ octreotide may be necessary<br><br>**<u>Sandostatin LAR</u>**<br>• Coverage for the non-preferred product **Sandostatin LAR** is provided when **ONE** of the following criteria is met:<br>o Currently receiving treatment with Sandostatin LAR, excluding when the product is obtained as samples or via manufacturer's patient assistance programs<br>o Documented inadequate response or intolerable adverse event with Lanreotide, Somatuline Depot, OR Somavert (pegvisomant; acromegaly only)<br><br>**<u>Lanreotide, Somatuline Depot</u>**<br>• GEP-NETs must use 120 mg injection<br><br>**<u>Reauthorization:</u>**<br>• **Acromegaly:** requires that the IGF-1 level is decreased or normalized<br>• **All other indications:** requires documentation of disease responsiveness to therapy |

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| **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 Approval: 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 |
| **Required Medical Information:** | Patient meets the following criteria for initiation of therapy:<br>• Clinical evidence of acromegaly,<br>• Pre-treatment high IGF-1 level for age/gender,<br>• Patient has had an inadequate or partial response to surgery and/or radiotherapy **OR** there is a clinical reason for why the patient has not had surgery or radiotherapy (e.g., medically unstable conditions, patient is at high risk for complications of anesthesia because of airway difficulties, lack of an available skilled surgeon, patient refuses surgery or prefers the medical option over surgery, major systemic manifestations of acromegaly including cardiomyopathy, severe hypertension and uncontrolled diabetes).<br>• For continuation of therapy, the IGF-1 level decreased or normalized. |
| **Appropriate Treatment Regimen & Other Criteria:** | |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |

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

POLICY NAME:
**SPESOLIMAB-SBZO**
Affected Medications: SPEVIGO (spesolimab-SBZO injection)

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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<br>• 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) |
| **Appropriate Treatment Regimen & Other Criteria:** | • 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 |

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| | • Drug-induced acute generalized exanthematous pustulosis |
| **Age Restriction:** | • Adults 18 years of age or 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 dermatologist |
| **Coverage Duration:** | • Authorization: One month with no reauthorization, unless otherwise specified |

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

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design<br>○ Indicated, in conjunction with an oral antidepressant, for the treatment of:<br>- Treatment-resistant depression (TRD) in adults<br>- Depressive symptoms in adults with major depressive disorder (MDD) with acute suicidal ideation or behavior |
| **Required Medical Information:** | **<u>Diagnosis of Treatment-Resistant Depression (TRD)</u>**<br>• Assessment of patient's risk for abuse or misuse<br>• Baseline Patient Health Questionnaire-9 (PHQ-9) score (or other standard rating scale)<br><br>**<u>Diagnosis of Major Depressive Disorder (MDD) with acute suicidal ideation or behavior:</u>**<br>• Assessment of patient's risk for abuse or misuse<br>• Montgomery-Asberg Depression Rating Scale (MADRS) total score greater than 28, PHQ-9 score greater than 15, or other standard rating scale indicating severe depression |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Treatment-Resistant Depression:</u>**<br>• Documentation of one of the following:<br>○ Failure to clinically respond to four trials of antidepressant drugs at highest tolerated doses for at least 6 weeks from two or more different classes during the current depressive episode as defined by a less than 50% reduction in symptom severity using a standard rating scale that reliably measures depressive symptoms (such as PHQ-9), and at least one trial must have used an augmentation strategy (aripiprazole, lithium, olanzapine, quetiapine, risperidone, thyroid hormone) OR<br>○ Demonstrated intolerance to four antidepressant drugs with distinct side effects<br>• 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 or similar rating scale for depressive symptoms

- Will use Spravato in addition to new oral antidepressant therapy
- **Reauthorization** (for TRD indication only) requires:
  - Documentation of treatment success defined as at least a 50% reduction in symptoms of depression compared to baseline using a standard rating scale that reliably measures depressive symptoms
  - Spravato continues to be used in addition to antidepressant therapy
- Dosing according to the approved label:

| | | Adults |
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| Induction Phase | Weeks 1 to 4 | Day 1 starting dose: 56 mg |
| | Administer twice per week | Subsequent doses: 56 mg or 84 mg |
| Maintenance Phase | Weeks 5 to 8 | |
| | Administer once weekly | 56 mg or 84 mg |
| | Week 9 and after | |
| | Administer every 2 weeks or once weekly* | 56 mg or 84 mg |

*Dosing frequency should be individualized to the least frequent dosing to maintain remission/response

**Major depressive disorder (MDD) with acute suicidal ideation or behavior:**

- Documentation of current inpatient psychiatric hospitalization OR documentation of why patient is not currently at inpatient level of care
- Newly initiated or optimized oral antidepressant (AD) (AD monotherapy or AD plus augmentation therapy)

Dosing: 84 mg twice weekly for 4 weeks maximum (No reauthorization unless requirements for TRD met)

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| **Exclusion Criteria:** | • 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:** | • REMS Program certified (others will be unable to order drug)<br>• Behavioral health specialist |
| **Coverage Duration:** | Initial authorization<br>• Major depressive disorder (MDD) with acute suicidal ideation or behavior: 1 month (limit #24 nasal spary 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>Reauthorization (TRD indication only): 6 months, unless otherwise specified |

POLICY NAME:
**STIMULANTS**
Affected Medications: ***All drugs used for treatment of ADHD***

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design.<br>• New starts only |
| **Required Medical Information:** | |
| **Appropriate Treatment Regimen & Other Criteria:** | • For patients 6-12 years old newly prescribed a stimulant medication, providers must schedule the following clinic visits:<br>  ○ One initial face-to-face visit to evaluate the safety & effectiveness of the medication within 30 days of the initial prescription<br>  • Two continuation and maintenance visits, with one being face-to-face, between 31–300 days.<br>  • Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • Criteria applies to ages 6-12 years |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**STIRIPENTOL**
Affected Medications: Diacomit capsules, Diacomit powder for suspension

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Patient 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<br>• Documented treatment and inadequate control of seizures with at least four guideline directed therapies including:<br>o Valproate and<br>o Onfi and<br>o Topiramate and<br>o Clonazepam, levetiracetam, or zonisamide |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing: 50 mg/kg/day, in 2 or 3 divided doses, not to exceed 3,000mg/day<br>• Reauthorization will require documentation of at least 50% reduction in generalized clonic or tonic-clonic seizure frequency |
| **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:** | • Initial Authorization: 3 months, unless otherwise specified<br>• Reauthorization: 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:** | • Baseline 6 minute walk test<br>• Bone density testing (such as DEXA scan)<br><br>**<u>Diagnosis of Perinatal/Infantile or Juvenile onset hypophosphatasia (HPP) with ALL of the following:</u>**<br><br>• Age of onset less than 18 years<br>• Clinical manifestations consistent with hypophospatasia at onset prior to age 18 including any of the following: vitamin B6 dependent seizures, skeletal abnormalities (such as rachitic chest deformity or bowed arms/legs), failure to thrive<br>• Radiographic imaging to support presence of skeletal abnormalities<br>• Molecular genetic test confirming mutations in the ALPL gene that encodes the tissue nonspecific isoenzyme of ALP (TNSALP)<br>• Low level of serum alkaline phosphatase (ALP) evidenced by lab result below lab standard for age and gender adjusted normal range<br>• One of the following:<br>  ○ elevated (urine or serum) concentration of phosphoethanolamine (PEA)<br>  ○ elevated serum concentration of pyridoxal 5'-phosphate (PLP) in the absence of vitamin supplements within one week prior to the test<br>  ○ elevated urinary inorganic pyrophosphate (PPi) |
| **Appropriate Treatment Regimen & Other Criteria:** | Weight based dosing according to package insert (following recommendations for appropriate vial size selection)<br><br>**<u>Perinatal/Infantile-Onset HPP</u>**<br>• Maximum dose – 9 mg/ kg per week |

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| | **<u>Juvenile-Onset HPP</u>**<br>• Maximum dose – 6 mg/ kg per week<br>**Please note 80mg/0.8ml vial is for patients greater than 40kg<br><br>**<u>Reauthorization requires documentation of</u>**:<br><br>• All of the above criteria at time of initiation<br>• Laboratory results confirming a decrease in urine concentration of phosphoethanolamine (PEA), serum concentration of pyridoxal 5'-phosphate (PLP) or urinary inorganic pyrophosphate (PPi)<br>• Chart notes showing one or more of the following<br>o Radiographic evidence of improvement in skeletal deformities or growth<br>o Improvement in 6 minute walk test<br>o Improved bone density<br>o Reduction in fractures |
| **Exclusion Criteria:** | • Adult-onset hypophosphatasia |
| **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 endocrinologist OR specialist experienced in the treatment of metabolic bone disorders |
| **Coverage Duration:** | • Initial approval: 4 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

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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, 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) |
| **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>• One of the following:<br>○ Documented 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 |

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| | - Two or more deep-seated infections including septicemia<br>• A documented deficiency in producing antibodies in response to vaccination<br>o Titers that were drawn before challenging with vaccination AND<br>o 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 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>o Partial motor conduction block in at least two motor nerves or in 1 nerve plus one other demyelination criterion listed here in at least 1 other nerve<br>o Distal compound muscle action potential (CMAP) duration increase in at least 1 nerve plus one other demyelination criterion listed here in at least 1 other nerve<br>o Abnormal temporal dispersion conduction must be present in at least 2 motor nerves<br>o Reduced conduction velocity in at least 2 motor nerves<br>o Prolonged distal motor latency in at least 2 motor nerves<br>o Absent F wave in at least two motor nerves plus one other demyelination criterion listed here in at least 1 other nerve<br>o Prolonged F wave latency in at least 2 motor nerves<br>• Cerebrospinal fluid (CSF) analysis indicates the following:<br>o CSF white cell count of less than 10 cells/$mm^3$ **AND** |

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| | ○ CSF protein is elevated |
| **Appropriate Treatment Regimen & Other Criteria:** | • Meets all criteria for IVIG approval<br>• Exceptions may be given for patients without prior intravenous (IV) or subcutaneous (SC) immune globulin use<br><br>**PID**<br>• Documentation of at least 3 months of IVIG therapy<br>**CIDP**<br>• Hizentra and Gamunex-c only<br>• Refractory to or intolerant of corticosteroids (prednisolone, prednisone) given in therapeutic doses over at least three months<br><br>**Renewal Criteria**<br>• PID: Renewal requires documented disease response defined as a decrease in the frequency or severity of infections<br>• CIDP: Renewal requires documentation of a beneficial clinical response to maintenance therapy, without relapses, based on an objective clinical measuring tool;<br>OR<br>• Re-initiating maintenance therapy after experiencing a relapse while on Hizentra; AND documented improvement and stability on IVIG treatment AND was NOT receiving maximum dosing of Hizentra prior to relapse |
| **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 rheumatologist with CIDP expertise |
| **Coverage Duration:** | Initial Authorization<br>• CIDP: 3 months, unless otherwise specified<br>• PID: 12 months, unless otherwise specified |

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| | Reauthorization: 12 months, unless otherwise specified |

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

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design<br>  ○ Cold Agglutinin Disease |
| **Required Medical Information:** | **<u>Cold Agglutinin Disease (CAD)</u>**<br>• Documentation of 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 total 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<br>• History of recent blood transfusion for Cold Agglutinin Disease in the past 6 months |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Cold Agglutinin Disease (CAD)</u>**<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>**<u>Reauthorization</u>**: documentation of disease responsiveness to therapy |
| **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 |

POLICY NAME:
**SYMDEKO**
Affected Medications: SYMDEKO (tezacaftor/ivacaftor tablets)

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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 cystic fibrosis (CF) diagnosis.<br>• Documentation of Homozygous for the F508 del mutation by Food and Drug Administration (FDA)-cleared CF mutation test on both alleles of the CFTR gene or who have at least one mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene that is responsive to tezacaftor/ivacaftor based on in vitro data and/or clinical evidence.<br>• Baseline forced expiratory volume in 1 second (FEV1)<br>• Documentation of baseline and follow-up liver function tests |
| **Appropriate Treatment Regimen & Other Criteria:** | • **Reauthorization** requires documentation of improvement in FEV1 from baseline, documentation of follow-up liver function tests |
| **Exclusion Criteria:** | • Concurrent use of strong CYP3A inducers: rifampin, rifabutin, phenobarbital, carbamazepine, phenytoin, and St. John's wort |
| **Age Restriction:** | • 6 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 approval: 6 months, unless otherwise specified<br>• Reauthorization: 12 months unless otherwise specified |

POLICY NAME:
**SYMLIN**
Affected Medications: SYMLINPEN, SYMLINPEN 120, SYMLINPEN 60

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications<br>• Patient has type 1 or 2 diabetes mellitus. |
| **Required Medical Information:** | |
| **Appropriate Treatment Regimen & Other Criteria:** | • If patient received Symlin in previous 3 months, patient demonstrated an expected reduction in HbA1c since starting Symlin therapy. **OR**<br>• The patient has inadequate glycemic control (HbA1c > 7%). **AND**<br>• Patient is currently receiving optimal mealtime insulin therapy.<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Severe hypoglycemia that required assistance during the past 6 months.<br>• Gastroparesis.<br>• Patient requires drug therapy to stimulate gastrointestinal motility.<br>• Hypoglycemia unawareness (i.e., inability to detect and act upon the signs or symptoms of hypoglycemia).<br>• HbA1c level greater than 9 percent.<br>• Weight loss treatment. |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**TAFAMIDIS**
Affected Medications: VYNDAQEL (tafamidis meglumine 20 mg), VYNDAMAX (tafamidis 61 mg)

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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 the cardiomyopathy of wild-type or hereditary transthyretin-mediated amyloidosis (ATTR-CM) in adults to reduce cardiovascular mortality and cardiovascular-related hospitalization. |
| **Required Medical Information:** | • Documented diagnosis of cardiomyopathy of wild-type (ATTRwt) or hereditary (ATTRm) transthyretin-mediated amyloidosis confirmed by<br>○ Presence of amyloid deposits on analysis of cardiac biopsy specimens<br>OR<br>○ Presence of grade 2 or 3 positive bone tracer cardiac scintigraphy in the absence of monoclonal protein (i.e., free light chain ratio is normal and serum and urine immunofixation results are both normal)<br>• Genetic test results identifying a mutation in the transthyretin (TTR) gene (Val122Ile or Thr60Ala mutation) or wild-type amyloidosis<br>○ For those with ATTRwt: documented presence of transthyretin precursor protein confirmed on immunohistochemical analysis, scintigraphy, or mass spectrometry is required<br>• Cardiac involvement has been confirmed by echocardiography or cardiac magnetic resonance imaging<br>• Diagnosis of heart failure with NYHA Class I to III symptoms |
| **Appropriate Treatment** | • Maximum dosing<br>○ Vyndaqel 80 mg (four 20 mg capsules) |

