2024 PacificSource Health Plans Prior Authorization Criteria

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(All criteria reviewed at least once per year)

PacificSource HEALTH PLANS

Contents

ACTIMMUNE 13
ADDYI & VYLEESI 15
ADENOSINE DEAMINASE (ADA) REPLACEMENT 17
ADZYNMA 19
AFAMELANOTIDE 21
AFINITOR 23
ALEMTUZUMAB 25
ALGLUCOSIDASE ALFA 27
ALOSETRON 29
ALPHA-1 PROTEINASE INHIBITORS 31
AMIFAMPRIDINE 33
ANIFROLUMAB 35
ANTIEMETICS 37
ANTIHEMOPHILIC FACTORS 40
ANTITHYMOCYTE GLOBULIN 44
ANTITHROMBIN ALFA 46
ANTI-AMYLOID MONOCLONAL ANTIBODY 48
ANTI-TUBERCULOSIS AGENTS 49
APOMORPHINE 51
ARIKAYCE 53
ASCIMINIB 55
ATIDARSAGENE AUTOTEMCEL 57
AVACOPAN 59
AVALGLUCOSIDASE ALFA-NGPT 61
AVATROMBOPAG 63
AVONEX 65
AZTREONAM 66
BELIMUMAB 67

BELZUTIFAN 69
BENRALIZUMAB 71
BEREMAGENE GEPERPAVEC-SVDT 73
BESREMI 75
BETAINE 76
BETASERON 77
BETIBEGLOGENE AUTOTEMCEL 78
BEVACIZUMAB 80
BEZLOTOXUMAB 82
BIRCH TRITERPENES 83
BOTOX 85
BUROSUMAB 89
CALCIFEDIOL 91
CANNABIDIOL 93
CANTHARIDIN 95
CAPLACIZUMAB-YHDP 97
CAPSAICIN KIT 99
CARGLUMIC ACID 101
CERLIPONASE ALFA 103
CFTR MODULATORS 105
CALCITONIN GENE-RELATED PEPTIDE (CGRP) INHIBITORS 106
CHELATING AGENTS 109
CHOLBAM 112
CHOLESTATIC LIVER DISEASE 114
CIALIS 116
CLADRIBINE 118
COAGADEX 120
COMPOUNDED MEDICATION 122
CONTINUOUS GLUCOSE MONITORS 123
CORLANOR 124

COVERAGE OF SELECT HIGH INTENSITY STATINS AT TIER 0 COPAY 126
CRIZANLIZUMAB 127
CROVALIMAB 128
CYSTARAN, CYSTADROPS 130
CYSTEAMINE 131
DANICOPAN 132
DASATINIB 133
DEFIBROTIDE 134
DEFLAZACORT 135
DELANDISTROGENE MOXEPARVOVEC-ROKL 137
DIFELIKEFALIN 139
DINUTUXIMAB 141
DIROXIMEL FUMARATE 143
DOJOLVI 145
DONISLECEL 147
DORNASE ALFA 149
DROXIDOPA 150
DUOPA 152
DUPILUMAB 154
ECULIZUMAB 157
EDARAVONE 162
EFLORNITHINE 163
ELAGOLIX 165
ELIVALDOGENE AUTOTEMCEL 167
ELTROMBOPAG DERIVATIVES 169
EMAPALUMAB 173
EMICIZUMAB 175
EMSAM 177
ENDOTHELIN RECEPTOR ANTAGONISTS 178
ENFUVIRTIDE 180

ENZYME REPLACEMENT THERAPY (ERT) FOR GAUCHER DISEASE TYPE 1 182
EPLONTERSEN, PATISIRAN, VUTRISIRAN 185
EPOPROSTENOL 187
ERECTILE DYSFUNCTION 189
ERGOT ALKALOIDS 191
ERYTHROPOIESIS STIMULATING AGENTS (ESAs) 193
ETELCALCETIDE 195
ETRANACOGENE 196
EVKEEZA and JUXTAPID 197
EVOLOCUMAB 199
EXAGAMGLOGENE AUTOTEMCEL 202
FABRY DISEASE AGENTS 205
FECAL MICROBIOTA 207
FENFLURAMINE 209
FIDANACOGENE 211
FINERENONE 213
FLUCYTOSINE 215
FLUOCINOLONE OCULAR IMPLANT 216
Food and Drug Administration (FDA) APPROVED DRUG – Drug or Indication Not Yet Reviewed By Plan for Formulary Placement 218
FOSTAMATINIB 219
FYARRO 221
GABA-A RECEPTOR MODULATORS 222
GANAXOLONE 225
GIVINOSTAT 226
GIVOSIRAN 228
GLUCAGON-LIKE PEPTIDE (GLP-1) RECEPTOR AGONIST 230
GONADOTROPIN 231
GOSERELIN ACETATE IMPLANT 232
GROWTH HORMONES 234

HEPATITIS C DIRECT-ACTING ANTIVIRALS 238
HISTRELIN 242
HEREDITARY ANGIOEDEMA 244
HEREDITARY TYROSINEMIA (HT-1) AGENTS 247
Hormone Supplementation under 18 years of age 248
HYDROCORTISONE ORAL GRANULES 250
HYFTOR 252
HYPOXIA-INDUCIBLE FACTOR PROLYL HYDROXYLASE (HIF PH) INHIBITORS 253
IBREXAFUNGERP 255
ILARIS 257
ILOPROST 260
IMMUNE GLOBULIN 262
INCLISIRAN 273
INEBILIZUMAB-CDON 276
INTRAVITREAL ANTI-VEGF THERAPY 278
INTRAVITREAL COMPLEMENT INHIBITORS 283
INTRON-A 285
ISAVUCONAZONIUM SULFATE 287
LAROTRECTINIB 289
LENACAPAVIR 290
LENIOLISIB 292
LETERMOVIR 294
LEUPROLIDE 296
LEVOKETOCONAZOLE 299
LIFILEUCEL 301
LONAFARNIB 303
LOTILANER 305
LOVOTIBEGLOGENE AUTOTEMCEL 306
LUSUTROMBOPAG 308
MANNITOL 309

MARIBAVIR 311
MAVACAMTEN 312
MAVORIXAFOR 314
MECASERMIN 315
MEDICAL NECESSITY 316
MEPOLIZUMAB 320
METHYLNALTREXONE 324
METRELEPTIN 326
MIACALCIN 328
MIGLUSTAT 330
MILTEFOSINE 331
MITAPIVAT 333
MOMETASONE SINUS IMPLANT 336
MONOMETHYL FUMARATE 337
MOTIXAFORTIDE 339
MUCOPOLYSACCHARIDOSIS (MPS) AGENTS 341
MUSCULAR DYSTROPHY RNA THERAPY 343
MYELOID GROWTH FACTORS 344
NAFARELIN 350
NALOXEGOL 352
NATALIZUMAB 353
NAXITAMAB 355
NEMOLIZUMAB-ILTO 357
NEONATAL FC RECEPTOR ANTAGONISTS 358
NILOTINIB 362
NIROGACESTAT 363
NON-Preferred HYALURONIC ACID DERIVATIVES 364
NON-PREFERRED MEDICAL DRUG CODES 366
NUEDEXTA 367
NULIBRY 368

NUPLAZID 370
NUSINERSEN 371
OCRELIZUMAB 373
OFEV 375
OLIPUDASE ALFA 377
OMALIZUMAB 379
OMAVELOXOLONE 383
OMIDUBICEL 384
ONASEMNOGENE ABEPARVOVEC XIOI 385
ONCOLOGY AGENTS 387
OPIOID QUANTITY ABOVE 90 MORPHINE MILLIGRAM EQUIVALENTS (MME) 389
OPZELURA 392
ORAL-INTRANASAL FENTANYL 393
ORAL TESTOSTERONE 395
ORENITRAM 397
ORGOVYX 399
OSILODROSTAT 400
OTESECONAZOLE 401
OXERVATE 403
OXYBATES 404
OZANIMOD 406
PALFORZIA 408
PALIVIZUMAB 410
PALOVAROTENE 412
PALYNZIQ 413
PARATHYROID HORMONE 414
PARATHYROID HORMONE ANALOGS 415
PEDMARK 417
PEGASYS 418
PEGLOTICASE 420

PEMIVIBART 422
PENICILLAMINE 424
PHENOXYBENZAMINE 426
PHESGO 427
PHOSPHODIESTERASE-5 (PDE-5) ENZYME INHIBITORS FOR PULMONARY ARTERIAL HYPERTENSION 428
PIRFENIDONE 430
PLEGRIDY 431
POMBILITI and OPFOLDA 432
PONVORY 434
POSACONAZOLE 435
POZELIMAB 436
PRAMLINTIDE 438
PROLIA 439
PROSTAGLANDIN IMPLANTS 440
PROXIMAL COMPLEMENT INHIBITOR 442
PRIMARY BILIARY CHOLANGITIS AGENTS 444
PYRIMETHAMINE 446
RAVICTI 447
RAVULIZUMAB-CWVZ 448
REBLOZYL 452
REBIF 454
RELYVRIO 455
REMODULIN 456
RESLIZUMAB 458
RESMETIROM 460
RETHYMIC 462
RILONACEPT 463
RIOCIGUAT 465
RISDIPLAM 467

RITUXIMAB 469
RNA INTERFERENCE DRUGS FOR PRIMARY HYPEROXALURIA 1 475
ROMIPLOSTIM 477
ROMOSOZUMAB 479
RUFINAMIDE 481
RYPLAZIM 482
SACROSIDASE 485
SAPROPTERIN 486
SATRALIZUMAB-MWGE 487
SEBELIPASE ALFA 489
SELF-ADMINISTERED DRUGS (SAD) 490
SELUMETINIB 491
SEROSTIM 493
SIGNIFOR 495
SIGNIFOR LAR 496
SILTUXIMAB 498
SIPONIMOD 499
SODIUM PHENYLBUTYRATE 501
SOLRIAMFETOL 503
SOMATOSTATIN ANALOGS 505
SOMAVERT 508
SOTATERCEPT-CSRK 510
SPARSENTAN 512
SPESOLIMAB 513
SPRAVATO 515
STIRIPENTOL 518
STRENSIQ 519
SUBCUTANEOUS IMMUNE GLOBULIN 521
SUTIMLIMAB 524
TAFAMIDIS 526

TAGRAXOFUSP-ERZS 528
TARGETED IMMUNE MODULATORS 530
TARPEYO 556
TASIMELTEON 558
TEDIZOLID 560
TEDUGLUTIDE 562
TENAPANOR 564
TENOFOVIR ALAFENAMIDE 566
TEPLIZUMAB-MZWV 568
TEPROTUMUMAB-TRBW 570
TESTOPEL 572
TEZEPELUMAB-EKKO 574
THALIDOMIDE 575
THYMOGLOBULIN 577
TOBRAMYCIN INHALATION 579
TOFERSEN 580
TOLVAPTAN 582
TOPICAL AGENTS FOR CUTANEOUS T-CELL LYMPHOMA (including Mycosis fungoides and Sézary syndrome) 584
TOPICAL DERMATITIS AND PSORIATIC AGENTS 586
TRALOKINUMAB 588
TRASTUZUMAB 590
TRIENTINE 592
TRIPTORELIN 594
TROFINETIDE 596
TROGARZO 597
TUCATINIB 599
TYVASO 601
UBLITUXIMAB-XIIY 604
UPNEEQ 606

VAGINAL PROGESTERONE 607
VALOCTOCOGENE ROXAPARVOVEC-RVOX 608
VAMOROLONE 610
VARIZIG 612
VELMANASE ALFA-TYCV 614
VERTEPORFIN 615
VIGABATRIN 617
VIJOICE 619
VISTOGARD 621
VMAT2 INHIBITORS 623
VOCLOSPORIN 625
VORETIGENE NEPARVOVEC 628
VOSORITIDE 630
XEOMIN, DYSPORT, MYOBLOC, and DAXXIFY 631
XGEVA 633
XIAFLEX 635
XIFAXAN 637
XURIDEN 641
YONSA 642
ZILUCOPLAN 643

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.
	  ○ Chronic Granulomatous Disease (CGD)
	  ○ Severe, malignant osteopetrosis (SMO)
	• 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.
	• Pediatrics with BSA less than 0.5 m^2: weight must be documented along with prescribed dose.
	
	<u>Chronic granulomatous disease</u>
	• Diagnosis established by a molecular genetic test identifying a gene-related mutation associated with CGD
	
	<u>Severe, malignant osteopetrosis</u>
	• Diagnosis of severe infantile osteopetrosis established by ONE of the following:
	  ○ Radiographic imaging consistent with osteopetrosis
	  OR
	  ○ Molecular genetic test identifying a gene-related mutation associated with SMO
	
	<u>Oncology indications</u>
	• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course

Appropriate Treatment Regimen & Other Criteria:	**<u>Chronic Granulomatous Disease</u>** • Patient is on a prophylactic regimen with an antibacterial agent and an antifungal agent **<u>All indications</u>**

	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced **<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 • **SMO:** prescribed by, or in consultation with, an endocrinologist • **Oncology indications:** prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	**<u>CGD and SMO</u>** Approval: 12 months, unless otherwise specified **<u>Oncology indications:</u>** Initial Authorization: 4 months, unless otherwise specified Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Premenopausal women with acquired, generalized hypoactive sexual desire disorder (HSDD) Acquired HSDD refers to HSDD that develops in a patient who previously had no problems with sexual desire 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 or arousal disorder:
	  ○ Lack of, or significantly reduced, sexual interest or arousal, as manifested by at least three of the following:
	  ▪ Absent or reduced interest in sexual activity
	  ▪ Absent or reduced sexual thoughts or fantasies
	  ▪ No or reduced initiation of sexual activity, and typically unreceptive to a partner's attempts to initiate
	  ▪ Absent or reduced sexual pleasure or sensation during sexual activity in 75% to 100% of sexual encounters
	  ▪ Absent or reduced sexual arousal in response to any sexual cues (e.g., written, verbal, visual)
	  ○ The above symptoms have persisted for a minimum duration of approximately 6 months
	  ○ The above symptoms cause clinically significant distress in the individual
	  ○ The sexual dysfunction is not
	  ▪ Better explained by a nonsexual mental disorder OR
	  ▪ A consequence of severe relationship distress (e.g., partner violence) or other significant stressors AND
	  ▪ It is not attributable to the effects of substance or medication use or another medical condition (such as a physical condition

Appropriate Treatment Regimen & Other Criteria:	• Addyi
	  ○ Documentation of appropriate patient counseling regarding alcohol use while taking Addyi
	• Vyleesi
	  ○ Documentation that patients who may become pregnant are using an effective form of contraception
	
	<u>Reauthorization</u> will require documentation of treatment success and a clinically significant response to therapy

Exclusion Criteria:	• Postmenopausal females • Males • Intended use is to enhance sexual performance
Age Restriction:	• Adult premenopausal women only
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a mental health provider • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 2 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of adenosine deaminase severe combined immune deficiency (ADA-SCID) in pediatric and adult patients

Required Medical Information:	• Diagnosis of ADA-SCID confirmed by genetic testing showing biallelic pathogenic variants in the *ADA* gene
	• Laboratory findings show the following:
	  ○ Absent ADA levels in lysed erythrocytes
	  ○ A marked increase in deoxyadenosine triphosphate (dATP) levels in erythrocyte lysates
	  ○ A significant decrease in ATP concentration in red blood cells
	  ○ Absent or extremely low levels of N adenosylhomocysteine hydrolase in red blood cells
	  ○ 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 • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced **<u>Reauthorization</u>** requires documentation of treatment success defined as disease stability and/or improvement as indicated by one or more of the following: • Increase in plasma ADA activity • Decrease in red blood cell dATP/dAXP level • Improvement in immune function with diminished frequency/complications of infections
Exclusion Criteria:	• Other forms of autosomal recessive SCIDs

	• All uses not listed under covered uses are considered experimental
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an immunologist or specialist experienced in the treatment of severe combined immune deficiency (SCID) • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 6 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Congenital thrombotic thrombocytopenic purpura (cTTP)

Required Medical Information:	• Diagnosis of severe cTTP confirmed by BOTH of the following:
	  ○ Molecular genetic testing confirming mutation in the ADAMTS13 gene
	  ○ ADAMTS13 activity testing showing less than 10% of normal activity
	• **For on-demand treatment:**
	  ○ Documentation of current or past acute event with 50% or greater drop in platelet count OR platelet count less than 100,000/microliter
	  ○ Lactase dehydrogenase elevation (LDH) is more than 2 times baseline or more than 2 times upper limit of normal (ULN) as defined by laboratory values
	• **For prophylactic use:**
	  ○ Must have history of at least one documented thrombotic thrombocytopenic purpura (TTP) event (past acute event or subacute event such as thrombocytopenia event or a microangiopathic hemolytic anemia event)

Appropriate Treatment Regimen & Other Criteria:	• **Dosing:**
	  ○ Prophylactic: 40 IU/kg once every other week
	  ▪ May be dosed weekly with documentation of appropriate prior dosing regimen or clinical response.
	  ○ On-demand therapy: 40 IU/kg on day 1, 20 IU/kg on day 2, and 15 IU/kg on day 3 and beyond until 2 days after the acute event is resolved.
	
	<u>Reauthorization</u>:

	• For prophylactic use: documentation of treatment success defined as an improvement in the number or severity of TTP events, platelet counts, or clinical symptoms
	• For on-demand use:
	  ○ Documentation that after previous on-demand therapy, platelet counts increased to at least 150,000/microliter or 25% from baseline platelet count
	  ○ Members without previous on-demand use must meet initial criteria

Exclusion Criteria:	• Diagnosis of other TTP-like disorder, such as acquired or immune-mediated TTP
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist, oncologist, intensive care specialist, or specialist in rare genetic hematologic diseases • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of patients with erythropoietic protoporphyria (EPP) with phototoxic reactions (including X-linked protoporphyria [XLP])
Required Medical Information:	• Documented symptoms of phototoxic reactions, resulting in dysfunction and significant impact on activities of daily living **<u>Erythropoietic Protoporphyria (EPP)</u>** • Documented diagnosis of EPP confirmed by biallelic loss-of-function mutation in the ferrochelatase (FECH) gene • Documented increase in total erythrocyte protoporphyrin, with at least 85% metal-free protoporphyrin **<u>X-Linked Erythropoietic Protoporphyria (XLP)</u>** • Documented diagnosis of XLP confirmed by gain-of-function mutations in the delta-aminolevulinic acid synthase (ALAS2) gene • Documented increase in total erythrocyte protoporphyrin, with at least 50% metal-free protoporphyrin
Appropriate Treatment Regimen & Other Criteria:	**<u>Reauthorization</u>:** • Documentation of treatment success and clinically significant response to therapy (e.g., decreased severity and number of phototoxic reactions, increased duration of sun exposure, increased quality of life, etc.) • Continued implementation of sun and light protection measures during treatment to prevent phototoxic reactions
Exclusion Criteria:	

Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist at a recognized Porphyria Center • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher

Required Medical Information:	**<u>Oncology Indications</u>**
	• Documentation of performance status, all prior therapies used, and prescribed treatment regimen
	
	<u>Tuberous Sclerosis Complex (TSC)</u>
	• Documentation of treatment resistant epilepsy, defined as lack of seizure control with 2 different antiepileptic regimens and meeting following criteria:
	  ○ Documentation of treatment failure with Epidiolex (cannabidiol solution) adjunct therapy
	  ○ Documentation that Afinitor Disperz (only form approved for TSC-seizures) is being used as adjunct therapy for seizures
	OR
	• Documentation of symptomatic subependymal giant cell tumors (SGCTs) or TSC-associated subependymal giant cell astrocytoma (SEGA) in a patient who is not a good candidate for surgical resection

Appropriate Treatment Regimen & Other Criteria:	**<u>Reauthorization</u>** requires documentation of disease responsiveness to therapy
Exclusion Criteria:	**<u>Oncology Indications</u>** • Karnofsky Performance Status less than or equal to 50% or ECOG performance score greater than or equal to 3
Age Restriction:	
Prescriber/Site of Care Restrictions:	• **Oncology Indication**: Prescribed by, or in consultation with, an oncologist • **TSC Indication**: Prescribed by, or in consultation with, a neurologist or specialist in the treatment of TSC

	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
ALEMTUZUMAB
Affected Medications: LEMTRADA (alemtuzumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive multiple sclerosis (SPMS)

Required Medical Information:	• Diagnosis confirmed with magnetic resonance imaging (MRI) per revised McDonald diagnostic criteria for MS ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS
Appropriate Treatment Regimen & Other Criteria:	• Documentation of inadequate response to Tysabri (natalizumab) AND one additional medication indicated for MS **Reauthorization** requires provider attestation of treatment success • Eligible for renewal 12 months after administration of last dose
Exclusion Criteria:	• Human immunodeficiency virus (HIV) infection • Active infection • Concurrent use of other disease-modifying medications indicated for the treatment of multiple sclerosis
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or a multiple sclerosis specialist • 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 • Reauthorization: 3 doses for 3 days, unless otherwise specified

POLICY NAME:
ALGLUCOSIDASE ALFA
Affected Medications: LUMIZYME

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ 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. • Patient weight and planned treatment regimen

Appropriate Treatment Regimen & Other Criteria:	• One or more clinical signs or symptoms of Pompe disease:
	  ○ Readily observed evidence of glycogen storage (macroglossia, hepatomegaly, normal or increased muscle bulk)
	  ○ Involvement of respiratory muscles manifesting as respiratory distress (such as tachypnea)
	  ○ Profound diffuse hypotonia
	  ○ Proximal muscle weakness
	  ○ Reduced forced vital capacity (FVC) in upright or supine position
	• Appropriate medical support is readily available when medication is administered in the event of anaphylaxis, severe allergic reaction, or acute cardiorespiratory failure.
	
	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. • All approvals are subject to utilization of the most cost-effective site of care

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

POLICY NAME:
ALOSETRON
Affected Medications: ALOSETRON, LOTRONEX (alosetron)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Women with severe diarrhea-predominant irritable bowel syndrome (IBS)

Required Medical Information:	• Female gender
	• Chronic IBS syndrome lasting at least 6 months
	• Diarrhea AND one or more of the following are present:
	  ○ Frequent and severe abdominal pain/discomfort
	  ○ Frequent bowel urgency or fecal incontinence
	  ○ Disability or restriction of daily activities due to IBS
	• 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:
	  ○ Dicyclomine
	  ○ Hyoscyamine
	  ○ Diphenoxylate-atropine
	  ○ Amitriptyline or nortriptyline
	• **Reauthorization** requires documentation of treatment success and a clinically significant response to therapy

Exclusion Criteria:	• History of chronic or severe constipation or sequelae from constipation, intestinal obstruction, stricture, toxic megacolon, gastrointestinal perforation, and/or adhesions, ischemic colitis, impaired intestinal circulation, thrombophlebitis, or hypercoagulable state, Crohn's disease or ulcerative colitis, diverticulitis, or severe hepatic impairment • Concomitant use of fluvoxamine
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a gastroenterologist • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 2 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. ○ 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:
	  ○ 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
	  ○ 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 ○ Has not smoked for a minimum of 6 consecutive months leading up to therapy initiation and will continue to abstain from smoking during therapy • Coverage of Aralast NP, Glassia, or Zemaira will require a documented intolerable adverse event to Prolastin-C • Dosing: 60 mg/kg intravenously once weekly **<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 • Patients with IgA deficiency or with the presence of IgA antibodies • Prior lung or liver transplant

Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a pulmonologist • 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)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Lambert-Eaton myasthenic syndrome (LEMS)

Required Medical Information:	• Documented diagnosis of LEMS confirmed by **ONE** of the following:
	  ○ Positive anti-P/Q-type voltage-gated calcium channel (VGCC) antibody test
	  ○ Repetitive nerve stimulation (RNS) abnormalities, such as an increase in compound muscle action potential (CMAP) amplitude at least 60 percent after maximum voluntary contraction (i.e., post-exercise stimulation) or at high frequency (50 Hz)
	• Documentation of clinical signs and symptoms consistent with LEMS, as follows: proximal muscle weakness (without atrophy), with or without autonomic features and areflexia

Appropriate Treatment Regimen & Other Criteria:	• Documentation of inadequate clinical response or intolerance to **ONE** of the following (except in active small cell lung carcinoma [SCLC]-LEMS):
	  ○ Combination oral prednisone and azathioprine therapy
	  ○ Combination intravenous immunoglobulin therapy with one of the following: oral prednisone or azathioprine
	
	<u>Reauthorization:</u> documentation of treatment success, confirmed by improved or sustained muscle strength on clinical assessments

Exclusion Criteria:	• Seizure disorder • Active brain metastases • Clinically significant long QTc interval on ECG in previous year OR history of additional risk factors for torsade de pointes

Age Restriction:	• 6 years of age or older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
ANIFROLUMAB
Affected Medications: SAPHNELO (anifrolumab)

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

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

POLICY NAME:
ANTIEMETICS
Affected Medications: AKYNZEO CAPSULES (netupitant-palonosetron), AKYNZEO INJECTION (fosnetupitant-palonosetron), VARUBI (rolapitant)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Prevention of delayed nausea and vomiting associated with initial and repeat courses of emetogenic cancer chemotherapy, including, but not limited to, highly emetogenic chemotherapy
	  ▪ **Varubi** (rolapitant)
	  ○ Prevention of acute and delayed nausea and vomiting associated with initial and repeat courses of highly emetogenic cancer chemotherapy.
	  ▪ **Akynzeo injection** (fosnetupitant-palonosetron)
	  ○ 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
	  ▪ **Akynzeo capsules** (netupitant-palonosetron)

Required Medical Information:	**<u>Chemotherapy Induced Nausea and Vomiting Prophylaxis</u>**
	• Documentation of planned chemotherapy regimen
	• **Varubi**
	  ○ Documentation of a highly OR moderately emetogenic chemotherapy regimen
	• **Akynzeo injection**
	  ○ Documentation of a highly emetogenic chemotherapy regimen
	• **Akynzeo capsule**
	  ○ Documentation of a highly OR moderately emetogenic chemotherapy regimen
	Highly Emetogenic Chemotherapy

	Any regimen that contains an anthracycline and cyclophosphamide	Cyclophosphamide	Fam-trastuzumab deruxtecan-nxki	Sacituzumab govitecan-hziy
	Carboplatin	Dacarbazine	Ifosfamide	Streptozocin
	Carmustine	Doxorubicin	Mechlorethamine	FOLFOX
	Cisplatin	Epirubicin	Melphalan	
	May be considered highly emetogenic in certain patients			
	Dactinomycin	Idarubicin	Methotrexate (250 mg/m^2 or greater)	Trabectedin
	Daunorubicin	Irinotecan	Oxaliplatin	
	Moderately Emetogenic Chemotherapy			
	Aldesleukin	Cytarabine	Idarubicin	Mirvetuximab soravtansine-gynx
	Amifostine	Dactinomycin	Irinotecan	Naxitamab-gqgk
	Bendamustine	Daunorubicin	Irinotecan (liposomal)	Oxaliplatin
	Busulfan	Dinutuximab	Lurbinectedin	Romidepsin
	Clofarabine	Dual-drug liposomal encapsulation of cytarabine and daunorubicin	Methotrexate (250 mg/m^2 or greater)	Temozolomide
	Trabectedin			
Appropriate Treatment Regimen & Other Criteria:	**<u>Chemotherapy induced Nausea and Vomiting Prophylaxis</u>** • **Varubi**: ○ Documented treatment failure with a 5-HT3 receptor antagonist (e.g., ondansetron, granisetron) in combination			

	with dexamethasone while receiving the current chemotherapy regimen
	• **Akynzeo injection and capsule**
	  ○ Documented treatment failure with both of the following while receiving the current chemotherapy regimen:
	  ▪ 5-HT3 receptor antagonist (e.g., ondansetron, granisetron or palonosetron)
	  ▪ NK1 receptor antagonist (e.g., aprepitant, fosaprepitant or rolapitant)
	
	Quantity Limit:
	• Varubi: 1 dose per 14 days
	• Akynzeo injection and capsule: 1 dose per 7 days
	
	Reauthorization requires documentation of treatment success and initial criteria to be met

Exclusion Criteria:	• Treatment of acute or breakthrough nausea and vomiting • Used in anthracycline or cyclophosphamide-based chemotherapy (Akynzeo injection only)
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 6 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design

Required Medical Information:	• Documentation of dose based on reasonable projections, current dose utilization, product labeling, diagnosis, baseline factor level, circulating factor activity (% of normal or units/dL), and rationale for use
	• Current weight
	• Documentation of Bethesda Titer level and number of bleeds in the past 3 months with severity and cause of bleed
	
	<u>Documentation of one of the following diagnostic categories:</u>
	• Hemophilia A or Hemophilia B
	  ○ Mild: factor levels greater than 5% and less than 30%
	  ○ Moderate: factor levels of 1% to 5%
	  ○ Severe: factor levels of less than 1%
	• Von Willebrand disease (VWD), which must be confirmed with plasma von Willebrand factor (VWF) antigen, plasma VWF activity, and factor VIII activity
	
	<u>Documentation of one of the following indications:</u>
	• Acute treatment of moderate to severe bleeding in patients with:
	  ○ Mild, moderate, or severe hemophilia A or B
	  ○ Severe VWD
	  ○ Mild to moderate VWD in clinical situations with increased risk of bleeding
	• Perioperative prophylaxis and/or treatment of acute, moderate to severe bleeding in patients with hemophilia A, hemophilia B,

	or VWD • Routine prophylaxis in patients with severe hemophilia A, severe hemophilia B, or severe VWD ○ For Wilate and Vonvendi for routine prophylaxis: documentation of severe Type 3 VWD

Appropriate Treatment Regimen & Other Criteria:	**<u>Hemophilia A (factor VIII deficiency)</u>**
	• Documentation indicates requested medication is to achieve or maintain but not to exceed maximum functional capacity in performing daily activities
	• For mild disease: treatment failure or contraindication to Stimate (desmopressin)
	• **Eloctate** and **Nuwiq** require documented inadequate response, or documented intolerable adverse event, with all preferred products (Kogenate FS, Kovaltry, Novoeight, Jivi, Adynovate)
	• **Helixate FS** requires documented treatment failure with Kogenate FS due to an intolerable adverse event and the prescriber has a compelling medical rationale for not expecting the same event to occur with Helixate FS
	• **Altuviiio** requires documentation of severe hemophilia or moderate hemophilia with a severe bleeding phenotype defined by frequent non-traumatic bleeds requiring prophylaxis
	
	<u>Hemophilia B (factor IX deficiency)</u>
	• For **Benefix**, **Idelvion,** and **Rebinyn**: documentation treatment failure or contraindication to Rixubis
	• For **Alprolix**: documentation of contraindication to Rixubis for perioperative management
	
	<u>von Willebrand disease (VWD)</u>
	• For **Vonvendi**:
	  ○ Documentation of treatment failure or contraindication to Humate P AND Alphanate for perioperative prophylaxis and/or treatment of acute, moderate to severe bleeding
	  ○ Documentation of treatment failure or contraindication to Wilate for routine prophylaxis

	<u>All Indications</u>
	• Approval based on necessity and laboratory titer levels
	• Coverage for a non-preferred product requires documentation of one of the following:
	  ○ Documented intolerable adverse event to all preferred products, and the adverse event was not an expected adverse event attributed to the active ingredient
	  ○ Currently receiving treatment with a non-preferred product, excluding via samples or manufacturer's patient assistance programs
	
	<u>Reauthorization</u>: requires documentation of planned treatment dose, number of acute bleeds since last approval (with severity and cause of bleed), past treatment history, and titer inhibitor level to factor VIII and IX as appropriate

Exclusion Criteria:	• Acute thrombosis, embolism, or symptoms of disseminated intravascular coagulation • Obizur for congenital hemophilia A or VWD • Tretten for congenital factor XIII B-subunit deficiency • Jivi and Adynovate for VWD • Idelvion for immune tolerance induction in patients with Hemophilia B • Vonvendi for congenital hemophilia A or hemophilia B • Afstyla and Nuwiq for VWD
Age Restriction:	• Subject to review of FDA label for each product • Jivi and Adynovate: 12 years of age and older • Vonvendi: 18 years of age and older • Wilate for routine prophylaxis with von Willebrand disease: 6 years and older
Prescriber Restrictions:	• Prescribed by, or in consultation with, a hematologist • Members who are on a State Based Drug List are required to utilize pharmacy benefits only • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Authorization: 12 months, unless otherwise specified • Perioperative management: 1 month, unless otherwise specified

POLICY NAME:
ANTITHYMOCYTE GLOBULIN
Affected Medications: ATGAM

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Management of allograft rejection in renal transplant patients
	  ○ Treatment of moderate to severe aplastic anemia in patients unsuitable for bone marrow transplantation
	• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better
	• 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
	  ○ 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
	  ○ MDS: 40 mg/kg once daily for 4 days
	  ○ 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 • Oncology: Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater • 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 • Specialist in oncology, hematology or transplant medicine

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

POLICY NAME:
ANTITHROMBIN ALFA
Affected Medications: ATRYN

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) • Can be given for prophylaxis if negative personal/family history of thromboembolic events in high risk-settings as in surgery and pregnancy. • Patient weight • 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 **<u>Peri-partum thromboembolic prophylaxis</u>** • 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 • If negative personal history of VTE, patient may need single dose of ATryn • ATryn use is limited to third trimester • If positive personal/family history of VTE, ATryn recommended • Can be concomitantly given with LMWH or heparin **<u>Peri-operative thromboembolic event prophylaxis</u>** • Used during warfarin interruption leading up to surgical procedure (with or without heparin) • Utilized until patient can resume warfarin therapy
Exclusion Criteria:	• Hypersensitivity to goats and goat milk protein • Administration within first two trimesters of pregnancy • Active thromboembolic event
Age Restriction:	• 18 – 65 years of age
Prescriber/Site of Care Restrictions:	• OB-GYN, MD • All approvals are subject to utilization of the most cost-effective site of care

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

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

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

POLICY NAME:
ANTI-TUBERCULOSIS AGENTS
Affected Medications: SIRTURO (bedaquiline), PRETOMANID

Covered Uses:	• All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design. ○ **Sirturo** ▪ Treatment of adult and pediatric patients with pulmonary tuberculosis (TB) due to *Mycobacterium tuberculosis* resistant to at least rifampin and isoniazid ○ **Pretomanid** ▪ Treatment of adults with pulmonary TB resistant to isoniazid, rifamycins, a fluoroquinolone and a second line injectable antibacterial drug ▪ Treatment of adults with pulmonary TB resistant to isoniazid and rifampin who are treatment-intolerant or nonresponsive to standard therapy
Required Medical Information:	**Sirturo** • Documented diagnosis of multidrug resistant TB (MDR-TB), defined as resistance to at least isoniazid and rifampin **Pretomanid** • Documented diagnosis of one of the following: ○ Extensively drug resistant TB (XDR-TB) ○ Treatment-intolerant or nonresponsive MDR-TB
Appropriate Treatment Regimen & Other Criteria:	**Sirturo** • Documentation that this drug has been prescribed as part of a combination regimen with other anti-tuberculosis agents • Documentation that this drug is being administered by directly observed therapy (DOT) **Pretomanid** • Documentation that this drug has been prescribed as part of a combination regimen with Sirturo (bedaquiline) and linezolid • Documentation that this drug is being administered by DOT

Exclusion Criteria:	• Drug-sensitive (DS) pulmonary TB • Latent infection due to *Mycobacterium tuberculosis* • Extra-pulmonary infection due to *Mycobacterium tuberculosis* • Infections caused by non-tuberculous mycobacteria
Age Restriction:	• **Sirturo**: 5 years of age and older • **Pretomanid**: 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an infectious disease specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	**Sirturo** • Authorization: 24 weeks, unless otherwise specified **Pretomanid** • Authorization: 26 weeks, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Acute, intermittent treatment of hypomobility, "off" episodes in patients with advanced Parkinson's disease (PD)
Required Medical Information:	• Diagnosis of advanced PD • Documentation of acute, intermittent hypomobility, "off" episodes occurring for at least 2 hours per day while awake despite an optimized oral PD treatment regimen

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:
	  ○ Monoamine oxidase-B (MAO-B) inhibitors (ex: selegiline, rasagiline)
	  ○ Dopamine agonists (ex: amantadine, pramipexole, ropinirole)
	  ○ Catechol-O-methyltransferase (COMT) inhibitors (ex: entacapone)
	
	Requests for Apokyn and apomorphine solution require documentation of treatment failure or contraindication to Kynmobi
	
	<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
Age Restriction:	

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

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of *Mycobacterium avium* complex (MAC) lung disease as part of a combination antibacterial drug regimen in adults who have limited or no alternative treatment options, and who do not achieve negative sputum cultures after a minimum of 6 consecutive months of a multidrug background regimen therapy

Required Medical Information:	• Diagnosis of MAC lung disease confirmed by BOTH of the following:
	  ○ A MAC-positive sputum culture obtained within the last 3 months
	  ○ Evidence of underlying nodular bronchiectasis and/or fibrocavity disease on a chest radiograph or chest computed tomography
	• The MAC isolate is susceptible to amikacin with a minimum inhibitory concentration (MIC) of less than or equal to 64 µg/mL
	• Documentation of failure to obtain a negative sputum culture after a minimum of 6 consecutive months of a multidrug background regimen therapy for MAC lung disease such as clarithromycin (or azithromycin), rifampin and ethambutol

Appropriate Treatment Regimen & Other Criteria:	• Documentation of BOTH of the following:
	  ○ This drug has been prescribed as part of a combination antibacterial drug regimen
	  ○ This drug will be used with the Lamira® Nebulizer System
	
	<u>Reauthorization</u> requires documentation of negative sputum culture obtained within the last 30 days.