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| **Regimen & Other Criteria:** | ○ Vyndamax 61 mg (one 61 mg capsule)<br>• Reauthorization: Documentation of treatment success |
| **Exclusion Criteria:** | • Heart Failure NYHA Class IV<br>• Presence of light-chain amyloidosis<br>• Prior liver or heart transplant<br>• Implanted cardiac device<br>• Concurrent use with Onpattro or Tegsedi |
| **Age Restriction:** | • 18 years and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a cardiologist or a physician who specializes in the treatment of amyloidosis<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:
**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 |
| **Required Medical Information:** | • Diagnosis of blastic plasmacytoid dendritic cell neoplasm (BPDCN) made by a board certified Hematopathologist or Dermatopathologist.<br>• If diagnosis of BPDCN is based on skin biopsy, clear plasmacytoid dendritic blast cells are present by morphology and confirmed by IHC and flow cytometry. Features excluding AML and Leukemia cutis must be present.<br>• If BPDCN presents as the leukemic form or it there is bone marrow involvement, AML, T-cell lymphoblastic leukemia, and NK-cell Leukemia must be excluded.<br>• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course. |
| **Appropriate Treatment Regimen & Other Criteria:** | • The recommended dose and schedule is 12 mcg/kg administered intravenously over 15 minutes once daily on day 1 to 5 of a 21-day cycle.<br><br><u>Reauthorization:</u> documentation of disease responsiveness to therapy |
| **Exclusion Criteria:** | • Renal toxicity: Withhold tagraxofusp until serum creatinine is less than or equal to 1.8 mg/dL or CrCl is greater than or equal to 60 mL/minute.<br>• Hepatotoxicity: Withhold tagraxofusp until AST and/or ALT are less than or equal to 2.5 times ULN<br>• Persistent clinically significant toxicities from prior chemotherapy<br>• Receiving immunosuppressive therapy<br>• Pregnancy |
| **Age Restriction:** | • For adults and pediatric patients 2 years and older only |

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| **Prescriber/Site of Care Restrictions:** | • Must be 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 approval: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**TALIGLUCERASE**
Affected Medications: ELELYSO (taliglucerase alfa)

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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 Gaucher Disease<br>• Diagnosis confirmed by enzyme assay demonstrating a deficiency of beta-glucocerebrosidase enzyme activity<br>• At least one of the following disease complications: anemia, thrombocytopenia, bone disease, hepatomegaly, or splenomegaly |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dosing: 60 units/kg every 2 weeks; dosing is individualized based on disease severity<br>  ○ Supplied as 200 unit vials<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Patients currently taking miglustat (Zavesca) or eliglustat (Cerdelga) |
| **Age Restriction:** | • 4 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subjects to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 months, unless otherwise specified |

POLICY NAME:
**TARGETED IMMUNE MODULATORS**
PA Policy Applicable to:
**Preferred Drugs:** Humira, Enbrel, Cosentyx, Otezla, Tremfya, Stelara, Xeljanz, Skyrizi, Rinvoq
**Preferred Medical Drugs:** Inflectra, Renflexis, Stelara, Simponi Aria Intravenous
**Non-preferred Medical Drugs:** Remicade, Entyvio, Orencia Intravenous, Actemra Intravenous, Avsola, Infliximab (J1745)

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| 1. Is the request for continuation of currently approved therapy? | Yes – Go to renewal criteria | No – Go to #2 |
| 2. Is the request for combined treatment with multiple targeted immune modulators (E.g., Humira plus Otezla) | Yes – Criteria not met, experimental | No – Go to #3 |
| 3. Is the request for Xeljanz, Xeljanz XR or Rinvoq | Yes – Go to #4 | No – Go to #5 |
| 4. Has there been an inadequate response or intolerance to one or more tumor necrosis factor (TNF) inhibitors? | Yes – Go to #5 | No – Criteria not met |
| 5. Is the diagnosis being treated with a preferred pharmacy drug or covered medical infusion drug according to one of the indications below? | Yes – Go to appropriate section below | No – Criteria not met |
| **Rheumatoid Arthritis (RA)**<br>**Preferred Pharmacy Drugs – Humira, Enbrel, Xeljanz, Rinvoq**<br>**Preferred Medical Drugs –Inflectra, Renflexis, Simponi Aria Intravenous**<br>**Non-Preferred Medical Drugs –Remicade, Actemra IV, Orencia IV, Infliximab (J1745), Avsola** | | |

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| 1. Is there documented current disease activity with one of the following (or equivalent objective scale)?<br>○ The Disease Activity Score derivative for 28 joints (DAS-28) greater than 3.2<br>○ The Clinical Disease Activity Index (CDAI) greater than 10<br>○ Weighted RAPID3 of at least 2.3 | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documented treatment failure with at least 12 weeks of combination disease-modifying antirheumatic drug (DMARD) therapy<br>○ Methotrexate plus sulfasalazine, methotrexate plus hydroxychloroquine, sulfasalazine plus hydroxychloroquine, leflunomide plus sulfasalazine, or leflunomide plus hydroxychloroquine | Yes – Go to #3 | No – Criteria not met |
| 3. Is the request for a non-preferred medical drug? | Yes – Go to #4 | No – Go to #5 |
| 4. Is there documented failure with one of the preferred pharmacy drugs (Humira, Enbrel, Xeljanz, Rinvoq) AND one of the preferred medical drugs (Inflectra, Renflexis, Simponi Aria)? | Yes – Document and Go to #5 | No – Criteria not met |
| 5. Is the drug prescribed by, or in consultation with, a rheumatology specialist? | Yes – 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 – Approve up to 6 months | No – Criteria not met |

| **Plaque Psoriasis (PP)**<br>**Preferred Pharmacy Drugs – Humira, Enbrel, Cosentyx, Otezla, Stelara, Skyrizi, Tremfya**<br>**Preferred Medical Drugs – Inflectra, Renflexis, Stelara**<br>**Non-Preferred Medical Drugs – Remicade, Infliximab (J1745), Avsola** | | |
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| 1. Is there documentation that the skin disease meets one of the following:<br>○ At least 10% body surface area involvement despite current treatment<br>○ Hand, foot or mucous membrane involvement | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is the request for Otezla? | Yes – Go to #3 | No – Go to #4 |
| 3. Is there documented clinical failure with at least one systemic therapy for a minimum of 12 weeks (methotrexate, cyclosporine, Acitretin, phototherapy [UVB, PUVA])? | Yes – Document and go to #7 | No – Criteria not met |
| 4. Is there documented treatment failure with 12 weeks of at least two systemic therapies (methotrexate, cyclosporine, Acitretin, phototherapy [UVB, PUVA])? | Yes – Document and go to #5 | No – Criteria not met |
| 5. Is the request for Remicade, Avsola or Infliximab (J1745)? | Yes – Go to #6 | No – Go to #7 |
| 6. Is there documented treatment failure or intolerable adverse event with the biosimilar drugs (Inflectra, Renflexis), and the adverse event was not an expected adverse event attributed to the active ingredient? | Yes – Go to #7 | No – Criteria not met; Remicade requires failure with the biosimilar infliximab products |

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| 7. Is the drug prescribed by, or in consultation with, a dermatology specialist? | Yes – Go to #8 | No – Criteria not met |
| 8. 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 |
| **Psoriatic Arthritis (PsA)**<br>**Preferred Pharmacy Drugs – Humira, Enbrel, Otezla, Cosentyx, Xeljanz, Stelara, Tremfya, Rinvoq, Skyrizi**<br>**Preferred Medical Drugs – Inflectra, Renflexis, Stelara, Simponi Aria**<br>**Non-Preferred Medical Drugs – Remicade, Orencia IV, Infliximab (J1745), Avsola** | | |
| 1. Is there documentation of CASPAR criteria score 3 or greater based on chart notes:<br>○ Skin psoriasis: present – two points, **OR** previously present by history – one point, **OR** a family history of psoriasis, if the patient is not affected – one point<br>○ Nail lesions (onycholysis, pitting): one point<br>○ Dactylitis (present or past, documented by a rheumatologist): one point<br>○ Negative rheumatoid factor (RF): one point<br>○ Juxtaarticular bone formation on radiographs (distinct from osteophytes): one point | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documented failure with at least 12 weeks of treatment with methotrexate, or if unable to tolerate methotrexate or contraindications apply, another disease modifying antirheumatic drug | Yes – Document and go to #3 | No – Criteria not met |

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| (sulfasalazine, cyclosporine, leflunomide)? | | |
| 3. Is the request for a non-preferred medical drug? | Yes – Go to #4 | No – Go to #5 |
| 4. Is there documented failure with one of the preferred pharmacy drugs (Humira, Enbrel, Cosentyx, Otezla, Stelara, Xeljanz, Tremfya, Rinvoq, Skyrizi) AND one of the preferred medical drugs (Inflectra, Renflexis, Simponi Aria)? | Yes – Go to #5 | No – Criteria not met |
| 5. Is the drug prescribed by, or in consultation with, a rheumatology specialist? | Yes – 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 – Approve up to 6 months | No – Criteria not met |
| **Ankylosing Spondylitis (AS) & Non-radiographic Axial Spondyloarthritis (nr-axSpA) & Psoriatic Arthritis with Axial Involvement**<br>**Preferred Pharmacy Drugs – Humira, Enbrel, Cosentyx, Xeljanz, Rinvoq**<br>**Preferred Medical Drugs –Inflectra, Renflexis, Simponi Aria**<br>**Non-preferred Medical Drugs –Remicade, Infliximab (J1745), Avsola** | | |
| 1. Is there a diagnosis of axial spondyloarthritis (SpA) confirmed by sacroiliitis on imaging AND at least 1 Spondyloarthritis (SpA) feature:<br>○ Inflammatory back pain (4 of 5 features met):<br>▪ Onset of back discomfort before the age of 40 years<br>▪ Insidious onset<br>▪ Improvement with exercise | Yes – Go to #2 | No – Criteria not met |

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| ▪ No improvement with rest<br>▪ Pain at night (with improvement upon arising)<br>○ Arthritis<br>○ Enthesitis<br>○ Uveitis<br>○ Dactylitis (inflammation of entire digit)<br>○ Psoriasis<br>○ Crohn's disease/ulcerative colitis<br>○ Good response to NSAIDs<br>○ Family history of SpA<br>○ Elevated CRP<br>**OR**<br>○ HLA-B27 genetic test positive AND at least 2 SpA features | | |
| 2. Is there documentation of active disease defined by Bath ankylosing spondylitis disease activity index (BASDAI) at least 4 or equivalent objective scale? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documented failure with two daily prescription strength nonsteroidal anti-inflammatory drugs (ibuprofen, naproxen, diclofenac, meloxicam, etc.) with minimum 1 month trial each?<br>OR<br>For isolated sacroiliitis, enthesitis, peripheral arthritis: documented treatment failure with locally administered parenteral glucocorticoid? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is the request for a non-preferred medical drug? | Yes – Go to #5 | No – Go to #6 |

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| 5. Is there documented failure with one of the preferred pharmacy drugs (Humira, Enbrel, Cosentyx, Xeljanz, Rinvoq) AND one of the preferred medical drugs (Inflectra, Renflexis, Simponi Aria)? | Yes – Go to #6 | No – Criteria not met |
| 6. Is the drug prescribed by, or in consultation with, a rheumatology 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 6 months | No – Criteria not met |
| **Crohn's Disease**<br>**Preferred Pharmacy Drugs – Humira, Stelara, Skyrizi**<br>**Preferred Medical Drugs – Inflectra, Renflexis, Stelara**<br>**Non-preferred Medical Drugs –Remicade, Entyvio, Infliximab (J1745), Avsola** | | |
| 1. Is there moderate to severely active disease despite current treatment? | Yes – Go to #2 | No – Criteria not met |
| 2. Is there documented failure with at least two oral treatments for a minimum of 12 weeks: corticosteroids, azathioprine, 6-mercaptopurine, methotrexate, sulfasalazine, balsalazide?<br>**OR**<br>Documentation of previous surgical intervention for Crohn's disease? | Yes – Document and go to #4 | No –Go to #3 |
| 3. Is there documented severe, high-risk disease on colonoscopy defined by one of the following:<br>○ Fistulizing disease<br>○ Stricture | Yes – Document and go to #4 | No – Criteria not met |

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| ○ Presence of abscess/phlegmon<br>○ Deep ulcerations<br>○ Large burden of disease including ileal, ileocolonic, or proximal gastrointestinal involvement | | |
| 4. Is the request for a non-preferred medical drug? | Yes – Go to #5 | No – Go to #6 |
| 5. Is there documented failure with one of the preferred pharmacy drugs (Humira, Stelara, Skyrizi) AND one of the preferred medical drugs (Inflectra, Renflexis)? | Yes – Go to #6 | No – Criteria not met |
| 6. Is the drug prescribed by, or in consultation with, a gastroenterology 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 6 months | No – Criteria not met |
| **Ulcerative Colitis (UC)**<br>**Preferred Pharmacy Drugs – Humira, Rinvoq, Xeljanz, Stelara**<br>**Preferred Medical Drugs –Inflectra, Renflexis, Stelara**<br>**Non-Preferred Medical Drugs –Remicade, Entyvio, Infliximab (J1745), Avsola** | | |
| 1. Is there a diagnosis supported by endoscopy/colonoscopy/sigmoidoscopy or biopsy with moderate to severely active disease despite current treatment? | Yes – Go to #2 | No – Criteria not met |
| 2. Is there severely active disease despite current treatment defined by greater than or equal to 6 bloody, loose stools per day with severe cramps and evidence of systemic toxicity (fever, tachycardia, | Yes – Document and got to #4 | No – Go to #3 |

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| anemia, and/or elevated CRP/ESR), or recent hospitalization for ulcerative colitis? | | |
| 3. Is there documented failure with at least two oral treatments for a minimum of 12 weeks: corticosteroids, sulfasalazine, azathioprine, mesalamine, balsalazide, cyclosporine, 6-mercaptopurine | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is the request for a non-preferred medical drug? | Yes – Go to #5 | No – Go to #6 |
| 5. Is there documented failure with one of the preferred pharmacy drugs (Humira, Xeljanz, Stelara, Rinvoq) AND one of the preferred medical drugs (Inflectra, Renflexis)? | Yes – Go to #6 | No – Criteria not met |
| 6. Is the drug prescribed by, or in consultation with, a gastroenterology 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 6 months | No – Criteria not met |
| **Juvenile Idiopathic Arthritis (JIA)**<br>**Preferred Pharmacy Drugs – Humira, Enbrel, Xeljanz**<br>**Preferred Medical Drug – Simponi Aria**<br>**Non-Preferred Medical Drugs – Orencia IV, Actemra IV** | | |
| 1. Is there documented current level of disease activity with physician global assessment (MD global score) or active joint count? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documented failure with glucocorticoid joint injections or oral | Yes – Go to #3 | No – Criteria not met |

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| corticosteroids AND<br>At least one of methotrexate or leflunomide for a minimum of 12 weeks? | | |
| 3. Is the request for a non-preferred medical drug? | Yes – Go to #4 | No – Go to #5 |
| 4. Is there documented failure with one of the preferred pharmacy drugs (Humira, Enbrel Xeljanz) AND a preferred medical drug (Simponi Aria)? | Yes – Go to #5 | No – Criteria not met |
| 5. Is the drug prescribed by, or in consultation with, a rheumatologist? | Yes – 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 – Approve up to 6 months | No – Criteria not met |
| **Uveitis - Humira** | | |
| 1. Is there a confirmed diagnosis of noninfectious uveitis? | Yes – Go to #2 | No – Criteria not met |
| 2. Is the diagnosis being treated intermediate or panuveitis? | Yes – Go to #5 | No – Go to #3 |
| 3. Is the diagnosis being treated posterior uveitis? | Yes – Go to #6 | No – Go to #4 |
| 4. Is the diagnosis being treated anterior uveitis? | Yes – Criteria not met | |

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| 5. Is there documented treatment failure with at least one immunosuppressive agent: methotrexate, azathioprine, mycophenolate AND at least one calcineurin inhibitor (cyclosporine, tacrolimus)? | Yes – Go to #7 | No – Criteria not met |
| 6. Is there documented treatment failure with Yutiq AND Retisert? | Yes – Go to #7 | No – Criteria not met |
| 7. Is the drug prescribed by, or in consultation with, an ophthalmology specialist? | Yes – Go to #8 | No – Criteria not met |
| 8. 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 |
| **Hidradenitis Suppurativa (HS)**<br>**Preferred Pharmacy Drugs – Humira**<br>**Preferred Medical Drugs –Inflectra, Renflexis**<br>**Non-Preferred Medical Drugs –Remicade, Infliximab (J1745), Avsola** | | |
| 1. Is there a diagnosis of moderate to severe Hidradenitis Suppurativa (HS) [Hurley Stage II or III disease] AND Documentation of baseline count of abscess and inflammatory nodules? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documented failure with at least a 90 day trial of oral antibiotics for treatment of HS (Doxycycline/tetracycline/minocycline or | Yes – Document and go to #3 | No – Criteria not met |

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| clindamycin plus rifampin) AND 8 weeks on a retinoid (Isotretinoin, Acitretin)? | | |
| 3. Is the request for a non-preferred medical drug? | Yes – Go to #4 | No- Go to #5 |
| 4. Is there documented failure with the preferred pharmacy drug (Humira) AND one of the preferred medical drugs (Inflectra, Renflexis)? | Yes – Go to #5 | No – Criteria not met |
| 5. Is the drug prescribed by, or in consultation with, a dermatology specialist? | Yes – Go to #6 | No – Criteria not met |
| 6. Is the age of the member and 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 |
| **Giant Cell Arteritis (GCA) & Cytokine Release Syndrome (CRS) – Actemra** | | |
| 1. Is there a confirmed diagnosis of Cytokine Release Syndrome (CRS)? | Yes – Go to #4 | No – Go to #2 |
| 2. Is there a confirmed diagnosis of Giant Cell Arteritis (GCA) based on temporal artery biopsy or color doppler ultrasound OR Large vessel GCA diagnosis by advanced imaging of the vascular tree with computed tomography (CT), magnetic resonance imaging(MRI), magnetic resonance | Yes – Go to #3 | No – Criteria not met |

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| angiography (MRA), positron emission tomography (PET) or PET with CT? | | |
| 3. Is there documentation of disease refractory to treatment with glucocorticoids? | Yes – Go to #4 | No – Criteria not met |
| 4. Is the drug prescribed by, or in consultation with, a rheumatology specialist? | Yes – Go to #5 | No – Criteria not met |
| 5. Is the age of the member and requested dose within the Food and Drug Administration (FDA)-approved label and PacificSource quantity limitations? | Yes – Approve up to 6 months (Maximum 4 doses for CRS) | No – Criteria not met |
| **Oral Ulcers Associated with Behcet's Disease – Otezla** | | |
| 1. Is there a diagnosis of Behcet's with documentation of recurrent oral aphthae at least 3 times in a year AND two of the following: Recurrent genital aphthae, Eye lesions, Skin lesions, Positive pathergy test defined by a papule 2 mm or greater? | Yes – Go to #2 | No – Criteria not met |
| 2. Is there documented clinical failure of at least 1 oral medication for Behcet's disease | Yes – Go to #3 | No – Criteria not met |

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| after at least 12 weeks (colchicine, prednisone, azathioprine)? | | |
| 3. Is the drug prescribed by, or in consultation with, a specialist with experience in treating Behcet's? | Yes – Go to #4 | No – Criteria not met |
| 4. Is the age of the member and 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 |
| **Acute Graft Versus Host Disease (GVHD) Prophylaxis – Orencia Intravenous** | | |
| 1. Is there documentation of a planned hematopoietic stem cell transplant (HSCT) including procedure date, patient weight, and planned dose? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documentation that the drug will be used in combination with a calcineurin inhibitor (tacrolimus, cyclosporine) AND methotrexate? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documentation of a prior allogeneic HSCT, HIV infection or any uncontrolled active infection (viral, bacterial, fungal, or protozoal)? | Yes – Criteria not met | No – Go to #4 |
| 4. Is the drug prescribed by, or in consultation with, a hematologist or oncologist? | Yes – Approve up to 1 month (4 days of treatment maximum) with no | No – Criteria not met |

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| | reauthorization, unless otherwise specified | |
| **Atopic Dermatitis (AD) - Rinvoq** | | |
| 1. Is the request for use in combination with a monoclonal antibody (Fasenra, Nucala, Xolair, Cinqair)? | Yes – Criteria not met, combination use is experimental | No – Go to #2 |
| 2. Is there 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)? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there a documented body surface area (BSA) effected of at least 10% OR hand, foot or mucous membrane involvement? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is there documented failure with at least 6 weeks of treatment with one of the following: tacrolimus ointment, pimecrolimus cream, Eucrisa? | Yes – Document and go to #5 | No – Criteria not met |
| 5. Is there documented treatment failure with one of the following for at least 12 weeks: phototherapy, cyclosporine, azathioprine, methotrexate, mycophenolate? | Yes – Document and go to #6 | No – Criteria not met |
| 6. Is the drug prescribed by or in consultation with a specialist in the treatment of atopic dermatitis (such as a dermatologist)? | Yes – Approve up to 6 months | No – Criteria not met |

| Enthesitis-Related Arthritis (ERA) & Juvenile Psoriatic Arthritis (JPsA) Preferred Drugs – Cosentyx | | |
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| 1. Is there diagnosis of ERA confirmed by presence of the following:<br>• Arthritis persisting at least 6 weeks AND enthesitis present<br>**OR**<br>• Arthritis or enthesitis with two of the following features:<br>○ Sacroiliac tenderness or inflammatory lumbosacral pain<br>○ Positive HLA-B27<br>○ Onset of arthritis in males greater than 6 years of age<br>○ Acute symptomatic anterior uveitis<br>○ First-degree relative with ERA, sacroilitis associated with inflammatory bowel disease, reactive arthritis, or acute anterior uveitis | Yes – Document and go to #2 | No – Go to #2 |
| 2. Is there diagnosis of JPsA confirmed by presence of:<br>• Arthritis and psoriasis<br>**OR**<br>• Arthritis and at least 2 of the following:<br>○ Dactylitis<br>○ Nail pitting or onycholysis<br>○ Psoriasis in a first-degree relative | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documented treatment failure with a nonsteroidal anti-inflammatory drug (ibuprofen, naproxen, celecoxib, meloxicam, etc.) with a minimum trial of 1 | Yes – Document and go to #4 | No – Criteria not met |

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| month? | | |
| 4. Is there documented treatment failure with at least one of the following disease-modifying antirheumatic drugs (DMARDs) with a minimum trial of 12 weeks: methotrexate, sulfasalazine, leflunomide. | Yes – Document and go to #5 | No – Criteria not met |
| 5. Is the drug prescribed by, or in consultation with, a rheumatologist? | 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 – Approve up to 6 months | No – Criteria not met |
| **Generalized Pustular Psoriasis Flare**<br>**Preferred Drugs – Inflectra, Renflexis**<br>**Non-Preferred Medical Drugs – Remicade, Avsola, Infliximab (J1745)** | | |
| 1. Is there documentation of a diagnosis of generalized pustular psoriasis (GPP) confirmed by the following:<br>a. The presence of widespread sterile pustules arising on erythematous skin<br>b. Pustulation is not restricted to psoriatic plaques | Yes – Document and go to #2 | No – Criteria not met |
| 2. Are there signs and symptoms of an acute GPP flare of moderate-to-severe intensity as follows:<br>a. A Generalized Pustular Psoriasis Physician Global Assessment (GPPGA) total score of greater than or equal to 3<br>b. A GPPGA pustulation category subscore of greater than or equal to 2 | Yes – Document and go to #3 | No – Criteria not met |

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| c. Greater than or equal to 5% body surface are (BSA) covered with erythema and the presence of pustules | | |
| 3. Is there documented 1-week treatment failure with cyclosporine? | Yes – Approve up to 6 months | No – Criteria not met |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider, with clinical documentation to support? | Yes – Go to #2 | No – Criteria not met |
| 2. Is the request for combined treatment with multiple targeted immune modulators? (E.g., Humira plus Otezla) | Yes – Criteria not met | No – Go to #3 |
| 3. 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** | | |
| • **Humira**<br>  ○ Induction<br>  ▪ Plaque Psoriasis/Uveitis: 160 mg in first 28 days<br>  ▪ Crohn's/Ulcerative Colitis/HS: 160 mg day 1, then 80 mg on day 15<br>  ○ Maintenance<br>  ▪ RA/Psoriasis/Psoriatic Arthritis/Crohn's/UC/AS/Uveitis/JIA: 40 mg every 14 days<br>  ▪ HS: 40 mg every week OR 80 mg every 14 days<br>• **Enbrel**<br>  ○ Induction | | |

- Plaque Psoriasis: 8 injections per 28 days for first 3 months
  - Maintenance (All indications):
    - 50 mg once weekly dosing: 4 injections per 28 days
    - 25 mg twice weekly dosing: 8 injections per 28 days
- **Cosentyx**
  - Induction
    - Adult Plaque Psoriasis: 4 two-packs (300 mg) in first 28 days
    - Pediatric Plaque Psoriasis/Pediatric Psoriatic Arthritis/Pediatric Enthesitis-Related Arthritis:
      - Less than 50 kg: four 75 mg doses in the first 28 days
      - Greater than or equal to 50 kg: four 150 mg doses in the first 28 days
  - Maintenance
    - Adult Plaque Psoriasis: 1 two-pack (300 mg) per 28 days
    - Pediatric Plaque Psoriasis/Pediatric Psoriatic Arthritis/Pediatric Enthesitis-Related Arthritis:
      - Less than 50 kg: 75 mg per 28 days
      - Greater than or equal to 50 kg: 150 mg per 28 days
    - Psoriatic arthritis without plaque psoriasis/AS/Nr-axSpA: 1 injection (150 mg) per 28 days
      - If a patient continues to have active disease, a dosage of 300 mg may be considered
- **Otezla**
  - Induction (All indications): Titration pack
  - Maintenance (All indications): 60 tablets per 30 days
- **Stelara**
  - Induction
    - Plaque Psoriasis: One 45 mg injection (0.5 mL) in first 28 days for those weighing 60 to 100 kg, one 90 mg injection (1 mL) in first 28 days for those weighing over 100 kg
      - For those under 60kg, the dose is 0.75 mg/kg
    - Psoriatic Arthritis: One 45 mg injection (0.5 mL) in the first 28 days
      - For coexistent moderate to severe PP and weight greater than 100kg: one 90 mg injection (1 mL) in first 28 days

- Crohn's Disease and Ulcerative Colitis: A single intravenous infusion per below
  - 55 kg or less: 260 mg
  - 55 kg to 85 kg: 390 mg
  - More than 85 kg: 520 mg
- Maintenance
  - Plaque Psoriasis: One 45 mg injection (0.5 mL) per 84 days for those weighing 100 kg or less; one 90 mg injection (1 mL) per 84 days for those weighing over 100 kg
  - Psoriatic Arthritis: 45 mg (0.5 mL) per 84 days
    - For coexistent moderate-to-severe plaque psoriasis weighing more than 100 kg: 90 mg (1 ml) per 84 days
  - Crohn's Disease and Ulcerative Colitis: 90 mg (1 mL) per 56 days starting 8 weeks after the initial IV dose

- **Tremfya**
  - Induction: 100 mg (One injection) in first 28 days
  - Maintenance: 100 mg (One injection) per 56 days
- **Skyrizi**
  - PP/PsA:
    - Induction: 150 mg in the first 28 days
    - Maintenance: 150 mg per 84 days
  - Crohn's Disease:
    - Induction: 600 mg intravenous at week 0, week 4, and week 8
    - Maintenance: 360 mg subcutaneously every 8 weeks, beginning week 12
- **Rinvoq**
  - RA/PsA/AS/nr-axSpA: 15 mg once daily (30 tablets per 30 days)
  - AD: 15 mg once daily, may increase to 30 mg once daily if inadequate response (30 tablets per 30 days)
  - UC: 45 mg once daily for 8 weeks then 15 mg once daily. May increase to 30 mg once daily if inadequate response (30 tablets per 30 days).