	• The American Thoracic Society/Infectious Diseases Society of America (ATS/IDSA) guidelines state that patients should continue to be treated until they have negative cultures for 1 year. Treatment beyond the first reauthorization (after 18 months) will require documentation of a positive sputum culture to demonstrate the need for continued treatment. Patients that have had negative cultures for 1 year will not be approved for continued treatment.
Exclusion Criteria:	• Diagnosis of non-refractory MAC lung disease
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an infectious disease specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
ASCIMINIB
Affected Medications: SCEMBLIX (asciminib)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design • 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 • Documentation of Philadelphia chromosome or BCR::ABL1-positive chronic myeloid leukemia (CML) in chronic phase (CP) OR advanced phase in accelerated phase (AP-CML)
Appropriate Treatment Regimen & Other Criteria:	**<u>Advanced phase chronic myeloid leukemia (CML)</u>** • Documentation of accelerated phase by 10 to 19 percent blasts in blood or bone marrow **<u>Philadelphia chromosome or BCR::ABL1- positive chronic myeloid leukemia (CML) in chronic phase (CP) meeting one of the following:</u>** • Previous treatment with imatinib [if used as initial tyrosine kinase inhibitor (TKI)] AND one or more additional tyrosine kinase inhibitor (TKI) such as: ○ A second generation TKI which includes: bosutinib, dasatinib, or nilotinib. (Note BCR:ABL1 kinase domain mutation status for selections and contraindications) • Documented resistance or intolerance to at least two prior TKIs • Documented T315I positive mutation and clinical failure with ponatinib **<u>Reauthorization</u>** requires documentation of disease responsiveness to therapy
Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater • Presence of either A337T, P465S, M244V, or F359V/I/C

	BCR::ABL1 kinase domain mutation
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design ○ Treatment of children with pre-symptomatic late-infantile (PSLI), pre-symptomatic early-juvenile (PSEJ), or early symptomatic early-juvenile (ESEJ) metachromatic leukodystrophy (MLD)

Required Medical Information:	• Diagnosis of metachromatic leukodystrophy (MLD) confirmed by the following:
	  ○ Arylsulfatase (ARSA) activity below the normal range in peripheral blood mononuclear cells or fibroblasts
	  ○ Presence of two disease-causing mutations of either known or novel alleles
	  ○ Presence of sulfatides in a 24-hour urine collection (to exclude MLD carriers and patients with ARSA pseudodeficiency)
	AND
	• Diagnosis of the late-infantile subtype of MLD confirmed by two out of three of the following:
	  ○ Age at onset of symptoms in the older sibling(s) less than or equal to 30 months
	  ○ Two null (0) mutant ARSA alleles
	  ○ Peripheral neuropathy as determined by electroneurographic study
	OR
	• Diagnosis of the early-juvenile subtype of MLD confirmed by two out of three of the following:
	  ○ Age at onset of symptoms (in the patient or in the older sibling) between 30 months and 6 years (has not celebrated their seventh birthday)
	  ○ One null (0) and one residual (R) mutant ARSA allele(s)
	  ○ Peripheral neuropathy as determined by electroneurographic study

Appropriate Treatment Regimen & Other Criteria:	
Exclusion Criteria:	• Allogeneic hematopoietic stem cell transplantation in the previous six months • Previous gene therapy • Documented HIV infection • Documented history of a hereditary cancer
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by or in consultation with a neurologist or hematologist/oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 2 months (for one time infusion), no reauthorization, unless otherwise specified

POLICY NAME:
AVACOPAN
Affected Medications: TAVNEOS 10mg capsule

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ 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:
	  ○ Tissue biopsy of kidney or other affected organs
	  ○ Positive ANCA, clinical presentation compatible with AAV, and low suspicion for secondary vasculitis
	  ○ Clinical presentation compatible with AAV, low suspicion for secondary vasculitis, and concern for rapidly progressive disease
	• 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)
	• Documentation of all prior therapies used and anticipated treatment course
	• Baseline liver test panel: serum alanine aminotransferase, aspartate aminotransferase, alkaline phosphatase, and total bilirubin
	• Current hepatitis B virus (HBV) status

Appropriate Treatment Regimen & Other Criteria:	• Will be used with a standard immunosuppressive regimen including glucocorticoids • Will be used during induction therapy only • Will be used in any of the following populations/scenarios:

	○ In patients unable to use glucocorticoids at appropriate doses ○ In patients with an estimated glomerular filtration rate less than 30 mL/min/1.73 m2 ○ In patients who have experienced relapse following treatment with two or more different induction regimens, including both rituximab- and cyclophosphamide-containing regimens (unless contraindicated) ○ During subsequent induction therapy in patients with refractory disease (failure to achieve remission with initial induction therapy regimen) • Dosing: 30 mg (three 10 mg capsules) twice daily (once daily when used concomitantly with strong CYP3A4 inhibitors) • Reauthorization: must meet criteria above (will not be used for maintenance treatment)
Exclusion Criteria:	• Treatment of eosinophilic-GPA (EGPA) • Active, untreated and/or uncontrolled chronic liver disease (e.g., chronic active hepatitis B, untreated hepatitis C virus infection, uncontrolled autoimmune hepatitis) and cirrhosis • Active, serious infections, including localized infections • History of angioedema while receiving Tavneos, unless another cause has been established • 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 • 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)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ 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. • Patient weight and planned treatment regimen.

Appropriate Treatment Regimen & Other Criteria:	• One or more clinical signs or symptoms of Late-Onset Pompe Disease:
	  ○ Progressive proximal weakness in a limb-girdle distribution
	  ○ Delayed gross-motor development in childhood
	  ○ Involvement of respiratory muscles causing respiratory difficulty (such as reduced forced vital capacity [FVC] or sleep disordered breathing)
	  ○ Skeletal abnormalities (such as scoliosis or scapula alata)
	  ○ Low/absent reflexes
	• Appropriate medical support is readily available when medication is administered in the event of anaphylaxis, severe allergic reaction, or acute cardiorespiratory failure.
	• Patients weighing less than 30 kilograms will require documented treatment failure or intolerable adverse event to Lumizyme.
	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced.
	
	<u>Reauthorization</u> will require documentation of treatment success and a clinically significant response to therapy.

Exclusion Criteria:	• Diagnosis of infantile-onset Pompe Disease • Concurrent treatment with Lumizyme
Age Restriction:	• 1 year of age or older

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)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Thrombocytopenia in adult patients with chronic liver disease (CLD) who are scheduled to undergo a procedure
	  ○ Thrombocytopenia in adult patients with chronic immune thrombocytopenia (ITP) who have had an insufficient response to a previous treatment

Required Medical Information:	**<u>Thrombocytopenia in patients with CLD undergoing a procedure</u>**
	• Documentation of planned procedure including date
	• Documentation of baseline platelet count of less than 50,000/microliter
	
	<u>Thrombocytopenia in patients with chronic ITP</u>
	• Documentation of **ONE** of the following:
	  ○ Platelet count less than 20,000/microliter
	  ○ Platelet count less than 30,000/microliter AND symptomatic bleeding
	  ○ Platelet count less than 50,000/microliter AND increased risk for bleeding (such as peptic ulcer disease, use of antiplatelets or anticoagulants, history of bleeding at higher platelet count, need for surgery or invasive procedure)

Appropriate Treatment Regimen & Other Criteria:	**<u>Thrombocytopenia in patients with chronic (ITP):</u>**
	• Documentation of inadequate response, defined as platelets did not increase to at least 50,000/microliter, to the following therapies:
	  ○ **ONE** of the following:
	  ▪ Inadequate response with at least 2 therapies for immune thrombocytopenia, including corticosteroids, rituximab, or immunoglobulin
	  ▪ Splenectomy
	  ○ Promacta

	<u>Reauthorization (chronic ITP only)</u> • Response to treatment with platelet count of at least 50,000/microliter (not to exceed 400,000/microliter) **OR** • The platelet counts have not increased to at least 50,000/microliter and the patient has NOT been on the maximum dose for at least 4 weeks
Exclusion Criteria:	• Use in combination with another thrombopoietin receptor agonist, spleen tyrosine kinase inhibitor, or similar treatments (Promacta, Nplate, Tavalisse)
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist or gastroenterology/liver specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• **Thrombocytopenia in patients with CLD undergoing a procedure**: o 1 month (for a one time 5-day regimen), unless otherwise specified • **Thrombocytopenia in patients with chronic ITP**: o Initial Authorization: 4 months, unless otherwise specified o Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
AVONEX
Affected Medications: AVONEX, AVONEX PEN

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.
	  ○ Treatment of relapsing forms of Multiple Sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive disease (SPMS)

Required Medical Information:	• Diagnosis confirmed with magnetic resonance imaging (MRI), per revised McDonald diagnostic criteria for MS ▪ 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 • Prescribed by, or in consultation with, a neurologist
Coverage Duration:	• Approval: 12 months, unless otherwise specified

POLICY NAME:
AZTREONAM
Affected Medications: CAYSTON (aztreonam)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design • Cystic fibrosis
Required Medical Information:	• Documentation of confirmed diagnosis of cystic fibrosis • Culture and sensitivity report confirming presence of Pseudomonas aeruginosa in the lungs • Baseline FEV1 greater than 25% but less than 75% predicted
Appropriate Treatment Regimen & Other Criteria:	• Documented failure, contraindication, or resistance to inhaled tobramycin. • **Dosing:** 28 days on and 28 days off **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 • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
BELIMUMAB
Affected Medications: BENLYSTA (belimumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Systemic Lupus Erythematosus (SLE)
	  ○ Lupus Nephritis (LN)

Required Medical Information:	• Documentation of current weight (intravenous requests only)
	
	<u>Systemic Lupus Erythematosus:</u>
	
	• Documentation of active SLE with moderate classification (significant but non-organ threatening disease including constitutional, cutaneous, musculoskeletal, or hematologic involvement)
	• Autoantibody-positive SLE, defined as positive for antinuclear antibodies (ANA) and/or anti-double-stranded DNA (anti-dsDNA) antibody
	• Baseline measurement of **ONE** or more of the following:
	  ○ SLE Responder Index-4 (SRI-4)
	  ○ Frequency of flares requiring corticosteroid use
	
	<u>Lupus Nephritis:</u>
	
	• Documentation of biopsy-proven active Class III, IV, and/or V disease
	• Baseline measurement of one or more of the following: urine protein-creatinine ratio (uPCR), urine protein, estimated glomerular filtration rate (eGFR), or frequency of flares or corticosteroid use

Appropriate Treatment Regimen & Other Criteria:	**<u>All uses:</u>**
	• Use of intravenous formulation requires:
	  ○ Documented inability to use subcutaneous formulation OR
	  ○ Currently receiving treatment with the intravenous formulation, excluding via samples or manufacturer's patient assistance programs

	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced (intravenous requests only) **<u>Systemic Lupus Erythematosus:</u>** • Failure with at least 12 weeks of combination therapy including hydroxychloroquine OR chloroquine with one of the following: ○ Cyclosporine, azathioprine, methotrexate, or mycophenolate mofetil **<u>Reauthorization</u>** requires documentation of treatment success defined as a clinically significant improvement in Systemic Lupus Erythematosus Responder Index-4 (SRI-4) **OR** decrease in flares or corticosteroid use **<u>Lupus Nephritis:</u>** • No dialysis in the past 12 months AND estimated glomerular filtration rate (eGFR) equal to or above 30 mL/min/1.73m^2 • Failure of at least 12 weeks of mycophenolate mofetil AND cyclophosphamide **<u>Reauthorization</u>** requires documentation of treatment success defined as ONE of the following: • Improvement in eGFR • Reduction in urinary protein-creatinine ratio or urine protein • Decrease in flares or corticosteroid use
Exclusion Criteria:	• Use in combination with other biologic therapies for LN or SLE • Use in severe active central nervous system lupus
Age Restriction:	• 5 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a nephrologist, rheumatologist, or specialist with experience in the treatment of systemic lupus erythematosus or lupus nephritis • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
BELZUTIFAN
Affected Medications: WELIREG (belzutifan)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better

Required Medical Information:	**<u>Von Hippel-Lindau (VHL) disease</u>**
	• Diagnosis documented by the following:
	  ○ Pathogenic VHL germline mutation diagnostic for VHL disease AND at least one of the following:
	  ▪ Presence of solid, locoregional tumor in kidney showing accelerated tumor growth (growth of 5 mm or more per year)
	  ▪ Presence of symptomatic and/or progressively enlarging central nervous system (CNS) hemangioblastomas not amenable to surgery
	  ▪ Presence of pancreatic solid lesion or pancreatic neuroendocrine tumor (pNET) with rapid tumor growth
	
	<u>Treatment-refractory advanced or metastatic clear cell renal carcinoma</u>
	• Advanced disease after use of the following treatments (per NCCN guidelines):
	  ○ A programmed death receptor-1 (PD-1) OR programmed death-ligand 1 (PD-L1) AND
	  ○ A vascular endothelial growth factor tyrosine kinase inhibitor (VEGF-TKI)
	
	• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course

Appropriate Treatment Regimen & Other Criteria:	**<u>Reauthorization:</u>** documentation of disease responsiveness to therapy

Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater • Metastatic pNET disease • Not to be used in combination with other oncologic agents for the treatment of VHL disease
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
BENRALIZUMAB
Affected Medications: FASENRA (benralizumab subcutaneous injection)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Add-on maintenance treatment of patients with severe asthma aged 6 years and older with an eosinophilic phenotype

Required Medical Information:	• Diagnosis of severe asthma with an eosinophilic phenotype, defined by both of the following:
	  ○ Baseline eosinophil count of at least 150 cells/μL **OR** dependent on daily oral corticosteroids
	  AND
	  ○ FEV1 less than 80% at baseline or FEV1/FVC reduced by at least 5% from normal

Appropriate Treatment Regimen & Other Criteria:	• Documented use of high-dose inhaled corticosteroid (ICS) plus a long-acting beta agonist (LABA) for at least three months with continued symptoms
	AND
	• Documentation of one of the following:
	  ○ 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
	  ○ Documentation that chronic daily oral corticosteroids are required
	
	<u>Reauthorization</u>: documentation of treatment success and a clinically significant response to therapy

Exclusion Criteria:	• Use in combination with another monoclonal antibody (e.g., Dupixent, Nucala, Xolair, Cinqair, Tezspire)
Age Restriction:	• 6 years of age and older

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an allergist, immunologist, or pulmonologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Dystrophic Epidermolysis Bullosa (DEB)

Required Medical Information:	• Diagnosis of recessive DEB confirmed by both of the following:
	  ○ Skin biopsy of an induced blister with immunofluorescence mapping (IFM) and/or transmission electron microscopy (TEM)
	  ○ Genetic test results documenting mutations in the COL7A1 gene
	• Clinical signs and symptoms of DEB such as skin fragility, blistering, scarring, nail changes, and milia formation in the areas of healed blistering

Appropriate Treatment Regimen & Other Criteria:	• Documentation of receiving standard of care preventative or treatment therapies for wound care, control of infection, nutritional support
	• Documented trial and failure of Filsuvez
	• **Dosing** is in accordance with FDA labeling and does not exceed the following:
	  ○ Maximum weekly volume of 2.5 mL (1.6 mL useable dose)
	  ○ Maximum of 12-week course per wound
	  ○ Maximum of 4 tubes per 28 days
	
	Reauthorization will require documentation of treatment success defined as complete wound healing on a previous site and need for treatment on a new site

Exclusion Criteria:	• Evidence or history of squamous cell carcinoma in the area that will undergo treatment • Concurrent use with Filsuvez (birch triterpenes topical gel) • Dominant DEB (DDEB)
Age Restriction:	• 6 months of age and older

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a dermatologist or a specialist experienced in the treatment of epidermolysis bullosa • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 3 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of adults with polycythemia vera • 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
	• Evidence of increased red cell volume such as abnormal hemoglobin, hematocrit, or red cell mass **AND** one of the following:
	  ○ Presence of JAK2 V617F or JAK2 exon 12 mutation
	  ○ Subnormal serum erythropoietin level

Appropriate Treatment Regimen & Other Criteria:	• Documentation of treatment failure, intolerance, or contraindication to hydroxyurea **Reauthorization** requires documentation of disease responsiveness to therapy
Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist or hematologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Homocystinuria

Required Medical Information:	• Diagnosis of homocystinuria associated with one of the following:
	  ○ Cystathionine beta-synthase (CBS) deficiency
	  ○ 5,10-methylenetetrahydrofolate reductase (MTHFR) deficiency
	  ○ Cobalamin cofactor metabolism (cbl) defect
	• Baseline plasma homocysteine levels

Appropriate Treatment Regimen & Other Criteria:	• Documented trial and failure of **ONE** of the following forms of supplementation:
	  ○ Vitamin B6 (pyridoxine)
	  ○ Vitamin B9 (folate)
	  ○ Vitamin B12 (cobalamin)
	
	Reauthorization will require documentation of treatment success and a clinically significant response to therapy shown by lowering of plasma homocysteine levels

Exclusion Criteria:	• Uncorrected vitamin B12 or folic acid levels
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a metabolic or genetic disease specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive disease (SPMS)

Required Medical Information:	• Diagnosis confirmed with magnetic resonance imaging (MRI), per revised McDonald diagnostic criteria for MS ○ 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 indicated for the treatment of multiple sclerosis
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or multiple sclerosis specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Approval: 24 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ 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:
	  ○ 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
	  ○ Confirmed genetic testing based on the presence of biallelic mutations at the beta-globin gene (*HBB* gene)
	• Clinically stable and eligible to undergo hematopoietic stem cell transplant (HSCT)
	• Used as single agent therapy (not applicable to lymphodepleting or bridging therapy while awaiting manufacture)
	• 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 • Severe iron overload warranting exclusion from therapy, as determined by the treating physician • Uncorrected bleeding disorder • Cardiac T2* less than 10 milliseconds by magnetic resonance imaging (MRI)

	• White blood cell count less than $3x10^9$/L and/or platelet count less than $100x10^9$/L that is unrelated to hypersplenism • 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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months (one-time infusion), unless otherwise specified

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

Covered Uses:	• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher
	• For the Treatment of Ophthalmic disorders:
	  ○ Neovascular (Wet) Age-Related Macular Degeneration (AMD)
	  ○ Macular Edema Following Retinal Vein Occlusion (RVO)
	  ○ Diabetic Macular Edema (DME)
	  ○ Diabetic Retinopathy (DR) in patients with Diabetes Mellitus

Required Medical Information:	• Documentation of disease staging, all prior therapies used, and anticipated treatment course

Appropriate Treatment Regimen & Other Criteria:	**<u>Stage III or IV Epithelial Ovarian, Fallopian Tube, or Primary Peritoneal Cancer following initial surgical resection</u>**
	• Approval will be limited for up to 22 cycles of therapy
	
	<u>All Indications</u>
	• Coverage for a non-preferred product (Avastin, Alymsys, Vegzelma) requires documentation of one of the following:
	  ○ Use for an ophthalmic condition (Avastin only)
	  ○ A documented intolerable adverse event to the preferred products, Mvasi and Zirabev, and the adverse event was not an expected adverse event attributed to the active ingredient
	
	<u>Reauthorization</u> requires documentation of disease responsiveness to therapy

Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater
Age Restriction:	

Prescriber/Site of Care Restrictions:	• Oncologic indication: prescribed by, on in consultation with, an oncologist • Ophthalmic indication: prescribed by, on in consultation with, an ophthalmologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
BEZLOTOXUMAB
Affected Medications: ZINPLAVA (bezlotoxumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Reduce recurrence of Clostridioides difficile infection (CDI) in patients who are receiving antibacterial drug treatment for CDI and are at a high risk for CDI recurrence

Required Medical Information:	• Diagnosis of CDI confirmed by both of the following:
	  ○ Presence of at least 3 unformed stools in 24 hours
	  ○ Positive stool test for toxigenic Clostridium difficile collected within 7 days prior to request
	• Patient must be receiving concurrent CDI treatment when infusion is administered

Appropriate Treatment Regimen & Other Criteria:	• Documentation of **ONE** of the following risk factors for CDI recurrence:
	  ○ Age greater than 65
	  ○ One or more episodes of CDI in the past 6 months prior to the current episode
	  ○ Immunocompromised status
	  ○ Clinically severe CDI (defined by Zar score greater than or equal to 2)
	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced

Exclusion Criteria:	• Previous treatment with Zinplava
Age Restriction:	• 1 year of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an infectious disease specialist or gastroenterologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 1 month (a single 10 mg/kg dose) with no reauthorization, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design
	  ○ Dystrophic Epidermolysis Bullosa (DEB)
	  ○ Junctional Epidermolysis Bullosa (JEB)

Required Medical Information:	• Diagnosis of recessive DEB or JEB confirmed by skin biopsy of an induced blister with immunofluorescence mapping (IFM) and/or transmission electron microscopy (TEM) • Genetic test results documenting mutations in one of the following genes: COL7A1, COL17A1, ITGB4, LAMA3, LAMB3, or LAMC2 • Clinical signs and symptoms of EB such as skin fragility, blistering, scarring, nail changes, and milia formation in the areas of healed blistering • Presence of open partial-thickness wounds that have been present for at least 21 days

Appropriate Treatment Regimen & Other Criteria:	• Documentation of receiving standard of care preventative or treatment therapies for wound care, control of infection, nutritional support.
	• **Dosing** does not exceed the following:
	  ○ Maximum of 1 mm layer to affected area(s)
	  ○ Maximum of 28 tubes per 28 days
	
	Reauthorization requires documentation of treatment success defined as complete wound healing on a previous site and need for continued treatment on a new site

Exclusion Criteria:	• Concurrent use with Vyjuvek (beremagene geperpavec-svdt) • Dominant DEB (DDEB)
Age Restriction:	• 6 months of age and older

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a dermatologist or a specialist experienced in the treatment of epidermolysis bullosa • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 3 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved or compendia-supported indications not otherwise excluded by plan design
	  ○ Spasticity
	  ○ Chronic migraine
	  ○ Overactive bladder (OAB) with symptoms of urge urinary incontinence, urgency, and frequency
	  ○ Neurogenic detrusor overactivity (NDO)
	  ○ Focal dystonia
	  ▪ Cervical dystonia
	  ▪ Blepharospasm
	  ▪ Laryngeal dystonia
	  ▪ Oromandibular dystonia
	  ▪ Severe brachial dystonia (writer's cramp)
	  ○ Strabismus
	  ○ Primary axillary hyperhidrosis
	  ○ Achalasia
	  ○ Anal fissure

Required Medical Information:	• Pertinent medical records and diagnostic testing • Complete description of the site(s) of injection • 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 plan design that are not listed below, failure of first-line recommended and conventional therapies is required • Approved first-line for: focal dystonia, hemifacial spasm, orofacial dyskinesia, upper/lower limb spasticity, or other conditions of focal spasticity wherein botulinum toxin is the preferred mode of therapy

	Overactive bladder (OAB)/Neurogenic detrusor overactivity (NDO):
	• Documentation of inadequate response or intolerance to at least two urinary incontinence anticholinergic agents (e.g., oxybutynin, solifenacin, tolterodine)
	
	Chronic migraine
	• Documentation of chronic migraine defined as headaches on at least 15 days per month, of which at least 8 days are with migraine
	• Documented failure with an adequate trial (at least 8 weeks) of a migraine preventative therapy, as follows:
	  ○ Candesartan 16 mg daily
	  ○ Antiepileptic (divalproex sodium 500 mg daily, valproic acid 500 mg daily, topiramate 50 mg daily)
	  ○ Beta-blocker (metoprolol 100 mg daily, propranolol 40 mg daily, timolol 20 mg daily, nadolol 80 mg daily)
	  ○ Antidepressant (amitriptyline 25 mg daily, nortriptyline 25 mg daily, venlafaxine 75 mg daily, duloxetine 60 mg daily)
	  ○ Anti-calcitonin gene-related peptide (CGRP) monoclonal antibody or CGRP receptor antagonist (when used for prevention)
	
	Primary Axillary Hyperhidrosis
	• Thyroid-stimulating hormone (TSH) level AND inadequate response to two or more alternative therapies (topical aluminum chloride 20%, iontophoresis, oral glycopyrrolate, oral oxybutynin)
	
	Achalasia (Cardiospasm) - must meet 1 of the following
	• Type I or II achalasia: Treatment failure with peroral endoscopic myotomy (POEM), laparoscopic Heller myotomy (LHM), and pneumatic dilation (PD)
	• Type III achalasia: Treatment failure with tailored POEM and LHM
	• Not a candidate for POEM, surgical myotomy, or pneumatic dilation due to high risk of complications

	<u>Anal fissure</u>
	• Documented failure or intolerance to an 8-week trial of each of the following:
	  ○ Rectiv ointment
	  ○ Topical diltiazem or topical nifedipine
	
	<u>Number of treatments must not exceed the following:</u>
	• OAB/NDO: 2 treatments/12 months
	• Chronic migraine: initial treatment limited to two injections given 3 months apart, subsequent treatment approvals limited to 4 treatments per 12 months
	• Primary axillary hyperhidrosis: 2 treatments/12 months
	• Anal fissure: 2 treatments/12 months
	• All other indications maximum of 4 treatments/12 months unless otherwise specified
	
	<u>Reauthorization:</u>
	• **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.
	• All other indications: Documentation of treatment success and a clinically significant response to therapy.

Exclusion Criteria:	• Cosmetic procedures
	• For intradetrusor injections: documented current/recent urinary tract infection or urinary retention
	• Possible medication overuse headache: headaches occurring 15 or more days each month in a patient with pre-existing headache-causing condition possibly due to
	  ○ Use of ergotamines, triptans, opioids, or combination analgesics at least 10 days per month for at least three months
	  ○ Use of simple analgesics (acetaminophen, aspirin, or an NSAID) at least 15 days per month for at least 3 months
	  ○ Combined use of any of the previously mentioned products without overuse of any one agent if no causative pattern can be established

	• Combined use with an anti-calcitonin gene-related peptide (CGRP) monoclonal antibody or an oral CGRP antagonist when used for migraine prevention
Age Restriction:	

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist for the following:
	  ○ Blepharospasm, strabismus: ophthalmologist, optometrist, or neurologist
	  ○ Chronic migraine: neurologist or headache specialist
	  ○ OAB/NDO: urologist or neurologist
	  ○ Anal fissure: gastroenterologist or colorectal surgeon
	• All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	**Chronic migraine:** • Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified **OAB/NDO:** • Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified **Anal Fissure:** • Authorization: 3 months (one treatment), unless otherwise specified **All other indications:** • Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ X-linked hypophosphatemia (XLH)
	  ○ FGF23-related hypophosphatemia in tumor induced osteomalacia (TIO) associated with phosphaturic mesenchymal tumors

Required Medical Information:	**<u>All Indications</u>**
	• Documentation of diagnosis by:
	  ○ A blood test demonstrating ALL of the following (in relation to laboratory reference ranges):
	  ▪ Low phosphate
	  ▪ Elevated FGF23
	  ▪ Low 1,25-(OH)2D
	  ▪ Normal calcium or parathyroid hormone (PTH)
	  ○ A urine test demonstrating decreased tubular reabsorption of phosphate corrected for glomerular filtration rate (TmP/GFR)
	  ○ Evidence of skeletal abnormalities, confirmed by radiographic evaluation
	
	<u>Tumor-Induced Osteomalacia</u>
	• Documentation that tumor cannot be located or is unresectable
	• Alternative renal phosphate-wasting disorders have been ruled out

Appropriate Treatment Regimen & Other Criteria:	**<u>All Indications</u>** • Documentation of treatment failure with at least 12 months of oral phosphate and calcitriol supplementation in combination, unless contraindicated or not tolerated • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced

	<u>Reauthorization</u> requires: • Documentation of normalization of serum phosphate levels • If established on therapy for 12 months or more, improvement in radiographic imaging of skeletal abnormalities
Exclusion Criteria:	
Age Restriction:	
Prescriber Restrictions:	• Prescribed by, or in consultation with, a nephrologist, endocrinologist, or a provider experienced in managing patients with metabolic bone disease • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of secondary hyperparathyroidism in adult patients with stage 3 or 4 chronic kidney disease (CKD) and serum total 25-hydroxyvitamin D levels less than 30 ng/mL

Required Medical Information:	• A confirmed diagnosis of secondary hyperparathyroidism with persistently elevated or progressively rising serum intact parathyroid hormone (iPTH) that is at least 2.3 times above the upper limit of normal for the assay used
	• Documentation of all of the following prior to treatment initiation:
	  ○ Stage 3 or 4 CKD
	  ○ Serum total 25-hydroxyvitamin D level is less than 30 ng/mL
	  ○ Corrected serum calcium is below 9.8 mg/dL

Appropriate Treatment Regimen & Other Criteria:	• Documentation of persistent vitamin D deficiency (level below 30 ng/mL), despite at least 12 weeks of adherent treatment with each of the following at an appropriate dose, unless contraindicated or not tolerated:
	  ○ Vitamin D2 (ergocalciferol) or vitamin D3 (cholecalciferol)
	  ○ Calcitriol
	  ○ Doxercalciferol
	  ○ Paricalcitol
	
	<u>Reauthorization</u> will require documentation of a clinically significant response to therapy, evidenced by increased serum total 25-hydroxyvitamin D level (to at least 30 ng/mL) and reduced plasma iPTH to goal therapeutic range (or an approximate 30% reduction compared to baseline)

Exclusion Criteria:	• A diagnosis of stage 1, 2, or 5 chronic kidney disease, or end-stage renal disease (ESRD) on dialysis

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

POLICY NAME:
CANNABIDIOL
Affected Medications: EPIDIOLEX (cannabidiol)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Lennox-Gastaut Syndrome (LGS)
	  ○ Dravet Syndrome (DS)
	  ○ Tuberous Sclerosis Complex (TSC)

Required Medical Information:	**<u>All Indications</u>**
	• Patient weight
	• Documentation that cannabidiol will be used as adjunctive therapy
	
	<u>Lennox-Gastaut Syndrome (LGS)</u>
	• Documentation of at least 8 drop seizures per month while on stable antiepileptic drug therapy
	• Documented treatment and inadequate seizure control with at least three guideline directed therapies including:
	  ○ Valproate **and**
	  ○ Lamotrigine **and**
	  ○ Rufinamide, topiramate, felbamate, or clobazam
	
	<u>Dravet Syndrome (DS)</u>
	• Documentation of at least 4 convulsive seizures in the last month while on stable antiepileptic drug therapy
	• Documented treatment and inadequate seizure control with at least four guideline directed therapies including:
	  ○ Valproate **and**
	  ○ Clobazam **and**
	  ○ Topiramate **and**
	  ○ Clonazepam, levetiracetam, or zonisamide
	