    ****45mg limited to 56 tablets (first 8 weeks of treatment)**

- **Xeljanz**

- RA/PsA/AS: 60 tablets per 30 days (5 mg IR) OR 30 tablets per 30 days (11 mg XR)
- UC: 60 tablets per 30 days (5 mg or 10 mg IR tablets) OR 30 tablets per 30 days (11 mg or 22 mg XR)
- JIA: 10 kg to less than 20 kg: 3.2 mg (3.2 mL oral solution) twice daily; 20 kg to less than 40 kg: 4 mg (4 mL oral solution) twice daily; 40 kg or greater: 5 mg (one 5 mg tablet or 5 mL oral solution) twice daily
  - Oral solution available as 240 mL bottle

- **Infliximab (Remicade, Inflectra, Renflexis, Avsola, Infliximab (J1745))***
  - Availability: 100 mg single-dose vials
  - Crohn's/UC/HS: 5 mg/kg at 0, 2 and 6 weeks followed by 5 mg/kg every 8 weeks thereafter. For those who respond and lose response, consideration may be given to treatment with 10 mg/kg
  - Psoriatic Arthritis/Plaque Psoriasis/Generalized Pustular Psoriasis: 5 mg/kg at 0, 2 and 6 weeks followed by 5 mg/kg every 8 weeks thereafter
  - RA: 3 mg/kg at 0, 2 and 6 weeks followed by 3 mg/kg every 8 weeks thereafter. For those with an incomplete response, consideration may be given for dosing up to 10 mg/kg or as often as every 4 weeks
  - AS: 5 mg/kg at 0, 2 and 6 weeks followed by 5 mg/kg every 6 weeks thereafter
- **Simponi Aria Intravenous***
  - Availability: 50 mg single-dose vials
  - RA/PsA/AS: 2 mg/kg at weeks 0 and 4, then every 8 weeks thereafter
  - Pediatric PsA and JIA: 80 mg/m2 at weeks 0 and 4, then every 8 weeks thereafter
- **Orencia Intravenous***
  - Availability: 250 mg single-use vials
  - RA/PsA: <60 kg: 500 mg, 60-100 kg: 750 mg, >100 kg: 1,000 mg at 0, 2, and 4 weeks followed by every 4 weeks thereafter
  - JIA: 6 years and older and <75 kg: 10 mg/kg; 75-100 kg: 750 mg; >100 kg: 1,000 mg (maximum dose) at 0, 2 and 4 weeks followed by every 4 weeks thereafter
  - Acute GVHD Prophylaxis:

- 2 to less than 6 years: 15 mg/kg on day -1 (day before transplantation) followed by 12 mg/kg on days 5, 14, and 28 post-transplant
- 6 years and older: 10 mg/kg on day -1 (day before transplantation) followed by 10 mg/kg on days 5, 14, and 28 post-transplant (maximum: 1,000 mg/dose)

- **Entyvio***
  - Availability: 300 mg single-use vials
  - Crohn's/UC: 300 mg at 0, 2 and 6 weeks followed by every 8 weeks thereafter
  - For Consideration of every 4 week dosing must meet all of the following:
    - Documented clinical failure to Entyvio at standard dosing for at least 6 months
      - Clinical failure defined as failure to achieve a clinical response (greater than or equal to 70 point improvement in CDAI score for Crohn's)
    - Documented failure to minimum of 12 weeks on two alternative Tumor necrosis factor–alpha (TNF) inhibitors
- **Actemra Intravenous***
  - Availability: 400 mg, 200 mg & 80 mg single-dose vials
  - RA: 4 mg/kg once every 4 weeks; may be increased to 8 mg/kg once every 4 weeks based on clinical response (maximum dose: 800 mg)
  - CRS: For patients less than 30kg, recommended dose is 12mg/kg; patients 30kg or greater recommended dose is 8mg/kg up to maximum of 800mg (Maximum 4 doses)
  - Polyarticular JIA: <30 kg: 10 mg/kg every 4 weeks; 30 kg or greater: 8 mg/kg every 4 weeks
  - Systemic JIA: <30 kg: 12 mg/kg every 2 weeks; 30 kg or greater: 8 mg/kg every 2 weeks

*Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced for all medical infusion drugs

| Drug Name | Ankylosing Spondylitis | Crohn's Disease | Juvenile Idiopathic Arthritis | Plaque Psoriasis | Psoriatic Arthritis | Rheumatoid Arthritis | Ulcerative Colitis | Other |
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| **Abatacept (Orencia)** | | | ≥2 yo | | ≥18 yo | ≥18 yo | | Acute GVHD prophylaxis: IV: ≥2 yo |
| **Adalimumab (Humira)** | ≥18 yo | ≥6 yo (Humira) ≥18 yo (biosimilars) | ≥2 yo (Humira) ≥4 yo (biosimilars) | ≥18 yo | ≥18 yo | ≥18 yo | ≥18 yo | Uveitis (noninfectious) ≥2 yo (Humira) HS ≥12 yo |
| **Anakinra (Kineret)** | | | | | | ≥18 yo | | NOMID |
| **Apremilast (Otezla)** | | | | ≥18 yo | ≥18 yo | | | Behçet's Disease |
| **Baricitinib (Olumiant)** | | | | | | ≥18 yo | | |
| **Brodalumab (Siliq)** | | | | ≥18 yo | | | | |
| **Canakinumab (Ilaris) [See standalone policy]** | | | ≥2 yo | | | | | FCAS ≥4 yo MWS ≥4 yo TRAPS ≥ 4yo HIDS≥ 4 yo MKD≥ 4 yo FMF≥ 4 yo |
| **Certolizumab (Cimzia)** | ≥18 yo | ≥18 yo | | ≥18 yo | ≥18 yo | ≥18 yo | | Nr-axSpA ≥18 yo |
| **Etanercept (Enbrel)** | ≥18 yo | | ≥2 yo | ≥4 yo (Enbrel) ≥18 yo (biosimilars) | ≥18 yo | ≥18 yo | | |
| **Golimumab (Simponi & Simponi Aria)** | ≥18 yo | | ≥2 yo | | ≥18 yo | ≥18 yo | ≥18 yo (Simponi) | |
| **Guselkumab (Tremfya)** | | | | ≥18 yo | ≥18 yo | | | |
| **Infliximab (J1745) Remicade Inflectra, Renflexis, Avsola** | ≥18 yo | ≥6 yo | | ≥18 yo | ≥18 yo | ≥18 yo | ≥6 yo | |
| **Ixekizumab (Taltz)** | ≥18 yo | | | ≥6 yo | ≥18 yo | | | Nr-axSpA ≥18 yo |
| **Rituximab (Rituxan)** | | | | | | ≥18 yo | | CLL ≥18 yo NHL ≥18 yo |

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| **[See standalone policy]** | | | | | | | | GPA ≥18 yo<br>Pemphigus Vulgaris ≥18 yo<br>RRMS ≥18 yo |
| **Risankizumab-rzaa (Skyrizi)** | | ≥18 yo | | ≥18 yo | ≥18 yo | | | |
| **Sarilumab (Kevzara)** | | | | | | ≥18 yo | | |
| **Secukinumab (Cosentyx)** | ≥18 yo | | | ≥18 yo | ≥18 yo | | | Nr-axSpA ≥18 yo<br><br>ERA ≥ 4 yo<br>JPsA ≥ 2 yo |
| **Tildrakizumab-asmn (Ilumya)** | | | | ≥18 yo | | | | |
| **Tocilizumab (Actemra)** | | | ≥2 yo | | | ≥18 yo | | CRS >2 yo<br>GCA >18 yo |
| **Tofacitinib (Xeljanz)** | ≥18 yo | | | | ≥18 yo | ≥18 yo | ≥18 yo | |
| **Upadacitinib (Rinvoq)** | | | | | ≥18 yo | ≥18 yo | | AD ≥12 yo |
| **Ustekinumab (Stelara)** | | ≥18 yo | | ≥6 yo | ≥18 yo | | ≥18 yo | |
| **Vedolizumab (Entyvio)** | | ≥18 yo | | | | | ≥18 yo | |

Yellow: Preferred Pharmacy Drugs Green: Medical Infusion Drugs

Abbreviations: AD = Atopic Dermatitis; CLL = Chronic Lymphocytic Leukemia; CRS = Cytokine Release Syndrome; ERA= Enthesitis-Related Arthritis; FCAS = Familial Cold Autoinflammatory Syndrome; FMF = Familial Mediterranean Fever; GCA = Giant Cell Arteritis; GPA = Granulomatosis with Polyangiitis (Wegener's Granulomatosis); HIDS: Hyperimmunoglobulin D Syndrome; HS = Hidradenitis Suppurativa; JPsA= Juvenile Psoriatic Arthritis; MKD = Mevalonate Kinase Deficiency; MPA = Microscopic Polyangitis; MWS = Muckle-Wells Syndrome; NHL = Non-Hodgkin's Lymphoma; NOMID = Neonatal Onset Multi-Systemic Inflammatory Disease; Nr-axSpA = nonradiographic Axial Spondyloarthritis; Still's dx = Adult-onset Still's disease; TRAPS = Tumor Necrosis Factor Receptor Associated Periodic Syndrome; RRMS = Relapsing-Remitting Multiple Sclerosis; yo = years

POLICY NAME:
**TARPEYO**
Affected Medications: **TARPEYO** (Budesonide Delayed Release Capsule 4 mg)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design.<br>  ○ Primary immunoglobulin A nephropathy (IgAN) |
| **Required Medical Information:** | • Diagnosis of primary immunoglobulin A nephropathy (IgAN) confirmed with biopsy.<br>• Documentation of risk of rapid disease progression with a urine protein-to-creatinine ratio (UPCR) equal to or greater than 1.5 g/g (labs current within 30 days of request) OR<br>• Proteinuria defined as equal to or greater than 1 g/day (labs current within 30 days of request). |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of treatment failure of a minimum of 12 weeks of angiotensin-converting enzyme (ACE) inhibitor or angiotensin receptor blocker (ARB) AND<br>• Documentation of treatment failure of glucocorticoid therapy with prednisone or methylprednisolone (treatment failure defined as proteinuria equal to or greater than 1g/day and a minimum of 8 weeks therapy, unless you have had an adverse effect to glucocorticoid therapy that is not associated with the corticosteroid class) OR<br>• Documentation of treatment failure of mycophenolate mofetil (treatment failure defined as proteinuria equal to or greater than 1g/day and a minimum of 12 weeks therapy)<br><br>**<u>No reauthorization</u>** – Recommended duration of therapy is 9 months followed by a 2-week dose taper prior to discontinuation. |
| **Exclusion Criteria:** | • Patients with other glomerulopathies or nephrotic syndrome. |
| **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 |

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| **Coverage Duration:** | Authorization: 10 months, unless otherwise specified. |

POLICY NAME:
**TASIMELTEON**
Affected Medications: HETLIOZ (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 distrubances in Smith-Magenis Syndrome (SMS) |
| **Required Medical Information:** | <u>Non-24</u><br>• Documentation of being legally blind with no light perception or in patients who are not blind but have abnormal self-selected light/darkness schedules.<br>• Diagnosis of Non-24 hour sleep wake disorder per International Classification of Sleep Disorders by ALL the following:<br>○ Documented history of insomnia, excessive daytime sleepiness, or both, that alternates with time periods of being asymptomatic, as the individual rotates between alignment and misalignment with the environmental light-dark schedule<br>○ Symptoms must be present for at least three months<br>○ Daily sleep logs and actigraphy for at least 4 weeks, demonstrating a gradual drift in rest-activity patterns<br>○ Symptoms not better explained by another current sleep, medical, neurologic, mental, or substance abuse disorder, or medication use<br><br><u>Smith-Magenis Syndrome (SMS)</u><br>• Diagnosis of Smith-Magenis Syndrome (SMS) confirmed by genetic test with significant nighttime sleep disturbances |
| **Appropriate Treatment Regimen & Other Criteria:** | <u>Non-24</u><br>• Documentation of treatment failure with at least 12 weeks of:<br>○ Melatonin<br>○ Ramelteon AND<br>• Failure with chronotherapy treatment<br>• Polysomnogram with documentation of treatment or having ruled out other sleep disorders: Insomnia, shift work disorder, jet lag |

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| | disorder, irregular sleep-wake rhythm disorder, delayed sleep-wake phase disorder, advanced sleep-wake rhythm disorder<br><br><u>Smith-Magenis Syndrome (SMS)</u><br>• Documented treatment failure with melatonin and acebutolol for at least 12 weeks<br><br><u>Reauthorization</u> will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Taking sedative or stimulant central nervous system-active drugs |
| **Age Restriction:** | • 18 years and older for Non-24<br>• 16 years and older for SMS, ages 3 to 15 for Hetlioz LQ solution |
| **Prescriber/Site of Care Restrictions:** | • Neurologist, Internist board certified in Sleep Medicine 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:
**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 |
| **Required Medical Information:** | • Colonoscopy results within 6 months.<br>• Bilirubin, alkaline phosphatase, lipase, amylase within 6 months.<br>• Recent fluid and electrolyte status and documented plan to assess.<br>• Serum Creatinine.<br>• Review of REMS criteria.<br>• Documentation of any malignancy and disclosure of increased risk of malignancy to patient with risk benefit consideration.<br>• Clinical justification of need for reduction in Parenteral Nutrition/IV fluid volume after at least 12 consecutive months of PN/IV fluid dependence AND three or more days per week of PN support (electrolytes and/or nutrients).<br>• Plan to assess weekly PN/IV volume and evaluation of success of treatment and continued need.<br>• Documentation of Short Bowel Syndrome (SBS) with current dependence on parenteral support. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Dose: 0.05 mg/kg SQ QD Dose: 50% reduction for CrCl less than 50 mL/min.<br>• Reauthorization: documentation of clinically significant benefit defined by parenteral support reduction of 1 day or greater a week. |
| **Exclusion Criteria:** | |
| **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 |

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| **Coverage Duration:** | • Approval: 6 months, unless otherwise specified |

POLICY NAME:
**TEDIZOLID**
Affected Medications: SIVEXTRO powder for IV 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: 200 mg once daily for 6 days<br><br>Trial and failure with either intravenous antibiotics or oral antibiotics per below:<br><br>**<u>Intravenous</u>**<br>• Documentation of treatment failure of intravenous Linezolid, or contraindication to therapy **AND**<br>• Documentation of treatment failure of at least 2 of the following drugs/drug classes, or contraindication to therapy:<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 |

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| | baseline, whichever is greater), without an alternative explanation<br>○ Daptomycin<br>○ Cephalosporin (Cefazolin)<br><br>**<u>Oral tablets</u>**<br>• Documentation of treatment failure of oral Linezolid, or contraindication to therapy **AND**<br>• Documentation of treatment failure of at least 2 of the following drugs/drug classes, or contraindication to therapy:<br>• Trimethoprim-Sulfamethoxazole<br>• Tetracycline (Doxycycline, Minocycline)<br>• Clindamycin |
| **Exclusion Criteria:** | |
| **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:** | • 1 month, unless otherwise specified. |

POLICY NAME:
**TEGSEDI**
Affected Medications: TEGSEDI (inotersen sodium)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise exclude by plan design.<br>  ○ Treatment of the polyneuropathy of hereditary transthyretin-mediated amyloidosis in adults. |
| **Required Medical Information:** | • Documented pathogenic mutation in transthyretin (TTR; e.g. V30M mutation)<br>• Diagnosis of hereditary transthyretin (hATTR) amyloidosis with polyneuropathy<br>• Documentation of baseline Neuropathy Impairment Score (NIS) of 10 to 130<br>• Documented amyloid deposits determined on biopsy<br>• Presence of clinical signs and symptoms of disease (e.g. peripheral/autonomic neuropathy, motor disability, cardiovascular dysfunction, and renal dysfunction)<br>• Complete blood count, basic metabolic panel prior to start |
| **Appropriate Treatment Regimen & Other Criteria:** | • Coverage of the non-preferred product, Tegsedi, is provided when there has been a documented inadequate response or intolerable adverse event to Onpattro.<br><br>**<u>Hereditary transthyretin-mediated (hATTR) amyloidosis</u>**<br>• Tegsedi 284 mg injected subcutaneously once weekly<br>• During treatment, monitor platelets weekly during treatment if values are 75 x $10^9$/L or greater, and more frequently if values are less than 75 x $10^9$/L<br>• During treatment, monitor kidney function every 2 weeks<br><br>• Do not initiate if urinary protein to creatinine ratio (UPCR) is 1000 mg/g or higher<br><br>• Reauthorization requires documentation of a positive clinical response to inotersen (e.g. improved neurologic impairment, motor function, cardiac function, quality of life assessment, serum TTR levels, etc) |
| **Exclusion Criteria:** | • Platelet count less than 100 x $10^9$/L prior to start of Tegsedi |

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| **Age Restriction:** | • Adults 18 years and older |
| **Prescriber/Site of Care Restrictions:** | • Physicians experienced in the management of amyloidosis |
| **Coverage Duration:** | • Initial approval: 4 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 12 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:** | • 12 years or older |
| **Prescriber Restrictions:** | • Must be 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 |

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

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

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| **Covered Uses:** | • All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design<br>  ○ 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 |
| **Required Medical Information:** | • Diagnosis of Stage 2 type 1 diabetes, confirmed by both of the following:<br>  ○ Positive for two or more of the following pancreatic islet cell autoantibodies within the past 6 months:<br>  ▪ Glutamic acid decarboxylase 65 (GAD) autoantibodies<br>  ▪ Insulin autoantibody (IAA)<br>  ▪ Insulinoma-associated antigen 2 autoantibody (IA-2A)<br>  ▪ Zinc transporter 8 autoantibody (ZnT8A)<br>  ▪ Islet cell autoantibody (ICA)<br>  ○ Dysglycemia on oral glucose tolerance testing (OGTT) within the past 6 months, as shown by one of the following:<br>  ▪ Fasting blood glucose between 110 mg/dL and 125 mg/dL<br>  ▪ 2 hour glucose greater than or equal to 140 mg/dL and less than 200 mg/dL<br>  ▪ 30, 60, or 90 minute value on OGTT greater than or equal to 200 mg/dL on two separate occasions<br>• Documentation that the patient has a first-degree or second-degree relative with type 1 diabetes and one of the following:<br>  ○ If first-degree relative (brother, sister, parent, offspring), patient must be between 8 and 45 years of age<br>  ○ If second-degree relative (niece, nephew, aunt, uncle, grandchild, cousin), patient must be between 8 and 20 years of age |

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

POLICY NAME:
**TEPTROTUMUMAB-TRBW**
Affected Medications: TEPEZZA (teptrotumumab-trbw)

<table>
<tr><td>Covered Uses:</td><td>• All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design.<br>o Thyroid Eye Disease</td></tr>
<tr><td>Required Medical Information:</td><td>• <u>Documentation of moderate to severe active thyroid eye disease (TED) with ALL of the following:</u><br>o Lid retraction at least 2 mm<br>o Moderate or severe soft tissue involvement<br>o Exophthalmos at least 3 mm above normal for race and gender<br>o Must be euthyroid with the baseline disease under control prior to starting therapy<br>o Must not have had previous orbital surgery or irradiation for TED prior to the start of therapy<br>o Clinical Activity Score (CAS) 4 or greater<br><br><table><tr><th>Component</th><th>Scoring if Present</th></tr><tr><td>Spontaneous retrobulbar pain</td><td>1</td></tr><tr><td>Pain on attempted upward or downward gaze</td><td>1</td></tr><tr><td>Redness of eyelids</td><td>1</td></tr><tr><td>Redness of conjunctiva</td><td>1</td></tr><tr><td>Swelling of eyelids</td><td>1</td></tr><tr><td>Swelling of caruncle or plica</td><td>1</td></tr><tr><td>Swelling of conjunctiva (chemosis)</td><td>1</td></tr></table><br>• <u>Documented failure to ALL to the following therapies:</u><br>o intravenous methylprednisolone over 12 weeks<br>o mycophenolate mofetil 500mg twice daily for 24 weeks</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td>Initial dose 10mg/kg followed by 20mg/kg every 3 weeks for 7 additional doses<br><br>Product Availability<br>Single-dose vials for injection: 500mg<br>• <u>Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced</u></td></tr>
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| **Exclusion Criteria:** | • Prior surgical treatment for TED |
| **Age Restriction:** | • 18 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • 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:
**TERIFLUNOMIDE**
Affected Medications: AUBAGIO (teriflunomide)

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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), to include clinically isolated syndrome, relapsing-remitting disease (RRMS) and active secondary progressive disease (SPMS) in adults. |
| **Required Medical Information:** | • Diagnosis of relapsing or active secondary progressive forms of multiple sclerosis (MS) confirmed by magnetic resonance imaging (MRI) (Revised McDonald diagnostic criteria for multiple sclerosis)<br>  ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS<br>• Transaminase, bilirubin, and complete blood count (CBC) within 6 months before initiation of teriflunomide |
| **Appropriate Treatment Regimen & Other Criteria:** | • **<u>Reauthorization:</u>** provider attestation of treatment success |
| **Exclusion Criteria:** | • Patients with severe hepatic impairment<br>• Pregnancy or trying to conceive (both genders)<br>• Concurrent use of medications indicated for treatment of relapsing-remitting multiple sclerosis<br>• Primary progressive multiple sclerosis |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or an MS specialist<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:
**TESTOPEL**
Affected Medications: TESTOPEL (testosterone pellets)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise exclude by plan design. |
| **Required Medical Information:** | • All therapies tried/failed for indicated diagnosis<br>• Dosage (in milligrams) or number of pellets to be administered and frequency<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<br>• Documented treatment failure with testosterone injection AND generic transdermal testosterone<br><br>**For member 65 years and above:**<br>• Yearly evaluation of need is completed discussing need for hormone replacement therapy<br>• Yearly documentation that provider has educated patient on risks of hormone replacement (heart attack, stroke)<br>• Yearly documentation that provider has discussed limited efficacy and safety for hormone replacement in patients experiencing age related decrease in testosterone levels<br><br>**Gender Dysphoria hormone supplementation under 18 years of age:**<br>• Documentation of current Tanner stage 2 or greater OR Documentation of baseline and current estradiol and testosterone levels to confirm onset of puberty.<br>• Documentation from a licensed mental health professional (LMHP) confirming diagnosis and addressing the patient's general identifying characteristics;<br>o The initial and evolving gender and any associated mental health concerns, and other psychiatric diagnoses;<br>o The duration of the referring licensed mental health professional's relationship with the client, including the type of evaluation and psychotherapy to date; |

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| | ○ The clinical rationale for supporting the client's request for cross-hormone therapy and statement that the client meets eligibility criteria;<br>○ Informed consent required from both patient and guardian documented by prescribing provider<br>○ Permission to contact the licensed mental health professional for coordination of care<br>• Comprehensive mental health evaluation should be provided in accordance with most current version of the World Professional Association for Transgender Health (WPATH) Standards of Care<br><br>• **Note:** For requests following pubertal suppression therapy, an updated or new comprehensive mental health evaluation must be provided prior to initiation of hormone supplementation |
| **Appropriate Treatment Regimen & Other Criteria:** | • Maximum of 450 mg per treatment<br>• Reauthorization: documentation of recent testosterone levels within normal limits<br>**Gender Dysphoria:**<br>• Reauthorization: documentation of treatment success |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Gender Dysphoria: Diagnosis made and prescribed by or in consultation with a specialist in gender dysphoria<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: maximum 4 treatments in 12 months, unless otherwise specified. |

POLICY NAME:
**TETRABENAZINE**
Affected Medications: XENAZINE, tetrabenazine

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. |
| **Required Medical Information:** | • Current complete medication list |
| **Appropriate Treatment Regimen & Other Criteria:** | • Check for CYP2D6 interactions - strong CYP2D6 inhibitors (such as quinidine or antidepressants e.g., fluoxetine, paroxetine) significantly increase exposure therefore the total daily dose should not exceed a maximum of 50 mg<br>• Reauthorization requires documentation of clinically significant response to therapy with no major adverse reactions to treatment |
| **Exclusion Criteria:** | • Comorbid untreated or inadequately treated depression or actively suicidal<br>• Combination use with an MAOI, or within a minimum of 14 days of discontinuing therapy with an MAOI<br>• Combination use with reserpine. At least 20 days should elapse after stopping reserpine before starting Xenazine<br>• Comorbid hepatic impairment, including mild impairment |
| **Age Restriction:** | • 18 years of age or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with neurologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**TEZEPELUMAB-EKKO**
Affected Medications: TEZSPIRE

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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 for use in combination with another monoclonal antibody (e.g., Fasenra, Nucala, Xolair, Cinqair, Dupixent)? | Yes – Criteria not met, combination use is experimental | No – Go to #3 |
| 3. Is the request to treat a diagnosis according to one of the Food and Drug Administration (FDA)-approved indications?<br><br>o Add-on maintenance treatment of patients with severe asthma aged 12 years and older | Yes – Go to appropriate section below | No – Criteria not met |
| **Severe Asthma** | | |
| 1. Is there documentation of severe asthma defined by the following:<br><br>For adults:<br>o FEV1 less than 80% at baseline or FEV1/FVC reduced by at least 5% from normal<br><br>For those between the age of 12-17:<br>o FEV1 less than 90% at baseline or FEV1/FVC reduced by at least 5% from normal | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there documented use of a high-dose inhaled corticosteroid (ICS) plus a long- | Yes – Document and go to #3 | No – Criteria not met |

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| acting beta agonist (LABA) for at least three months with continued symptoms? | | |
| 3. Is there a documented history of 2 or more asthma exacerbations requiring oral or systemic corticosteroid treatment in the past 12 months while on combination inhaled treatment and at least 80% adherence? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is the drug prescribed by or in consultation with an Allergist, Immunologist, or Pulmonologist? | Yes – Approve up to 6 months | No – Criteria not met |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider? | 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** | | |
| • **Tezspire**<br>○ Availability: 210 mg/1.91 ml prefilled syringe; 210 mg/1.91 ml single-dose vial<br>○ Dosing: 210 mg every 4 weeks | | |

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>• 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:** | **<u>Multiple Myeloma</u>**<br>• Used in combination with dexamethasone in newly diagnosed MM<br><br>**<u>Erythema Nodosum Leprosum</u>**<br>• **Acute treatment** of the cutaneous manifestations of moderate to severe ENL<br>  ○ Not indicated as monotherapy in the presence of moderate to severe neuritis<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 or older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, an oncologist or infectious disease specialist |

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| | • 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:
**THYMOGLOBULIN**
Affected Medications: THYMOGLOBULIN

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.<br>• Renal transplant acute rejection treatment and induction therapy<br>• Off-label uses:<br>  ○ Heart transplant<br>  ○ Intestinal and multivisceral transplantation<br>  ○ Lung transplant<br>  ○ Chronic graft-versus-host disease prevention |
| **Required Medical Information:** | • For prophylaxis: 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: donor cold ischemia for more than 24 hours, donor age older than 50 years old, donor without a heartbeat, donor with ATN, donor requiring high-dose inotropic support. Recipient risk factors include: repeated transplantation, panel-reactive antibody value exceeding 20% before transplant, black race, and one or more HLA antigen mismatches with the donor. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Treatment of acute renal graft rejection-No PA required for this diagnosis<br>• Prophylaxis: 1.5mg/kg of body weight administered daily for 4-7 days<br>• Clinical rationale for avoiding Simulect (basiliximab) in prophylaxes |
| **Exclusion Criteria:** | • Active acute or chronic infections that contraindicates any additional immunosuppression |
| **Age Restriction:** | |

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| **Prescriber/Site of Care Restrictions:** | • Physicians experienced in immunosuppressive therapy for the management of renal transplant patients.<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 1 Month, unless otherwise specified<br>• Reauthorization: 1 Month, unless otherwise specified |

POLICY NAME:
**TISAGENLECLEUCEL**
Affected Medications: KYMRIAH (tisagenlecleucel)