	<u>Tuberous Sclerosis Complex (TSC)</u>
	• Documentation of monotherapy failure for seizure control with two antiepileptic regimens AND
	• Documentation of failure with at least one adjunctive therapy for seizure control

Appropriate Treatment	Dosing:

Regimen & Other Criteria:	• Lennox-Gastaut Syndrome or Dravet Syndrome: Not to exceed 20 mg/kg per day • Tuberous Sclerosis Complex: Not to exceed 25 mg/kg per day **Reauthorization** 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:	• 1 year of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
CANTHARIDIN
Affected Medications: YCANTH

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Molluscum contagiosum (MC)

Required Medical Information:	• Diagnosis of MC confirmed by one of the following:
	  ○ Presence of lesions that are consistent with MC (small, firm, pearly, with pitted centers, 2-5 millimeters in diameter, not associated with systemic symptoms such as fever)
	  ○ For lesions with unclear cause or otherwise not consistent with MC, confirmation of diagnosis using dermoscopy, microscopy, histological examination, or biopsy
	• Documentation of persistent itching or pain AND one of the following:
	  ○ Concomitant bacterial infection of the lesion
	  ○ Concomitant atopic dermatitis
	  ○ Significant concern for contagion (such as daycare setting) and prevention cannot be reasonably prevented through good hygiene and covering lesions with bandages or clothing
	  ○ Continued presence of lesions after 12 months

Appropriate Treatment Regimen & Other Criteria:	• Trial of at least two cycles of one of the following procedures for the removal of MC lesions:
	  ○ Cryotherapy
	  ○ Curettage
	  ○ Laser therapy
	• Adequate trial and failure of one additional treatment for MC that has evidence supporting use, such as:
	  ○ Topical podofilox for at least 1 month
	  ○ Oral cimetidine for at least 2 months

	• **Dosing:** Two applicators per treatment every 21 days, limit to 4 total treatments
Exclusion Criteria:	
Age Restriction:	• 2 years of age or older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a dermatologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 3 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of adult patients with acquired thrombotic thrombocytopenic purpura (aTTP), in combination with plasma exchange and immunosuppressive therapy

Required Medical Information:	• Diagnosis, or suspected diagnosis, of aTTP, meeting all of the following:
	  ○ Severe thrombocytopenia (platelet count less than 100 x 10^9/L)
	  ○ Microangiopathic hemolytic anemia (MAHA) confirmed by red blood cell fragmentation (e.g., schistocytes) on peripheral blood smear
	  ○ Baseline ADAMTS13 activity level of less than 10%
	• Documentation of **ONE** of the following:
	  ○ Failure of at least one initial treatment for aTTP, such as therapeutic plasma exchange (TPE), glucocorticoids, or rituximab
	  ○ Documentation of high-risk disease meeting **ONE** of the following:
	  ▪ Neurologic abnormalities (seizures, focal weakness, aphasia, dysarthria, confusion, coma)
	  ▪ Altered mental status
	  ▪ Elevated serum troponin levels
	• Documentation that Cablivi will be used in combination with standard-of-care treatment for aTTP (TPE and glucocorticoid)

Appropriate Treatment Regimen & Other Criteria:	• Total treatment duration will be limited to 58 days beyond the last TPE treatment **Reauthorization** requires documented signs of ongoing disease (such as suppressed ADAMTS13 activity levels) and no more than 2 recurrences of aTTP while on Cablivi. Recurrence is defined as thrombocytopenia after initial recovery of platelet count (platelet

	count greater than or equal to 150,000) that requires re-initiation of daily plasma exchange.
Exclusion Criteria:	• Use for other causes of thrombocytopenia, such as other TTP-like disorders (congenital or hereditary TTP)
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 3 months (for new episode), unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Neuropathic pain associated with postherpetic neuralgia (PHN)
	  ○ Neuropathic pain associated with diabetic peripheral neuropathy (DPN) of the feet

Required Medical Information:	

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure with at least 12 weeks of ALL of the following:
	  ○ gabapentin
	  ○ pregabalin
	  ○ carbamazepine, oxcarbazepine, or valproic acid/divalproex sodium
	  ○ amitriptyline or nortriptyline
	  ○ topical lidocaine
	
	• Dose limited to a single treatment (up to 4 patches) once every 90 days
	
	Reauthorization: requires documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider

Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a pain management specialist • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 3 months (single treatment), unless otherwise specified • Reauthorization: 12 months (up to 4 treatments), unless otherwise specified

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

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:
	  ○ N-Acetylglutamate Synthase (NAGS) deficiency
	  ○ Propionic Acidemia (PA) or Methylmalonic Acidemia (MMA)
	• Chronic hyperammonemia due to N-Acetylglutamate Synthase (NAGS) deficiency

Appropriate Treatment Regimen & Other Criteria:	**<u>Acute hyperammonemia</u>** • Ammonia level greater than 100 micromol/L • 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) • Prescribed treatment course not to exceed 7 days **<u>Chronic hyperammonemia due to N-Acetylglutamate Synthase (NAGS) deficiency</u>** • Ammonia level greater than or equal to 50 micromol/L • NAGS deficiency confirmed by enzymatic, biochemical, or genetic testing • Prescribed in combination with a protein-restricted diet **<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:
	  ○ Carbamyl phosphate synthetase I (CPSI) deficiency
	  ○ Ornithine transcarbamylase (OTC) deficiency
	  ○ Argininosuccinate synthetase (ASS) deficiency
	  ○ Argininosuccinate lyase (ASL) deficiency
	  ○ Arginase deficiency

Age Restriction:	

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

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ To slow the loss of ambulation in pediatric patients with neuronal ceroid lipofuscinosis type 2 (CLN2), also known as tripeptidyl peptidase-1 (TPP1) deficiency

Required Medical Information:	• Diagnosis of CLN2 disease confirmed by BOTH of the following:
	  ○ Enzyme assay demonstrating deficient TPP1 activity
	  ○ Genetic testing that has detected two pathogenic variants/mutations in the TPP1/CLN2 gene (one on each parental allele of the TPP1/CLN2 gene)
	• Documentation of mild to moderate functional impairment at baseline using the CLN2 Clinical Rating Scale, defined as ALL the following:
	  ○ Combined score of 3 to 6 in the motor and language domains
	  ○ Score of at least 1 in the motor domain
	  ○ Score of at least 1 in the language domain

Appropriate Treatment Regimen & Other Criteria:	• Dosing is in accordance with FDA labeling **<u>Reauthorization</u>:** • Documentation of clinical responsiveness to therapy defined as disease stabilization OR a score of at least 1 in the motor domain of the CLN2 Clinical Rating Scale
Exclusion Criteria:	• Any sign or symptom of acute or unresolved localized infection on or around the device insertion site (e.g., cellulitis or abscess); or suspected or confirmed CNS infection (e.g., cloudy CSF or positive CSF gram stain, or meningitis) • Any acute intraventricular access device-related complication (e.g., leakage, extravasation of fluid, or device failure) • Other forms of neuronal ceroid lipofuscinosis • Patients with ventriculoperitoneal shunts
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist with expertise in the diagnosis of CLN2

	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 6 months, unless otherwise specified

POLICY NAME:
CFTR MODULATORS
Affected Medications: ORKAMBI (lumacaftor/ivacaftor), KALYDECO (ivacaftor), TRIKAFTA (elexacaftor, tezacaftor and ivacaftor; ivacaftor), SYMDEKO (tezacaftor/ivacaftor tablets)

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

POLICY NAME:
CALCITONIN GENE-RELATED PEPTIDE (CGRP) INHIBITORS
Affected Medications: AJOVY (fremanezumab), EMGALITY (galcanezumab), NURTEC ODT (rimegepant), QULIPTA (atogepant), VYEPTI (eptinezumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Preventative treatment of migraine in adults
	  ○ Episodic cluster headaches (Emgality only)
	  ○ Acute treatment of migraine in adult (Nurtec ODT only)

Required Medical Information:	**<u>Chronic migraine prevention:</u>**
	• 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
	
	<u>Episodic migraine prevention:</u>
	• Diagnosis of episodic migraine with at least 4 migraines per month at baseline
	
	<u>Episodic cluster headaches (Emgality Only):</u>
	• History of episodic cluster headache with at least two cluster periods lasting from 7 days to 1 year (when untreated) separated by pain-free remission periods of at least one month
	
	Headaches are not due to medication overuse: headaches occurring 15 or more days each month in a patient with pre-existing headache-causing condition possibly due to:
	  ○ Use of ergotamines, triptans, opioids, or combination analgesics at least 10 days per month for at least three months
	  ○ Use of simple analgesics (acetaminophen, aspirin, or an NSAID) at least 15 days per month for at least 3 months
	  ○ Use of combination of any previously mentioned products without overuse of any one agent if no causative pattern can be established

Appropriate Treatment	**<u>Chronic or Episodic migraine:</u>** • Documented treatment failure with an adequate trial (at least 8 weeks) of ONE oral migraine preventive therapy as follows:

Regimen & Other Criteria:	○ Candesartan 16 mg daily ○ Propranolol 40 mg daily, metoprolol 100 mg daily, timolol 20 mg daily, nadolol 80 mg daily ○ Amitriptyline 25 mg daily, nortriptyline 25 mg daily, venlafaxine 75 mg daily, duloxetine 60 mg daily ○ Topiramate 50 mg daily, valproic acid 500 mg daily, divalproex sodium 500 mg daily • Requests for Vyepti: Documented treatment failure to an adequate 8-week trial of an oral preventive therapy **AND** a minimum 12-week trial with each of the following: ○ One preferred drug: Nurtec (in migraine prevention), Ajovy, Emgality, Qulipta ○ Botox (chronic migraine only) **Episodic cluster headaches (Emgality Only):** • Documented treatment failure with an adequate trial of verapamil (dose of at least 480 mg daily for a minimum of 3 weeks), or if unable to tolerate verapamil or contraindications apply, another oral preventative therapy (lithium, topiramate) **Acute treatment of migraine (Nurtec ODT only):** • Documented treatment failure with one of the following: eletriptan, naratriptan, sumatriptan, rizatriptan, rizatriptan ODT, zolmitriptan, zolmitriptan ODT **Reauthorization:** • (Preventative treatment): documentation of treatment success defined as a 50% reduction in monthly headache frequency since starting therapy • (Acute treatment): documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Combined use with Botox • Combined use with another anti-calcitonin gene-related peptide (CGRP) monoclonal antibody or CGRP receptor antagonist (acute or preventive)
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 Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

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 **Preferred Drugs –** deferasirox soluble tablet, deferasirox tablet **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

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 **Preferred Drugs –** deferasirox soluble tablet, deferasirox tablet **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 **Preferred Drugs –**deferasirox soluble tablet, deferasirox tablet		
1. Documentation of liver iron (Fe) concentration (LIC) levels consistently greater than or equal to 5 mg Fe per gram of dry weight	Yes – Document and go to #2	No – Criteria not met

2. Documentation of serum ferritin levels consistently greater than 300 mcg/L prior to initiation of treatment	Yes – Document and go to #3	No – Criteria not met
3. Is the requested dose within the Food and Drug Administration (FDA) approved label?	Yes – Approve up to 12 months	No – Criteria not met
Renewal Criteria		
1. Is there documentation of treatment success and a clinically significant response to therapy defined as a reduction from baseline liver iron concentration (LIC) or serum ferritin level (LIC and serum ferritin must still be above 3 mg Fe per gram of dry weight and 500 mcg/L, respectively)	Yes – Go to #2	No – Criteria not met
2. Is the requested dose within the Food and Drug Administration (FDA)-approved label and PacificSource quantity limitations?	Yes – Approve up to 12 months	No – Criteria not met
Quantity Limitations		

• **Exjade (deferasirox soluble tablet) – available in 125mg, 250mg, 500mg tablets**
  ○ **20-40 mg/kg/day**
• **Jadenu (deferasirox tablet or granules) – available in 90mg, 180mg, 360mg tablets**
  ○ **14-28 mg/kg/day**
• **Ferriprox (deferiprone) – 100mg/ml oral solution, 500mg, 1000mg tablets**
  ○ **75-99 mg/kg/day**
  ○ **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)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of bile acid synthesis disorders due to single enzyme defects (SEDs)
	  ○ Adjunctive treatment of peroxisomal disorders, including Zellweger spectrum disorders, in patients who exhibit manifestations of liver disease, steatorrhea, or complications from decreased fat-soluble vitamin absorption

Required Medical Information:	• Documentation of all prior therapies, patient weight and anticipated treatment course • Baseline liver function tests (AST, ALT, GGT, ALP, total bilirubin, INR) **<u>Bile acid synthesis disorder</u>** • Diagnosis confirmed by assessment of serum or urinary bile acid levels using mass spectrometry (Fast Atom Bombardment ionization - Mass Spectrometry (FAB-MS) analysis) **<u>Peroxisomal disorders including Zellweger spectrum disorders</u>** • Diagnosis confirmed by clinical features, elevated very long-chain fatty acid (VLCFA) levels, peroxisomal biomarkers, genetic testing • Prothrombin time (vitamin K), serum levels of vitamins A, D, and E • Hepatic injury or at risk of liver injury (elevations in liver enzymes or atypical bile acids) OR • If normal liver function tests, must show manifestations of liver disease, steatorrhea, or complications from decreased fat-soluble vitamin absorption

Appropriate Treatment Regimen & Other Criteria:	• Will not be used for treatment of extrahepatic manifestations (such as neurologic symptoms) of bile acid synthesis disorders **<u>Reauthorization</u>** requires documentation of clinically significant improvement in liver function as determined by meeting TWO of the following criteria: • Improvement in abnormal liver chemistries (AST, ALT, bilirubin) • Reduction or stabilization of hepatic inflammation and fibrosis • Reduced levels of the toxic C27-bile acid intermediates dihydroxycholestanoic acid (DHCA) and trihydroxycholestanoic acid (THCA) in plasma and urine • Improvement in prothrombin time (as a result of improved vitamin K absorption) and serum levels of vitamins A, D, and E • No evidence of cholestasis on liver biopsy • Body weight increased or stabilized • Treatment should be discontinued if liver function does not improve after 3 months of start of treatment
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hepatologist, gastroenterologist, or metabolic specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Pruritus due to progressive familial intrahepatic cholestasis (PFIC)
	  ○ Cholestatic pruritus in patients with Alagille syndrome (ALGS)

Required Medical Information:	• Documentation of experiencing moderate to severe pruritis associated with PFIC or ALGS
	• Documentation of serum bile acid concentration above the upper limit of normal (ULN) reference range for the reporting laboratory
	
	<u>PFIC</u>
	• Documentation of confirmed molecular diagnosis of PFIC type 1 or type 2
	  ○ Documentation of absence of ABCB11 gene variant if PFIC type 2
	
	<u>ALGS</u>
	• Documentation of ALGS confirmed by:
	  ○ Genetic test detecting a JAG1 or NOTCH2 mutation **OR**
	  ○ Liver biopsy and at least three clinical features:
	  ▪ Chronic cholestasis
	  ▪ Cardiac disease
	  ▪ Ocular or skeletal abnormalities
	  ▪ Characteristic facial features
	  ▪ Renal and vascular disease

Appropriate Treatment Regimen & Other Criteria:	• Documentation of current weight and dosing in accordance with FDA labeling • Documented treatment failure with **<u>ALL</u>** of the following for at least 30 days: ○ Rifampin

	○ Ursodiol ○ Cholestyramine (or colesevelam if requesting for ALGS) **<u>Reauthorization</u>** requires documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Prior hepatic decompensation events • Decompensated cirrhosis (such as ALT or total bilirubin greater than 10-times the ULN) • Concomitant liver disease (e.g., biliary atresia, liver cancer, non-PFIC related cholestasis) • Prior liver transplant
Age Restriction:	• Age is in accordance with FDA labeling
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hepatologist or a specialist with experience in the treatment of PFIC or ALGS • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of symptomatic benign prostatic hyperplasia (BPH)
	  ○ Mental health diagnosis of erectile disorder (ED) meeting sexual dysfunction criteria

Required Medical Information:	• Diagnosis of benign prostatic hyperplasia (BPH)
	
	• Mental health diagnosis for the sexual dysfunction of erectile dysfunction, meeting the following Diagnostic and Statistical Manual of Mental Disorders, fifth edition (DSM-5) diagnostic criteria:
	  ○ At least one of the three following symptoms must be experienced with 75% to 100% of occasions of sexual activity:
	  ▪ Marked difficulty in obtaining an erection during sexual activity
	  ▪ Marked difficulty in maintaining an erection until the completion of sexual activity
	  ▪ Marked decrease in erectile rigidity
	  ○ The above symptoms have persisted for a minimum duration of approximately 6 months AND
	  ○ The above symptoms cause clinically significant distress in the individual AND
	  ○ The sexual dysfunction is not:
	  ▪ Better explained by a nonsexual mental disorder OR
	  ▪ A consequence of severe relationship distress or other significant stressors AND
	  ▪ It is not attributable to the effects of substance or medication use or another medical condition (such as a physical condition)

Appropriate Treatment Regimen & Other Criteria:	**Benign Prostate Hyperplasia (BPH)** • Treatment failure of at least two of the following: alfuzosin ER, doxazosin, finasteride, prazosin, tamsulosin

	<u>Reauthorization</u> requires documentation of treatment success and a clinically significant response to therapy • Limited to 1 tablet per day
Exclusion Criteria:	• Erectile dysfunction unrelated to a mental health diagnosis of sexual dysfunction according to the DSM-5 diagnostic criteria
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Mental health diagnosis of sexual dysfunction: prescribed by, or in consultation with, a mental health provider • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
CLADRIBINE
Affected Medications: MAVENCLAD (cladribine)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive disease (SPMS)

Required Medical Information:	• Diagnosis confirmed with magnetic resonance imaging (MRI) per revised McDonald diagnostic criteria for MS ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS
Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure with (or intolerance to) a minimum 12-week trial of at least two disease-modifying therapies for MS **<u>Reauthorization (one time only):</u>** provider attestation of treatment success • Eligible to initiate second treatment cycle 43 weeks after last dose was administered
Exclusion Criteria:	• Current malignancy • Human immunodeficiency virus (HIV) infection • Active chronic infections (e.g., hepatitis, tuberculosis) • Pregnancy • Treatment beyond 2 years
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or MS specialist • All approved are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 2 months, unless otherwise specified • Reauthorization: 2 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Perioperative management of bleeding in patients with mild, moderate, and severe hereditary Factor X deficiency
	  ○ Routine prophylaxis to reduce the frequency of bleeding episodes
	  ○ On-demand treatment and control of bleeding episodes

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
	• Patient weight
	• Documentation with one of the following diagnostic categories:
	  ○ On-demand treatment and control of bleeding episodes
	  ○ Perioperative management of bleeding in patients with mild, moderate, and severe hereditary Factor X deficiency
	  ○ Routine prophylaxis to reduce the frequency of bleeding episodes
	
	<u>Reauthorization (routine prophylaxis only):</u> 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:	
Age Restriction:	

Prescriber/Site of Care Restrictions:	• Hematologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial approval: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified • Perioperative management: 1 month, unless otherwise specified

POLICY NAME:
COMPOUNDED MEDICATION
Affected Medications: ALL COMPOUNDED MEDICATIONS

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 • In the case of payable claim, only compound ingredients that are covered on the applicable formulary will be reimbursed under this policy ○ 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 • Compounds containing non-Food and Drug Administration (FDA) approved ingredients will not be covered • 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
Preferred Products: Freestyle Libre, Freestyle Libre 2, Freestyle Libre 2 Plus, Freestyle Libre 3, Freestyle Libre 3 Plus, Dexcom G6, Dexcom G7
Non-Preferred Products: Medtronic Products (Enlite, Guardian, Minimed Guardian, Sof-sensor), Eversense Products

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
Required Medical Information:	• Documentation of diabetes mellitus diagnosis • Currently on insulin treatment of at least 3 subcutaneous (SubQ) injections daily OR on an insulin pump • Performing at least 4 blood glucose tests per day with a home blood glucose monitoring device • Requiring frequent insulin dose adjustments based on home blood glucose monitoring readings
Appropriate Treatment Regimen & Other Criteria:	**Coverage for non-preferred continuous glucose monitoring devices and supplies (receiver, transmitter, sensor) must meet the following criteria:** • Current use of insulin pump that is only compatible with a non-preferred continuous glucose monitor
Exclusion Criteria:	• Type 2 diabetes not on intensive insulin therapy • Use of continuous glucose monitor while on dialysis
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Must utilize pharmacy benefits only for coverage of all continuous glucose monitoring systems • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 2 years, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Heart failure with reduced ejection fraction (adjunctive agent)
	  ○ Heart failure due to dilated cardiomyopathy (DCM) in pediatric patients 6 months and older
	• Inappropriate sinus tachycardia

Required Medical Information:	<u>Chronic heart failure</u> • Documentation of chronic heart failure with left ventricular ejection fraction (LVEF) 35% or less AND • Resting heart rate of at least 70 beats per minute (bpm) <u>Heart failure in pediatric patients</u> • Documentation of stable symptomatic disease due to DCM • Currently in sinus rhythm with an elevated heart rate <u>Inappropriate sinus tachycardia</u> • 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) • Symptomatic (palpitations, shortness of breath, dizziness, and/or decreased exercise capacity) • 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 **<u>Chronic heart failure</u>** • Documented treatment failure with a beta blocker (metoprolol succinate extended release, carvedilol, or carvedilol extended release) at the maximally tolerated dose for heart failure treatment OR • Documentation of contraindication to beta-blocker use

	<u>Heart failure in pediatric patients</u> • Treatment failure with beta blocker or digoxin, or contraindication to beta blocker and digoxin use. **<u>Reauthorization</u>** 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 • Blood pressure less than 90/50 mm Hg • Sick sinus syndrome, sinoatrial block, third-degree atrioventricular block (unless stable with functioning demand pacemaker) • Severe hepatic impairment (Child-Paugh class C) • Heart rate maintained exclusively by pacemaker
Age Restriction:	• Heart failure due to DCM: 6 months to less than 18 years of age
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a cardiologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Primary prevention of cardiovascular disease

Required Medical Information:	**<u>Primary prevention of cardiovascular disease (must meet all of the following):</u>**
	• 40 to 75 years of age
	• Presence of at least one cardiovascular risk factor such as:
	  ○ Dyslipidemia
	  ○ Diabetes
	  ○ Hypertension
	  ○ Smoking
	• Estimated 10-year risk of cardiovascular event of at least 10% or higher

Appropriate Treatment Regimen & Other Criteria:	
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ To reduce the frequency of vaso-occlusive crises (VOCs) in adults and pediatric patients aged 16 years and older with sickle cell disease
Required Medical Information:	• Two or more sickle cell-related crises in the past 12 months • Therapeutic failure of 6-month trial on maximum tolerated dose of hydroxyurea or intolerable adverse event to hydroxyurea
Appropriate Treatment Regimen & Other Criteria:	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced **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 • Hemoglobin is less than 4.0 g/dL • Chronic anticoagulation therapy (such as warfarin, heparin) other than aspirin • History of stroke within the past 2 years • 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, a hematologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
CROVALIMAB
Affected Medications: PIASKY (crovalimab)

Covered Uses:	• All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design ○ Paroxysmal nocturnal hemoglobinuria (PNH)

Required Medical Information:	• Detection of PNH clones of at least 5% by flow cytometry diagnostic testing
	  ○ Presence of at least 2 different glycosylphosphatidylinositol (GPI) protein deficiencies (e.g., CD55, CD59, etc.) within at least 2 different cell lines (e.g., granulocytes, monocytes, erythrocytes)
	• Baseline lactate dehydrogenase (LDH) levels greater than or equal to 2 times the upper limit of normal range
	• One of the following PNH-associated clinical findings:
	  ○ Presence of a thrombotic event
	  ○ Presence of organ damage secondary to chronic hemolysis
	  ○ History of 4 or more blood transfusions required in the previous 12 months
	• Body weight

Appropriate Treatment Regimen & Other Criteria:	• Documented inadequate response, contraindication, or intolerance to ravulizumab-cwvz (Ultomiris) • Dosing is in accordance with FDA labeling and most recent body weight **<u>Reauthorization</u>** requires documentation of treatment success defined as a decrease in serum LDH, stabilized/improved hemoglobin, decreased transfusion requirement, and reduction in thromboembolic events compared to baseline
Exclusion Criteria:	• Concurrent use with other biologics for PNH (Soliris, Ultomiris, Empaveli, Fabhalta) • Current meningitis infection or other unresolved serious infection caused by encapsulated bacteria
Age Restriction:	• 13 years of age and older

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • 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)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. ○ Ocular Cystinosis
Required Medical Information:	• Diagnosis of ocular cystinosis • 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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Nephropathic cystinosis

Required Medical Information:	• Diagnosis of nephropathic cystinosis confirmed by **ONE** of the following:
	  ○ Molecular genetic testing showing mutations in the CTNS gene
	  ○ Leukocyte cystine concentration above the laboratory reference range
	  ○ Presence of cysteine corneal crystals by slit lamp examination

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure or intolerable adverse event with Cystagon
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
DANICOPAN
Affected Medications: VOYDEYA (danicopan)

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

POLICY NAME:
DASATINIB
Affected Medications: SPRYCEL (dasatinib)

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 • Documentation of Philadelphia chromosome or BCR::ABL1-positive mutation status
Appropriate Treatment Regimen & Other Criteria:	• For patients with Chronic Myeloid Leukemia (CML) and low risk score, documented clinical failure with imatinib **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:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

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

POLICY NAME:
DEFLAZACORT
Affected Medications: EMFLAZA (deflazacort), DEFLAZACORT

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ 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
	• Baseline motor function assessment from one of the following:
	  ○ 6-minute walk test
	  ○ North Star Ambulatory Assessment (NSAA)
	  ○ Motor Function Measure (MFM)
	  ○ Hammersmith Functional Motor Scale (HFMS)

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure with a 6-month trial of prednisone, or intolerable adverse event causing one of the following:
	  ○ Clinically significant weight gain defined as greater than or equal to 10% of body weight gain over a 6-month period
	  ○ Psychiatric/behavioral issues (e.g., abnormal behavior, aggression, irritability) that persists beyond the first six weeks of prednisone treatment
	
	Reauthorization requires a documented improvement from baseline or stabilization of motor function demonstrated by a motor function assessment tool

Exclusion Criteria:	
Age Restriction:	• 2 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• Some Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of ambulatory pediatric patients ages 4 and up with Duchenne muscular dystrophy (DMD)
Required Medical Information:	• Confirmed mutation of DMD gene between exons 18-58 • Documentation of being ambulatory without needing an assistive device such as a wheelchair, walker, or cane • North Star Ambulatory Assessment (NSAA) scale total score of 17 or more • Receiving physical and/or occupational therapy • Baseline anti-AAVrh74 total binding antibody titer of less than 1:400 as measured by ELISA • Current weight
Appropriate Treatment Regimen & Other Criteria:	• Documentation of being on a stable dose of an oral corticosteroid such as prednisone for at least 12-weeks, and will continue prior to and following Elevidys infusion, according to FDA approved labeling • Does not exceed FDA approved dosing based on weight and maximum of 70 vials Number of vials needed = patient body weight (kg) rounded to nearest number of vials
Exclusion Criteria:	• Exon 8 and/or exon 9 deletion in DMD gene • Concomitant therapy or within the past 6 months with DMD-directed antisense oligonucleotides such as golodirsen, casimersen, viltolarsen, eteplirsen • Current active infection • Previous Elevidys treatment in their lifetime • Acute liver disease or impaired liver function

	• Treatment in non-ambulatory patients – at this time, this indication is not considered medically necessary due to insufficient available evidence of therapeutic value
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 1 month (one-time dose, no reauthorization), unless otherwise specified

POLICY NAME:
DIFELIKEFALIN
Affected Medications: KORSUVA (difelikefalin)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ 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 • Documentation of moderate to severe pruritus associated with HD • Documentation of normal serum parathyroid hormone (PTH), phosphate, calcium, and magnesium levels • Documentation of patient's current dry body weight

Appropriate Treatment Regimen & Other Criteria:	• Documentation of inadequate relief with trial of all of the following therapies (minimum 1 month trial each):
	  ○ A topical agent (such as an emollient or analgesic)
	  ○ An oral antihistamine (such as hydroxyzine or diphenhydramine)
	  ○ Gabapentin or pregabalin
	
	Reauthorization will require documentation of clinically significant improvement or stabilization in pruritus from baseline and continued hemodialysis use

Exclusion Criteria:	• Peritoneal dialysis • Severe hepatic impairment
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a nephrologist • All approvals are subject to utilization of the most cost-effective site of care

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

POLICY NAME:
DINUTUXIMAB
Affected Medications: UNITUXIN (dinutuximab)

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
	• Documentation of high-risk neuroblastoma diagnosis as defined per the International Neuroblastoma Response Criteria (INRC):
	  ○ 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] OR
	  ○ Evidence of metastases to bone marrow on an aspirate or trephine biopsy with concomitant elevation of urinary or serum catecholamines or their metabolites
	• Evidence of high-risk neuroblastoma, including:
	  ○ Stage 2/3/4/4S disease with amplified MYCN gene (any age)
	  ○ Stage 4 disease in patients greater than 18 months of age
	• Disease is evaluable in the bone and/or bone marrow, as documented by histology and/or appropriate imaging [e.g., metaiodobenzylguanidine (MIBG) scan or PET scan if MIBG is negative]
	• Documented history of previous treatment with at least a partial response to prior first-line multi-agent, multimodality therapy

Appropriate Treatment Regimen & Other Criteria:	• Maximum duration: 5 cycles • Must be used in combination with granulocyte-macrophage colony-stimulating factor [GM-CSF; sargramostim], interleukin-2 [IL-2; aldesleukin], and 13-cis-retinoic acid [RA; isotretinoin]) **Reauthorization** will require documentation of disease responsiveness to therapy

Exclusion Criteria:	• Hold therapy if Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater
Age Restriction:	• Under 18 years of age
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 5 months, unless otherwise specified

POLICY NAME:
DIROXIMEL FUMARATE
Affected Medications: VUMERITY (diroximel fumarate)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive disease (SPMS)

Required Medical Information:	• Diagnosis confirmed with magnetic resonance imaging (MRI), per revised McDonald diagnostic criteria for MS ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS

Appropriate Treatment Regimen & Other Criteria:	**<u>Relapsing forms of MS</u>**
	• Coverage of Vumerity (diroximel fumarate) requires documentation of one of the following:
	  ○ Documented disease progression or intolerable adverse event with one of the following: teriflunomide, dimethyl fumarate or fingolimod
	  ○ Currently receiving treatment with Vumerity (diroximel fumarate), excluding via samples or manufacturer's patient assistance program
	
	<u>Reauthorization</u> requires provider attestation of treatment success

Exclusion Criteria:	• Concurrent use of other disease-modifying medications indicated for the treatment of multiple sclerosis
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or a multiple sclerosis specialist

	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. ○ A source of calories and fatty acids for the treatment of pediatric and adult patients with molecularly confirmed long-chain fatty acid oxidation disorders

Required Medical Information:	• Diagnosis of long chain fatty acid oxidation disorder (LC-FAOD) confirmed by molecular genetic testing or enzyme assay
	• Documentation of total prescribed daily caloric intake
	• Documentation of severe disease as evidenced by one of the following:
	  ○ Hypoglycemia after short periods of fasting
	  ○ Evidence of functional cardiomyopathy with poor ejection fraction requiring ongoing management
	  ○ Frequent severe major medical episodes requiring emergency room visits, acute care, or hospitalization (3 events within the past year, or 5 events within the past 2 years)
	  ○ Elevated creatinine kinase (chronic or episodic)

Appropriate Treatment Regimen & Other Criteria:	• Documentation of persistent symptoms despite dietary management and use of an over the counter (OTC) medium-chain triglyceride (MCT) product • Dose not to exceed 35% of daily caloric intake **Reauthorization** will require documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Concurrent use of another medium chain triglyceride product
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist or provider experienced in the management of metabolic disorders • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design

Required Medical Information:	• Diagnosis of type 1 diabetes for 5 or more years
	• Documentation of inability to achieve target HbA1c despite adherence to intensive insulin management with all the following:
	  ○ Multiple daily injections of prandial and basal insulin or on an insulin pump
	  ○ Performing at least four blood glucose tests per day or using a continuous glucose monitor
	• Documentation of 2 or more episodes of severe hypoglycemia (blood glucose level less than 50 mg/dL) in the past three years requiring assistance of another person with either an oral carbohydrate, intravenous glucose, or glucagon administration
	• Documentation of hypoglycemia unawareness, defined by the absence of adequate autonomic symptoms during an episode of severe hypoglycemia

Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** requires documentation of not achieving exogenous insulin independence within one year of infusion or within one year of losing independence from exogenous insulin (maximum of three infusions per lifetime)
Exclusion Criteria:	• Pregnancy • Malignancy • Active infection • Previous kidney or pancreas transplant • Prior portal vein thrombosis
Age Restriction:	• 18 years of age and older

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 3 months (single treatment), unless specified otherwise

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

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 • 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 • Reauthorization will require documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	
Age Restriction:	• 1 month of age or older
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 24 months, unless otherwise specified

POLICY NAME:
DROXIDOPA
Affected Medications: DROXIDOPA

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of orthostatic dizziness with symptomatic neurogenic orthostatic hypotension (nOH) caused by:
	  ▪ Primary autonomic failure (Parkinson's disease [PD], multiple system atrophy [MSA], pure autonomic failure [PAF])
	  ▪ Dopamine beta-hydroxylase deficiency
	  ▪ Non-diabetic autonomic neuropathy

Required Medical Information:	• Diagnosis of nOH caused by one of the following:
	  ○ Primary autonomic failure (such as PD, MSA, PAF)
	  ○ Dopamine beta-hydroxylase deficiency
	  ○ Non-diabetic autonomic neuropathy
	• Documentation of severe symptomatic orthostatic hypotension, demonstrated by **both** of the following:
	  ○ Minimum 20 mmHg decrease in systolic blood pressure **OR** minimum 10 mmHg decrease in diastolic blood pressure within 3 minutes of standing
	  ○ Documentation of significant symptoms affecting activities of daily living

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure or intolerable adverse event with a minimum 30-day trial to both fludrocortisone and midodrine **Reauthorization** requires documentation of treatment success as determined by treating provider
Exclusion Criteria:	
Age Restriction:	• 18 years of age or older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or cardiologist • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 1 month, unless otherwise specified • Reauthorization: 3 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of motor fluctuations in patients with advanced Parkinson's disease (PD)

Required Medical Information:	• Documentation of all the following:
	  ○ Diagnosis of advanced PD
	  ○ Clear response to levodopa treatment with evidence of "On" periods
	  ○ Persistent motor fluctuations with "Off" time occurring 3 hours or more per day while awake despite an optimized PD treatment regimen
	  ○ Has undergone or has planned placement of a nasojejunal (NJ) tube for temporary administration of Duopa OR gastrostomy-jejunostomy (PEG-J) tube for long-term administration of Duopa

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure with both of the following:
	  ○ Oral levodopa/carbidopa
	  ○ Two additional agents from different anti-PD drug classes:
	  ▪ Monoamine oxidase-B (MAO-B) inhibitors (ex: selegiline, rasagiline)
	  ▪ Dopamine agonists (ex: amantadine, pramipexole, ropinirole)
	  ▪ Catechol-O-methyltransferase (COMT) inhibitors (ex: entacapone)
	
	Reauthorization requires documentation of treatment success and a clinically significant response to therapy

Exclusion Criteria:	• Atypical Parkinson's syndrome ("Parkinson's Plus" syndrome) or secondary Parkinson's • Non-levodopa responsive PD • Contraindication to percutaneous endoscopic gastro-jejunal (PEG-J) tube placement or long-term use of a PEG-J • Concomitant use with nonselective MAO inhibitors or have recently (within 2 weeks) taken a nonselective MAO inhibitor
Age Restriction:	

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)–approved indications not otherwise excluded by plan design
	  ○ Moderate to severe eosinophilic phenotype or oral corticosteroid dependent asthma
	  ○ Moderate to severe atopic dermatitis (AD)
	  ○ Chronic rhinosinusitis with nasal polyposis (CRSwNP)
	  ○ Eosinophilic esophagitis (EoE)
	  ○ Prurigo nodularis (PN)

Required Medical Information:	**AD:**
	• 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)
	• Body surface area (BSA) involvement greater than or equal to 10% or hand, foot, or mucous membrane involvement
	
	Asthma:
	• Documentation of BOTH of the following:
	  ○ Baseline eosinophil count at least 150 cells/μL
	  ○ Forced expiratory volume (FEV1) less than 80% at baseline or FEV1/FVC reduced by at least 5% from normal
	
	CRSwNP:
	• Documentation of both of the following:
	  ○ Diagnosis of chronic rhinosinusitis and has undergone prior bilateral total ethmoidectomy
	  ○ Indicated for revision sinus endoscopic sinus surgery due to recurrent symptoms of nasal polyps (such as nasal obstruction/congestion, bilateral sinus obstruction)

	<u>EoE:</u>
	• Diagnosis confirmed by endoscopic biopsy with greater than or equal to 15 eosinophils per high power field (HPF)
	• Documentation of TWO or more dysphagia episodes per week despite current treatment
	
	<u>PN:</u>
	• Documentation of all of the following:
	  ○ Diagnosis confirmed by skin biopsy
	  ○ Presence of at least 20 PN lesions for at least 3 months
	  ○ Severe itching

Appropriate Treatment Regimen & Other Criteria:	• Requested dosing according to the FDA label based on diagnosis
	
	<u>AD:</u>
	• Documented treatment failure with at least 12 weeks of two of the following (1 in each category):
	  ○ Tacrolimus ointment or pimecrolimus cream or Eucrisa
	  ○ Phototherapy or cyclosporine or azathioprine or methotrexate or mycophenolate
	
	<u>Asthma:</u>
	• Use of high-dose inhaled corticosteroid (ICS) plus a long-acting beta agonist (LABA) for at least three months with continued symptoms
	• Documentation of one of the following:
	  ○ Documented history of 2 or more asthma exacerbations requiring oral or systemic corticosteroid treatment in the past 12 months while on combination inhaler treatment with at least 80% adherence
	  ○ Documentation that chronic daily oral corticosteroids are required
	
	<u>CRSwNP:</u>
	• Documented treatment failure with at least 1 intranasal corticosteroid (such as fluticasone) after ethmoidectomy
	• Documented treatment failure with Sinuva implant

	<u>EoE:</u>
	• Documented treatment failure with at least 12 weeks of **BOTH** of the following:
	  ○ High dose (twice daily dosing) proton pump inhibitor (e.g., omeprazole or esomeprazole)
	  ○ Swallowed corticosteroid therapy (such as fluticasone or budesonide)
	
	<u>PN:</u>
	• Documented treatment failure with at least 2 weeks of a super high potency topical corticosteroid (such as clobetasol propionate 0.05%, halobetasol propionate 0.05%)
	• Documentation of treatment failure with at least 12 weeks of one of the following: phototherapy, methotrexate, cyclosporine
	
	<u>Reauthorization</u> requires documentation of treatment success as determined by treating provider

Exclusion Criteria:	• Concurrent use with another therapeutic immunomodulator agent utilized for the same indication
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a dermatologist, pulmonologist, otolaryngologist, gastroenterologist, allergist, or immunologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
ECULIZUMAB
Affected Medications: SOLIRIS (eculizumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis
	  ○ Atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy
	  ○ Generalized myasthenia gravis (gMG) in adults who are anti-acetylcholine receptor (AChR) antibody positive
	  ○ Neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive

Required Medical Information:	**<u>PNH</u>**
	• Detection of PNH clones of at least 5% by flow cytometry diagnostic testing
	  ○ Presence of at least 2 different glycosylphosphatidylinositol (GPI) protein deficiencies (e.g., CD55, CD59, etc.) within at least 2 different cell lines (e.g., granulocytes, monocytes, erythrocytes)
	• Baseline lactate dehydrogenase (LDH) levels greater than or equal to 1.5 times the upper limit of normal range
	• One of the following PNH-associated clinical findings:
	  ○ Presence of a thrombotic event
	  ○ Presence of organ damage secondary to chronic hemolysis
	  ○ History of 4 or more blood transfusions required in the previous 12 months
	
	<u>aHUS</u>
	• Clinical presentation of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury
	• Patient shows signs of thrombotic microangiopathy (TMA) (e.g., changes in mental status, seizures, angina, dyspnea, thrombosis, increasing blood pressure, decreased platelet count, increased serum creatinine, increased LDH, etc.)
	• ADAMTS13 activity level greater than or equal to 10%

	• Shiga toxin E. coli related hemolytic uremic syndrome (ST-HUS) has been ruled out
	• History of 4 or more blood transfusions required in the previous 12 months
	
	<u>gMG</u>
	• Diagnosis of gMG confirmed by **ONE** of the following:
	  ○ A history of abnormal neuromuscular transmission test
	  ○ A positive edrophonium chloride test
	  ○ Improvement in gMG signs or symptoms with an acetylcholinesterase inhibitor
	• Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV
	• Positive serologic test for AChR antibodies
	• Documentation of **ONE** of the following:
	  ○ MG-Activities of Daily Living (MG-ADL) total score of 6 or greater
	  ○ Quantitative Myasthenia Gravis (QMG) total score of 12 or greater
	
	<u>NMOSD</u>
	• Diagnosis of seropositive aquaporin-4 immunoglobulin G (AQP4-IgG) NMOSD confirmed by all the following:
	  ○ Documentation of AQP4-IgG-specific antibodies on cell-based assay
	  ○ Exclusion of alternative diagnoses (such as multiple sclerosis)
	  ○ At least **ONE** core clinical characteristic:
	  ▪ Acute optic neuritis
	  ▪ Acute myelitis
	  ▪ Acute area postrema syndrome (episode of otherwise unexplained hiccups or nausea/vomiting)
	  ▪ Acute brainstem syndrome
	  ▪ Symptomatic narcolepsy **OR** acute diencephalic clinical syndrome with NMOSD-typical diencephalic

<table>
<tr><td></td><td>lesion on magnetic resonance imaging (MRI) [see table below]
▪ Acute cerebral syndrome with NMOSD-typical brain lesion on MRI [see table below]
<table><tr><th>Clinical presentation</th><th>Possible MRI findings</th></tr><tr><td>Diencephalic syndrome</td><td>• Periependymal lesion
• Hypothalamic/thalamic lesion</td></tr><tr><td>Acute cerebral syndrome</td><td>• Extensive periependymal lesion
• Long, diffuse, heterogenous, or edematous corpus callosum lesion
• Long corticospinal tract lesion
• Large, confluent subcortical or deep white matter lesion</td></tr></table></td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td><u>PNH</u>
• Documented inadequate response, contraindication, or intolerance to ravulizumab-cwvz (Ultomiris)

<u>aHUS</u>
• Failure to respond to plasma therapy within 10 days
○ Trial of plasma therapy not required if one of the following is present:
▪ Life-threatening complications of HUS such as seizures, coma, or heart failure
▪ Confirmed presence of a high-risk complement genetic variant (e.g., CFH or CFI)
• Documented inadequate response, contraindication, or intolerance to ravulizumab-cwvz (Ultomiris)

<u>gMG</u>
• Documentation of one of the following:
○ Treatment failure with an adequate trial (one year or more) of at least 2 immunosuppressive therapies (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)
○ Has required three or more courses of rescue therapy (plasmapheresis/plasma exchange and/or intravenous</td></tr>
</table>

	immunoglobulin), while on at least one immunosuppressive therapy, over the last 12 months • Documented inadequate response, contraindication, or intolerance to each of the following: ○ Efgartigimod-alfa (Vyvgart) ○ Ravulizumab-cwvz (Ultomiris) **NMOSD** • Documented inadequate response, contraindication, or intolerance to **ALL** of the following: ○ Rituximab (preferred products: Riabni, Ruxience) ○ Satralizumab-mwge (Enspryng) ○ Inebilizumab-cdon (Uplizna) ○ Ravulizumab-cwvz (Ultomiris) **Reauthorization:** • gMG: documentation of treatment success defined as an improvement in MG-ADL and QMG scores from baseline • NMOSD: documentation of treatment success defined as the stabilization or improvement in neurological symptoms as evidenced by a decrease in acute relapses, Expanded Disability Status Scale (EDSS) score, hospitalizations, or plasma exchange treatments • PNH: documentation of treatment success defined as a decrease in serum LDH, stabilized/improved hemoglobin, decreased transfusion requirement, and reduction in thromboembolic events compared to baseline • aHUS: documentation of treatment success defined as a decrease in serum LDH, stabilized/improved serum creatinine, increased platelet count, and decreased plasma exchange/infusion requirement compared to baseline
Exclusion Criteria:	• Concurrent use with other disease-modifying biologics for requested indication, unless otherwise indicated by the FDA for combination use with Soliris • Current meningitis infection
Age Restriction:	• PNH, gMG and NMOSD: 18 years of age and older • aHUS: 2 months of age and older

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist
	  ○ PNH: hematologist
	  ○ aHUS: hematologist or nephrologist
	  ○ gMG: neurologist
	  ○ NMOSD: neurologist or neuro-ophthalmologist
	• All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

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

POLICY NAME:
EFLORNITHINE
Affected Medications: IWILFIN (eflornithine)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Maintenance therapy in patients with high-risk neuroblastoma who achieve at least a partial response to prior systemic agents and have completed maintenance immunotherapy with an anti-GD2 antibody • 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
	• Diagnosis of neuroblastoma as defined per the International Neuroblastoma Response Criteria (INRC):
	  ○ 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] OR
	  ○ Evidence of metastases to bone marrow on an aspirate or trephine biopsy with concomitant elevation of urinary or serum catecholamines or their metabolites
	• Evidence of high-risk neuroblastoma, including:
	  ○ Stage 2/3/4/4S disease with amplified MYCN gene (any age)
	  ○ Stage 3 disease with MYCN gene NOT amplified in patients at least 18 months of age with International Neuroblastoma Pathology Classification (INPC) as unfavorable histology (UH)
	  ○ Stage 4 disease in patients greater than 12 months of age
	• Staging studies documented by histology and/or appropriate imaging as follows:
	  ○ Computed tomography (CT) or magnetic resonance imaging (MRI) scan of the primary site and nodal sites of metastatic disease

	○ Bone imaging (preferably with a metaiodobenzylguanidine [MIBG] scan and positron emission topography (PET) scan (if MIBG is negative)
Appropriate Treatment Regimen & Other Criteria:	• Documentation of a partial response to prior systemic agents and completed maintenance immunotherapy with an anti-GD2 antibody (Dinutuximab, Naxitamab) **<u>Reauthorization</u>:** documentation of disease responsiveness to therapy up to a total of 2 years of treatment
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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial authorization: 4 months, unless otherwise specified • Reauthorization: One time reauthorization of 20 months to complete 2 years of treatment, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Moderate to severe endometriosis-associated pain (Orilissa)
	  ○ Heavy menstrual bleeding associated with uterine leiomyomas (Oriahnn)

Required Medical Information:	**<u>Pain due to endometriosis</u>**
	• Documentation of both of the following:
	  ○ Diagnosis of moderate to severe pain associated with endometriosis
	  ○ Attestation that patient is premenopausal
	
	<u>Heavy menstrual bleeding due to uterine leiomyomas</u>
	• Documentation of both of the following:
	  ○ Diagnosis of heavy menstrual bleeding associated with uterine leiomyomas
	  ○ Attestation that patient is premenopausal

Appropriate Treatment Regimen & Other Criteria:	**<u>Pain due to endometriosis</u>**
	• Documentation of a trial and inadequate relief (or contraindication) after at least 3 months of both of the following first-line therapies:
	  ○ Nonsteroidal anti-inflammatory drugs (NSAIDs)
	  ○ Continuous (no placebo pills) hormonal contraceptives
	
	<u>Reauthorization</u> requires documentation of treatment success and a clinically significant response to therapy

Exclusion Criteria:	• History of osteoporosis • Pregnancy • Severe (Child-Pugh Class C) hepatic impairment (Orilissa)

	• Mild, moderate, and severe (Child-Pugh Class A, B, and C) hepatic impairment (Oriahnn)
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist in obstetrics/gynecology or reproductive endocrinology • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 18 months (Orilissa 150 mg once daily* and Oriahnn only), unless otherwise specified • *Maximum treatment duration for Orilissa 150 mg once daily in patients with moderate hepatic impairment (Child-Pugh Class B) and Orilissa 200 mg twice daily is 6 months. Reauthorization not allowed

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Early, active cerebral adrenoleukodystrophy (CALD) in male patients

Required Medical Information:	• Confirmed diagnosis of CALD with all of the following:
	  ○ Confirmed *ABCD1* gene mutation
	  ○ Elevated very-long-chain fatty acid (VLCFA) values for ALL of the following:
	  ▪ Concentration of C26:0
	  ▪ Ratio of C24:0 to C22:0
	  ▪ Ratio of C26:0 to C22:0
	  ○ Neurologic function score (NFS) less than or equal to 1 (asymptomatic or mildly symptomatic disease)
	  ○ Active central nervous system disease established by central radiographic review of brain magnetic resonance imaging (MRI) demonstrating both of the following:
	  ▪ Gadolinium enhancement on MRI of demyelinating lesions
	  ▪ 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 **Approved for one-time single infusion only**
Exclusion Criteria:	• Female gender • Previously received an allogeneic transplant or gene therapy
Age Restriction:	• 4 to 17 years of age

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist, endocrinologist, or hematologist/oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified (one infusion only)

POLICY NAME:
ELTROMBOPAG DERIVATIVES
Affected Medications: PROMACTA (eltrombopag olamine), PROMACTA PACKET, ALVAIZ (eltrombopag choline)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of thrombocytopenia in patients with persistent or chronic immune thrombocytopenia (ITP)
	  ○ Treatment of thrombocytopenia in patients with hepatitis C infection
	  ○ Treatment of severe aplastic anemia

Required Medical Information:	**<u>Thrombocytopenia in patients with chronic ITP</u>**
	• Documentation of **ONE** of the following:
	  ○ Platelet count less than 20,000/microliter
	  ○ Platelet count less than 30,000/microliter AND symptomatic bleeding
	  ○ Platelet count less than 50,000/microliter AND increased risk for bleeding (such as peptic ulcer disease, use of antiplatelets or anticoagulants, history of bleeding at higher platelet count, need for surgery or invasive procedure)
	
	<u>Thrombocytopenia in patients with chronic hepatitis C</u>
	• Documentation of plan to initiate interferon-based therapy
	• Documentation of platelet count less than 75,000/microliter
	
	<u>Severe aplastic anemia</u>
	• Diagnosis confirmed by bone marrow biopsy
	• Documentation of at least two of the following:
	  ○ Absolute reticulocyte count (ARC) less than 60,000/microliter
	  ○ Platelet count less than 20,000/microliter
	  ○ Absolute neutrophil count (ANC) less than 500/microliter

Appropriate Treatment	• Promacta packet formulation requires documented medical inability to use oral tablet formulation

Regimen & Other Criteria:	**<u>Thrombocytopenia in patients with persistent or chronic ITP</u>**
	• Documentation of inadequate response, defined as platelets did not increase to at least 50,000/microliter, to the following therapies:
	  ○ **ONE** of the following:
	  ▪ Inadequate response with at least 2 therapies for immune thrombocytopenia, including corticosteroids, rituximab, or immunoglobulin
	  ▪ Splenectomy
	
	<u>Reauthorization</u>:
	• Response to treatment with platelet count of at least 50,000/microliter (not to exceed 400,000/microliter)
	OR
	• The platelet counts have not increased to a platelet count of at least 50,000/microliter and the patient has NOT been on the maximum dose for at least 4 weeks
	
	<u>Thrombocytopenia in patients with chronic hepatitis C</u>
	
	<u>Reauthorization</u>:
	• Response to treatment with platelet count of at least 90,000/microliter (not to exceed 400,000/microliter) and eltrombopag is used in combination with antiviral therapy
	
	<u>Severe aplastic anemia</u>
	• Documentation of refractory severe aplastic anemia as indicated by insufficient response to at least one prior immunosuppressive therapy
	OR
	• For those less than 40 years of age without a rapidly available matched related donor (MRD) or 40 years of age and older: documentation that eltrombopag is being used as first line treatment in combination with standard immunosuppressive therapy (Atgam and cyclosporine)
	
	<u>Reauthorization</u> (refractory severe aplastic anemia only):
	Requires hematologic response to treatment defined as meeting **ONE** or more of the following criteria:

	• Platelet count increases to 20,000/microliter above baseline, or stable platelet counts with transfusion independence for a minimum of 8 weeks • Hemoglobin increases by greater than 1.5 g/dL or a reduction in greater than or equal to 4 units red blood cell (RBC) transfusions for 8 consecutive weeks • ANC increase of 100% or an ANC increase greater than 500/microliter
Exclusion Criteria:	• Use in combination with another thrombopoietin receptor agonist, spleen tyrosine kinase inhibitor, or similar treatments (Doptelet, Nplate, Tavalisse)
Age Restriction:	**<u>Thrombocytopenia in patients with ITP</u>** • 1 year of age and older (Promacta) • 6 years of age and older (Alvaiz) **<u>Thrombocytopenia in patients with chronic hepatitis C and patients with severe aplastic anemia</u>** • 18 years of age and older (Promacta and Alvaiz) **<u>Severe Aplastic Anemia (initial therapy)</u>** • 2 years of age and older • 18 years of age and older (Alvaiz)
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist or gastroenterology/liver specialist • All approvals are subjects to utilization of the most cost-effective site of care
Coverage Duration:	**<u>Thrombocytopenia in patients with ITP</u>** • Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified **<u>Thrombocytopenia in patients with chronic hepatitis C</u>** • Initial Authorization: 2 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified **<u>Severe aplastic anemia</u>** • Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

	<u>Severe aplastic anemia in combination with cyclosporine and Atgam</u> • Authorization: 6 months, no reauthorization, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ 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:	• Diagnosis confirmed by presence of a genetic mutation known to cause primary HLH (e.g., PRF1, UNC13D, STX11, STXBP2) OR documentation showing at least 5 of the following are present:
	  ○ Prolonged fever (lasting over 7 days)
	  ○ Splenomegaly
	  ○ **Two** of the following cytopenias in the peripheral blood:
	  ▪ Hemoglobin less than 9 g/dL
	  ▪ Platelet count less than 100,000/mcL
	  ▪ Neutrophils less than 100/mcL
	  ○ **One** of the following:
	  ▪ Hypertriglyceridemia defined as fasting triglycerides 3 mmol/L or higher OR 265 mg/dL or higher
	  ▪ Hypofibrinogenemia defined as fibrinogen 1.5 g/L or lower
	  ○ Hemophagocytosis in bone marrow, spleen, or lymph nodes (with no evidence of malignancy)
	  ○ Low or absent natural killer cell activity (according to local laboratory reference)
	  ○ Ferritin 500 mg/L or higher
	  ○ Soluble CD25 (i.e., soluble IL-2 receptor) 2,400 U/ml or higher
	• Documentation confirming status as a hematopoietic stem cell transplant (HCST) candidate

Appropriate Treatment Regimen & Other Criteria:	• Documentation of refractory, recurrent, or progressive disease (or intolerable adverse event) on conventional HLH therapy (e.g., dexamethasone, etoposide, methotrexate, hydrocortisone) • Must be used in combination with dexamethasone (if established on the following, patient may instead continue: oral cyclosporine A; intrathecal methotrexate and/or glucocorticoids)

	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced **<u>Reauthorization</u>:** documentation of disease responsiveness to therapy AND patient has not received HSCT
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist, oncologist, transplant specialist, or provider with experience in the management of HLH • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 2 months, unless otherwise specified • Reauthorization: 4 months, unless otherwise specified

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

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 • 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 • Baseline factor level 1% to 3% AND a documented history of at least two episodes of spontaneous bleeding into joints • Prophylactic agents must be discontinued ▪ Factor VIII Inhibitors: after the first week of HEMLIBRA ▪ Bypassing Agents: one day before starting HEMLIBRA **Loading Dose:** • 3 mg/kg once every week for 4 weeks • Maximum 1,380 mg per 28 day supply **Maintenance dose:** • 1.5 mg/kg once every week or • 3 mg/kg once every 2 weeks or • 6 mg/kg once every 4 weeks • 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.) **Product Availability** • Single-dose vials for injection: 30 mg/mL, 60 mg/0.4 mL, 105 mg/0.7 mL, 150 mg/mL • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced Reauthorization requires documentation of treatment success defined as a reduction in spontaneous bleeds requiring treatment, as well as documentation of bleed history since last approval

Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Hematologist • 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)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
Required Medical Information:	• Diagnosis of major depressive disorder (MDD)

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure to an adequate trial (clinically sufficient doses for a minimum 6-week duration) to each of the following:
	  ○ A selective serotonin reuptake inhibitor (SSRI)
	  ○ A serotonin/norepinephrine reuptake inhibitor (SNRI)
	  ○ A tricyclic or tetracyclic antidepressant
	  ○ Bupropion
	OR
	• Documentation of inability to take any oral preparations (including commercially available liquid antidepressants)
	
	Reauthorization will require documentation of treatment success and a clinically significant response to therapy

Exclusion Criteria:	• Pheochromocytoma
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a psychiatrist • 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, AMBRISENTAN, OPSUMIT (macitentan), OPSYNVI (macitentan and tadalafil)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Pulmonary artery hypertension (PAH) World Health Organization (WHO) Group 1

Required Medical Information:	• Documentation of Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1 confirmed by right heart catheterization meeting the following criteria:
	  ○ Mean pulmonary artery pressure of at least 20 mm Hg
	  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg
	  ○ Pulmonary vascular resistance of at least 2.0 Wood units
	• New York Heart Association (NYHA)/WHO Functional Class II or higher symptoms
	• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blocker), unless there are contraindications:
	  ○ Low systemic blood pressure (systolic blood pressure less than 90)
	  ○ Low cardiac index
	  ○ 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 • Documentation of inadequate response or intolerance to oral calcium channel blocking agents if positive Acute Vasoreactivity Test • For Opsumit (macitentan) and Opsynvi (macitentan and tadalafil) requests: documentation of inadequate response or intolerance to ambrisentan AND bosentan for 12 weeks is required **<u>Reauthorization</u>** requires documentation of treatment success defined as one or more of the following: • Improvement in walking distance • Improvement in exercise ability

	• Improvement in pulmonary function • Improvement or stability in WHO functional class
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a cardiologist or pulmonologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
ENFUVIRTIDE
Affected Medications: FUZEON (enfuvirtide)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of human immunodeficiency virus type 1 (HIV-1) infection in combination with other antiretroviral agents in treatment-experienced patients with evidence of HIV-1 replication despite ongoing antiretroviral therapy

Required Medical Information:	• Documented weight greater than or equal to 11 kg
	• Documentation of current (within past 30 days) HIV-1 RNA viral load of at least 200 copies/mL
	• Documented treatment failure with minimum 12-weeks of antiretroviral therapy with at least one antiretroviral agent from three different classes (unless contraindicated or clinically significant adverse effects are experienced):
	  ○ Nucleoside reverse-transcriptase inhibitors (NRTIs)
	  ○ Non-nucleoside reverse-transcriptase inhibitors (NNRTIs)
	  ○ Integrase strand transfer inhibitors (INSTIs)
	  ○ Protease inhibitors (PIs)

Appropriate Treatment Regimen & Other Criteria:	• Prescribed in combination with an optimized background antiretroviral regimen
	
	Reauthorization requires documentation of all of the following:
	• Treatment plan including continued use of optimized background antiretroviral regimen
	• Documentation of treatment success as evidenced by one of the following:
	  ○ Reduction in viral load from baseline or maintenance of undetectable viral load
	  ○ Absence of postbaseline emergence of enfuvirtide resistance-associated mutations confirmed by resistance testing

Exclusion Criteria:	• Initial therapy in patients who are antiretroviral naïve
Age Restriction:	• 6 years of age and older

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an infectious disease or HIV specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 24 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Vpriv: Gaucher disease type 1 (GD1)
	  ○ Elelyso: GD1 for ages 4 years and older
	  ○ Cerdelga: GD1 in adults who are CYP2D6 extensive metabolizers (EMs), intermediate metabolizers (IMs), or poor metabolizers (PMs) as detected by an FDA-cleared test
	  ○ Cerezyme: GD1 for ages 2 years and older that results in one or more of the following conditions:
	  ▪ Anemia
	  ▪ Thrombocytopenia
	  ▪ Bone disease
	  ▪ Hepatomegaly or splenomegaly

Required Medical Information:	• Diagnosis confirmed by enzyme assay showing deficiency of beta-glucocerebrosidase glucosidase enzyme activity **OR** genetic testing indicating mutation of two alleles of the glucocerebrosidase genome
	  ○ For Cerdelga, must also have documentation of cytochrome P450 2D6 (CYP2D6) genotype by an FDA-approved test indicating CYP2D6 EM, IM, or PM status
	• Documentation of baseline tests such as hemoglobin level, platelet count, liver function tests, renal function tests
	• Documentation of at least one clinically significant disease complication of GD1:
	  ○ Anemia (low hemoglobin and hematocrit levels)
	  ○ Thrombocytopenia (platelet count less than 120,000 mm^3)
	  ○ Bone disease (T-score less than -2.5 or bone pain)
	  ○ Hepatomegaly or splenomegaly

	○ For symptomatic children: symptoms of early presentation, such as malnutrition, growth retardation, impaired psychomotor development, and/or fatigue
Appropriate Treatment Regimen & Other Criteria:	**<u>Cerdelga</u>** <u>Extensive or Intermediate Metabolizers of CYP2D6</u> • Quantity limit - 84 mg capsules #60 per 30 days <u>Poor Metabolizers of CYP2D6</u> • Quantity limit - 84 mg capsules #30 per 30 days **<u>Elelyso, Vpriv, and Cerezyme</u>** • Dosing is in accordance with FDA labeling and patient's most recent weight • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced **<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Concomitant use with another ERT for GD1 or with miglustat **<u>Cerdelga</u>:** • CYP2D6 ultrarapid metabolizers • Moderate or severe hepatic impairment • Pre-existing cardiac disease (congestive heart failure, myocardial infarction, bradycardia, heart block, arrhythmias, and long QT syndrome) • Presence of moderate to severe renal impairment or end stage renal disease
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist in the management of Gaucher disease (hematologist, oncologist, hepatologist, geneticist or orthopedic specialist) • All approvals are subjects to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of hereditary transthyretin amyloidosis with polyneuropathy (hATTR-PN) in adults

Required Medical Information:	• Documented diagnosis of hATTR confirmed by **BOTH** of the following:
	  ○ Amyloid deposition on biopsy
	  ○ Presence of pathogenic transthyretin (TTR) variant on genetic testing
	• Presence of clinical manifestations of the disease, confirmed by presence of peripheral neuropathy on nerve conduction studies **OR** 2 of the following:
	  ○ Autonomic dysfunction (bladder/urinary tract infections, gastrointestinal disturbances, erectile dysfunction, orthostatic hypotension)
	  ○ Documented symptoms of sensorimotor polyneuropathy (e.g., paresthesia, balance issues, weakness/numbness in the hands/feet, or loss of sensation for pain, temperature, proprioception)
	  ○ Cardiomyopathy, ocular involvement, or renal involvement
	• Documentation of **ONE** of the following:
	  ○ Baseline polyneuropathy disability (PND) score of less than or equal to IIIb
	  ○ Baseline neuropathy impairment score (NIS) between 10 and 130
	  ○ Baseline familial amyloid polyneuropathy (FAP) stage 1 or 2

Appropriate Treatment Regimen & Other Criteria:	• **Onpattro:** Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced **<u>Reauthorization</u>:** • Documentation of a positive clinical response (e.g., stabilized or

	improved neurologic impairment, motor function, cardiac function, quality of life assessment, serum TTR levels)
Exclusion Criteria:	• Prior or planned liver transplantation • New York Heart Association (NYHA) Functional Class III or IV • Combined use with TTR-lowering or stabilizing therapy
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or specialist experienced in the treatment of amyloidosis • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1

Required Medical Information:	**<u>Pulmonary Arterial Hypertension (PAH) WHO Group 1</u>**
	• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:
	  ○ Mean pulmonary artery pressure of at least 20 mm Hg
	  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg
	  ○ Pulmonary vascular resistance of at least 2.0 Wood units
	• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class III or higher symptoms
	• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blockers) unless there are contraindications:
	  ○ Low systemic blood pressure (systolic blood pressure less than 90)
	  ○ Low cardiac index
	  OR
	  ○ Presence of severe symptoms (functional class IV)
	• Documentation of current patient weight
	• Documentation of a clear treatment plan

Appropriate Treatment Regimen & Other Criteria:	• Documentation of inadequate response or intolerance to the following therapy classes is required:
	  ○ PDE5 inhibitors **AND**
	  ○ Endothelin receptor antagonists (exception WHO Functional Class IV)
	
	<u>Reauthorization</u> requires documentation of treatment success defined as one or more of the following:
	• Improvement in walking distance
	• Improvement in exercise ability
	• Improvement in pulmonary function
	• Improvement or stability in WHO functional class

Exclusion Criteria:	• Congestive heart failure due to severe left ventricular systolic dysfunction • Long-term use in patients who develop pulmonary edema during dose initiation
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a cardiologist or pulmonologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment for a mental health diagnosis of erectile dysfunction (ED), also known as erectile disorder, meeting sexual dysfunction criteria

Required Medical Information:	• Mental health diagnosis according to Diagnostic and Statistical Manual of Mental Disorders, fifth edition (DSM-5) diagnostic criteria for sexual dysfunction and erectile disorder:
	  ○ At least one of the three following symptoms must be experienced with 75% to 100% of occasions of sexual activity:
	  ▪ Marked difficulty in obtaining an erection during sexual activity
	  ▪ Marked difficulty in maintaining an erection until the completion of sexual activity
	  ▪ Marked decrease in erectile rigidity
	  ○ The above symptoms have persisted for a minimum duration of approximately 6 months AND
	  ○ The above symptoms cause clinically significant distress in the individual AND
	  ○ The sexual dysfunction is not:
	  ▪ Better explained by a nonsexual mental disorder OR
	  ▪ A consequence of severe relationship distress or other significant stressors AND
	  ▪ It is not attributable to the effects of substance or medication use or another medical condition (such as a physical condition)

Appropriate Treatment Regimen & Other Criteria:	• Documentation of treatment failure with tadalafil 2.5 mg or 5 mg tablets
Exclusion Criteria:	• Erectile dysfunction unrelated to a mental health diagnosis of sexual dysfunction according to the DSM-5 diagnostic criteria

Prescriber/Site of Care Restrictions	• Prescribed by, or in consultation with, a mental health provider • All approvals are subject to utilization of the most cost-effective site of care
Age Restriction:	
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

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

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

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design **Epogen & Aranesp & Procrit & Mircera** • 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 **Epogen & Procrit & Aranesp** • 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 **Epogen & Procrit only** • 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 • 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 **Compendia-supported uses** • Symptomatic anemia in Myelodysplastic syndrome • Allogenic bone marrow transplantation • Anemia associated with Hepatitis C (HCV) treatment • 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: ○ Anemia associated with chronic renal failure ○ Anemia secondary to chemotherapy with a minimum of two additional months of planned chemotherapy ○ Anemia secondary to zidovudine-treated Human Immunodeficiency Virus (HIV) patients ○ Anemia in patients scheduled to undergo elective, non-cardiac, nonvascular surgery ○ Symptomatic anemia in Myelodysplastic syndrome

	○ Allogenic bone marrow transplantation ○ Anemia associated with Hepatitis C (HCV) treatment ○ 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 the following criteria is met: ○ 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
Exclusion Criteria:	• Use in combination with another erythropoiesis stimulating agent (ESA)
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist, oncologist, or nephrologist
Coverage Duration:	• Authorization: 6 months, unless otherwise specified

POLICY NAME:
ETELCALCETIDE
Affected Medications: PARSABIV (etelcalcetide)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Secondary hyperparathyroidism in adults with chronic kidney disease (CKD) on dialysis