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| **Covered Uses:** | • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | **All indications:**<br>• Documentation of patient's body weight<br>• Documentation of patient's CAR-positive viable T-cells<br>• Documentation that Black Box Warnings (Cytokine release syndrome, neurological toxicities) have been fully reviewed and patient understands and accepts risks<br>• Documentation of disease staging, all prior therapies used, and anticipated treatment course<br><br>**Pediatric and Young Adult Relapsed or Refractory (r/r) B-cell Acute Lymphoblastic Leukemia (ALL)**<br>• Documentation of patient being less than 25 years old<br>• Documentation of Hepatitis B vaccination or protected titer status<br>• Documentation of relapsed (second or later relapse) or refractory B-cell precursor acute lymphoblastic leukemia AND<br>• Philadelphia chromosome status<br><br>**Adult Relapsed or Refractory (r/r) Diffuse Large B-Cell Lymphoma (DLBCL)**<br>• Documentation of patient being at least 18 years old<br>• Documentation that treatment is not for primary central nervous system lymphoma<br>• Documentation of treatment failure with at least two lines of systemic therapy<br>• Documentation of relapsed (second or later relapse) or refractory diffuse large B-cell lymphoma not otherwise specified OR<br>• High grade B-cell lymphoma ORDLBCL arising from follicular lymphoma<br><br>**Adult Relapsed or Refractory (r/r) Follicular Lymphoma**<br>• Documentation of patient being at least 18 years old |

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| | • Documentation of relapsed (second or later relapse) or refractory disease<br>• Documentation of treatment failure with at least two lines of systemic therapy |
| **Appropriate Treatment Regimen & Other Criteria:** | **Pediatric and Young Adult Relapsed or Refractory (r/r) B-cell Acute Lymphoblastic Leukemia (ALL)**<br><br>• Completion of lymphodepleting therapy before initiation of Kymriah. Fludarabine (30 mg/m2 intravenous daily for 4 days) and cyclophosphamide (500 mg/m2 intravenous daily for 2 days starting with the first dose of fludarabine).<br>• Infuse Kymriah 2 to 14 days after completion of lymphodepleting chemotherapy.<br>• Dosing for patients 50 kg or less: administer 0.2 to 5.0 x $10^6$ CAR positive viable T cells per KG of body weight.<br>• Dosing for patients above 50 kg: administer 0.1 to 2.5 x $10^8$ CAR positive viable T cells<br>• Chimeric antigen receptor (CAR)-positive viable T cells based on the patient weight reported at the time of leukapheresis.<br><br>**Adult Relapsed or Refractory (r/r) Diffuse Large B-Cell Lymphoma (DLBCL) and Adult Relapsed or Refractory (r/r) Follicular Lymphoma**<br><br>• Completion of lymphodepleting therapy 2 to 11 days before initiation of Kymriah. Fludarabine (25 mg/m2 IV daily for 3 days) and Cyclophosphamide (250 mg/m2 IV daily for 3 days starting with the first dose of fludarabine)<br>• Alternative pre-treatment: Bendamustine 90 mg/m2 IV daily for 2 days<br>• Pretreatment with lymphodepleting chemotherapy may be omitted if patient's WBC is 1 x $10^9$/L or less within 1 week prior to tisagenlecleucel<br>• Premedication, give acetaminophen and diphenhydrAMINE or other H1-antihistamine 30 to 60 minutes before infusion. Avoid use of prophylactic systemic corticosteroids, as it may interfere with the activity of tisagenlecleucel<br>• A single dose of KYMRIAH contains 0.6 to 6.0 x $10^8$ CAR-positive viable T cells suspended in one or more patient-specific infusion bag(s) for i.v. infusion. |

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| | **<u>Reauthorization</u>** for all indications not supported by compendia |
| **Exclusion Criteria:** | • Concomitant use of granulocyte colony-stimulating factors.<br>• Unresolved serious adverse reactions from chemotherapy, active uncontrolled infection, active GVHD, or increasing leukemia burden following lymphodepleting chemotherapy. |
| **Age Restriction:** | **<u>ALL</u>**<br>• Safety and effectiveness in patients 25 years of age and older has not been established<br><br>**<u>DLBCL and Follicular Lymphoma</u>**<br>Safety and effectiveness in patients under 18 years of age has not been established |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by an oncologist |
| **Coverage Duration:** | • Initial approval: 2 months, unless otherwise specified<br>• Reauthorization: None |

POLICY NAME:
**TIVDAK**
Affected Medications: TIVDAK (tisotumab)

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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 previous failure of systemic therapy for metastatic disease<br>• Documentation of PD-L1 levels<br>• Documentation of testing for mismatch repair deficiency (dMMR) and high levels of metastatic microsatellite instability (MSI-H) |
| **Appropriate Treatment Regimen & Other Criteria:** | **PD-L1 positive, MSI-H, or dMMR tumors:**<br>• Documented clinical failure with immunotherapy<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:** | • 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:
**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. |
| **Required Medical Information:** | • Diagnosis of Cystic Fibrosis (phenotyping not required).<br>• Culture and sensitivity report confirming presence of pseudomonas aeruginosa in the lungs<br>• For Tobi Podhaler: Baseline forced expiratory volume in 1 second (FEV1) equal to or greater than 25% but equal to or less than 80%<br>• For Bethkis: Baseline FEV1 equal to or greater than 40% but equal to or less than 80%<br>• For Kitabis Pak: Baseline FEV1 equal to or greater than 25% but equal to or less than 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>• Reauthorization requires documentation of improved respiratory symptoms and need for long-term use |
| **Exclusion Criteria:** | • For Tobi Podhaler: Baseline FEV1 less than 25% or greater than 80%<br>• For Bethkis: Baseline FEV1 less than 40% or greater than 80%<br>• For Kitabis Pak: Baseline FEV1 less than 25% or greater than 75% |
| **Age Restriction:** | • Age greater than or equal to 6 years |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 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>• Age 18 to 55 years old and estimated glomerular filtration rate (eGFR) greater than or equal to 25 mL/min/1.73m2<br>• High risk for rapid progression determined by Mayo imaging class 1C, 1D, or 1E |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Hyponatremia</u>**<br>• Patients should be in hospital for initiation and re-initiation of therapy<br>• Maximum dose 60 mg once daily<br>• Do not administer for more than 30 days<br><br>**<u>ADPKD</u>**<br>• Documentation of intensive blood pressure control with an |

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| | angiotensin-converting enzyme (ACE) inhibitor or angiotensin receptor blocker (ARB), unless contraindicated<br>• Dosing:<br>  ○ Initial: 60 mg/day in divided doses (given as 45 mg upon awakening and 15 mg 8 hours later)<br>  ○ May titrate at intervals of at least 7 days up to maximum 120 mg/day (given as 90 mg upon awakening and 30 mg 8 hours later)<br>• **Reauthorization:** will require documentation of treatment success and a clinically significant response to therapy. |
| **Exclusion Criteria:** | • Use in patients with ADPKD outside of FDA-approved REMS (Risk Evaluation and Mitigation Strategies)<br>• 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>• Concomitant use with strong CYP3A inhibitors<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 |
| **Coverage Duration:** | **<u>Hyponatremia</u>**<br>• Authorization: 1 month, 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 ANTIPSORIATICS**
Affected Medications: VTAMA (tapinarof 1% cream), Zoryve (roflumilast)

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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 chronic plaque psoriasis<br>• Documentation that the skin disease meets one of the following:<br>○ At least 10% body surface area involvement despite current treatment<br>○ Hand, foot, or mucous membrane involvement |
| **Appropriate Treatment Regimen & Other Criteria:** | Documented failure with ALL of the following:<br>• A high or super-high potency topical corticosteroid (such as betamethasone dipropionate, clobetasol, fluocinonide, or halobetasol)<br>• Calcipotriene cream or calcitriol ointment<br>• Tazarotene cream<br>• Vtama also requires documented treatment failure with 8 weeks of Zoryve<br><br>**Reauthorization** will require documentation of disease responsiveness to therapy defined as BSA reduction when compared to baseline |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • Vtama: 18 years of age and older<br>• Zoryve: 12 years of age and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a specialist (for example: 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:
**TRALOKINUMAB**
Affected Medications: ADBRY (tralokinumab)

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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?<br>- Treatment of moderate to severe atopic dermatitis in adults | Yes – Go to appropriate section below | No – Criteria not met |
| **Moderate to Severe Atopic Dermatitis** | | |
| 1. Is there 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)? | Yes – Document and go to #2 | No – Criteria not met |
| 2. Is there a documented body surface area (BSA) effected of at least 10% OR hand, foot or mucous membrane involvement? | Yes – Document and go to #3 | No – Criteria not met |
| 3. Is there documented failure with at least 6 weeks of treatment with one of the following: tacrolimus ointment, pimecrolimus cream, Eucrisa? | Yes – Document and go to #4 | No – Criteria not met |
| 4. Is there documented treatment failure with two of the following for at least 12 weeks: Phototherapy, cyclosporine, azathioprine, methotrexate, mycophenolate? | Yes – Document and go to #5 | No – Criteria not met |

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| 5. Is the drug prescribed by or in consultation with a specialist in the treatment of atopic dermatitis (Such as a dermatologist)? | Yes – Approve up to 6 months | No – Criteria not met |
| **Renewal Criteria** | | |
| 1. Is there documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider? | 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** | | |
| • **Adbry**<br>  ○ Availability: 150 mg/ml prefilled syringes<br>  ○ Dosing: 600 mg as single dose, then 300 mg every 2 weeks.<br>  ▪ If less than 100 kg and clear/almost clear is achieved dosing may be reduced to 300 mg every 4 weeks | | |

POLICY NAME:
**TRASTUZUMAB**
Affected Medications: HERCEPTIN IV (trastuzumab), HERCEPTIN HYLECTA, KANJINTI (trastuzumab-anns), OGIVRI(trastuzumab-dkst), TRAZIMERA (trastuzumab-qyyp), HERZUMA (trastuzumab-pkrb), ONTRUZANT (trastuzumab-dttb)

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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, disease staging, all prior therapies used, and prescribed dosing regimen<br>• Baseline evaluation of left ventricular function<br>• Documentation of HER2 positivity based on 3+ score on immunohistochemistry (IHC) testing or positive gene amplification by fluorescence in situ hybridization (FISH) test |
| **Appropriate Treatment Regimen & Other Criteria:** | • Max duration for adjuvant breast cancer therapy is 12 months<br>• Reauthorization requires documentation of disease responsiveness to therapy<br>All Indications<br>• Coverage for a non-preferred product (Trazimera, Herzuma, Ontruzant, or Herceptin) requires documentation of one of the following:<br>○ A documented intolerable adverse event to the preferred products Kanjinti and Ogivri 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 |
| **Exclusion Criteria:** | • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |

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| **Coverage Duration:** | • For new starts to adjuvant breast cancer therapy – approve 12 months with no reauthorization<br>• For all other clinical scenarios:<br>• Initial approval: 4 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**TRIENTINE**
Affected Medications: trientine

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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:** | • Documentation confirming a diagnosis of Wilson's disease |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented treatment failure with a minimum 6 month trial, or intolerable adverse event, with penicillamine<br><br>• **Reauthorization**: Documentation of treatment success with normalization of free serum copper (non-ceruloplasmin bound copper) to less than 15 mcg/dL |
| **Exclusion Criteria:** | • Rheumatoid arthritis<br>• Cystinuria<br>• Biliary cirrhosis |
| **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 Approval: 6 months, unless otherwise specified<br>• Reauthorization:12 months, unless otherwise specified |

POLICY NAME:
**TRIKAFTA**
Affected Medications: TRIKAFTA (elexacaftor, tezacaftor and ivacaftor; ivacaftor)

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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 cystic fibrosis (CF) diagnosis.<br>• Documentation of confirmed diagnosis by appropriate genetic or diagnostic testing (FDA approved CF mutation test).<br>• Documentation of at least one F508del mutation in the CFTR gene OR a mutation in the CFTR gene that is responsive based on in vitro data.<br>• Please provide the diagnostic testing report and/or Cystic Fibrosis Foundation Patient Registry Report.<br>• Liver function tests prior to initiation, every 3 months during the first year of treatment, and annually thereafter |
| **Appropriate Treatment Regimen & Other Criteria:** | Adults and pediatric patients ages 12 years and older:<br>• Morning dose: two elexacaftor 100 mg, tezacaftor 50 mg and ivacaftor 75 mg tablets<br>• Evening dose: one ivacaftor 150 mg tablet<br><br>Pediatric patients ages 6 years and older weighing less than 30 kg:<br>• Morning dose: two elexacaftor 50 mg, tezacaftor 25 mg and ivacaftor 37.5 mg tablets<br>• Evening dose: one ivacaftor 75 mg tablet<br><br>• Reauthorization will require documentation of treatment success |
| **Exclusion Criteria:** | • Concurrent use of strong CYP3A inducers: rifampin, rifabutin, phenobarbital, carbamazepine, phenytoin, and St. John's wort |
| **Age Restriction:** | • Approved in patients ages 6 years 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 subjects 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 benefit design. (For non-cancer use only) |
| **Required Medical Information:** | **Prostate cancer**<br>• Documentation of performance status, disease staging, all prior therapies used, and prescribed treatment regimen<br>• Documentation that Trelstar is being used as NCCN 2A level of evidence regimen<br><br>**Central Precocious Puberty (CPP)**<br>• Documentation of central precocious puberty (CPP) confirmed by basal luteinizing hormone (LH), follicle-stimulating hormone (FSH), and either estradiol or testosterone concentrations<br>• Documented clinical rationale for avoiding Lupron depot-ped and Supprelin LA |
| **Appropriate Treatment Regimen & Other Criteria:** | • Triptorelin QL: 22.5 mg every 6 months<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Use as neoadjuvant ADT for radical prostatectomy |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Oncology: prescribed by or in consultation with Oncologist<br>• CPP: prescribed by or in consultation with pediatric endocrinologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Oncology initial approval: 4 months, unless otherwise specified<br>• CPP Approval/Oncology 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 benefit design. |
| **Required Medical Information:** | • Documentation of all prior therapies used<br>• Documentation of active antiretroviral therapy for at least 6 months<br>• Documentation of multidrug resistant HIV-1 with resistance to at least one antiretroviral medication from each of the following classes: Nucleoside Reverse Trancriptase Inhibitors (NRTIs), Non-Nucleoside Reverse Transcriptase Inhibitors, and Protease Inhibitors (PIs).<br>• Failure with current regimen or not on current antiretroviral therapy and failure with most recent regimen (viral load greater than 1,000 copies/mL) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Loading dose 2000mg<br>• Maintenance dose 800mg every 2 weeks<br>• Initial reauthorization will require documentation of greater than or equal to a 0.5 $\log_{10}$ reduction in viral load<br>• Reauthorization: Continued authorization will require undetectable viral load |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • 18 years and older |
| **Prescriber/Site of Care Restrictions:** | • Infectious Disease or specialist in HIV treatment |
| **Coverage Duration:** | • Initial approval: 3 months, unless otherwise specified<br>• Reauthorization 12 months, unless otherwise specified |