Required Medical Information:	• Documentation of both of the following:
	  ○ Currently on dialysis
	  ○ Intact parathyroid hormone (iPTH) level greater than 300 pg/mL
	• Documentation of iPTH that is persistently elevated above target range despite at least 12 weeks of adherent treatment with each of the following at an appropriate dose, unless contraindicated or not tolerated:
	  ○ Calcitriol
	  ○ Doxercalciferol
	  ○ Paricalcitol
	  ○ Cinacalcet

Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Diagnosis of parathyroid carcinoma, primary hyperparathyroidism or with chronic kidney disease who are not on hemodialysis
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist or nephrologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Hemophilia B (congenital factor IX deficiency)

Required Medical Information:	• Documentation of diagnosis of Hemophilia B
	• Documentation of baseline circulating level of factor IX less than or equal to 2% as attested by the managing physician AND requiring prophylactic Factor IX treatment
	• Documentation of negative Factor IX inhibitor titers (if test result is positive, re-test within 2 weeks with negative result)
	• Baseline lab values (less than 2 times upper limit of normal):
	  ○ ALT
	  ○ AST
	  ○ Total bilirubin
	  ○ Alkaline phosphatase (ALP)

Appropriate Treatment Regimen & Other Criteria:	• Documentation of plan to discontinue Factor IX prophylaxis therapy upon achieving circulating factor IX levels of 5% **Dosing:** • 2×10^{13} genome copies (gc) per kilogram of body weight
Exclusion Criteria:	• Prior gene therapy administration
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care • Prescribed by, or in consultation with, a hematologist or specialist with experience in the treatment of hemophilia
Coverage Duration:	• Authorization: 2 months (one-time infusion only), unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Homozygous familial hypercholesterolemia (HoFH)

Required Medical Information:	• Documentation of baseline untreated low-density lipoprotein cholesterol (LDL-C)
	• Diagnosis confirmed by **ONE** of the following:
	  ○ Baseline LDL-C greater than 500 mg/dL
	  ○ Baseline LDL-C of 400 mg/dL and at least 1 parent with familial hypercholesterolemia
	  ○ Baseline LDL-C of 400 mg/dL with aortic valve disease or xanthoma in ages less than 20 years
	  ○ Presence of two abnormal LDL-C-raising gene defects

Appropriate Treatment Regimen & Other Criteria:	• History of statin intolerance requires documentation of the following:
	  ○ Minimum of two different statin trials
	  ○ Documentation of statin-associated muscle symptoms, which stopped when statin therapy was discontinued and restarted when re-challenged
	• History of statin-associated rhabdomyolysis requires documentation of elevation in creatinine kinase (CK) level to at least 10 times the upper limit of normal, in concurrence with statin use
	• 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:
	  ○ Maximally tolerated statin therapy
	  ○ Ezetimibe
	  ○ PCSK9 monoclonal antibody, unless double-null or LDLR activity 15% or less
	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced

	<u>Reauthorization</u> requires documentation of treatment success and a clinically significant response to therapy defined by an LDL-C level at goal or decreased by at least 30% from baseline
Exclusion Criteria:	• Combination therapy with Juxtapid and Evkeeza is considered experimental and is not a covered benefit
Age Restriction:	• Juxtapid: 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist, cardiologist, or lipid specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
EVOLOCUMAB
Affected Medications: REPATHA (evolocumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Secondary prevention in clinical atherosclerotic cardiovascular disease (ASCVD)
	  ○ Primary hyperlipidemia (including heterozygous familial hypercholesterolemia [HeFH])
	  ○ Homozygous familial hypercholesterolemia (HoFH)

Required Medical Information:	**<u>All Indications</u>**
	• Documentation of current complete lipid panel within last 3 months
	• Documentation of baseline (untreated) low-density lipoprotein cholesterol (LDL-C)
	• Documentation of dietary measures being undertaken to lower cholesterol
	
	<u>Clinical ASCVD</u>
	• Documentation of established ASCVD, confirmed by at least **ONE** of the following:
	  ○ Acute coronary syndromes (ACS)
	  ○ History of myocardial infarction (MI)
	  ○ Stable or unstable angina
	  ○ Coronary or other arterial revascularization
	  ○ Stroke or transient ischemic attack
	  ○ Peripheral artery disease (PAD) presumed to be of atherosclerotic origin
	
	<u>Primary Hyperlipidemia/HeFH</u>
	• Diagnosis confirmed by **ONE** of the following:
	  ○ Minimum baseline LDL-C of 160 mg/dL in adolescents or 190 mg/dL in adults **AND** 1 first-degree relative affected
	  ○ Presence of one abnormal LDL-C-raising gene defect (e.g., LDL receptor [LDLR], apolipoprotein B [apo B], proprotein

	convertase subtilisin kexin type 9 [PCSK9] gain-of-function mutation, LDL receptor adaptor protein 1 [LDLRAP1]) ○ World Health Organization (WHO)/Dutch Lipid Network criteria score of at least 8 points ○ Definite FH diagnosis per the Simon Broome criteria **HoFH** • Diagnosis confirmed by **ONE** of the following: ○ Baseline LDL-C greater than 500 mg/dL ○ Baseline LDL-C of 400 mg/dL and at least 1 parent with familial hypercholesterolemia ○ Baseline LDL-C of 400 md/dL with aortic valve disease or xanthoma in ages < 20 years ○ Presence of two abnormal LDL-C-raising gene defect (excluding double-null LDLR mutations)
Appropriate Treatment Regimen & Other Criteria:	**All Indications** • Documented intent to take alongside maximally tolerated statin, unless otherwise contraindicated • History of statin intolerance requires documentation of the following: ○ Minimum of two different statin trials ○ Documentation of statin-associated muscle symptoms, which stopped when statin therapy was discontinued and restarted when re-challenged • History of statin-associated rhabdomyolysis requires documentation of elevation in creatinine kinase (CK) level to at least 10 times the upper limit of normal, in concurrence with statin use **Clinical ASCVD** • Documented treatment failure with minimum 12 weeks of consistent statin therapy at maximally tolerated dose, as shown by **ONE** of the following: ○ Current LDL-C of at least 70 mg/dL ○ Current LDL-C of at least 55 mg/dL in patients at very high risk of future ASCVD events (based on history of

<table>
<tr><td></td><td>multiple major ASCVD events OR 1 major ASCVD event + multiple high-risk conditions)

<table><tr><th>Major ASCVD Events</th><th>High-Risk Conditions</th></tr><tr><td>• ACS within the past 12 months
• History of MI (distinct from ACS event)
• Ischemic stroke
• Symptomatic PAD</td><td>• Age 65 years and older
• HeFH
• Prior coronary artery bypass or percutaneous intervention (outside of major ASCVD events)
• Diabetes
• Hypertension
• Chronic kidney disease
• Currently smoking
• History of congestive heart failure</td></tr></table>
<u>Primary Hyperlipidemia/HeFH/HoFH</u>
• Documented treatment failure with minimum 12 weeks of consistent statin therapy at maximally tolerated dose</td></tr>
<tr><td>Exclusion Criteria:</td><td></td></tr>
<tr><td>Age Restriction:</td><td></td></tr>
<tr><td>Prescriber/Site of Care Restrictions:</td><td>• All approvals are subject to utilization of the most cost-effective site of care</td></tr>
<tr><td>Coverage Duration:</td><td>• Authorization: 12 months, unless otherwise specified</td></tr>
</table>

POLICY NAME:
EXAGAMGLOGENE AUTOTEMCEL
Affected Medications: CASGEVY (exagamglogene autotemcel)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of sickle cell disease in adults and pediatric patients at least 12 years of age with recurrent vaso-occlusive crises.
	  ○ Treatment of transfusion-dependent beta-thalassemia in adults and pediatric patients at least 12 years of age.

Required Medical Information:	**<u>SICKLE CELL DISEASE</u>**
	
	• Documentation of sickle cell disease confirmed by genetic testing to show the presence of βS/βS, βS/β0 or βS/β+ genotype as follows:
	  ○ Identification of significant quantities of HbS with or without an additional abnormal β-globin chain variant by hemoglobin assay
	  OR
	  ○ Identification of biallelic *HBB* pathogenic variants where at least one allele is the p.Glu6Val or p.Glu7Val pathogenic variant on molecular genetic testing
	  AND
	  ○ Patient does NOT have disease with more than two α-globin gene deletions
	• Documentation of severe disease defined as 2 or more severe vaso-occlusive crises (VOCs) or vaso-occlusive events (VOEs) within the previous year (4 events over 2 years will also meet this requirement) VOC/VOEs defined as:
	  ○ Acute pain event requiring a visit to a medical facility and administration of pain medications (opioids or IV NSAIDs) or RBC transfusions
	  ○ Acute chest syndrome
	  ○ Priapism lasting more than 2 hours and requiring visit to medical facility
	  ○ Splenic sequestration
	• Clinically stable and eligible to undergo hematopoietic stem cell transplant (HSCT) but unable to find a human leukocyte antigen (HLA) matched, related donor

	• Adequate bone marrow, lung, heart, and liver function to undergo myeloablative conditioning regimen **<u>TRANSFUSION DEPENDENT BETA THALASSEMIA</u>** • Documented diagnosis of homozygous beta thalassemia or compound heterozygous beta thalassemia including β-thalassemia/hemoglobin E (HbE) (excludes alpha-thalassemia and hemoglobin S/ß-thalassemia variants) as outlined by the following: ○ Patient diagnosis is confirmed by HBB sequence gene analysis showing biallelic pathogenic variants **OR** ○ Patient has severe microcytic hypochromic anemia, anisopoikilocytosis with nucleated red blood cells on peripheral blood smear, and hemoglobin analysis that reveals decreased amounts or complete absence of hemoglobin A and increased amounts of hemoglobin F • Documented transfusion-dependent disease defined as a history of transfusions of at least 100 mL/kg/year of packed red blood cells (pRBCs) or with 10 or more transfusions of pRBCs *per year* in the 2 years preceding therapy • Clinically stable and eligible to undergo hematopoietic stem cell transplant (HSCT) but unable to find a human leukocyte antigen (HLA) matched, related donor
Appropriate Treatment Regimen & Other Criteria:	• Must weigh a minimum of 6 kilograms and able to provide a minimum number of cells (3×10^6 CD34+ cells/kg) • Documentation that cardiac iron overload has been evaluated and there is no evidence of severe iron overload. (cardiac T2* less than 10 msec by magnetic resonance imaging [MRI] or left ventricular ejection fraction [LVEF] less than 45% by echocardiogram) • No evidence of advanced liver disease [i.e., AST or ALT more than 3 times the upper limit of normal (ULN), or direct bilirubin value more than 2.5 times the ULN, or if a liver biopsy demonstrated bridging fibrosis or cirrhosis]
Exclusion Criteria:	• Prior HSCT or other gene therapy

Age Restriction:	• 12 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 6 months (one time infusion), unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Fabry disease

Required Medical Information:	• Diagnosis of Fabry disease confirmed by one of the following:
	  ○ Males: enzyme assay demonstrating undetectable (less than 3 percent) alpha-galactosidase A enzyme activity
	  ○ Males: deficiency of alpha-galactosidase A enzyme activity (less than 35 percent) and genetic testing showing a mutation in the galactosidase alpha (GLA) gene
	  ○ Females: genetic testing showing a mutation in the GLA gene
	• For Galafold: Genetic testing confirming the presence of at least one amenable GLA variant
	
	• Clinical signs and symptoms of Fabry disease, such as:
	  ○ Severe neuropathic pain
	  ○ Dermatologic manifestations (telangiectasias and angiokeratomas)
	  ○ Corneal opacities
	  ○ Kidney manifestations (proteinuria, polyuria, polydipsia)
	  ○ Cardiac involvement (left ventricular hypertrophy, myocardial fibrosis, heart failure)
	  ○ Cerebrovascular involvement (transient ischemic attacks, ischemic strokes)
	  ○ Other manifestations common in Fabry disease (sweating abnormalities, hearing loss, or intolerance to heat, cold, or exercise)

Appropriate Treatment Regimen & Other Criteria:	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced **Reauthorization** requires documentation of treatment success and a clinically significant response to therapy

Exclusion Criteria:	• Concurrent use with another agent on this policy (Galafold or enzyme replacement therapy for Fabry disease) • For Galafold: Severe renal impairment (eGFR less than 30) or end-stage renal disease requiring dialysis
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a geneticist or a specialist experienced in the treatment of Fabry disease • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Prophylaxis of *Clostridioides difficile* (C.diff) infection recurrence following antibiotic treatment

Required Medical Information:	• Documentation confirming a current diagnosis of recurrent C.diff infection (CDI) with a history of at least 2 recurrent episodes (initial episode + a minimum of 2 recurrences)
	  ○ Recurrent CDI is defined as a resolution of CDI symptoms while on appropriate therapy, followed by a reappearance of symptoms within 8 weeks of discontinuing treatment
	• Current episode of CDI must be controlled (less than 3 unformed or loose stools per day for 2 consecutive days)
	• Administration will occur following completion of antibiotic course for CDI treatment
	  ○ Within 24 to 72 hours for Rebyota
	  ○ Within 2 to 4 days for Vowst
	• Positive stool test for C.diff within the 30 days prior to request

Appropriate Treatment Regimen & Other Criteria:	• Previous treatment with each of the following in the setting of CDI recurrence:
	  ○ Vancomycin OR fidaxomicin (Dificid)
	  ○ Zinplava OR fecal microbiota transplantation (FMT)
	
	For Vowst requests: Documented treatment failure with all of the above agents AND Rebyota

Exclusion Criteria:	• Retreatment with Rebyota or Vowst
Age Restriction:	• 18 years of age and older

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an infectious disease specialist or gastroenterologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 1 month with no reauthorization, unless otherwise specified

POLICY NAME:
FENFLURAMINE
Affected Medications: FINTEPLA (fenfluramine)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of seizures associated with Dravet syndrome (DS)
	  ○ Treatment of seizures associated with Lennox-Gastaut syndrome (LGS)

Required Medical Information:	• Documented diagnosis of Dravet syndrome (DS) or Lennox-Gastaut Syndrome (LGS) • Current weight • Documentation that therapy is being used as adjunct therapy for seizures **<u>Dravet Syndrome</u>** • Documentation of at least 6 convulsive seizures in the last 6 weeks while on stable antiepileptic drug therapy **<u>Lennox-Gastaut Syndrome (LGS)</u>** • Documentation of at least 8 drop seizures per month while on stable antiepileptic drug therapy

Appropriate Treatment Regimen & Other Criteria:	**<u>Dravet Syndrome</u>**
	• Documented treatment and inadequate control of seizures with Epidiolex AND at least four of the following therapies:
	  ○ Valproate, clobazam, clonazepam, levetiracetam, zonisamide, or topiramate
	
	<u>Lennox-Gastaut Syndrome (LGS)</u>
	• Documented treatment and inadequate control of seizures with Epidiolex AND at least three guideline directed therapies:
	  ○ Valproate, lamotrigine, rufinamide, topiramate, felbamate, or clobazam
	
	• **Dosing:** not to exceed 26 mg daily

	<u>Reauthorization</u> requires documentation of treatment success and a reduction in seizure severity, frequency, or duration
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
FIDANACOGENE
Affected Medications: BEQVEZ (fidanacogene elaparvovec-dzkt)

Covered Uses:	• All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design ○ Hemophilia B (congenital factor IX deficiency)

Required Medical Information:	• Documentation of diagnosis of Hemophilia B
	• Documentation of baseline circulating level of factor IX less than or equal to 2% of normal AND requiring prophylactic factor IX treatment for at least 6 months
	• Documentation of negative factor IX inhibitor titers (less than 0.6 Bethesda units)
	• Documentation of negative antibodies to AAVRh74var capsid per FDA approved diagnostic test
	• Baseline lab values (less than 2 times upper limit of normal):
	  ○ ALT
	  ○ AST
	  ○ Alkaline phosphatase (ALP)
	  ○ Bilirubin

Appropriate Treatment Regimen & Other Criteria:	• Documentation of plan to discontinue factor IX prophylaxis therapy upon achieving circulating factor IX levels of 5% **<u>Dosing</u>** • 5×10^{11} vector genomes per kilogram of body weight
Exclusion Criteria:	• Prior gene therapy administration • Unstable liver or biliary disease • Active Hepatitis B or C infection • HIV infection with CD4 cell count less than 200 mm^3 or viral load greater than 20 copies/mL
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation, with a hematologist or specialist with experience in treatment of hemophilia • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Authorization: 2 months (one-time infusion)

POLICY NAME:
FINERENONE
Affected Medications: KERENDIA (finerenone)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Chronic kidney disease associated with type 2 diabetes to reduce the risk of:
	  ▪ Sustained estimated glomerular filtration rate (eGFR) decline
	  ▪ End-stage kidney disease
	  ▪ Cardiovascular death
	  ▪ Non-fatal myocardial infarction
	  ▪ Hospitalization for heart failure

Required Medical Information:	• Documentation of all the following:
	  ○ eGFR greater than or equal to 25 mL/min/1.73 m^2
	  ○ Urine albumin-to-creatinine ratio (UACR) greater than or equal to 30 mg/g
	  ○ Serum potassium level less than or equal to 5.0 mEq/L

Appropriate Treatment Regimen & Other Criteria:	• Currently receiving maximally tolerated dosage of an angiotensin converting enzyme (ACE) inhibitor or angiotensin receptor blocker (ARB), unless intolerant or contraindicated • Documented treatment failure or intolerable adverse event to at least 12 weeks of sodium-glucose cotransporter 2 (SGLT2) inhibitor therapy **<u>Reauthorization</u>** requires documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a nephrologist, endocrinologist, or cardiologist • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
FLUCYTOSINE
Affected Medications: FLUCYTOSINE

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Candida endocarditis
	  ○ Candidiasis
	  ○ Candidiasis of urogenital site
	  ○ Cryptococcosis
	• Compendia-supported uses that will be covered (if applicable)
	  ○ Candida endophthalmitis
	  ○ Central nervous system candidiasis
	  ○ Cryptococcal meningitis – HIV infection
	  ○ HIV infection – Pulmonary cryptococcosis

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:	• Prescribed by, or in consultation with, an infectious disease specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 8 weeks, or lesser requested duration, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Diabetic macular edema (DME)
	  ○ Chronic, non-infectious posterior uveitis

Required Medical Information:	**Iluvien** • Diagnosis of clinically significant diabetic macular edema • Documentation of past treatment with corticosteroids without a clinically significant rise in intraocular pressure **Retisert and Yutiq** • Diagnosis of chronic, non-infectious posterior uveitis confirmed by slit lamp and fundoscopic examination

Appropriate Treatment Regimen & Other Criteria:	**Iluvien**
	• Documentation of inadequate response or intolerance to an intravitreal vascular endothelial growth factor (VEGF) inhibitor (preferred products: Avastin, Byooviz, Cimerli)
	• Documentation of inadequate response to laser photocoagulation
	
	Retisert and Yutiq
	• Documentation of inadequate response or intolerance to all of the following:
	  ○ Minimum 12-week trial with oral systemic corticosteroid
	  ○ At least one corticosteroid-sparing immunosuppressive therapy (methotrexate, azathioprine, or mycophenolate mofetil)
	  ○ At least one calcineurin inhibitor (cyclosporine, tacrolimus)
	• **Retisert**: Documentation of treatment failure with Yutiq

Exclusion Criteria:	• Active or suspected ocular or periocular infections • Concurrent use of intravitreal implants or injections (corticosteroid, anti-VEGF) • **Iluvien:** Glaucoma (with cup to disc ratios greater than 0.8)
Age Restriction:	

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an ophthalmologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• **Iluvien**: 36 months, unless otherwise specified • **Retisert**: 30 months, unless otherwise specified • **Yutiq**: 36 months, unless otherwise specified

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

Covered Uses:	• Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
Required Medical Information:	• Documentation of disease state, level of control, and therapies failed • Documentation of failure with all available formulary products for treatment of disease state • 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 • All approvals are subject to utilization of the most cost effective site of care
Coverage Duration:	• Case by case based on member need

POLICY NAME:
FOSTAMATINIB
Affected Medications: TAVALISSE (fostamatinib)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Thrombocytopenia in adults with chronic immune thrombocytopenia (ITP) who have had an insufficient response to a previous treatment

Required Medical Information:	**<u>Thrombocytopenia in patients with chronic ITP</u>**
	• Documentation of **ONE** of the following:
	  ○ Platelet count less than 20,000/microliter
	  ○ Platelet count less than 30,000/microliter AND symptomatic bleeding
	  ○ Platelet count less than 50,000/microliter AND increased risk for bleeding (such as peptic ulcer disease, use of antiplatelets or anticoagulants, history of bleeding at higher platelet count, need for surgery or invasive procedure)

Appropriate Treatment Regimen & Other Criteria:	**<u>Thrombocytopenia in patients with chronic ITP</u>**
	• Documentation of inadequate response, defined as platelets did not increase to at least 50,000/microliter, to the following therapies:
	  ○ **ONE** of the following:
	  ▪ Inadequate response with at least 2 therapies for immune thrombocytopenia, including corticosteroids, rituximab, or immunoglobulin
	  ▪ Splenectomy
	  ○ Promacta
	
	<u>Reauthorization</u>:
	• Response to treatment with platelet count of at least 50,000/microliter or above (not to exceed 400,000/microliter)

Exclusion Criteria:	• Use in combination with a thrombopoietin receptor agonist, spleen tyrosine kinase inhibitor, or similar treatment for thrombocytopenia (such as Promacta, Doptelet, or Nplate)

Age Restriction:	
Prescriber Restrictions:	• Prescribed by, or consultation with, a hematologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. • 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)**
	• Presence of malignant locally advanced unresectable or metastatic disease confirmed by pathology.
	• History of intolerable adverse event with trial of each of the following agents:
	  ○ Sirolimus oral tablet
	  ○ Everolimus or temsirolimus
	
	Reauthorization: documentation of disease responsiveness to therapy

Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater • 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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial approval: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of postpartum depression (PPD)

Required Medical Information:	• Documentation of major depressive episode as diagnosed by DSM-5 Criteria
	  ○ 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
	  ▪ Depressed mood most of the day, nearly every day, as indicated by either subjective report (e.g., feels sad, empty, hopeless) or observations made by others (e.g., appears tearful). (NOTE: In children and adolescents, can be irritable mood.)
	  ▪ 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)
	  ▪ Significant weight loss when not dieting or weight gain (e.g., 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.)
	  ▪ Insomnia or hypersomnia nearly every day
	  ▪ Psychomotor agitation or retardation nearly every day (observable by others, not merely subjective feelings of restlessness or being slowed down)
	  ▪ Fatigue or loss of energy nearly every day
	  ▪ Feelings of worthlessness or excessive or inappropriate guilt (which may be delusional) nearly every day (not merely self-reproach or guilt about being sick)
	  ▪ Diminished ability to think or concentrate, or indecisiveness, nearly every day (either by their subjective account or as observed by others)

	▪ 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 AND ○ Symptoms cause clinically significant distress or impairment in social, occupational, or other important areas of functioning AND ○ Episode is not attributable to the direct physiological effects of a substance or to another condition • Major depressive episode began no earlier than the third trimester and no later than the first 4 weeks following delivery • Moderate to severe postpartum depression documented by one of the following rating scales: ○ Hamilton Rating Scale for Depression (HAM-D) score of greater than 17 ○ Patient Health Questionnaire-9 (PHQ-9) score of greater than 10 ○ Montgomery-Åsberg Depression Rating Scale (MADRS) greater than 20 points ○ Edinburgh Postnatal Depression Scale (EPDS) score of greater than 13
Appropriate Treatment Regimen & Other Criteria:	• Documented trial with an oral antidepressant for at least 8 weeks unless contraindicated or documentation shows that the severity of the depression would place the health of the mother or infant at significant risk • For Zulresso requests: Documented treatment failure with Zurzuvae
Exclusion Criteria:	• Greater than 6 months postpartum
Age Restriction:	• 15 years of age and older for Zulresso • 18 years of age and older for Zurzuvae
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a psychiatrist • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Authorization: 1 month, one time approval per pregnancy, unless otherwise specified

POLICY NAME:
GANAXOLONE
Affected Medications: ZTALMY (ganaxolone)

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

POLICY NAME:
GIVINOSTAT
Affected Medications: DUVYZAT (givinostat)

Covered Uses:	• All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design ○ Duchenne muscular dystrophy (DMD) in patients 6 years of age and older

Required Medical Information:	• Genetically confirmed diagnosis of DMD
	• Documentation of being ambulatory without needing an assistive device such as a wheelchair, walker, or cane
	• North Star Ambulatory Assessment (NSAA) scale total score of 17 or more
	• Baseline motor function assessment from one of the following:
	  ○ 4-stair climb (4SC) test
	  ○ Time to Stand Test (TTSTAND)
	  ○ 6-minute walk test (6MWT)
	  ○ North Star Ambulatory Assessment (NSAA)
	  ○ Motor Function Measure (MFM)
	  ○ Hammersmith Functional Motor Scale (HFMS)
	• Current weight and planned treatment regimen

Appropriate Treatment Regimen & Other Criteria:	• Documentation of being on a stable dose of an oral corticosteroid such as prednisone for at least 6 months, and will continue while on Duvyzat unless contraindicated **<u>Reauthorization</u>** requires a documented improvement from baseline or stabilization of motor function demonstrated by a motor function assessment tool
Exclusion Criteria:	• Concomitant therapy or within the past 6 months with DMD-directed antisense oligonucleotides such as golodirsen, casimersen, viltolarsen, eteplirsen • Platelet, white blood cell, or hemoglobin counts less than the lower limit of normal • QTc is greater than 500 ms or the change from baseline is greater than 60 ms. • History of additional risk factors for torsades de pointes (e.g. heart failure, hypokalemia, or family history of long QT syndrome)

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

POLICY NAME:
GIVOSIRAN
Affected Medications: GIVLAARI (givosiran)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of adults with acute hepatic porphyria (AHP)
Required Medical Information:	• Documentation of elevated urine porphobilinogen (PBG) levels based on specific lab test utilized • Diagnosis confirmed based on Porphyria Genomic testing • Documentation of baseline acute attack frequency • Evaluation and elimination of exacerbating factors including medications, smoking, drinking, and infections

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
	• For women:
	  ○ Documented 12-week trial and failure of gonadotropin releasing hormone analogue (ex. leuprolide) OR
	  ○ Documentation that attacks are not related to the luteal phase of the menstrual cycle
	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced
	
	<u>Reauthorization</u> will require documentation of greater than 50% reduction in baseline acute attack frequency

Exclusion Criteria:	• Active HIV, hepatitis C, or hepatitis B infection(s) • History of pancreatitis • Concomitant use with prophylactic hemin
Age Restriction:	• 12 years of age or older

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, specialist in the treatment of acute hepatic porphyria • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design ○ Diabetes Mellitus, Type 2
Required Medical Information:	• Available information is reviewed, including previous fill history • Diagnosis of Type 2 diabetes with a recent hemoglobin A1c greater than or equal to 7% despite current therapy • Documented treatment failure with minimum of 12-week trial with metformin or metformin extended release 2000 mg daily (or if unable to tolerate 2000 mg daily, the maximum tolerated dose) defined as failure to achieve or maintain A1c less than 7% ○ If intolerant to immediate release metformin, 12-week trial with metformin extended release must be trialed
Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** requires documentation of disease responsiveness to therapy
Exclusion Criteria:	• Use for weight loss or other excluded diagnosis • Dosing above Food and Drug Administration (FDA) approved label for treatment of diabetes • 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:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Hypogonadotropic hypogonadism secondary to a pituitary deficiency in males
	  ○ Prepubertal cryptorchidism not caused by anatomic obstruction
	• Perioperative use in male infants/toddlers with hypospadias and chordee OR total epispadias and bladder exstrophy

Required Medical Information:	**<u>Hypogonadotropic hypogonadism secondary to a pituitary deficiency in males:</u>** • Documentation confirming the diagnosis
Appropriate Treatment Regimen & Other Criteria:	**<u>Reauthorization</u>** 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) • Obesity • Prevention of recurrent or habitual miscarriage • Treatment or prevention of breast cancer
Age Restriction:	• Prepubertal cryptorchidism: generally, between 4 and 9 years of age • Hypospadias or epispadias: infant or toddler
Prescriber/Site of Care Restrictions:	• All approvals are subjects to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Endometriosis
	  ○ Endometrial thinning
	• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better

Required Medical Information:	**<u>Endometriosis:</u>** • Documentation of moderate to severe pain due to endometriosis

Appropriate Treatment Regimen & Other Criteria:	**<u>Endometriosis:</u>**
	
	• Documentation of a trial and inadequate relief (or contraindication) after at least 3 months of both of the following first-line therapies:
	  ○ Nonsteroidal anti-inflammatory drugs (NSAIDs)
	  ○ Continuous (no placebo pills) hormonal contraceptives
	
	<u>Endometrial thinning:</u>
	
	• Documentation of both of the following:
	  ○ Diagnosis of dysfunctional uterine bleeding
	  ○ Planning to use as an endometrial-thinning agent prior to endometrial ablation
	
	<u>Reauthorization for oncologic uses</u> require documentation of disease responsiveness to therapy

Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater • For endometriosis, prior use of Zoladex for a 6-month period
Age Restriction:	• 18 years of age and older

Prescriber/Site of Care Restrictions:	• For oncologic uses: Prescribed by, or in consultation with, an oncologist • For gynecologic uses: Prescribed by, or in consultation with, a gynecologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	**Oncologic uses:** • Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified **Endometriosis:** • Authorization: 6 months with no reauthorization, unless otherwise specified **Endometrial thinning:** • Authorization: 4 months (up to 2 doses only), unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design

Required Medical Information:	**<u>All indications:</u>**
	• Documentation of baseline height, height velocity, bone age (pediatrics), and patient weight
	
	<u>Growth hormone deficiency or Pituitary dwarfism</u>
	• For initial approval, documentation of the following is required:
	  ○ Diagnosis of growth hormone deficiency or pituitary dwarfism AND
	  ○ Low serum values for GH stimulation test, IGF-1, and IGFBP-3 with delayed bone age AND
	  ▪ Height standard deviation score (SDS) of -2.5 (0.6^{th} percentile)
	  OR
	  ▪ Height velocity impaired AND
	  ▪ Height SDS of -2 (2.3rd percentile) for bone age
	
	<u>Turner's syndrome</u>
	• For initial approval, documentation of the following is required:
	  ○ Diagnosis of Turner Syndrome done through genetic testing AND
	  ▪ For patients less than 2 years of age:
	  • Documented 50% delay in growth from projected based on World Health Organization (WHO) growth curves at equivalent age, AND
	  • No secondary factor present that would explain observed growth delays
	  ▪ For patients greater than or equal to 2 years of age:
	  • Height below the 5th percentile for bone age, AND
	  • No secondary factor present that would explain observed growth delays

	Noonan's syndrome
	• For initial approval, documentation of the following is required:
	  ○ Diagnosis of Noonan's syndrome done through genetic testing AND
	  ▪ Height standard deviation score (SDS) of -2.5 (0.6th percentile)
	  OR
	  ▪ Height velocity impaired AND
	  ▪ Height SDS of -2 (2.3rd percentile) for bone age
	
	Short stature homeobox-containing gene (SHOX) deficiency
	• For initial approval, documentation of the following is required:
	  ○ Diagnosis of SHOX deficiency done through genetic testing
	  ▪ Height standard deviation score (SDS) of -2.5 (0.6th percentile)
	  OR
	  ▪ Height velocity impaired AND
	  ▪ Height SDS of -2 (2.3rd percentile) for bone age
	
	Chronic kidney disease stage 3 and greater OR kidney transplant
	• For initial approval, documentation of the following is required:
	  ○ Diagnosis of chronic kidney disease stage 3 or higher (CrCl less than 60mL/min)
	  ○ Height velocity (SDS) less than -1.88 for bone age.
	
	Prader-Willi syndrome
	• For initial approval, documentation of the following is required:
	  ○ Diagnosis of Prader-Willi syndrome through genetic testing **AND**
	  ○ Height velocity impaired
	
	Short Stature born small for gestational age (SGA) with no catch-up growth by 2 years to 4 years of age
	• Birth weight and/or length of at least 2 standard deviations (-2 SD) from the mean for gestational age and sex
	• Height standard deviation score (SDS) of -2.5 (0.6th percentile)
	• Age at start of growth hormone therapy cannot be greater than 10 years

	• Exclusion of other causes of short stature including growth-inhibiting medication, chronic disease, endocrine disorders
	
	<u>Adult Growth Hormone Deficiency:</u>
	• For initial approval, documentation of the following is required:
	  ○ Dose and frequency are appropriate **AND**
	  ○ Documented Growth Hormone Deficiency **AND**
	  ○ Documented IGF-1 outside reference range for patient's sex and age, AND the patient has failed one growth hormone stimulation test (insulin tolerance test-ITT or Glucagon stimulation test when ITT is contraindicated)
	
	<u>Reauthorization:</u>
	• **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
	• **Adult Growth Hormone Deficiency:** requires documented clinical improvement and IGF-I within normal reference range for age and sex

Appropriate Treatment Regimen & Other Criteria:	• Documented trial and failure of at least 12 weeks of Norditropin prior to any other daily growth hormone • For Skytrofa and Sogroya: ○ Documented trial and failure of at least 12 weeks of Norditropin and one additional daily growth hormone
Exclusion Criteria:	• Pregnancy • Elderly adults with age-adjusted low IGF-1 levels and no history of pituitary or hypothalamic disease. • Growth Hormone (GH) replacement to enhance athletic performance • Diagnosis of: Idiopathic Short Stature (ISS), height standard deviation score (SDS) less than -2.25, and associated with growth rates unlikely to permit attainment of adult height in the normal range
Age Restriction:	

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist • All approvals are subjects to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. • 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
	• Current HIV status
	• Current Hepatitis B status
	• Baseline HCV RNA level within last 3 months with genotyping
	• Documentation that patient is one of the following:
	  ○ Treatment-naïve
	  ○ Treatment experienced, including documentation of previous treatment regimen and outcome
	• 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
	• 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) • Vosevi is not recommended in patients with moderate or severe hepatic impairment (Child-Pugh class B or C) • 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 • 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 Mavyret x 8 weeks
	Compensated Cirrhosis	SOF/VEL x 12 weeks Mavyret x 8 weeks
	Decompensated Cirrhosis	SOF/VEL + RBV x 12 weeks SOF/VEL x 24 weeks (if ribavirin ineligible*)
Treatment Experienced (Genotype 1-6)		
Sofosbuvir based regimen treatment failures, including: - Sofosbuvir + ribavirin - Ledipasvir/sofosbuvir (Harvoni) - SOF/VEL	Non-cirrhotic or compensated cirrhosis	Vosevi x 12 weeks 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 Vosevi x 12 weeks (plus RBV if compensated cirrhosis)

Multiple DAA treatment failures, including: - Vosevi - Mavyret + sofosbuvir	Non-cirrhotic or compensated cirrhosis	Mavyret + SOF + RBV x 16-24 weeks Vosevi + RBV x 24 weeks
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 Mavyret x 8 weeks
	Decompensated Cirrhosis	SOF/VEL + RBV x 12 weeks
Treatment Experienced		
Efficacy and safety is extremely limited in treatment experienced patients in this population. Can consider recommended treatment regimens in adults if FDA approved for pediatric use. Recommend consulting with hepatologist.		
Abbreviations: DAA = direct-acting antiviral; PEG = pegylated interferon; RBV = ribavirin; SOF/VEL = sofosbuvir/velpatasvir		

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

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

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

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

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Central precocious puberty (CPP) • Gender dysphoria

Required Medical Information:	**<u>Central Precocious Puberty:</u>**
	
	• Documentation of CPP confirmed by basal luteinizing hormone (LH), follicle-stimulating hormone (FSH), and either estradiol or testosterone concentrations
	
	<u>Gender Dysphoria:</u>
	
	• Documentation of all of the following:
	  ○ Current Tanner stage 2 or greater OR baseline and current estradiol and testosterone levels to confirm onset of puberty
	  ○ Confirmed diagnosis of gender dysphoria that is persistent
	  ○ The patient has the capacity to make a fully informed decision and to give consent for treatment
	  ○ Any significant medical or mental health concerns are reasonably well controlled
	  ○ A comprehensive mental health evaluation has been completed by a licensed mental health professional (LMHP) and provided in accordance with the most current version of the World Professional Association for Transgender Health (WPATH) Standards of Care

Appropriate Treatment Regimen & Other Criteria:	**<u>All Indications:</u>** • Approval requires documented treatment failure with leuprolide **<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy

Exclusion Criteria:	
Age Restriction:	• 2 years of age or older
Prescriber/Site of Care Restrictions:	• Central Precocious Puberty: Prescribed by, or in consultation with, an endocrinologist • Gender dysphoria: Diagnosis made and prescribed by, or in consultation with, a specialist in the treatment of gender dysphoria • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
HEREDITARY ANGIOEDEMA
Affected Medications: Berinert, Icatibant Acetate, Sajazir, Ruconest, Kalbitor, Cinryze, Haegarda, Takhzyro, Orladeyo

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Hereditary angioedema attacks, prophylaxis (Cinryze, Haegarda, Takhzyro, Orladeyo)
	  ○ Hereditary angioedema attacks, acute treatment (Berinert, icatibant acetate, Sajazir, Kalbitor, Ruconest)

Required Medical Information:	**Diagnosis of hereditary angioedema (HAE) classified as one of the following:**
	• Type I or II HAE confirmed by low C4 levels AND one of the following:
	  ○ Low C1 inhibitor functional or antigenic level less than 50% of the lower limit of normal as defined by the laboratory performing test
	• "Type III" HAE confirmed by normal C4, C1 inhibitor (functional and antigenic) with one of the following:
	  ○ Genetic testing confirming presence of HAE causing mutation such as mutation of coagulation factor XII gene (F12 mutation), mutation in the angiopoietin-1 gene, mutation in the plasminogen gene, mutation in the kininogen 1 gene, mutation in the myoferlin gene, mutation in the heparan sulfate 3-Osulfotransferase 6 gene
	  ○ Family history of HAE AND documented recurring angioedema attacks that are refractory to high dose antihistamines (four times the usual dose)
	• Documented full treatment plan and current body weight
	• Documentation of number of attacks requiring treatment in the past year

Appropriate Treatment	**<u>Acute Treatment:</u>** • Documented history of one of the following: ○ Non-inflammatory subcutaneous angioedema (without

Regimen & Other Criteria:	hives) which is recurrent and lasts greater than 12 hours ○ Abdominal pain without a clear organic cause lasting greater than 6 hours Coverage for non-preferred products (Berinert, Kalbitor, Ruconest) requires documentation of one of the following: • Documented treatment failure to one of the preferred products: icatibant acetate or Sajazir • Currently receiving treatment with a non-preferred product, excluding via samples or manufacturer's patient assistance programs For requests to treat more than 3 attacks per month: • Documentation of current treatment with, or failure, intolerance, or clinical rationale for avoidance of, prophylactic therapies • Authorization for acute treatment will provide a sufficient quantity to treat the average number of acute attacks per month plus 1 additional dose **<u>Prophylaxis Treatment:</u>** • History of TWO or more severe attacks per month for the past 3 months (airway swelling, debilitating cutaneous or gastrointestinal episodes) despite short term treatment and at least one of the following: ○ Disabling symptoms for at least 5 days per month ○ History of at least one laryngeal attack caused by HAE • Avoidance of possible triggers for HAE attacks such as ○ estrogen containing oral contraceptives/hormone replacement ○ angiotensin-converting-enzyme (ACE) inhibitors ○ dipeptidyl peptidase IV (DPP-4) inhibitors ○ Neprilysin inhibitor Coverage for non-preferred products (Cinryze, Orladeyo) requires documentation of one of the following: • Documented treatment failure to the preferred products

	Haegarda and Takhzyro • Currently receiving treatment with a non-preferred product, excluding via samples or manufacturer's patient assistance programs **<u>Reauthorization</u>** requires documentation of number of acute HAE attacks treated in the past year AND documentation of treatment success defined as reduction of frequency and severity of HAE attack episodes requiring acute therapy by greater than or equal to 50% from baseline. • Requested dose within the Food and Drug Administration (FDA)-approved label • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced for all medical infusion drugs
Exclusion Criteria:	• Concurrent use of multiple HAE prophylactic treatments (Orladeyo, Haegarda, Takhzyro, Cinryze) • Concurrent use of multiple HAE acute treatments (Berinert, Kalbitor, Runconest, icatibant acetate, Sajazir)
Age Restriction:	• Product specific per FDA labeled indication
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an allergist, immunologist, or pulmonologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Hereditary tyrosinemia type 1 (HT-1)

Required Medical Information:	• Diagnosis of hereditary tyrosinemia type 1 confirmed by:
	  ○ Presence of succinylacetone (SA) in urine or blood
	  ○ Genetic testing showing a mutation in the gene encoding fumarylacetoacetate hydrolase (FAH)
	• Current patient weight

Appropriate Treatment Regimen & Other Criteria:	• Use as an adjunct to dietary restriction of tyrosine and phenylalanine • Orfadin requires: ○ A documented intolerable adverse event to Nityr and the adverse event was not an expected adverse event attributed to the active ingredient **<u>Reauthorization</u>**: documentation of treatment success confirmed by: • Reduction in urine or plasma succinylacetone from baseline • Documentation of dietary restriction of tyrosine and phenylalanine
Exclusion Criteria:	• Use without dietary restriction of tyrosine and phenylalanine
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist in the treatment of hereditary tyrosinemia or related disorders • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • 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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design • Gender dysphoria ○ Applies to patients under 18 years of age

Required Medical Information:	**<u>Gender dysphoria</u>**
	• Documentation of all of the following:
	  ○ Current Tanner stage 2 or greater OR baseline and current estradiol and testosterone levels to confirm onset of puberty
	  ○ Confirmed diagnosis of gender dysphoria that is persistent
	  ○ The patient has the capacity to make a fully informed decision and to give consent for treatment
	  ○ Any significant medical or mental health concerns are reasonably well controlled
	  ○ A comprehensive mental health evaluation has been completed by a licensed mental health professional (LMHP) and provided in accordance with the most current version of the World Professional Association for Transgender Health (WPATH) Standards of Care
	• **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:	**<u>Reauthorization</u>** requires 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 the treatment of gender dysphoria • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 24 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Glucocorticoid replacement therapy in pediatric patients with adrenocortical insufficiency
Required Medical Information:	• Diagnosis of adrenal insufficiency confirmed with an adrenal stimulation test • Current body surface area (or height and weight to calculate) • Current height and weight velocity • For adolescents, evaluation of epiphyses (growth plates) documenting they remain open • Complete treatment plan including dose in mg/m^2/day

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure with a 6-month trial of two or more of the following:
	  ○ Hydrocortisone tablets
	  ○ Cortisone acetate tablets
	  ○ Prednisolone or prednisone tablets
	  ○ Compounded hydrocortisone oral capsules or solution
	• **Dosing** is in accordance with FDA labeling and does not exceed the following:
	  ○ Starting dose: 8-10 mg/m^2/day in 3 divided doses
	  ○ When switching from other oral hydrocortisone formulations, use the same total hydrocortisone dosage
	  ○ Infants with Congenital Adrenal Hyperplasia may start at a dose of 8-15 mg/m^2/day in 3 divided doses
	
	Reauthorization requires documentation of treatment success and a clinically significant response to therapy

Exclusion Criteria:	• Use in adolescents who have achieved their adult height • Use for stress dosing • Use in acute treatment of adrenal crisis or acute adrenal insufficiency

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

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ For the treatment of facial angiofibroma (FA) associated with tuberous sclerosis complex (TSC)

Required Medical Information:	• Documented diagnosis of FA associated with TSC which are:
	  ○ Rapidly changing in size and/or number
	  ○ Causing functional interference, pain or bleeding
	  ○ Inhibiting social interactions
	• Current and baseline description of FA including lesion count, associated symptoms and complications, and overall severity

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure with laser therapy and/or surgery (such as shave excision, cryotherapy, radiofrequency ablation, or dermabrasion), unless contraindicated **Reauthorization** requires documentation of a positive clinical response to therapy (decrease in size and/or redness of facial angiofibromas)
Exclusion Criteria:	• Concurrent use of systemic mammalian target of rapamycin (mTOR) inhibitors • Treatment of non-facial angiofibroma
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a dermatologist, oncologist, or neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Anemia due to chronic kidney disease (CKD) in adults who have been receiving dialysis

Required Medical Information:	• Diagnosis of anemia due to CKD
	• Documentation of dialysis use for:
	  ○ Jesduvroq: 4 or more months
	  ○ Vafseo: 3 or more months
	• Documentation of pretreatment hemoglobin level greater than 8 g/dL and less than 12 g/dL
	• Adequate iron stores as indicated by current (within the last three months) serum ferritin level greater than or equal to 100 mcg/L or serum transferrin saturation greater than or equal to 20%

Appropriate Treatment Regimen & Other Criteria:	• Documentation of **ONE** of the following:
	  ○ Documented hypo-responsiveness to an erythropoiesis stimulating agent (ESA), defined as the need for **ONE** of the following:
	  ▪ Greater than 300 IU/kg per week of epoetin alfa
	  ▪ Greater than 1.5 mcg/kg per week of darbepoetin
	  ○ Intolerance to **BOTH** preferred ESA products epoetin alfa-epbx (Retacrit) and darbepoetin alfa (Aranesp)
	
	<u>Reauthorization</u> requires documentation of treatment success and hemoglobin of greater than 8 g/dL and less than 12 g/dL

Exclusion Criteria:	• Use in combination with ESAs • Current uncontrolled hypertension • Active malignancy • For Jesduvroq: Major adverse cardiac events (such as myocardial infarction, acute coronary syndrome, stroke,

	transient ischemic attack, venous thromboembolism) within 3 months prior to starting treatment
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist, such as a hematologist or nephrologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
IBREXAFUNGERP
Affected Medications: BREXAFEMME (ibrexafungerp)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of vulvovaginal candidiasis (VVC)
	  ○ Reduction in the incidence of recurrent vulvovaginal candidiasis (RVVC)

Required Medical Information:	**<u>All Indications</u>** • Documented presence of signs/symptoms of current acute vulvovaginal candidiasis with a positive potassium hydroxide (KOH) test • Documentation confirming that the patient is not pregnant and is on contraceptive for length of planned treatment **<u>RVVC</u>** • Documentation of three or more episodes of symptomatic vulvovaginal candidiasis infection within the past 12 months

Appropriate Treatment Regimen & Other Criteria:	**<u>VVC</u>**
	• Documented treatment failure with both of the following for the current VVC episode:
	  ○ Vaginally administered treatment (such as clotrimazole cream, miconazole cream, terconazole cream or suppository)
	  ○ A 7-day course of fluconazole taken orally every third day for a total of 3 doses (days 1, 4, and 7)
	
	<u>RVVC</u>
	• 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
	
	<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:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	Authorization (VVC): 3 months, unless otherwise specified Authorization (RVVC): 6 months, unless otherwise specified

POLICY NAME:
ILARIS
Affected Medications: ILARIS (canakinumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS), Hyperimmunoglobulin D syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD), Familial Mediterranean Fever (FMF), Adult-Onset Still's Disease (AOSD), Systemic Juvenile Idiopathic Arthritis (SJIA), Cryopyrin-Associated Periodic Syndromes (CAPS), Gout Flares

Required Medical Information:	**<u>Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS)</u>**
	• Confirmed diagnosis of TRAPS with frequent and/or severe recurrent disease (such as recurrent fevers, prominent myalgias, migratory rash, periorbital edema) AND documented genetic defect of TNFRSF1A gene
	
	<u>Hyperimmunoglobulin D syndrome (HIDS)/ Mevalonate Kinase Deficiency (MKD)</u>
	• Confirmed diagnosis with one of the following:
	  ○ Elevated serum IgD with or without elevated IgA
	  ○ Genetic testing showing presence of heterozygous or homozygous mutation in the mevalonate kinase (MVK) gene
	• Documentation of 3 or more febrile acute flares within a 6-month period
	
	<u>Still's Disease</u>
	• Confirmed diagnosis of Still's Disease, including Adult-Onset Still's Disease (AOSD) and Systemic Juvenile Idiopathic Arthritis (SJIA) in patients 2 years of age and older
	• Documented clinical signs and symptoms including fever, rash, arthritis, arthralgia, myalgia, pharyngitis, pulmonary disease, elevated liver enzymes, C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), serum ferritin

	<u>Cryopyrin-Associated Periodic Syndromes (CAPS)</u>
	• Confirmed diagnosis of CAPS in patients 4 years and older including Familial Cold Autoinflammatory Syndrome (FCAS) or Muckle-Wells Syndrome (MWS) with one of the following:
	  ○ Elevated inflammatory markers such as CRP and serum amyloid A with two of the following manifestations:
	  ▪ Urticaria-like rash, cold-triggered episodes, sensorineural hearing loss, musculoskeletal symptoms, chronic aseptic meningitis, skeletal abnormalities
	  ○ Genetic testing showing presence of NALP3 mutations
	
	<u>Gout Flares</u>
	• Confirmed diagnosis of gout that is refractory to standard therapies
	• Documentation of having 3 or more gout flares in the past 12 months

Appropriate Treatment Regimen & Other Criteria:	**<u>TRAPS</u>** • Documented clinical failure to episodic treatment with nonsteroidal anti-inflammatory drugs (NSAIDs), glucocorticoids (prednisone or prednisolone), and a minimum 12-week trial with Enbrel **<u>HIDS/MKD</u>** • Documented treatment failure to episodic treatment with nonsteroidal anti-inflammatory drugs (NSAIDs), glucocorticoids, and anakinra **<u>FMF</u>** • Documented treatment failure with maximal tolerable dose of colchicine (3 mg daily in adults and 2 mg daily in children) • Documentation of frequent and/or severe recurrence disease despite adequate treatment with at least 12 weeks of anakinra **<u>Still's Disease</u>** • Documentation of frequent and/or severe recurrent disease despite adequate treatment with a minimum 12-week trial with each of the following: ○ NSAIDs or glucocorticoids

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

POLICY NAME:
ILOPROST
Drug Name: VENTAVIS (iloprost)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1

Required documentation:	**<u>Pulmonary Arterial Hypertension (PAH) WHO Group 1</u>**
	• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:
	  ○ Mean pulmonary artery pressure of at least 20 mm Hg
	  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg
	  ○ Pulmonary vascular resistance of at least 2.0 Wood units
	• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class III or higher symptoms
	• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blockers) unless there are contraindications:
	  ○ Low systemic blood pressure (systolic blood pressure less than 90)
	  ○ Low cardiac index
	  OR
	  ○ Presence of severe symptoms (functional class IV)

Appropriate Treatment Regimen:	• Documentation of inadequate response or intolerance to the following therapy classes is required:
	  ○ PDE5 inhibitors **AND**
	  ○ Endothelin receptor antagonists (exception WHO Functional Class IV)
	
	<u>Reauthorization</u> requires documentation of treatment success defined as one or more of the following:
	• Improvement in walking distance
	• Improvement in exercise ability
	• Improvement in pulmonary function
	• Improvement or stability in WHO functional class

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

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved and compendia-supported uses not otherwise excluded by plan design as follows: ○ Primary immunodeficiency (PID)/Wiskott - Aldrich syndrome ○ Idiopathic thrombocytopenia purpura (ITP) ○ Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) ○ Guillain-Barre Syndrome (Acute inflammatory polyneuropathy) ○ Multifocal Motor Neuropathy ○ Pediatric HIV: Bacterial control or prevention ○ Myasthenia Gravis ○ Dermatomyositis/Polymyositis ○ Complications of transplanted solid organ (kidney, liver, lung, heart, pancreas) and bone marrow transplant ○ Stiff-Person Syndrome ○ Allogeneic Bone Marrow or Stem Cell Transplant ○ Kawasaki's disease (Pediatric) ○ Fetal alloimmune thrombocytopenia (FAIT) ○ Hemolytic disease of the newborn ○ Auto-immune Mucocutaneous Blistering Diseases ○ Chronic lymphocytic leukemia with associated hypogammaglobulinemia (CLL) ○ Toxic Shock Syndrome ○ Pediatric Acute-Onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS)
Initial Approval Criteria:	**<u>Primary immunodeficiency (PID)/Wiskott - Aldrich syndrome:</u>** Includes but not limited to: X-linked agammaglobulinemia, common variable immunodeficiency (CVID), transient hypogammaglobulinemia of infancy, IgG subclass deficiency with or without IgA deficiency, antibody deficiency with near normal

	immunoglobulin levels) and combined deficiencies (severe combined immunodeficiencies, ataxia-telangiectasia, x-linked lymphoproliferative syndrome)
	• Documentation of one of the following:
	  ○ IgG level less than 200
	  ○ 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:
	  ▪ Four or more ear infections within 1 year
	  ▪ Two or more serious sinus infections within 1 year
	  ▪ Two or more months of antibiotics with little effect
	  ▪ Two or more pneumonias within 1 year
	  ▪ Recurrent or deep skin abscesses
	  ▪ Need for intravenous antibiotics to clear infections
	  ▪ Two or more deep-seated infections including septicemia
	AND
	• Documentation showing a deficiency in producing antibodies in response to vaccination including all the following:
	  ○ Titers that were drawn before challenging with vaccination
	  ○ Titers that were drawn between 4 and 8 weeks after vaccination
	
	Idiopathic thrombocytopenia purpura (ITP):
	
	For Acute disease state:
	• Documented use to manage acute bleeding due to severe thrombocytopenia (platelet counts less than 30,000/microliter)
	OR
	• To increase platelet counts prior to invasive surgical procedures, such as splenectomy (platelet count less than 100,000/microliter)
	OR
	• Documented severe thrombocytopenia (platelet count less than 20,000/microliter) and is considered to be at risk for intracerebral hemorrhage
	
	Chronic Immune Thrombocytopenia (CIT):
	• Documentation of increased risk for bleeding as indicated by a platelet count less than 30,000/microliter

	• History of failure, contraindication, or intolerance with corticosteroids • Duration of illness more than 6 months **<u>Chronic Inflammatory Demyelinating Polyneuropathy (CIDP):</u>** • Documented baseline in strength/weakness using objective clinical measuring tool (INCAT, Medical Research Council (MRC) muscle strength, 6 MWT, Rankin, Modified Rankin) • Documented disease course is progressive or relapsing and remitting for 2 months or longer • Abnormal or absent deep tendon reflexes in upper or lower limbs • Electrodiagnostic testing indicating demyelination with one of the following: o Motor distal latency prolongation in 2 nerves o Reduction of motor conduction velocity in 2 nerves o Prolongation of F-wave latency in 2 nerves o Absence of F-waves in at least 1 nerve o Partial motor conduction block of at least 1 motor nerve o Abnormal temporal dispersion in at least 2 nerves o Distal CMAP duration increase in at least 1 nerve • Cerebrospinal fluid (CSF) analysis indicates all the following (if electrophysiologic findings are nondiagnostic): o CSF white cell count of less than 10 cells/mm3 o CSF protein is elevated (greater than 45 mg/dL) • Refractory to or intolerant of corticosteroids (prednisolone, prednisone) given in therapeutic doses over at least three months **<u>Guillain-Barre Syndrome (Acute inflammatory polyneuropathy):</u>** • Documentation that the disease is severe (aid required to walk) • Onset of symptoms are recent (less than 1 month) **<u>Multifocal Motor Neuropathy (MMN):</u>** • Slowly progressive or stepwise progressive, focal, asymmetric limb weakness over at least one month

	• Partial conduction block or abnormal temporal dispersion conduction must be present in at least 2 nerves
	• Absence of upper motor neuron signs and bulbar involvement
	• Baseline in strength/weakness has been documented using objective clinical measuring tool (e.g., Inflammatory Neuropathy Cause and Treatment (INCAT) Disability Score, Medical Research Council (MRC) muscle strength, 6 Minute walk test, Rankin, Modified Rankin
	
	<u>Pediatric HIV: Bacterial control or prevention:</u>
	
	• Approved for those 13 years of age and younger with HIV diagnosis
	• Documented hypogammaglobulinemia (IgG less than 400 mg/dL)
	OR
	• Functional antibody deficiency as demonstrated by either poor specific antibody titers or recurrent bacterial infections
	
	<u>Myasthenia Gravis:</u>
	
	• Documented myasthenic crisis (impending respiratory or bulbar compromise)
	• Documented use for an exacerbation (difficulty swallowing, acute respiratory failure, functional disability leading to discontinuation of physical activity)
	• Documented failure with conventional therapy alone (azathioprine, cyclosporine and/or cyclophosphamide)
	
	<u>Dermatomyositis/Polymyositis:</u>
	
	• Documented severe active disease state on physical exam
	• Documentation of at least two of the following:
	  ○ Proximal muscle weakness in all upper and/or lower limbs
	  ○ Elevated serum creatine kinase (CK) or aldolase level
	  ○ Interstitial lung disease (ILD)
	  ○ Skin findings such as Gottron papules, Gottron sign, heliotrope eruption, poikiloderma
	  ○ Nailfold abnormalities
	  ○ Hyperkeratosis and fissuring of palms and lateral fingers

	• Documented failure with a trial of corticosteroids (such as prednisone)
	• Documented failure with a trial of an immunosuppressant (methotrexate, azathioprine, cyclophosphamide)
	
	Complications of transplanted solid organ (kidney, liver, lung, heart, pancreas) and bone marrow transplant:
	
	Coverage is provided for one or more of the following:
	• Suppression of panel reactive anti-HLA antibodies prior to transplantation
	• Treatment of antibody mediated rejection of solid organ transplantation
	• Prevention of cytomegalovirus (CMV) induced pneumonitis
	
	Stiff-Person Syndrome:
	
	• Documented anti-GAD antibodies
	• Documented failure with at least 2 of the following treatments: benzodiazepines, baclofen, phenytoin, clonidine and/or tizanidine
	
	Allogeneic Bone Marrow or Stem Cell Transplant:
	
	• Approved in use for prevention of acute Graft- Versus- Host Disease (GVHD) or infection (such as cytomegalovirus)
	• Documentation that the bone marrow transplant (BMT) was allogeneic
	• Transplant was less than 100 days ago
	
	Kawasaki's Disease (Pediatric):
	
	• Diagnosis or suspected diagnosis of Kawasaki's disease
	• 13 years of age and under
	
	Fetal alloimmune thrombocytopenia (FAIT):
	
	• Documentation of one or more of the following:
	  ○ Previous FAIT pregnancy
	  ○ Family history of the disease
	  ○ Screening reveals platelet alloantibodies

	• Authorization is valid until delivery date only
	
	<u>Hemolytic disease of the newborn:</u>
	
	• Diagnosis or suspected diagnosis of hemolytic disease in newborn patient
	
	<u>Auto-immune Mucocutaneous Blistering Diseases:</u>
	
	• Diagnosis confirmed by biopsy of one of the following:
	  ○ Pemphigus vulgaris
	  ○ Pemphigus foliaceus
	  ○ Bullous Pemphigoid
	  ○ Mucous Membrane Pemphigoid (Cicatricial Pemphigoid)
	  ○ Epidermolysis bullosa aquisita
	  ○ Pemphigus gestationis (Herpes gestationis)
	  ○ Linear IgA dermatosis
	• Documented severe disease that is extensive and debilitating
	• Disease is progressive and refractory to a trial of conventional combination therapy with corticosteroids and immunosuppressive treatment (azathioprine, cyclophosphamide, mycophenolate mofetil)
	
	<u>Chronic lymphocytic leukemia (CLL) with associated hypogammaglobulinemia:</u>
	
	• Documentation of an IgG level less than 500 mg/dL
	• Documented history of recurrent or chronic infections that have required intravenous antibiotics or hospitalization
	
	<u>Toxic Shock Syndrome:</u>
	
	• Diagnosis or suspected diagnosis of toxic shock syndrome
	
	<u>Pediatric Acute-Onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS):</u>
	
	• A clinically appropriate trial of two or more less-intensive treatments was either not effective, not tolerated, or did not

	result in sustained improvement in symptoms, as measured by a lack of clinically meaningful improvement on a validated instrument directed at the patient's primary symptom complex. Treatments may be given concurrently or sequentially and may include: ○ Selective-serotonin reuptake inhibitor SSRI (e.g., fluoxetine, fluvoxamine, sertraline) ○ Behavioral therapy ○ Nonsteroidal anti-inflammatory (NSAID) (e.g., naproxen, diclofenac, ibuprofen) ○ Oral and IV corticosteroids (e.g., prednisone, methylprednisolone) • Documentation of a consultation with a pediatric subspecialist (or adult subspecialist for adolescents) and the consulted subspecialist and the patient's primary care provider recommend the treatment
Renewal Criteria:	**Primary immunodeficiency (PID)** • Renewal requires disease response as evidenced by a decrease in the frequency and/or severity of infections **Chronic Immune Thrombocytopenia (Chronic ITP or CIT)** • 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 **Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)** • Renewal requires documentation of a documented clinical response to therapy based on an objective clinical measuring tool (e.g., INCAT, Medical Research Council (MRC) muscle strength, 6 Minute walk test, Rankin, Modified Rankin) **Multifocal Motor Neuropathy (MMN)** • Renewal requires documentation that there has been a demonstrated clinical response to therapy based on an objective clinical measuring tool (INCAT, Medical Research Council (MRC) muscle strength, 6 Minute walk test, Rankin, Modified Rankin) **Pediatric HIV: Bacterial control or prevention** • 13 years of age or less **Dermatomyositis/Polymyositis** • Renewal requires documentation that CPK (Creatine

	phosphokinase) levels are lower and documentation of clinically significant improvement above baseline per physical exam
	Complications of transplanted solid organ (kidney, liver, lung, heart, pancreas) and bone marrow transplant
	• Renewal requires documentation of clinically significant disease response
	Stiff Person Disease
	• Renewal requires documentation of a clinically significant improvement over baseline per physical exam
	Allogeneic Bone Marrow or Stem Cell Transplant
	• Renewal requires documentation that the IgG is less than or equal to 400mg/dL; AND
	• Therapy does not exceed one year past date of allogeneic bone marrow transplantation
	Auto-immune mucocutaneous blistering diseases:
	• Renewal requires a documented clinically significant improvement over baseline per physical exam
	Chronic lymphocytic leukemia (CLL) with associated hypogammaglobulinemia
	• Renewal requires disease response as evidenced by a decrease in the frequency and/or severity of infections
	Pediatric Acute-Onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS)
	• Renewal requires all of the following:
	  ○ Documentation of a clinical reevaluation at three months after treatment initiation
	  ○ Documentation of clinically meaningful improvement in the results of clinical testing with a validated instrument (which must be performed pretreatment and posttreatment)

Dosing and Coverage Duration:

- Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced
- Authorization durations are as stated below, unless otherwise specified

Indication	Dose	Approval Duration
PID	Up to 800 mg/kg every 3 to 4 weeks	Initial: up to 3 months Reauthorization: up to 12 months
CIDP	2 g/kg divided over 2-5 days for one dose then maintenance dosing of 1 g/kg every 21 days	Initial: up to 3 months Reauthorization: up to 12 months
ITP	1 g/kg once daily for 1-2 days May be repeated monthly for chronic ITP	Acute ITP: • Approval: 1 month only Chronic ITP: • Initial: up to 3 months • Reauthorization: up to 12 months
FAIT	1 g/kg/week until delivery	Authorization is valid until delivery date only
Kawasaki's Disease (pediatric patients)	Up to 2 g/kg x 1 single dose	Approval: 1 month only
MMN	2 g/kg divided over 2-5 days in a 28-day cycle May be repeated monthly	Initial approval: 1 month Reauthorization: up to 12 months
CLL	400 mg/kg every 3 to 4 weeks	Approval: up to 6 months
Pediatric HIV	400 mg/kg every 28 days	Initial: up to 3 months Reauthorization: up to 12 months
Guillain-Barre	400 mg/kg once daily for 5 days	Approval: maximum of 2 rounds of therapy within 6 weeks of onset; 2 months maximum

	Myasthenia Gravis	Up to 2 g/kg x 1 dose (acute attacks)	Approval: 1 month (one course of treatment)
	Auto-immune blistering diseases	Up to 2 g/kg divided over 5 days in a 28-day cycle	Approval: up to 6 months
	Dermatomyositis /Polymyositis	Up to 2 g/kg given over 2-5 days in a 28-day cycle	Initial: up to 3 months Reauthorization: up to 6 months
	Allogeneic Bone Marrow or Stem Cell Transplant	500 mg/kg/week x 90 days, then 500 mg/kg/month up to one-year post-transplant	Initial: up to 3 months Reauthorization: until up to one-year post-transplant
	Complications of transplanted solid organ: (kidney, liver, lung, heart, pancreas) transplant	2 g/kg divided over 5 days in a 28-day cycle	Initial: up to 3 months Reauthorization: up to 12 months
	Stiff Person Syndrome	2 g/kg divided over 5 days in a 28-day cycle	Initial: up to 3 months Reauthorization: up to 12 months
	Toxic shock syndrome	1 g/kg on day 1, followed by 500 mg/kg once daily on days 2 and 3	Approval: 1 month (one course of treatment)
	Hemolytic disease of the newborn	1 g/kg x 1 dose, may be repeated once if needed	Approval: 1 month (one course of treatment)
	PANS/PANDAS	Each dose: Up to 2 g/kg divided over 2-5 days	Initial: up to 3 months (3 monthly doses) Reauthorization: up to 3 months (3 monthly doses) Total 6 monthly doses only
Prescriber/Site of Care Restrictions:	• Must be prescribed by a specialist for the condition being treated (such as neurologist, rheumatologist, immunologist, hematologist)		

	• All approvals are subject to utilization of the most cost-effective site of care

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Primary hyperlipidemia (including heterozygous familial hypercholesterolemia [HeFH])
	  ○ Secondary prevention in atherosclerotic cardiovascular disease (ASCVD)

Required Medical Information:	• Documentation of baseline (untreated) low-density lipoprotein cholesterol (LDL-C)
	
	<u>Primary Hyperlipidemia/HeFH</u>
	• Diagnosis confirmed by **ONE** of the following:
	  ○ Minimum baseline LDL-C of 160 mg/dL in adolescents or 190 mg/dL in adults **AND** 1 first-degree relative affected
	  ○ Presence of one abnormal LDL-C-raising gene defect (e.g., LDL receptor [LDLR], apolipoprotein B [apo B], proprotein convertase subtilisin kexin type 9 [PCSK9] loss-of-function mutation, or LDL receptor adaptor protein 1 [LDLRAP1])
	  ○ World Health Organization (WHO)/Dutch Lipid Network criteria score of at least 8 points
	  ○ Definite FH diagnosis per the Simon Broome criteria
	
	<u>Clinical ASCVD</u>
	• Documentation of established ASCVD, confirmed by at least **ONE** of the following:
	  ○ Acute coronary syndromes (ACS)
	  ○ History of myocardial infarction (MI)
	  ○ Stable or unstable angina
	  ○ Coronary or other arterial revascularization
	  ○ Stroke or transient ischemic attack
	  Peripheral artery disease (PAD) presumed to be of atherosclerotic origin

PacificSource HEALTH PLANS

<table>
<tr>
<td>Appropriate Treatment Regimen & Other Criteria:</td>
<td>
<u>All Indications</u>
• History of statin intolerance requires documentation of the following:
  ○ Minimum of two different statin trials
  ○ Documentation of statin-associated muscle symptoms, which stopped when statin therapy was discontinued and restarted when re-challenged
• History of statin-associated rhabdomyolysis requires documentation of elevation in creatinine kinase (CK) level to at least 10 times the upper limit of normal, in concurrence with statin use

<u>Primary Hyperlipidemia/HeFH</u>
• Documented treatment failure with minimum 12-week trial with ALL of the following, shown by inability to achieve LDL-C reduction of 50% or greater OR LDL-C less than 100 mg/dL:
  ○ Maximally tolerated statin therapy
  ○ Repatha