POLICY NAME:
**TURALIO**
Affected Medications: TURALIO (pexidartinib oral capsules)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design<br>  ○ Symptomatic tenosynovial giant cell tumor (TGCT) |
| **Required Medical Information:** | • A diagnosis of TGCT that has been histologically confirmed either by a pathologist at the treating institution or a central pathologist, and where surgical resection would be associated with potentially worsening functional limitation or severe morbidity (locally advanced disease), with morbidity determined consensually by qualified personnel (Two surgeons or a multi-disciplinary tumor board)<br>• Measurable disease of at least 2 cm, assessed from MRI scans by a central radiologist<br>• Symptomatic disease because of active TGCT, defined as one or more of the following:<br>  ○ A worst pain of at least 4 at any time during the week preceding the Screening Visit (based on scale of 0 to 10, with 10 representing "pain as bad as you can imagine"<br>  ○ A worst stiffness of at least 4 at any time during the week preceding the Screening Visit (based on a scale of 0 to 10, with 10 representing "stiffness as bad as you can imagine") |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented failure or contraindication of imatinib<br>• Reauthorization requires documentation of treatment success |
| **Exclusion Criteria:** | • Liver Disease<br>• Pregnancy |
| **Age Restriction:** | • Age greater than or equal to 18 years |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care<br>• Prescribers enrolled in REMS program |
| **Coverage Duration:** | • Initial approval: 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 benefit design. |
| **Required Medical Information:** | **<u>Pulmonary arterial hypertension (PAH) WHO Group 1</u>**<br>• Documentation of PAH confirmed by right-heart catheterization<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>• Documentation of acute vasoreactivity testing (positive result requires trial/failure to calcium channel blocker)<br>• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class III symptoms<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:** | • For initiation of therapy patient must have a mean pulmonary artery pressure of at least 20 mmHg at rest, an elevated pulmonary vascular resistance (PVR) of at least 3.0 Wood units, and a mean pulmonary capillary wedge pressure less than 15 mmHg **AND**<br>• The pulmonary hypertension has progressed despite maximal medical and/or surgical treatment of the identified condition |

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| | • Treatment with oral calcium channel blocking agents has been tried and failed, or has been considered and ruled out (not required for WHO group 3)<br>• Documentation that treprostinil is used as a single route of administration (Remodulin, Tyvaso, Orenitram should not be used in combination)<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 symptoms (not required for WHO group 3)<br>  ○ Ambrisentan and tadalafil<br>  ○ Bosentan and riociguat<br>  ○ Macitentan and sildenafil<br>• Subsequent approval requires documentation of treatment success: exercise endurance, echocardiographic testing, hemodynamic testing, BNP, 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:** | • 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 coverage: 6 months unless otherwise specified<br>• Subsequent coverage: 12 months unless otherwise specified |

POLICY NAME:
**UPLIZNA**
Affected Medications: UPLIZNA (inebilizumab-cdon)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by plan design<br>  ○ Neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive |
| **Required Medical Information:** | • Testing for serum immunoglobulins levels<br><br>Neuromyelitis Optica Spectrum Disorder (NMOSD)<br>• Diagnosis of NMOSD with AQP4-IgG requiring all of the following:<br>  ○ At least one core clinical characteristic:<br>  • Optic neuritis<br>  • Acute myelitis<br>  • Area postrema syndrome: episode of otherwise unexplained hiccups or nausea and vomiting<br>  • Acute brainstem syndrome<br>  • Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMSOD-typical diencephalic MRI lesions<br>  • Symptomatic cerebral syndrome with NMOSD-typical brain lesions<br>  ○ Positive test for AQP4-IgG using best available detection method<br>  ○ Exclusion for alternative diagnoses<br>• History of at least 1 attack in the past year, or at least 2 attacks in the past 2 years, requiring rescue therapy<br>• Expanded Disability Status Scale (EDSS) score of 8 or less<br>• Documented treatment failure with 12 weeks of at least 2 of the following immunosuppressive therapies: azathioprine, mycophenolate, methotrexate<br>• Documented treatment failure with 12 weeks of at least 1 of the following: mitoxantrone (authorization required), rituximab (authorization required)<br>• Documented treatment failure with Enspryng (authorization required) |

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| | **<u>Reauthorization</u>** requires documentation of treatment success. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Initial dosing: 300 mg, followed by a second 300mg dose 2 weeks later<br>• Subsequent doses (starting 6 months after the first infusion): 300mg every 6 months |
| **Exclusion Criteria:** | • Active Hepatitis B Virus (HBV) infection<br>• Active or untreated latent tuberculosis<br>• Concurrent use with other monoclonal antibodies (rituximab, eculizumab, tocilizumab, etc.) or IVIG |
| **Age Restriction:** | • 18 years of age and older |
| **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:
**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 100,200, or 400mg (vaginal progesterone)

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| **Covered Uses:** | • Prevention of preterm birth in pregnant women with a singleton pregnancy and prior history of preterm delivery before 37 weeks gestation or short cervical length |
| **Required Medical Information:** | • Singleton pregnancy<br>• History of singleton spontaneous preterm birth before 37 weeks gestation or short cervical length defined as less than 20 mm |
| **Appropriate Treatment Regimen & Other Criteria:** | **History of singleton spontaneous preterm birth (HSPB)**<br>• May initiate therapy beginning at 16 to 20 weeks gestation and continue until 36+6 weeks gestation<br><br>**Short cervical length (SCL)**<br>• May initiate therapy beginning at 0 to 24 weeks gestation (with pregnancy confirmed by positive test) and continue until 36+6 weeks gestation |
| **Exclusion Criteria:** | • Treatment of infertility |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with gynecologist or obstetrician<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • HSPB: up to 20 weeks, unless otherwise specified<br>• SCL: up to 36 weeks and 6 days, 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:** | |

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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:** | • Approval: 6 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>○ Predominantly classic subfoveal choroidal neovascularization (CNV) due to age-related macular degeneration (AMD), pathologic myopia or presumed ocular histoplasmosis |
| **Required Medical Information:** | • Subfoveal choroidal neovascularization (CNV) lesions caused by age-related macular degeneration (AMD); or<br>• Chronic (greater than 4 months) central serous chorioretinopathy; or<br>• Ocular histoplasmosis; or<br>• Pathologic myopia<br><br>Note: Most individuals treated with verteporfin will need to be re-treated every 3 months. All individuals having a re-treatment will need to have a flourescein angiogram or ocular coherence tomography (OCT) performed prior to each treatment. Re-treatment is necessary if fluorescein angiograms or OCT show any signs of recurrence or persistence of leakage |
| **Appropriate Treatment Regimen & Other Criteria:** | • Coverage for the non-preferred product Visudyne is provided when one of the following criteria is met:<br>○ Currently receiving treatment with Visudyne, 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 all of the preferred products (Avastin, Byooviz, and Cimerli).<br>• Dosing: 6 mg/m2 body surface area given intravenously; may repeat at 3-month intervals (if evidence of choroidal neovascular leakage)<br>○ Available as 15 mg vials<br><br>• **Reauthorization** requires documented treatment success and continued need for treatment with the non-preferred product |

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| **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: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**VESTRONIDASE ALFA**
Affected Medications: MEPSEVII (vestronidase alfa-vjbk)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design |
| **Required Medical Information:** | • Definitive diagnosis of Mucopolysaccharidosis VII (MPS VII; Sly Syndrome) confirmed by BOTH of the following:<br>  ○ Beta-glucuronidase enzyme deficiency in peripheral blood leukocytes **AND**<br>  ○ Detection of pathogenic mutations in the GUSB gene by molecular genetic testing<br>• Baseline value for one or more of the following:<br>  ○ Bruininks-Oseretsky Test of Motor Proficiency<br>  ○ 6 minute walk test<br>  ○ Liver and/or spleen volume<br>  ○ Pulmonary function tests |
| **Appropriate Treatment Regimen & Other Criteria:** | • 4 mg/kg infusion every 2 weeks<br>• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced<br>• Reauthorization will require:<br>  ○ Documentation of absence of unacceptable toxicity (ex. anaphylaxis or severe allergic reactions) AND<br>  ○ Patient has responded to therapy compared to pretreatment baseline in one or more of the following:<br>  ▪ Improvement in Bruininks-Oseretsky Test of Motor Proficiency<br>  ▪ Improvement in 6 minute walk test<br>  ▪ Reduction in liver and/or spleen volume<br>  ▪ Stability or improvement in pulmonary function tests |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • Age 8 - 25 years |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care<br>• Prescriber with experience in treating MPS |
| **Coverage Duration:** | • Initial approval: 2 months, unless otherwise specified<br>• Reauthorization: 6 months, unless otherwise specified |

POLICY NAME:
**VIGABATRIN**
Affected Medications: Vigabatrin, Vigabatrin Packet

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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 baseline vision assessment (no later than 4 weeks after starting vigabatrin) by an ophthalmologist<br>• Documentation that the potential benefits outweigh the risk of vision loss<br>• Proof that the patient is blind or formally exempt from vision assessments in the Support, Help, And Resources for Epilepsy (SHARE) program<br><br>**<u>Refractory complex partial seizures</u>**<br>• Documentation the patient has tried at least 2 alternative therapies: carbamazepine, phenytoin, levetiracetam, topiramate, oxcarbazepine, or lamotrigine |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Infantile Spasm</u>**<br>• Use as monotherapy for pediatric patients (1 month to 2 years of age)<br><br>**<u>Refractory Complex Partial Seizures</u>**<br>• As adjunctive therapy for patients who have inadequately responded to several alternative treatments<br><br><u>Reauthorization:</u><br>• Vision assessment by an ophthalmologist with no documented vision loss from baseline<br>• Documented planned reassessments every 3 months during therapy<br>• Documentation of substantial clinical benefit (within 3 months of initiation; within 2-4 weeks of initiation for patients with infantile spasms or sooner if treatment failure becomes obvious) |
| **Exclusion Criteria:** | • Use as a first line agent for Complex Partial Seizures |
| **Age Restriction:** | • Infantile Spasms: 1 month to 2 years of age<br>• Refractory Complex Partial Seizures: greater than 2 years of age |

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| **Prescriber/Site of Care Restrictions:** | • Prescriber certified with the SHARE program |
| **Coverage Duration:** | • Initial approval: 3 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**VIJOICE**
Affected Medications: VIJOICE (alpelisib)

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| **Covered Uses:** | • All FDA-approved indications not otherwise excluded by benefit design<br>  ○ PIK3CA-related overgrowth spectrum (PROS) |
| **Required Medical Information:** | • Diagnosis of PIK3CA-Related Overgrowth Spectrum (PROS) with severe clinical manifestations of lesions as assessed by the treating provider (such as those associated with CLOVES, Megalencephaly-Capillary Malformation Polymicrogyria [MCAP], Klippel-Trenaunay Syndrome [KTS], Facial Infiltrating Lipomatosis [FIL])<br>• Documentation of PIK3CA gene mutation<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:** | • Documentation that severe 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>• 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 |

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| **Age Restriction:** | • Must be 2 years of age or older |
| **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 benefit design. |
| **Required Medical Information:** | • Documented current use with fluorouracil or capecitabine, and experiencing life-threatening adverse effects |
| **Appropriate Treatment Regimen & Other Criteria:** | • To be used as an antidote for fluorouracil or capecitabine overdose or to treat severe adverse effects following treatment<br>• Ensure dosing according to Food and Drug Administration (FDA) approved regimen<br>• Ensure use is within 96 hours of fluorouracil/capecitabine treatment |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Oncologist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 5 days, 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 |

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| 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 6 months | No – Criteria not met |
| **Renewal Criteria** | | |
| 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 6 months (lifetime maximum 12 months of therapy) | No – Criteria not met |
| **Quantity Limitations** | | |

- **Lupkynis***
  - Starting dose: 23.7 mg twice daily (BID)
  - Starting dose must be reduced in the below situations as follows:
    - eGFR 45 mL/min/1.73 m2 or less at initiation: 15.8mg BID
    - Mild-to-moderate hepatic impairment (Child-Pugh A or B): 15.8mg BID
    - Concomitant use with moderate CYP3A4 inhibitors: 15.8mg in morning and 7.9mg in afternoon.

* Lifetime maximum 12 months of therapy.