<u>Clinical ASCVD</u>
• Documented treatment failure with minimum 12 weeks of consistent statin therapy at maximally tolerated dose, as shown by ONE of the following:
  ○ Current LDL-C of at least 70 mg/dL
  ○ Current LDL-C of at least 55 mg/dL in patients at very high risk of future ASCVD events, based on history of multiple major ASCVD events OR 1 major ASCVD event + multiple high-risk conditions (see below)
• Documented treatment failure or intolerance to minimum 12-week trial of Repatha

<table>
<tr><th>Major ASCVD Events</th><th>High-Risk Conditions</th></tr>
<tr><td>• ACS within the past 12 months
• History of MI (distinct from ACS event)</td><td>• Age 65 years and older
• HeFH
• Prior coronary artery bypass or percutaneous</td></tr>
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	<table><tr><td>• Ischemic stroke • Symptomatic PAD</td><td>intervention (outside of major ASCVD events) • Diabetes • Hypertension • Chronic kidney disease • Current smoking • History of congestive heart failure</td></tr></table> **<u>Reauthorization</u>** requires documentation of treatment success and a clinically significant response to therapy as assessed by the prescribing provider
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:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
INEBILIZUMAB-CDON
Affected Medications: UPLIZNA (inebilizumab-cdon)

<table>
<tr><td>Covered Uses:</td><td>• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
o Neuromyelitis optica spectrum disorder (NMOSD) in adults who are anti-aquaporin-4 (AQP4) antibody positive</td></tr>
<tr><td>Required Medical Information:</td><td><u>NMOSD</u>
• Diagnosis of seropositive aquaporin-4 immunoglobulin G (AQP4-IgG) NMOSD confirmed by all the following:
o Documentation of AQP4-IgG-specific antibodies on cell-based assay
o Exclusion of alternative diagnoses (such as multiple sclerosis)
o At least one core clinical characteristic:
▪ Acute optic neuritis
▪ Acute myelitis
▪ Acute area postrema syndrome (episode of otherwise unexplained hiccups or nausea/vomiting)
▪ Acute brainstem syndrome
▪ Symptomatic narcolepsy OR acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesion on magnetic resonance imaging (MRI) [see table below]
▪ Acute cerebral syndrome with NMOSD-typical brain lesion on MRI [see table below]
<table><tr><th>Clinical presentation</th><th>Possible MRI findings</th></tr><tr><td>Diencephalic syndrome</td><td>• Periependymal lesion
• Hypothalamic/thalamic lesion</td></tr><tr><td>Acute cerebral syndrome</td><td>• Extensive periependymal lesion
• Long, diffuse, heterogenous, or</td></tr></table></td></tr>
</table>

	<table><tr><td></td><td>edematous corpus callosum lesion • Long corticospinal tract lesion • Large, confluent subcortical or deep white matter lesion</td></tr></table> • History of at least 1 attack in the past year, or at least 2 attacks in the past 2 years, requiring rescue therapy

Appropriate Treatment Regimen & Other Criteria:	• Documentation of inadequate response, contraindication, or intolerance to each of the following:
	  ○ Rituximab (preferred products: Riabni, Ruxience)
	  ○ Satralizumab-mwge (Enspryng)
	
	<u>Reauthorization</u> requires documentation of treatment success

Exclusion Criteria:	• Active Hepatitis B Virus (HBV) infection • Active or untreated latent tuberculosis • Concurrent use with other disease-modifying biologics for requested indication
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or neuro-ophthalmologist. • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved, or compendia supported, indications not otherwise excluded by plan design ○ Neovascular (Wet) Age-Related Macular Degeneration (AMD) ▪ Eylea, Eylea HD, Lucentis, Susvimo, Beovu, Vabysmo, Byooviz, Cimerli ○ Macular Edema Following Retinal Vein Occlusion (RVO) ▪ Eylea, Lucentis, Byooviz, Cimerli, Vabysmo ○ Diabetic Macular Edema (DME) ▪ Eylea, Eylea HD, Lucentis, Vabysmo, Beovu, Cimerli ○ Diabetic Retinopathy (DR) in patients with Diabetes Mellitus ▪ Eylea, Eylea HD, Lucentis, Cimerli ○ Myopic Choroidal Neovascularization (mCNV) ▪ Lucentis, Byooviz, Cimerli ○ Retinopathy of Prematurity (ROP) ▪ Eylea, 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>** • **Coverage for the non-preferred product Eylea is provided when one of the following criteria is met:** ○ Currently receiving treatment with Eylea, excluding when the product is obtained as samples or via manufacturer's patient assistance programs. ○ A documented inadequate response or intolerable adverse event with all the preferred products (Avastin, AND Byooviz or Cimerli)

	○ Documentation of treatment-naïve retinopathy of prematurity (ROP) in a preterm infant 32 weeks or younger • **AMD –** 2 mg (0.05 mL) every 4 weeks for the first 3 injections followed by 2 mg (0.05 mL) every 8 weeks ○ Continued every 4-week dosing requires documented clinical failure to every 8-week maintenance dosing • **RVO -** 2 mg (0.05 mL) every 4 weeks • **DME and DR –** 2 mg (0.05 mL) every 4 weeks for the first 5 injections followed by 2 mg (0.05 mL) every 8 weeks • **ROP** – 0.4 mg (0.01 mL) as a single injection per affected eye(s); dose may be repeated up to 2 times with a minimum treatment interval between doses of at least 10 days (maximum of 3 doses total) **<u>Eylea HD Dosing</u>** • **Coverage for the non-preferred product Eylea HD is provided when one of the following criteria is met:** ○ Currently receiving treatment with Eylea HD, excluding when the product is obtained as samples or via manufacturer's patient assistance programs. ○ A documented inadequate response or intolerable adverse event with all the preferred products (Avastin AND Byooviz or Cimerli) • **AMD and DME** – 8 mg (0.07 mL) every 4 weeks for the first 3 injections, followed by 8 mg (0.07 mL) every 8 to 16 weeks ○ Every 4-week dosing is limited to the first 3 injections only • **DR -** 8 mg (0.07 mL) every 4 weeks for the first 3 injections, followed by 8 mg (0.07 mL) every 8 weeks to 12 weeks ○ Every 4-week dosing is limited to the first 3 injections only **<u>Lucentis Dosing</u>** • **Coverage for the non-preferred product Lucentis is provided when the following criteria is met:**

	○ A documented inadequate response or intolerable adverse event with all of the preferred products (Avastin, Byooviz, and Cimerli) • **AMD and RVO –** maximum 0.5 mg every 4 weeks • **DME and DR –** 0.3 mg every 4 weeks • **mCNV-** 0.5 mg every 4 weeks for up to 3 months • **ROP** – 0.1 to 0.3 mg as a single injection in the affected eye(s); dose may be repeated up to 2 times with a minimum treatment interval between doses of 28 days (maximum of 3 doses total) **<u>Beovu Dosing</u>** • **Coverage for the non-preferred product Beovu is provided when either of the following criteria is met:** ○ Currently receiving treatment with Beovu, excluding when the product is obtained as samples or via manufacturer's patient assistance programs. ○ A documented inadequate response or intolerable adverse event with all the preferred products (Avastin, AND Byooviz or Cimerli) • **AMD** – 6 mg every month for the first three doses followed by 6 mg every 8 to 12 weeks • **DME** – 6 mg every six weeks for the first five doses followed by 6 mg every 8 to 12 weeks **<u>Susvimo Dosing</u>** • **Coverage for the non-preferred product Susvimo is provided when the following criteria is met:** ○ A documented inadequate response or intolerable adverse event with all of the preferred products (Avastin, Byooviz, and Cimerli) • Must be established on ranibizumab (Lucentis, Byooviz, or Cimerli) injections with response to treatment for a minimum of 6 months at standard dosing (0.5 mg every 4 weeks) • **AMD** – 2 mg administered continuously via ocular implant with refills every 24 weeks. **<u>Vabysmo Dosing</u>** • **Coverage for the non-preferred product Vabysmo is**

	provided when either of the following criteria is met: ○ Currently receiving treatment with Vabysmo, excluding when the product is obtained as samples or via manufacturer's patient assistance programs. ○ A documented inadequate response or intolerable adverse event with all the preferred products (Avastin, AND Byooviz or Cimerli) • **AMD** – 6 mg every 4 weeks for the first 4 injections followed by 6 mg every 8 to 16 weeks ○ Some patients may require continued every 4-week injections following the initial doses • **DME** ○ Fixed interval regimen: 6 mg every 4 weeks for the first 6 injections followed by 6 mg every 8 weeks ○ 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) ○ Some patients may require continued every 4-week injections following the initial doses • **RVO** - 6 mg (0.05 mL) every 4 weeks for up to 6 months **Reauthorization** 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 • Active intraocular inflammation
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an ophthalmologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	**Macular Edema Following Retinal Vein Occlusion (RVO) for Vabysmo** • Authorization: 6 months with no reauthorization, unless otherwise specified

	<u>Retinopathy of Prematurity (ROP)</u> • Authorization: 3 months with no reauthorization, unless otherwise specified **<u>All other indications</u>** • Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of geographic atrophy (GA) secondary to age-related macular degeneration (AMD)

Required Medical Information:	• Diagnosis of geographic atrophy (GA) secondary to age-related macular degeneration (AMD) confirmed by all the following:
	  ○ Fundus Autofluorescence (FAF) imaging showing:
	  ▪ Total GA area size between 2.5 and 17.5 mm^2
	  ▪ If GA is multifocal, at least 1 focal lesion that is 1.25 mm^2 or greater
	• Best-corrected visual acuity (BCVA) using Early Treatment Diabetic Retinopathy Study (ETDRS) charts
	  ○ Must be 24 letters or greater (approximately 20/320 Snellen equivalent)

Appropriate Treatment Regimen & Other Criteria:	• Dosing not to exceed:
	  ○ Every 25-day dosing for Syfovre
	  ○ Every 30-day dosing with a maximum duration of 12 months for Izervay
	
	<u>Reauthorization</u>:
	• **Syfovre** requires:
	  ○ Documentation of treatment success as determined by treating provider
	  ○ BCVA remains 24 letters or greater
	• **Izervay**:
	  ○ No reauthorization - maximum duration up to 12 months

Exclusion Criteria:	• Presence of choroidal neovascularization in the eye(s) receiving treatment
Age Restriction:	• 60 years of age and older for Syfovre • 50 years of age and older for Izervay

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an ophthalmologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher • 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. • 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. ○ Non-lymphocytic variants of HES will also require documented failure with at least 12 weeks of hydroxyurea prior to interferon-alfa approval. • Recent liver function tests, comprehensive metabolic panel, complete blood count with differential, TSH (within past 3 months) • Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course • Reauthorization: documentation of disease responsiveness to therapy
Appropriate Treatment Regimen & Other Criteria:	• Patients with preexisting cardiac abnormalities and/or advanced cancer: recent electrocardiogram • Chest X ray for patients with pulmonary disorders • Recent ophthalmologic exam at baseline for all patients • Uncontrolled severe mental health illness should be addressed before use and monitored during treatment
Exclusion Criteria:	• Autoimmune hepatitis • Decompensated liver disease
Age Restriction:	• Hepatitis B: greater than or equal to 1 year of age • Hepatitis C: greater than or equal to 3 years of age • All other indications greater than or equal to 18 years of age
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial approval: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Invasive aspergillosis
	  ○ Invasive mucormycosis

Required Medical Information:	• Diagnosis of invasive aspergillosis or invasive mucormycosis confirmed by one or more of the following:
	  ○ Sputum fungal staining and culture
	  ○ Biopsy showing aspergillosis or mucormycosis organisms
	  ○ Serum biomarkers such as galactomannan, beta-D-glucan assays, or polymerase chain reaction (PCR) testing

Appropriate Treatment Regimen & Other Criteria:	**<u>Aspergillosis</u>**
	• Documented treatment failure or intolerable adverse event with at least a 6-week trial of all of the following:
	  ○ Voriconazole
	  ○ Posaconazole
	
	<u>Mucormycosis</u>
	• Documented treatment failure or intolerable adverse event with at least a 6-week trial of one of the following:
	  ○ Amphotericin B (if request is for initial therapy)
	  ○ Posaconazole (if request is for oral step-down therapy after initial therapy)
	
	<u>Reauthorization</u> 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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 3 months, unless otherwise specified

POLICY NAME:
LAROTRECTINIB
Affected Medications: VITRAKVI (larotrectinib)

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 • Documentation of positive neurotrophic tyrosine receptor kinase (NTRK) gene-fusion without a known acquired resistance mutation, as determined by an FDA approved test
Appropriate Treatment Regimen & Other Criteria:	• Documentation of an intolerance to, or clinical rationale for avoidance of Rozlytrek (entrectinib) **Reauthorization** requires documentation of disease responsiveness to therapy
Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
LENACAPAVIR
Affected Medications: SUNLENCA (lenacapavir)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of human immunodeficiency virus type 1 (HIV-1) infection, in combination with other antiretrovirals, in heavily treatment-experienced adults with multidrug resistant HIV-1 infection failing their current antiretroviral regimen due to resistance, intolerance, or safety considerations

Required Medical Information:	• Documentation of multidrug resistance within at least 3 of the 4 following antiretroviral classes (as defined by resistance to at least 2 agents within each of the 3 classes), unless contraindicated or clinically significant adverse effects are experienced:
	  ○ Nucleoside reverse-transcriptase inhibitors (NRTIs)
	  ○ Non-nucleoside reverse-transcriptase inhibitors (NNRTIs)
	  ○ Protease inhibitors (PIs)
	  ○ Integrase strand transfer inhibitors (INSTIs)
	• Documentation of current (within the past 30 days) HIV-1 RNA viral load of at least 200 copies/mL

Appropriate Treatment Regimen & Other Criteria:	• Must be used in combination with an optimized background antiretroviral regimen that contains at least one agent demonstrating full viral susceptibility, as confirmed by resistance testing
	
	<u>Reauthorization</u> requires all of the following:
	• Treatment plan includes continued use of optimized background antiretroviral regimen
	• Documentation of treatment success, as evidenced by one of the following:
	  ○ Reduction in viral load from baseline or maintenance of undetectable viral load
	  ○ Absence of postbaseline emergence of lenacapavir resistance-associated mutations confirmed by resistance testing

Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an infectious disease or HIV specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Oral Tablet Initial Authorization: 1 month, unless otherwise specified • Injection Initial Authorization: 6 months, unless otherwise specified • Injection Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
LENIOLISIB
Affected Medications: JOENJA (leniolisib)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Activated phosphoinositide 3-kinase delta syndrome (APDS)

Required Medical Information:	• Documentation of an APDS-associated *PIK3CD/PIK3R1* mutation without concurrent use of immunosuppressive medication
	• Presence of at least one measurable nodal lesion on a CT or MRI scan
	• Documentation of both of the following:
	  ○ Nodal and/or extranodal lymphoproliferation
	  ○ History of repeated oto-sino-pulmonary infections and/or organ dysfunction (e.g., lung, liver)
	• Current weight (must be at least 45 kg)

Appropriate Treatment Regimen & Other Criteria:	• Females of reproductive potential should have pregnancy ruled out and use effective contraception during therapy **Reauthorization** will require documentation of treatment success as shown by both of the following: • Improvement in lymphoproliferation as measured by a change from baseline in lymphadenopathy • Normalization of immunophenotype as measured by the percentage of naïve B cells out of total B cells
Exclusion Criteria:	
Age Restriction:	• 12 to 75 years of age
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an immunologist, hematologist/oncologist, or specialist with experience in the treatment of APDS • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
LETERMOVIR
Affected Medications: PREVYMIS (letermovir)

Covered Uses:	• All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design
	  ○ Prophylaxis of cytomegalovirus (CMV) infection and disease in adult CMV-seropositive recipients [R+] of an allogeneic hematopoietic cell transplant (HSCT)
	  ○ Prophylaxis of CMV disease in high-risk adult patients undergoing kidney transplant

Required Medical Information:	• Has received an allogeneic hematopoietic stem cell transplant (HSCT) • Is cytomegalovirus CMV-seropositive OR • Has received a kidney transplant and is at high risk (Donor CMV-seropositive/Recipient CMV-seronegative [D+/R-] of CMV infection
Appropriate Treatment Regimen & Other Criteria:	• Documented trial and failure (or intolerable adverse event) with an adequate trial (at least 14 days) of at least one of the following: ganciclovir, valganciclovir, Foscarnet (HSCT only) **HSCT Dosing**: 480 mg (or 240 mg) once daily beginning between Day 0 and Day 28 post-transplantation and continued through Day 100 post-transplantation **Kidney transplant Dosing**: 480mg once daily beginning between Day 0 and Day 7 post kidney transplant for high-risk recipients (donor CMV-seropositive/recipient CMV-seronegative) and continue through day 200 post transplantation
Exclusion Criteria:	

Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an infectious disease provider or a specialist with experience in the prevention and treatment of CMV infection • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	**HSCT** • Authorization: 4 months, unless otherwise specified **Kidney Transplant** • Authorization: 7 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Endometriosis
	  ○ Uterine leiomyomata (fibroids)
	  ○ Central precocious puberty (CPP)
	• NCCN (National Comprehensive Cancer Network) indications level 2A or higher
	• Gender dysphoria

Required Medical Information:	**<u>Endometriosis:</u>**
	
	• Documentation of moderate to severe pain due to endometriosis
	
	<u>Uterine leiomyomata (fibroids):</u>
	
	• Documentation of all of the following:
	  ○ Preoperative anemia due to uterine leiomyomata (fibroids)
	  ○ Planning to undergo leiomyomata-related surgery in the next 6 months or less
	  ○ Planning to use in combination with iron supplements
	
	<u>Gender dysphoria:</u>
	
	• Documentation of all the following:
	  ○ Current Tanner stage 2 or greater OR baseline and current estradiol and testosterone levels to confirm onset of puberty
	  ○ Confirmed diagnosis of gender dysphoria that is persistent
	  ○ The patient has the capacity to make a fully informed decision and to give consent for treatment
	  ○ Any significant medical or mental health concerns are reasonably well controlled

	○ A comprehensive mental health evaluation has been completed by a licensed mental health professional (LMHP) and provided in accordance with the most current version of the World Professional Association for Transgender Health (WPATH) Standards of Care **<u>Central precocious puberty:</u>** • Documentation of CPP confirmed by basal luteinizing hormone (LH), follicle-stimulating hormone (FSH), and either estradiol or testosterone concentrations
Appropriate Treatment Regimen & Other Criteria:	**<u>Endometriosis:</u>** • Documentation of a trial and inadequate relief (or contraindication) after at least 3 months of both of the following first-line therapies: ○ Nonsteroidal anti-inflammatory drugs (NSAIDs) ○ Continuous (no placebo pills) hormonal contraceptives **<u>Central precocious puberty:</u>** • Approval of Fensolvi requires rationale for avoidance of Lupron and Supprelin LA
Exclusion Criteria:	• Undiagnosed abnormal vaginal bleeding • Management of uterine leiomyomata without intention of undergoing surgery. • Pregnancy or breastfeeding • Use for infertility (if benefit exclusion)
Age Restriction:	• Endometriosis and preoperative uterine leiomyomata: 18 years of age and older • Central precocious puberty (CPP): 11 years of age or younger (females), 12 years of age or younger (males)

Prescriber/Site of Care Restrictions:	• Gender Dysphoria: Diagnosis made and prescribed by, or in consultation with, a specialist in the treatment of gender dysphoria • All other indications: prescribed by, or in consultation with, an oncologist, endocrinologist, or gynecologist as appropriate for diagnosis • 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 • Endometriosis: 6 months, unless otherwise specified • All other diagnoses: 12 months, unless otherwise specified

POLICY NAME:
LEVOKETOCONAZOLE
Affected Medications: RECORLEV (levoketoconazole)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Cushing syndrome

Required Medical Information:	• Diagnosis of Cushing's syndrome due to one of the following:
	  ○ Adrenocorticotropic hormone (ACTH)-secreting pituitary adenoma (Cushing's disease)
	  ○ Ectopic ACTH secretion by a non-pituitary tumor
	  ○ Cortisol secretion by an adrenal adenoma
	• Mean 24-hour urine free cortisol (mUFC) greater than 1.5 times the upper limit of normal (ULN) for the assay (at least two measurements)

Appropriate Treatment Regimen & Other Criteria:	• Documentation confirming surgery is not an option **OR** previous surgery has not been curative
	• Documentation of **ONE** of the following:
	  ○ Clinical failure to maximally tolerated dose of oral ketoconazole for at least 8 weeks
	  ○ Intolerable adverse event to oral ketoconazole, and the adverse event was not an expected adverse event attributed to the active ingredient
	
	<u>Reauthorization</u> requires documentation of treatment success defined as mUFC normalization (i.e., less than or equal to the ULN)

Exclusion Criteria:	• Adrenal or pituitary carcinoma
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist, neurologist, or adrenal surgeon • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
LIFILEUCEL
Affected Medications: AMTAGVI (lifileucel)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Diagnosis of unresectable or Stage IV metastatic melanoma • 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 • ECOG PS of 0 or 1 • Left ventricular ejection fraction (LVEF) greater than 45% • Forced expiratory volume (FEV1) greater than 60% • New York Heart Association (NYHA) classification not more than Class I

Appropriate Treatment Regimen & Other Criteria:	• At least one resectable lesion (or aggregate of lesions resected) of 1.5 cm or more in diameter post-resection to generate tumor-infiltrating lymphocytes (TILs)
	• Disease progression after 1 or more prior systemic therapy including:
	  ○ a PD-1–blocking antibody; and
	  ○ if BRAF V600 mutation–positive, a BRAF inhibitor or BRAF inhibitor plus a MEK inhibitor

Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater • Melanoma of uveal or ocular origin • Untreated or active brain metastasis
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist. • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Authorization: 6 months (one dose per patient's lifetime), unless otherwise specified

POLICY NAME:
LONAFARNIB
Affected Medications: ZOKINVY (lonafarnib)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ To reduce risk of mortality in Hutchinson-Gilford Progeria Syndrome
	  ○ 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)
	OR
	• A diagnosis of processing-deficient Progeroid Laminopathies with one of the following:
	  ○ Heterozygous LMNA mutation with progerin-like protein accumulation
	  ○ Homozygous or compound heterozygous ZMPSTE24 mutations

Appropriate Treatment Regimen & Other Criteria:	• Documented height and weight, or body surface area (BSA) • Documentation of medication review and avoidance of drugs that significantly affect the metabolism of lonafarnib (e.g. strong or moderate CYP3A4 inhibitors/inducers) • Females of reproductive potential should have pregnancy ruled out and use effective contraception during treatment <u>Labs</u>: • Absolute Phagocyte Count (sum of absolute neutrophil count, bands, and monocytes) greater than 1,000/microliters • Platelets greater than 75,000/microliters (transfusion independent) • Hemoglobin greater than 9g/dl. <u>Dosing</u>: • Available as oral capsules: 50 mg, 75 mg • Initial, 115 mg/m2/dose twice daily for 4 months, then increase to 150 mg/m2/dose twice daily

	○ Do not exceed 115 mg/m2/dose twice daily when used in combination with a weak CYP3A4 inhibitor ○ Round all total daily doses to the nearest 25 mg increment <u>Reauthorization</u>: • Documentation of treatment success and initial criteria to be met.
Exclusion Criteria:	• Use for other progeroid syndromes or processing-proficient progeroid laminopathies • Concomitant use with strong or moderate CYP3A4 inhibitors/inducers, midazolam, lovastatin, atorvastatin, or simvastatin • Overt renal, hepatic, pulmonary disease or immune dysfunction • 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 • Reauthorization: 12 months

POLICY NAME:
LOTILANER
Affected Medications: XDEMVY

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Demodex blepharitis (DB)

Required Medical Information:	• Diagnosis of DB meeting both of the following criteria:
	  ○ Presence of erythema of the upper eyelid margin
	  ○ Presence of mites upon examination of eyelashes by light microscopy OR presence of collarettes on slit lamp examination
	• Documented trial and failure to oral ivermectin, 200 mcg/kg in a single dose and repeated at least once after 7 days

Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** may be given at least 12 months after the first treatment and will require documentation of treatment success and returned presence of mites or collarettes requiring retreatment
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an optometrist or ophthalmologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design ○ Treatment of sickle cell disease in adults and pediatric patients at least 12 years of age with a history of recurrent vaso-occlusive crises

Required Medical Information:	• Documentation of sickle cell disease confirmed by genetic testing to show the presence of βS/βS, βS/β0 or βS/β+ genotype as follows:
	  ○ Identification of significant quantities of HbS with or without an additional abnormal β-globin chain variant by hemoglobin assay
	  OR
	  ○ Identification of biallelic *HBB* pathogenic variants where at least one allele is the p.Glu6Val or p.Glu7Val pathogenic variant on molecular genetic testing
	  AND
	  ○ Patient does NOT have disease with more than two α-globin gene deletions
	• Documentation of severe disease defined as 2 or more severe vaso-occlusive crises (VOCs) or vaso-occlusive events (VOEs) within the previous year (4 events over 2 years will also meet this requirement)
	  ○ VOC/VOEs defined as an event requiring a visit to a medical facility for evaluation AND necessitating subsequent interventions such as opioid pain management, non-steroidal anti-inflammatory drugs, red blood cell (RBC) transfusions, which results in a diagnosis of such being documented due to one (or more) of the following:
	  ▪ Acute pain event
	  ▪ Acute chest syndrome
	  ▪ Priapism lasting more than 2 hours
	  ▪ Acute splenic sequestration
	  ▪ Acute hepatic sequestration
	• For patients under 18 years of age, the patient does not have a known and suitable (10/10) human leukocyte antigen (HLA)

	matched related donor willing to participate in an allogeneic hematopoietic stem cell transplant (HSCT) • Adequate bone marrow, lung, heart, and liver function to undergo myeloablative conditioning regimen • Confirmed HIV negative as confirmed by a negative HIV test prior to mobilization
Appropriate Treatment Regimen & Other Criteria:	• Able to provide the minimum recommended dose of Lyfgenia- 3 × 10^6 CD34+ cells/kg.
Exclusion Criteria:	• Previous treatment with gene therapy for sickle cell disease • Prior hematopoietic stem cell transplant (HSCT) • History of hypersensitivity to dimethyl sulfoxide (DMSO) or dextran 40
Age Restriction:	• 12 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months (one-time infusion), unless otherwise specified

POLICY NAME:
LUSUTROMBOPAG
Affected Medications: MULPLETA (lusutrombopag)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Thrombocytopenia in adult patients with chronic liver disease who are scheduled to undergo a procedure

Required Medical Information:	• Documentation of ALL the following:
	  ○ Planned procedure including date
	  ○ Baseline platelet count of less than 50,000/microliter

Appropriate Treatment Regimen & Other Criteria:	• Approved for one time 7-day dosing regimen
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist or gastroenterology/liver specialist
Coverage Duration:	• Authorization: 1 month (7 days of treatment), based on planned procedure date, unless otherwise specified

POLICY NAME:
MANNITOL
Affected Medications: BRONCHITOL (mannitol)

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? a. Additional testing should include evaluation of overall clinical lung status and respiratory function (e.g. 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: a. Increase in pulmonary exacerbations from baseline? 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		

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:
MARIBAVIR
Affected Medications: LIVTENCITY (maribavir)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ 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 • 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 **<u>Reauthorization:</u>** • 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)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. ○ Hypertrophic cardiomyopathy with left ventricular outflow tract obstruction
Required Medical Information:	• Documented diagnosis of obstructive hypertrophic cardiomyopathy (OHCM) • New York Heart Association (NYHA) class II or III symptoms • Left ventricular ejection fraction (LVEF) of 55% or greater prior to starting therapy • Valsalva left ventricular outflow tract (LVOT) peak gradient of 50 mmHg or greater at rest or with provocation, prior to starting therapy • Documentation of negative pregnancy test in females of reproductive potential
Appropriate Treatment Regimen & Other Criteria:	• Use of effective contraception in females of reproductive potential • Documented treatment failure with trial of a beta blocker, or if unable to tolerate (or contraindication to) beta blockers, trial with verapamil. **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 of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by a cardiologist or a specialist with experience in the treatment of obstructive hypertrophic cardiomyopathy • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
MAVORIXAFOR
Affected Medications: XOLREMDI (mavorixafor)

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

POLICY NAME:
MECASERMIN
Affected Medications: INCRELEX (mecasermin)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Severe primary insulin-like growth factor-1 (IGF-1) deficiency (Primary IGFD)
	  ○ 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. • 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. • Maintenance: Up to 0.12 mg/kg subcutaneously twice daily. • **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. • 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 • All approvals are subjects to utilization of the most cost-effective site of care
Coverage Duration:	• Approval: 12 months, unless otherwise specified

POLICY NAME:

MEDICAL NECESSITY

Affected Medications: Abilify MyCitea, Abrilada, Absorica, Absorica LD, Acanya, Aciphex, Actemra SQ, Acthar Gel, Acuvail, Acyclovix, Aczone, Adalimumab-adbm, Adalimumab-fkjp, Adalimumab-ryvk, Adapalene pads, Adcirca, Adlarity, Adlyxin, Admelog, Advicor, Adzenys ER, Adzenys XR, Aerospan, Afrezza, Aimovig, AirDuo, AirDuo Digihaler, Airsupra, Aklief, Allopurinol 200 mg tablet, Allzital, Alprazolam Dispersible, Alprazolam Intensol, Altoprev, Alvesco, Ameluz, Amitiza, Amjevita, Amphetamine ER suspension, Ampyra, Amrix, Amturnide, Amzeeq, Ancobon, Androgel, Androxy, Apadaz, APAP-Caff-Dihydrocodeine, Apidra, Aplenzin, Arazlo, Aripiprazole Dispersible, Armonair Digihaler, Armonair Respiclick, Arymo ER, Asacol HD, Asmanex, Asmanex HFA, Aspruzyo, Astepro solution, Atorvaliq, Aubagio, Auvelity, Aveed, Azathioprine tablet (75 mg, 100 mg), Azelex, Azesco, Azstarys, Baclofen Oral Suspension, Basaglar, Basaglar Tempo pen, Baxdela, Beconase, Belbuca, Beser, Bevespi Aerophere, Bexagliflozin, BiDil, Biifenac, Bimzelx, Bismuth Subcitrate-Metronidazole-Tetracycline, Brenzavvy, Breztri, Bridion, Brisdelle, Briviact, Bryhali, Budesonide 9 mg ER tablet, Bunavail, Bupap, Buphenyl, Bupropion XL 450 mg, Butisol, Butrans patch, Bydureon, Bydureon BCise, Byetta, Bynfezia, Byvalson, Cabtreo, Calcipotriene-Betamethasone Dipropionate suspension, Cambia, Capex shampoo, Capital-Codeine, Carac, Carbinoxamine 6 mg tablet, Carisoprodol-ASA, Carisoprodol-ASA-Codeine, CaroSpir, Carticel implant, Cataflam, Cephalexin 750 mg capsule, Cephalexin tablet, Cequa, Chlorpheniramine-Codeine, Chlorzoxazone 250 mg tablet, Cibinqo, Cimzia, Ciprodex OTIC, Cipro HC Otic, Clemastine syrup, Clindamycin Phosphate-Benzoyl Peroxide gel 1.2-2.5 %, Clindavix, Clobetasol ophthalmic suspension, Clobetex, Clonidine ER 0.17 mg tablet, Codar AR, Colazal, Conjupri, Consensi, Conzip, Copaxone, Coreg CR, Cosopt PF, Cotempla XR ODT, Coxanto, Crexont, Crinone, Cuprimine, Cuvposa, Cyanocobalamin Nasal Spray, Cyclobenzaprine ER, Cyclosporine in Klarity, Cyltezo, Dapagliflozin, Dapagliflozin-Metformin ER, Dartisla ODT, Debacterol, Degludec, Delzicol, Demser, Depen, DermacinRx Lexitral cream pack, Dermalid, Desonate gel, Desonide gel, Desonide lotion, DesRx gel, Dexilant, Dhivy, Dichlorphenamide, Diclofenac 1.3 % patch, Diclofenac Potassium capsule, Diclofenac Potassium packet, Diclofenac Potassium 25 MG tablet, Diclofenac Sod soln 1.5 % & Capsaicin cream 0.025 % 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, Emflaza Suspension, Enalapril oral solution, Enstilar foam, Entadfi, Entyvio SQ, Eohilia, Epaned, Epanova, Epclusa, Eprontia, Equetro, Ergomar, Esbriet, Eskata, Evzio, Exjade, Exservan, Extavia, Extina foam 2 %, Fabior

foam, Faslodex, Fenofibrate 120 mg, Fenortho, Firazyr, First-lansoprazole, Flector patch, Fleqsuvy, Flolipid, Flowtuss, Fluopar kit, Fluorouracil 0.5 % cream, Flurandrenolide, Forfivo XL, Fortamet, Fortesta gel, Fosamax Plus D, Fulyzaq, Furoscix, Gabacaine pak, Gabapal, Giazo, Gilenya, Gimoti, Gleevec, Gloperba, Glumetza, Glycate, Glycopyrrolate 1.5 mg tablet, Gocovri, Gonitro, GPL pak, Halog, Halcinonide cream, Harvoni, Harvoni pak, Helidac, Hemady, Hemangeol, Hetlioz capsule, Hulio, Humalog, Humalog Junior KwikPen, Humatin, Humira, Humulin, Humulin 70/30 KwikPen, Humulin N, Humulin R-100, Hycofenix, Hyrimoz (Sandoz), Ibsrela, Ibuprofen-Famotidine, Idacio, Igalmi, Iheezo, Ilumya, Imbruvica 70 mg capsule, Imbruvica 140 mg & 280 mg tablet, Imiquimod 3.75 %, Impeklo, Impoyz, Imvexxy, Inbrija, Indocin suppository, Indomethacin 20 mg capsule, Inflatherm kit, Inflatherm pak, Infugem, Ingrezza, Ingrezza Sprinkle, Innolet Insulin, Inpefa, Insulin Aspart, Insulin Aspart Protamine & Aspart 70/30, Insulin Degludec, Insulin Glargine, Insulin Glargine-yfgn, Insulin Lispro, Intrarosa, Invega ER, Invokamet, Invokamet XR, Invokana, Isordil Titradose, Isosorbide Dinitrate-Hydralazine, Isotretinoin 25 mg and 35 mg capsule, Iyuzeh, Jadenu, Jadenu sprinkle packet, Jentadueto, Jentadueto XR, Jublia, Jylamvo, Karbinal ER, Katerzia, Kazano, K-bicarb, Kenalog aerosol, Kenalog susp, Keragel, KeragelT, Kerydin, Kesimpta, Ketek, Ketorolac nasal spray, Keveyis, Kevzara, Kineret, Kisqali, Kisqali-Femara co-pak, Klisyri, Kombiglyze XR, Konvomep, Korlym, Lampit, Latuda, Lescol XL, Letairis, Levamlodipine, Levorphanol Tartrate, Lexette, Lexuss, Lialda, Libervant, Licart, Lido GB 300 kit, Lidostream, Lidotin Pak, Lifems, Likmez, Lipritin Pak, Liptruzet, Liraglutide, Lithostat, LMR Plus Lidocaine, Lodoco, Lofena, Lonhala Magnair, Loreev XR, Lucemyra, Luzu, Lybalvi, Lyrica, Lyrica CR tablet, Lyumjev, Lyumjev Kwikpen, Lyvispah, Meclofen, Meloxicam capsule, Mentax cream 1 %, Mesalamine DR 800 mg tablet, Metaclopramide disintegrating tablet, Metaxall, Metaxall CP, Metformin ER (OSM), Metformin solution, Methadone Intensol, MethylTESTOSTERone capsule, Metyrosine, Miebo, Mifepristone, Migraine pack, Minocycline ER, Minolira, Mitigare, Monocycline ER, MorphaBond, MorphaBond ER, Motegrity, Motofen, Motpoly XR, Mycapssa, Myfembree, Myhibbin, Myrbetriq, Mytesi, Nalocet, Namenda XR, Namzaric, Naprelan, Naproxen-Esomeprazole, Nascobal, Natesto gel, Neo-Synalar cream, Nesina, Nexiclon XR, 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, Noxafil, NuDiclo Solupak, Nuvakaan kit, Nuvakaan II kit, Nuvigil, Nuzyra, Ofloxacin tablet, Ohtuvayre, Olpruva, Olumiant, Olysio, Omeprazole-Sodium Bicarb, Omnaris, Omvoh SQ, Ondansetron 24 mg tablet, Onexton, Onfi, Onglyza, Onmel, Onyda XR, Onzetra Xsail, Oracea, Oralair, Orencia SQ, Ormalvi, Orphenadrine-Aspirin-Caffeine tablet, Orphengesic Forte, Ortikos, Oseni, Otrexup, Oxaprozin capsule, 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 tablet, Pennsaid solution, Pentican pak, Percocet, Pertzye, Pheburane,