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) |

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| **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:** | • Approval: 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 FDA-approved indications not otherwise excluded by plan design<br>  ○ To increase linear growth in pediatric patients with achondroplasia who are 5 years of age and older with open epiphyses. |
| **Required Medical Information:** | • Genetic test results confirming achondroplasia.<br>• Baseline height, growth velocity, and patient weight. |
| **Appropriate Treatment Regimen & Other Criteria:** | • For initial approval, documentation of the following is required:<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:** | • Age 5 to 18 years |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with a pediatric orthopedist, endocrinologist, or a provider with experience in treating skeletal dysplasias<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:
**VOXELOTOR**
Affected Medications: OXBRYTA (voxelotor)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design.<br>○ Treatment of sickle cell disease (SCD) in adults and pediatric patients 4 years of age and older. |
| **Required Medical Information:** | • Two or more sickle cell-related crises in the past 12 months (defined as acute painful crisis or acute chest syndrome for which there are no explanation other than vaso-occlusive crisis).<br>• Therapeutic failure of 6 month trial on maximum tolerated dose of hydroxyurea or intolerable adverse event to hydroxyurea<br>• Baseline hemoglobin (Hb) greater than or equal to 5.5 or less than or equal to 10.5 g/dL<br>• Current Weight |
| **Appropriate Treatment Regimen & Other Criteria:** | For requests for tablets for oral suspension, must be unable to swallow tablets.<br><br>**Reauthorization** requires documentation of treatment success defined by an increase in hemoglobin of more than 1 gm/dL from baseline, or a decrease in the number of sickle cell-related crises |
| **Exclusion Criteria:** | • Receiving regular red-cell transfusion therapy or have received a transfusion in the past 60 days<br>• Have been hospitalized for vaso-occlusive crisis within 14 days of request<br>• Combined use with anti-P selectin monoclonal antibody (crizanlizumab) |
| **Age Restriction:** | • Ages 4 years and older |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or in consultation with hematologist.<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Initial approval: 6 months, unless otherwise specified<br>• Reauthorization: 12 months, unless otherwise specified |

POLICY NAME:
**VELAGLUCERASE ALFA**
Affected Medications: VPRIV (velaglucerase alfa)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design. |
| **Required Medical Information:** | • Patient has a diagnosis of type 1 Gaucher disease.<br>• Diagnosis of Gaucher disease is confirmed by an enzyme assay demonstrating a deficiency of beta-glucocerebrosidase enzyme activity.<br>• Therapy is initiated for a patient with one or more of the following conditions: anemia, thrombocytopenia, bone disease, hepatomegaly, or splenomegaly. |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented inadequate response or an intolerable adverse event with imiglucerase (Cerezyme)<br>• Dosing: 60 units/kg every 2 weeks; dosing is individualized based on disease severity (range: 15-60 units/kg evaluated in clinical trials)<br>  ○ Supplied as 400 unit vials<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Concomitant therapy with miglustat |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • 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:
**VUMERITY**
Affected Medications: VUMERITY (diroximel fumarate)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded from plan benefits.<br>○ Treatment of relapsing forms of multiple sclerosis (MS), to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease, in adults |
| **Required Medical Information:** | • Diagnosis of relapsing or active secondary progressive forms of Multiple Sclerosis (MS) confirmed with MRI (Revised McDonald diagnostic criteria for multiple sclerosis)<br>○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS<br>• Complete blood count with lymphocyte count, and liver function tests (within 6 months) before initiating treatment, then CBC annually and as clinically indicated |
| **Appropriate Treatment Regimen & Other Criteria:** | • Initial dose of 231 mg twice daily for 7 days, then increasing to 462 mg twice daily thereafter<br>• Hold therapy for four weeks if lymphocyte count is less than 500/mm3 for greater than 6 months<br>• No concurrent use of disease-modifying therapy for the treatment of multiple sclerosis<br>• Not approved for primary progressive multiple sclerosis<br><br>**Reauthorization:** provider attestation of treatment success |
| **Exclusion Criteria:** | • Pre-existing low lymphocyte counts (less than 500/mm3) |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • Prescribed by or after consultation with a neurologist or an MS specialist.<br>• All approvals are subject to utilization of the most cost-effective site of care |

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

POLICY NAME:
**VUTRISIRAN**
Affected Medications: 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 the polyneuropathy of hereditary transthyretin-mediated amyloidosis in adults |
| **Required Medical Information:** | • Documented pathogenic mutation in transthyretin (TTR)<br>• Diagnosis of hereditary transthyretin (hATTR) amyloidosis with polyneuropathy<br>• Documentation of baseline polyneuropathy disability (PND) score less than or equal to IIIb<br>• Presence of clinical signs and symptoms of disease (e.g., peripheral/autonomic neuropathy, motor disability, cardiovascular dysfunction, renal dysfunction) |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documented failure with diflunisal<br>• Dosing: 25 mg subcutaneous once every 3 weeks<br><br>**<u>Reauthorization:</u>**<br>• Documentation of continued PND score less than or equal to IIIb AND documentation of the patient experiencing positive clinical response to vutrisiran (e.g., improved neurologic impairment, motor function, cardiac function, quality of life assessment, serum TTR levels, etc.) |
| **Exclusion Criteria:** | • Prior or planned liver transplantation<br>• NYHA class III or IV<br>• Diagnosis of other (non-hATTR) forms of amyloidosis or leptomeningeal amyloidosis<br>• Combined use with TTR-lowering therapy, including inotersen or patisiran<br>• Combined use with TTR-stabilizing therapy, including diflunisal, tafamidis, or tafamidis meglumine |
| **Age Restriction:** | • Adults aged 18 to 85 years old |

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| **Prescriber/Site of Care Restrictions:** | • Prescribed by, or in consultation with, a neurologist or provider with experience in the management of amyloidosis<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:
**XEOMIN, DYSPORT and MYOBLOC**
Affected Medications: XEOMIN (incobotulinum toxin A), DYSPORT (abobotulinumtoxinA), MYOBLOC (rimabotulinumtoxinB), JEUVEAU (prbotulinumtoxinA-xvfs)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by benefit design |
| **Required Medical Information:** | • Pertinent medical records and diagnostic testing<br>• Complete description of the site(s) of injection<br>• Strength and dosage of botulinum toxin used |
| **Appropriate Treatment Regimen & Other Criteria:** | **<u>Dysport</u>**<br>• Approved first-line for focal dystonia, hemifacial spasm, orofacial dyskinesia, blepharospasm, upper or lower limb spasticity<br><br>**<u>Xeomin</u>**<br>• Approved first-line for the uses of cervical dystonia, upper limb spasticity, blepharospasm and chronic sialorrhea<br><br>**<u>Myobloc</u>**<br>• Cervical Dystonia<br>  ○ Documented failure with Botox, Xeomin and Dysport is required<br>• Overactive Bladder, urinary incontinence due to spinal cord injury or axillary hyperhidrosis<br>  ○ Documented failure with Botox is required<br>• Chronic Sialorrhea<br>  ○ Documented failure with glycopyrrolate oral tablets<br><br>**<u>Jeuveau</u>**<br>• Jeuveau is only indicated in the treatment of cosmetic conditions and is excluded from coverage<br><br>**<u>Other Criteria</u>**<br>• Reauthorization requires documented treatment success<br>• All indications not listed are considered experimental/investigational and are not a covered benefit<br>• Maximum of 4 treatments per 12 months (2 treatments for Myobloc in overactive bladder) |

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| **Exclusion Criteria:** | • Cosmetic procedures<br>• For intradetrusor injections: documented current/recent urinary tract infection or urinary retention<br>• Current aminoglycoside use (or current use of other agents interfering with neuromuscular transmission)<br>• Possible medication overuse headache: headaches occurring 15 or more days each month in a patient with pre-existing headache-causing condition possibly due to<br>  ○ Use of ergotamines, triptans, opioids, or combination analgesics at least 10 days per month for at least three months<br>  ○ Use of simple analgesics (acetaminophen, aspirin, or an NSAID) at least 15 days per month for at least 3 months<br>  ○ Use of combination of any previously mentioned products without overuse of any one agent if no causative pattern can be established<br>• Combined use with Calcitonin Gene-Related Peptide (CGRP) Receptor Antagonists (Ajovy, Aimovig or Emgality) for the treatment of migraine |
| **Age Restriction:** | • Ages 18 years or older for Myobloc |
| **Prescriber/Site of Care Restrictions:** | • Blepharospasm: ophthalmologist or optometrist<br>• Overactive bladder or urinary incontinence due to neurologic condition: urologist or neurologist<br>• Documentation of consultation with any of the above specialists mentioned<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | Overactive Bladder<br>• Initial approval: 3 months<br>• Reauthorization: 12 months, unless otherwise specified<br><br>All other indications<br>• Approval: 12 months, unless otherwise specified |

POLICY NAME:
**XGEVA**
Affected Medications: XGEVA (denosumab)

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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 |
| **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:** | • For treatment of breast cancer with bony metastases or castration resistant prostate cancer with bony metastases: Approval is limited to monthly dosage for the first 12 months of therapy followed by quarterly doses thereafter (not to exceed 4 dosages within a 12 month time)<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | |
| **Age Restriction:** | • Giant Cell Tumor of the Bone: 12 years of age and older, AND skeletally mature<br>• All other indications: 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 |

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| **Coverage Duration:** | • Approval: 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 plan design<br>• Dupuytren's contracture with a palpable cord<br>• Peyronie's disease |
| **Required Medical Information:** | **Peyronie's Disease**<br>• Documented diagnosis of Peyronie's disease with a palpable plaque<br>• Curvature deformity is at least 30 degrees at the start of therapy and results in pain<br>• Symptoms have been present for at least 12 months |
| **Appropriate Treatment Regimen & Other Criteria:** | **Dupuytren's**<br>• Authorization will be limited per joint as follows: One injection per month for a maximum of three injections per cord<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy<br>**Peyronie's**<br>• One treatment cycle consisting of two Xiaflex injection procedures<br>• Subsequent authorization(s) 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 |
| **Exclusion Criteria:** | • Prior intolerance or allergic reaction to requested medication |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • **Peyronie's**: Urologist<br>• All approvals are subjects to utilization of the most cost-effective site of care |

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| **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>• Treatment of complex Clostridium difficile infection in select populations |
| **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>For C. difficile disease</u>**<br>• Patient must have failed oral vancomycin for coverage to be considered<br><br>**<u>For recurrent or persistent hepatic encephalopathy</u>**<br>• Patient has failed or has contraindication to 30 day attempt of lactulose therapy, with documentation of continued altered mental status or elevated ammonium levels despite adequate upward titration of lactulose.<br><br>**<u>For Travelers' Diarrhea</u>**<br>• Documentation of travelers' diarrhea caused by noninvasive strains of E. coli (no systemic signs of infection) and returning from an area of high fluoroquinolone resistance.<br>• Documented contraindication or allergy to fluoroquinolone, and azithromycin.<br><br>**<u>For Small Intestinal Bacterial Overgrowth</u>**<br>• Patient must have a diagnosis of small intestinal bacterial overgrowth confirmed by a carbohydrate breath test<br>• Documented treatment failure with trial of at least two of the following antibiotics: amoxicillin/clavulanic acid, ciprofloxacin, metronidazole<br><br>**<u>For Irritable Bowel Syndrome with Diarrhea (IBS-D)</u>**<br>• Patient must have a Rome IV diagnosis: recurrent abdominal pain associated with at least two of the following: related to defecation, associated with a change in stool frequency, associated |

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| | with a change in stool form; for the last 3 months with symptom onset over six months prior to diagnosis<br>• Patient must have tried and failed at least 3 of the following: loperamide, dicyclomine, tricyclics (amitriptyline/nortriptyline), and probiotics prior to the approval of Xifaxan.<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:** | **<u>For C. difficile disease</u>**<br>• Xifaxan exceeding 400 mg three times per day for 20 days<br><br>**<u>For recurrent or persistent hepatic encephalopathy</u>**<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>**<u>For Travelers' Diarrhea</u>**<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>**<u>For Small Intestinal Bacterial Overgrowth</u>**<br>• Xifaxan exceeding 550 mg three times per day for 14 days<br><br>**<u>For IBS</u>** |

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| | • Mild cases 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 or older |
| **Prescriber/Site of Care Restrictions:** | • All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Clostridium difficile infection: 20 days, unless otherwise specified<br>• Hepatic encephalopathy: 12 months, unless otherwise specified<br>• Travelers' Diarrhea: 7 days, unless otherwise specified<br>• Small intestinal bacterial overgrowth: 14 days, unless otherwise specified (Once per lifetime)<br>• Irritable Bowel Syndrome: 14 days, unless otherwise specified (maximum 3 fills 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 |
| **Required Medical Information:** | • Diagnosis of hereditary orotic aciduria<br>• Urine orotic acid levels<br>• Patient weight |
| **Appropriate Treatment Regimen & Other Criteria:** | • Documentation of weight based dosing<br>• Reauthorization requires documentation of treatment success based on laboratory values |
| **Exclusion Criteria:** | |
| **Age Restriction:** | |
| **Prescriber/Site of Care Restrictions:** | • In consultation with geneticist specialist<br>• All approvals are subject to utilization of the most cost-effective site of care |
| **Coverage Duration:** | • Approval: 12 months |

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 benefit design.<br>• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher |
| **Required Medical Information:** | • Documentation of trial and failure to generic abiraterone acetate or clinical reason for avoiding generic abiraterone acetate |
| **Appropriate Treatment Regimen & Other Criteria:** | • Reauthorization will require 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:** | |
| **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, an oncologist |
| **Coverage Duration:** | • Initial approval: 4 months (2 week initial partial fill), unless otherwise specified<br>• Approval: 12 months, unless otherwise specified. |

POLICY NAME:
**ZAVESCA**
Affected Medications: ZAVESCA (miglustat)

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| **Covered Uses:** | • All Food and Drug Administration (FDA)-approved indications |
| **Required Medical Information:** | • Diagnosis of Type 1 Gaucher disease<br>• Mild to moderate disease<br>• Diagnosis of Gaucher disease confirmed by an enzyme assay demonstrating a deficiency of beta-glucocerebrosidase enzyme activity |
| **Appropriate Treatment Regimen & Other Criteria:** | • Enzyme replacement therapy (ERT) is not a therapeutic option for the patient (e.g. due to allergy, hypersensitivity, or poor venous access)<br>• The patient will use adequate contraception throughout Zavesca therapy and for 3 months thereafter<br>• Reauthorization will require documentation of treatment success and a clinically significant response to therapy |
| **Exclusion Criteria:** | • Female of childbearing potential who is pregnant or planning a pregnancy |
| **Age Restriction:** | • 18 years 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:** | • Initial approval: 4 months, unless otherwise specified.<br>• Reauthorization: 12 months, unless otherwise specified. |

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

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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 short bowel syndrome (SBS). |
| **Appropriate Treatment Regimen & Other Criteria:** | • Patients must be receiving specialized nutritional support (e.g., TPN, IPN, PPN, rehydration solutions, electrolyte replacement, high complex-carbohydrate, low-fat diet) in conjunction with optimal management of SBS. |
| **Exclusion Criteria:** | • Active malignancy (newly diagnosed or recurrent).<br>• Acute critical illness due to complications following open heart or abdominal surgery, accidental trauma or acute respiratory failure.<br>• Active proliferative or severe non-proliferative diabetic retinopathy |
| **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:** | • Approval: 4 weeks with no reauthorization, unless otherwise specifiied. |