Picato, Pioglitazone-Glimepiride, Pirfenidone 534 mg tablet, Pradaxa, Praluent, Prevacid SoluTab, Prevpac, Prialt, Prilo Patch, Prilopentin, Primlev, Primsol, Pristiq, ProAir Digihaler, Prolate, Prudoxin, Purified Cortrophin gel, Purixan, Qbrelis, Qbrexza, Qdolo, Qelbree, Qmiiz, QNASL, Qtern, Qudexy XR, QuilliChew ER, Quillivant XR, Quinixil, Quinosone, Qwo, Ranexa, Rasuvo, Rayos, Recarbrio, Reditrex, Relexxii, Relion Insulins, Relprevv, Reltone, Retin-A Micro pump gel (0.06 %, 0.08 %), Revatio, Reyvow, Rezvoglar, Rhofade, Ribasphere, Ridaura, Riomet, Riomet ER, Rocklatan, Ryaltris, Ryvent, Ryzodeg 70/30, Sabril, Samsca, Saphris, Sarafem, Savaysa, Saxagliptin-Metformin ER, Seconal, Seebri Neohaler, Seglentis, Segluromet, Semglee, Sensipar, Sernivo, Seysara, Siklos, Silenor, Sila III pak, Siliq subcutaneous injection, Simlandi, Simponi, Simvastatin suspension, Skelaxin, Skelid, Soaanz, Sofdra, Soliqua, Solodyn, Solosec, Soolantra, Sorilux, Sotyktu, Sovaldi, Sovaldi pak, Spevigo Subcutaneous, Spironolactone suspension, Sporanox solution, Spritam, Sprix, Steglatro, Steglujan, Striant, Striant buccal, Suboxone, Sumatriptan-Naproxen, Sure Result DSS premium pack, Symbyax, Sympazan, Symproic, Synalar, Syndros, Syprine, Taclonex suspension, Talicia, Taltz, Tanzeum, Targadox, Tascenso ODT, Tasoprol, Tavaborole, Tazarotene foam, Tecfidera, Technivie, Thalitone, Thiola, Thiola EC, Thyquidity, Ticlopidine, Tiglutik, Tiopronin, Tivorbex, Tolak, Tolsura, Topiramate ER, Tosymra, Tovet kit, Tracleer, Tradjenta, Tramadol oral solution, Tretinoin Microsphere Gel 0.08 %, Treximet, Tri-Luma, Trixylitral kit, Trokendi XR, Trudhesa, Trulance, Tudorza Pressair, Twyneo, Tyrvaya, Tyzeka, Tyzine, Ultravate, Ultresa, Uptravi, Ursodiol capsule (200 mg, 400 mg), Utibron Neohaler, Uzedy, Valsartan oral solution, Vanatol LQ, Vanos, Varophen, Vasotec, Vecamyl, Vectical, Velsipity, Veltassa, Venlafaxine Besylate ER, Veozah, Veramyst, Veregen, Verkazia, Versacloz, Vesicare LS, Vevye, Vexasyn, Vexasyn gel, Vfend oral suspension, V-Go, Viberzi, Vibramycin, Victrelis, Viekira, Vigafyde, Viibryd, Viibryd Starter Pack, Vimovo, Viokace, Vivlodex, Vogelxo, Voquezna dual pak, Voriconazole oral suspension, Vtol LQ solution, Vyzulta, Wakix, Wegovy, Winlevi, Wynorza, Xaciato, Xadago, Xartemis XR, Xatmep, Xcopri, Xelitral pack, Xeloda, Xelstrym, Xenazine, Xenleta, Xerese, Xermelo, Xhance, Ximino, Xtampza ER, Xultophy, Xyosted, Yosprala, Yuflyma, Yupelri, Yusimry, Zanaflex capsule, Zayzpret, Zcort, Zebutal, Zecuity, Zelnorm, Zembrace, Zenevix, Zepatier, Zetonna, Zileuton ER, Zinbryta, Zipsor, Zituvimet, Zituvio, Zolpak, Zolpidem capsule, Zolpimist, Zonalon, Zonisade, Zorvolex, ZTLido, Z-Tuss, Zyclara, Zymfentra, Zypitamag, Zytiga

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:	
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:
MEPOLIZUMAB
Affected Medications: NUCALA (mepolizumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Add-on maintenance treatment of patients with severe asthma aged 6 years and older with an eosinophilic phenotype
	  ○ Treatment of adult patients with eosinophilic granulomatosis with polyangiitis (EGPA)
	  ○ Treatment of patients aged 12 years and older with hypereosinophilic syndrome (HES)
	  ○ 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)

Required Medical Information:	**<u>Eosinophilic asthma</u>**
	• Diagnosis of severe asthma with an eosinophilic phenotype, defined by both of the following:
	  ○ Baseline eosinophil count of at least 150 cells/µL **OR** dependent on daily oral corticosteroids
	  AND
	  ○ FEV1 less than 80% at baseline or FEV1/FVC reduced by at least 5% from normal
	
	<u>EGPA</u>
	• Diagnosis of relapsing or refractory EGPA confirmed by all of the following:
	  ○ Chronic rhinosinusitis
	  ○ Asthma
	  ○ Blood eosinophilia (at least 1,500 cells/mcL and/or 10% eosinophils on differential) at baseline
	  ○ Diagnosis must be confirmed by a second clinical opinion
	• Documented relapsing disease while on the highest tolerated oral corticosteroid dose

	HES
	• Diagnosis of HES with all of the following:
	  ○ Blood eosinophil count greater than or equal to 1,000 cells/mcL
	  ○ Disease duration greater than 6 months
	  ○ At least 2 flares within the past 12 months
	  ○ Lab work showing Fip1-like1-platelet-derived growth factor receptor alpha (FIP1L1-PDGFRα) mutation negative disease
	  ○ Non-hematologic secondary HES (e.g., drug hypersensitivity, parasitic helminth infection, HIV infection, non-hematologic malignancy) has been ruled out
	• Documentation that disease is currently controlled on the highest tolerated glucocorticoid dose (defined as an improvement in clinical symptoms and a decrease in eosinophil count by at least 50% from baseline)
	
	CRSwNP
	• Documentation of both of the following:
	  ○ Diagnosis of chronic rhinosinusitis and has undergone prior bilateral total ethmoidectomy
	  ○ Indicated for revision sinus endoscopic sinus surgery due to recurrent symptoms of nasal polyps (such as nasal obstruction/congestion, bilateral sinus obstruction)

Appropriate Treatment Regimen & Other Criteria:	**Eosinophilic asthma** • Documented use of high-dose inhaled corticosteroid (ICS) plus a long-acting beta agonist (LABA) for at least three months with continued symptoms • Documentation of one of the following: ○ Documented history of 2 or more asthma exacerbations requiring oral or systemic corticosteroid treatment in the past 12 months while on combination inhaler treatment with at least 80% adherence

	○ Documentation that chronic daily oral corticosteroids are required **<u>EGPA</u>** • Documented treatment failure or contraindication to at least two oral immunosuppressant drugs (azathioprine, methotrexate, mycophenolate) for at least 12 weeks each **<u>HES</u>** • Documented treatment failure or contraindication to at least 12 weeks of hydroxyurea (not required if patient has a lymphocytic variant of HES [L-HES]) • Documented treatment failure with interferon alfa **<u>CRSwNP</u>** • Documented treatment failure with at least 1 intranasal corticosteroid (such as fluticasone) after ethmoidectomy • Documented treatment failure with Sinuva implant **<u>Reauthorization</u>:** documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Use in combination with another monoclonal antibody (e.g., Dupixent, Fasenra, Xolair, Cinqair, Tezspire)
Age Restriction:	• **<u>Eosinophilic asthma</u>**: 6 years of age and older • **<u>EGPA</u>**: 18 years of age and older • **<u>HES</u>**: 12 years of age and older • **<u>CRSwNP</u>**: 18 years of age and older
Prescriber/Site of Care Restrictions:	• **<u>Eosinophilic asthma</u>**: prescribed by, or in consultation with, an allergist, immunologist, or pulmonologist • **<u>EGPA</u>**: prescribed by, or in consultation with, a specialist in the treatment of EGPA (such as an immunologist or rheumatologist) • **<u>HES</u>**: prescribed by, or in consultation with, a specialist in the treatment of HES (such as an immunologist or hematologist) • **<u>CRSwNP</u>**: prescribed by, or in consultation with, an otolaryngologist

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

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

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

Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial approval: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
METRELEPTIN
Affected Medications: MYALEPT (metreleptin)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Congenital or acquired generalized lipodystrophy as a result of leptin deficiency
Required Medical Information:	• Current weight • Baseline serum leptin levels, hemoglobin A1c (HbA1c), fasting glucose, fasting triglycerides, fasting serum insulin • Prior Myalept use will require testing for anti-metrepeptin antibodies
Appropriate Treatment Regimen & Other Criteria:	• Documented leptin deficiency and at least ONE of the following: **<u>Generalized lipodystrophy with concurrent hypertriglyceridemia</u>** • Triglycerides of 500 mg/dL or higher despite optimized therapy with at least two triglyceride-lowering agents from different classes (e.g., fibrates, statins) at maximum tolerated doses **<u>Generalized lipodystrophy with concurrent diabetes</u>** • Persistent hyperglycemia (HbA1c 7 percent or greater) despite dietary intervention and optimized insulin therapy at maximally tolerated doses **<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy documented by increased metabolic control defined by improvement in HbA1c, fasting glucose, and fasting triglyceride levels
Exclusion Criteria:	• Partial lipodystrophy • General obesity not associated with leptin deficiency • HIV-related lipodystrophy • Metabolic disease, including diabetes mellitus and hypertriglyceridemia, without concurrent documentation of generalized lipodystrophy

Age Restriction:	• 1 year of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist • All approvals are subjects to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Paget's disease of bone
	  ○ Hypercalcemia

Required Medical Information:	**<u>Hypercalcemia</u>** • Documented calcium level greater than or equal to 14 mg/dL (3.5 mmol/L) **<u>Paget's disease of bone</u>** • Documented baseline radiographic findings of osteolytic bone lesions • Abnormal liver function test (LFT), including alkaline phosphatase • Documented lack of malignancy within the past 3 months

Appropriate Treatment Regimen & Other Criteria:	**<u>Hypercalcemia</u>**
	• Documentation that additional methods for lowering calcium (such as intravenous fluids) did not result in adequate efficacy
	OR
	• Clinical judgement necessitated immediate administration without waiting for other methods to show efficacy
	
	<u>Paget's disease of bone</u>
	• Documented trial and failure (or intolerable adverse event) with an adequate trial of both of the following:
	  ○ Zoledronic acid (at least one dose)
	  ○ Oral bisphosphonate (e.g., alendronate, risedronate) for at least 8 weeks
	OR
	• Documentation that the patient has severe renal impairment (e.g., creatinine clearance less than 35 mL/min)
	AND
	• Documentation of all of the following:
	  ○ Normal vitamin D and calcium levels and/or supplementation

	○ Symptoms that necessitate treatment with medication (e.g., bone pain, bone deformity) **<u>Reauthorization – Paget's disease of bone:</u>** • 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 ○ History of a skeletal malignancy or bone metastases ○ Concurrent use of zoledronic acid or oral bisphosphonates ○ Asymptomatic Paget's Disease of the bone • Treatment or prevention of osteoporosis
Age Restriction:	• 18 years of age or older - for Paget's disease of bone only
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
MIGLUSTAT
Affected Medications: MIGLUSTAT

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of adult patients with mild to moderate type 1 Gaucher disease

Required Medical Information:	• Diagnosis of Gaucher disease confirmed by **ONE** of the following:
	  ○ An enzyme assay demonstrating a deficiency of beta-glucocerebrosidase enzyme activity
	  ○ Detection of biallelic pathogenic variants in the GBA gene by molecular genetic testing
	• Enzyme replacement therapy is not a therapeutic option (e.g., due to allergy, hypersensitivity, or poor venous access)

Appropriate Treatment Regimen & Other Criteria:	**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:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist in the management of Gaucher disease (hematologist, oncologist, hepatologist, geneticist or orthopedic specialist) • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
MILTEFOSINE
Affected Medications: IMPAVIDO (miltefosine)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of the following in adults and pediatric patients 12 years of age and older weighing greater than or equal to 30 kg (66 lbs):
	  ▪ Visceral leishmaniasis caused by *Leishmania donovani*
	  ▪ Cutaneous leishmaniasis caused by *Leishmania braziliensis*, *Leishmania guyanensis*, and *Leishmania panamensis*
	  ▪ Mucosal leishmaniasis caused by *Leishmania braziliensis*

Required Medical Information:	**<u>All Indications</u>** • Current weight **<u>Visceral Leishmaniasis</u>** • Documentation of diagnosis confirmed by smear or culture in tissue (usually bone marrow or spleen) **<u>Cutaneous and Mucosal Leishmaniasis</u>** • Documentation of diagnosis confirmed by histology, culture, or molecular analysis via polymerase chain reaction (PCR)
Appropriate Treatment Regimen & Other Criteria:	**<u>Dosing:</u>** • 30 to 44 kg: 50 mg twice daily for 28 days • 45 kg or greater: 50 mg three times daily for 28 days
Exclusion Criteria:	• Pregnancy • Sjögren-Larsson syndrome • Weight less than 30 kg (66 lbs) • Treatment of leishmaniasis outside of the visceral, cutaneous, or mucosal settings • Treatment of other *Leishmania* species

Age Restriction:	• 12 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an infectious disease specialist • All approvals are subjects to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 1 month, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Hemolytic anemia

Required Medical Information:	• Diagnosis of pyruvate kinase deficiency (PKD), defined by **ALL** the following:
	  ○ Presence of at least two mutant alleles in the pyruvate kinase liver and red blood cell (PKLR) gene
	  ○ At least one of the mutant alleles is a missense mutation
	• Documentation of ONE of the following:
	  ○ Receiving regular transfusions:
	  - A minimum of 6 transfusion episodes in the 12-month period prior to treatment AND
	  - Baseline transfusion amount, including date of transfusion and number of red blood cell (RBC) units transfused
	OR
	  ○ Not receiving regular transfusions:
	  - No more than 4 transfusions in the 12-month period prior to treatment and no transfusions in the 3-month period prior to treatment AND
	  - Baseline hemoglobin (Hb) must be less than or equal to 10 g/dL

Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy, defined as: • For patients receiving regular transfusions at baseline: must document greater than or equal to a 33% reduction in RBC units transfused compared to baseline • 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 • Discontinue therapy after 6 months if no benefit in transfusion

<table>
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<td>requirement or Hb has been observed

• Dose: Approve 5 mg, 20 mg, and 50 mg tablets (QL of 56 per 28 days) per dosing schedule below

Table 1: Dose Titration Schedule
<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:
• Increase to 20 mg twice daily and maintain for 4 weeks.
If Hb is within normal range and patient has not required a transfusion within the last 8 weeks:
• 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:
• Increase to 50 mg twice daily and maintain thereafter.
If Hb is within normal range and patient has not required a transfusion within the last 8 weeks:
• 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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<tr>
<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
• Splenectomy scheduled during treatment or have undergone within the 12-month period prior to starting treatment
• Previous bone marrow or stem cell transplant
• Receiving hematopoietic stimulating agents or anabolic steroids (including testosterone preparations) within 28 days prior to treatment</td>
</tr>
</table>

Age Restriction:	• Must be 18 years or older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of chronic rhinosinusitis with nasal polyps in patients who have had ethmoid sinus surgery
Required Medical Information:	• Documentation of a diagnosis of chronic rhinosinusitis and has undergone prior bilateral total ethmoidectomy • Indication for revision endoscopic sinus surgery due to recurrent symptoms of nasal polyps (such as nasal obstruction/congestion, bilateral sinus obstruction)
Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure to an adequate trial (minimum of 3 months each) with two nasal corticosteroid sprays • Documented treatment failure of a minimum 14-day trial with an oral corticosteroid **Reauthorization:** documented presence of ethmoid sinus polyps, grade 1 or higher, at least 90 days after previous treatment with Sinuva
Exclusion Criteria:	• Known history of resistant or poor response to oral steroids • Acute bacterial or invasive fungal sinusitis • Immune deficiency (including cystic fibrosis)
Age Restriction:	• 18 years of age or older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an otolaryngologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 1 month, unless otherwise specified • Reauthorization: 1 month, unless otherwise specified

POLICY NAME:
MONOMETHYL FUMARATE
Affected Medications: BAFIERTAM (monomethyl fumarate)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive disease (SPMS)

Required Medical Information:	• Diagnosis confirmed with magnetic resonance imaging (MRI), per revised McDonald diagnostic criteria for MS ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS

Appropriate Treatment Regimen & Other Criteria:	**<u>Relapsing forms of MS</u>**
	• Coverage of Bafiertam (monomethyl fumarate) requires documentation of one of the following:
	  ○ Documented disease progression or intolerable adverse event with one of the following: teriflunomide, dimethyl fumarate or fingolimod
	  ○ Currently receiving treatment with Bafiertam (monomethyl fumarate), excluding via samples or manufacturer's patient assistance program
	
	<u>Reauthorization</u> requires provider attestation of treatment success

Exclusion Criteria:	• Concurrent use of other disease-modifying medications indicated for the treatment of multiple sclerosis
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or a multiple sclerosis specialist

	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified.

POLICY NAME:
MOTIXAFORTIDE
Affected Medications: APHEXDA (motixafortide)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan ○ In combination with filgrastim (granulocyte colony-stimulating factor [G-CSF]) to mobilize hematopoietic stem cells (HSCs) to the peripheral blood circulation to facilitate their collection for subsequent autologous stem cell transplantation (ASCT) in patients with multiple myeloma (MM) • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better (autologous HSCT must be NCCN recommended)
Required Medical Information:	• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course • Documentation of diagnosis of multiple myeloma in first or second remission • Eligible for Autologous stem cell transplantation (ASCT) • At least 7 days from most recent high dose induction therapy • No single agent chemotherapy or maintenance therapy within 7 days • Eastern Cooperative Oncology Group (ECOG) performance status (PS) of 0 or 1

Appropriate Treatment Regimen & Other Criteria:	• Inadequate stem cell collection amount despite previous trial with ALL the following:
	  ○ Single agent granulocyte colony stimulating factor (G-CSF)
	  ○ G-CSF in combination with plerixafor
	
	<u>No reauthorization</u>

Exclusion Criteria:	• Karnofsky Performance Status 50% or less or Eastern Cooperative Oncology Group (ECOG) performance status (PS) of 2 or greater

Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 2 months unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Vimizim: Mucopolysaccharidosis type IVA (MPS IVA; Morquio A syndrome) ○ Naglazyme: Mucopolysaccharidosis type VI (MPS VI, Maroteaux-Lamy syndrome) ○ Mepsevii: Mucopolysaccharidosis VII (MPS VII; Sly Syndrome) ○ Aldurazyme: ▪ Hurler Mucopolysaccharidosis type I (MPS I H) ▪ Herler-Scheie Mucopolysaccharidosis type I (MPS I H/S) ▪ Scheie form of Mucopolysaccharidosis (MPS I S) with moderate to severe symptoms ○ Elaprase: Mucopolysaccharidosis type II (MPS II; Hunters syndrome)
Required Medical Information:	• Diagnosis of specific MPS type confirmed by enzyme assay showing deficient activity of the relevant enzyme **OR** detection of pathogenic mutations in the relevant gene by molecular genetic testing, as follows: ○ For Vimizim: N-acetylgalactosamine 6-sulfatase (GALNS) enzyme or GALNS gene ○ For Naglazyme: N-acetylgalactosamine 4-sulfatase (ASB) enzyme or Arylsulfatase B (ARSB) gene ○ For Mepsevii: beta-glucuronidase (GUSB) enzyme or GUSB gene ○ For Aldurazyme: alpha-L-iduronidase (IDUA) enzyme or IDUA gene ○ For Elaprase: iduronate 2-sulfatase (I2S or IDS) enzyme or IDS gene • Documented clinical signs and symptoms of MPS, such as soft tissue abnormality, skeletal abnormality, joint abnormality, respiratory disease, gait abnormality, motor issues, or cardiac

	abnormality
	• Baseline value for one or more of the following:
	  ○ Function test such as the Bruininks-Oseretsky Test of Motor Proficiency (BOT-2), 6-minute walk test (6MWT), three-minute stair climb test (3-MSCT), or pulmonary function tests (PFTs)
	  ○ Liver and/or spleen volume
	  ○ Urinary glycosaminoglycan (GAGs) level

Appropriate Treatment Regimen & Other Criteria:	• Dose does not exceed the recommended dosing according to the FDA label • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced **Reauthorization** requires documentation of treatment success defined as **ONE** or more of the following: • Stability or improvement in function tests such as BOT-2, 6MWT, 3-MSCT, **or** PFTs • Reduction in liver and/or spleen volume • Reduction in urinary GAG level • Other clinically significant improvement in MPS signs and symptoms
Exclusion Criteria:	• Treatment of central nervous system manifestation of the disorder • Severe, irreversible cognitive impairment
Age Restriction:	• Vimizim and Naglazyme: 5 years of age and older • Elaprase: 16 months of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist in the treatment of inherited metabolic disorders, such as a geneticist or endocrinologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

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

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

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

	Patients With Severe Chronic Neutropenia • 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. **Patients Acutely Exposed to Myelosuppressive Doses of Radiation (Hematopoietic Syndrome of Acute Radiation Syndrome) (Neupogen)** • Indicated to increase survival in patients acutely exposed to myelosuppressive doses of radiation. **Leukine** **Use Following Induction Chemotherapy in Acute Myelogenous Leukemia** • 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. **Use in Mobilization and Following Transplantation of Autologous Peripheral Blood Progenitor Cells** • Indicated for the mobilization of hematopoietic progenitor cells into peripheral blood for collection by leukapheresis. **Use in Myeloid Reconstitution After Autologous Bone Marrow Transplantation** • 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). **Use in Myeloid Reconstitution After Allogeneic Bone Marrow Transplantation** • Indicated for acceleration of myeloid recovery in patients undergoing allogeneic BMT from human leukocyte antigen (HLA)-matched related donors.

	<u>Use in Bone Marrow Transplantation Failure or Engraftment Delay</u> • Indicated in patients who have undergone allogeneic or autologous BMT in whom engraftment is delayed or has failed. **Neulasta, Fulphila, Udenyca, Ziextenzo, Nyvepria, Fylnetra, Stimufend, and Rolvedon** **<u>Patients with Cancer Receiving Myelosuppressive Chemotherapy</u>** • 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. **<u>Patients with Hematopoietic Subsyndrome of Acute Radiation Syndrome (Neulasta, Udenyca, Ziextenzo)</u>** • Indicated to increase survival in patients acutely exposed to myelosuppressive doses of radiation **Granix** • 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. **Compendia supported uses that will be covered (if applicable)** **Neupogen/Granix/Zarxio/Nivestym/Leukine:** • Treatment of chemotherapy-induced febrile neutropenia in patients with non-myeloid malignancies • Treatment of anemia in patients with myelodysplastic syndromes (MDS) • Treatment of neutropenia in patients with MDS • Following chemotherapy for acute lymphocytic leukemia (ALL) • Stem cell transplantation-related indications • Agranulocytosis • Aplastic anemia • Neutropenia related to human immunodeficiency virus (HIV) • 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 • Documentation of therapy intention (curative, palliative) for prophylaxis of febrile neutropenia • Documentation of patient specific risk factors for febrile neutropenia • Documentation of febrile neutropenia risk associated with the chemotherapy regimen • Documentation of planned treatment course • Documentation of current patient weight
Appropriate Treatment Regimen & Other Criteria:	**<u>Filgrastim products: Neupogen, Nivestym, Releuko, Zarxio, Granix</u>** **When requested via the MEDICAL benefit:** Coverage for the non-preferred products, Neupogen, Releuko and Granix, is provided when the member meets the following criteria: • Documented treatment failure or intolerable adverse event to **Zarxio and Nivestym** **When requested through the specialty PHARMACY benefit:** Coverage for the non-preferred products, Neupogen, Zarxio, Releuko and Granix, is provided when the member meets the following criteria: • Documented treatment failure or intolerable adverse event to **Nivestym** **<u>Sargramostim product: Leukine</u>** Coverage for the non-preferred product, Leukine, is provided when the member meets one of the following criteria: • Leukine will be used for myeloid reconstitution after autologous or allogenic bone marrow transplant or bone marrow transplant engraftment delay or failure • A documented treatment failure or intolerable adverse event to preferred products listed above **<u>Pegfilgrastim products: Neulasta, Fulphila, Udenyca, Ziextenzo, Nyvepria, Fylnetra, Stimufend, Rolvedon</u>**

	Coverage for the non-preferred products, Neulasta, Fylnetra, Rolvedon, Stimufend, Ziextenzo and Nyvepria is provided when the member meets the following criteria: • Documented treatment failure or intolerable adverse event to **Fulphila and Udenyca** **<u>Eflapegrastim product: Rolvedon</u>** Coverage for the non-preferred product, Rolvedon, is provided when the member meets the following criteria: • Documented treatment failure or intolerable adverse event to the preferred pegfilgrastim products **Fulphila and Udenyca** **<u>For prophylaxis of febrile neutropenia (FN) or other dose-limiting neutropenic events for patients receiving myelosuppressive anticancer drugs:</u>** Meets **ONE** of the following: • **Curative Therapy:** ○ High risk (greater than 20% risk) for febrile neutropenia based on chemotherapy regimen **OR** ○ Intermediate risk (10-20% risk) for febrile neutropenia based on chemotherapy regimen with documentation of significant patient risk factors for serious medical consequences **OR** ○ Has experienced a dose-limiting neutropenic event on a previous cycle of current chemotherapy to be continued • **Palliative Therapy:** ○ Myeloid growth factors will not be approved upfront for prophylaxis of febrile neutropenia in the palliative setting. Per the NCCN (National Comprehensive Cancer Network), chemotherapy regimens with a 20% or greater risk of neutropenic events should not be used. If however, a dose limiting neutropenic event occurs on a previous cycle of chemotherapy, and the effectiveness of chemotherapy will be reduced with dose reduction, growth factor will be approved for secondary prophylaxis on a case by case basis.

	<u>For Treatment of Severe Chronic Neutropenia</u> Must meet **<u>ALL</u>** the following: • Congenital neutropenia, cyclic neutropenia, OR idiopathic neutropenia • Current documentation of absolute neutrophil count (ANC) less than 500 cells/microliter • Neutropenia symptoms (fever, infections, oropharyngeal ulcers)
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist or hematologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 6 months, unless otherwise specified

POLICY NAME:
NAFARELIN
Affected Medications: SYNAREL (nafarelin)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Central Precocious Puberty (CPP)
	  ○ Endometriosis

Required Medical Information:	**<u>Central Precocious Puberty:</u>** • Documentation of CPP confirmed by basal luteinizing hormone (LH), follicle-stimulating hormone (FSH), and either estradiol or testosterone concentrations **<u>Endometriosis:</u>** • Documentation of moderate to severe pain due to endometriosis

Appropriate Treatment Regimen & Other Criteria:	**<u>Endometriosis:</u>**
	
	• Documentation of a trial and inadequate relief (or contraindication) after at least 3 months of both of the following first-line therapies:
	  ○ Nonsteroidal anti-inflammatory drugs (NSAIDs)
	  ○ Continuous (no placebo pills) hormonal contraceptives
	• Maximum treatment duration 6 months total

Exclusion Criteria:	• Use for infertility (if benefit exclusion) • Undiagnosed abnormal vaginal bleeding
Age Restriction:	• Endometriosis: 18 years of age and older • Central precocious puberty (CPP): 11 years of age or younger (females), 12 years of age or younger (males)
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist or gynecologist • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Endometriosis: 6 months (no reauthorization), unless otherwise specified • CPP: 12 months, unless otherwise specified

POLICY NAME:
NALOXEGOL
Affected Medications: MOVANTIK (naloxegol)

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

POLICY NAME:
NATALIZUMAB
Affected Medications: TYSABRI (natalizumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive multiple sclerosis (SPMS)
	  ○ Crohn's disease (CD)

Required Medical Information:	• Screening for anti-JC virus (JCV) antibodies prior to initiating Tysabri therapy **<u>Relapsing Forms of MS</u>** • Diagnosis confirmed with magnetic resonance imaging (MRI), per revised McDonald diagnostic criteria for MS ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS **<u>Crohn's disease</u>** • Moderate to severely active disease despite current treatment

Appropriate Treatment Regimen & Other Criteria:	**<u>Relapsing Forms of MS</u>**
	• Documentation of treatment failure (or documented intolerable adverse event) to:
	  ○ Rituximab (preferred biosimilar products: Riabni and Ruxience) **OR**
	  ○ Ocrevus (ocrelizumab) if previously established on treatment **OR**
	  ○ Documentation of pregnancy and severe disease
	
	<u>Crohn's disease</u>
	• 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** • Documented clinical failure with at least 12 weeks of infliximab (preferred biosimilar products: Inflectra and Renflexis) **Reauthorization:** • **Anti-JCV antibody negative**: documentation of positive clinical response to therapy • **Anti-JCV antibody positive**: documentation of positive clinical response to therapy and periodic MRI to monitor for progressive multifocal leukoencephalopathy (PML)
Exclusion Criteria:	• Current or prior history of PML • MS: concurrent use of other disease-modifying medications indicated for the treatment of multiple sclerosis • CD: concurrent use of other targeted immune modulators for the treatment of Crohn's disease
Age Restriction:	
Prescriber/Site of Care Restrictions:	• MS: prescribed by, or in consultation with, a neurologist or a MS specialist • CD: prescribed by, or in consultation with, a gastroenterologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	**Relapsing Forms of MS:** • Authorization: 12 months, unless otherwise specified **Crohn's Disease:** • Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
NAXITAMAB
Affected Medications: DANYELZA (naxitamab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of relapsed or refractory high-risk neuroblastoma in the bone or bone marrow (in combination with granulocyte-macrophage colony-stimulating factor [GM-CSF]) in patients who have demonstrated a partial response, minor response, or stable disease to prior therapy • 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.
	• Diagnosis of neuroblastoma as defined per the International Neuroblastoma Response Criteria (INRC):
	  ○ 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]
	  OR
	  ○ Evidence of metastases to bone marrow on an aspirate or trephine biopsy with concomitant elevation of urinary or serum catecholamines or their metabolites
	• Evidence of high-risk neuroblastoma, including:
	  ○ Stage 2/3/4/4S disease with amplified MYCN gene (any age)
	  ○ Stage 4 disease in patients greater than 18 months of age
	• Disease is evaluable in the bone and/or bone marrow, as documented by histology and/or appropriate imaging [e.g., metaiodobenzylguanidine (MIBG) scan and positron emission topography (PET) scan if MIBG is negative]
	• Documented history of previous treatment with at least one systemic therapy to treat disease outside of the bone or bone marrow

	• Documentation of clinical rationale for avoiding use of dinutuximab plus chemotherapy (if under 18 years of age)
Appropriate Treatment Regimen & Other Criteria:	• Must be used in combination with granulocyte-macrophage colony-stimulating factor (GM-CSF). **Reauthorization** will require documentation of disease responsiveness to therapy
Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater • 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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design ○ Prurigo nodularis (PN)

Required Medical Information:	• Documentation of all the following:
	  ○ Diagnosis confirmed by skin biopsy
	  ○ Presence of at least 20 PN lesions for at least 3 months
	  ○ Severe itching

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure with at least 2 weeks of a super high potency topical corticosteroid (such as clobetasol propionate 0.05%, halobetasol propionate 0.05%) • Documentation of treatment failure with at least 12 weeks of one of the following: phototherapy, methotrexate, cyclosporine • Documented treatment failure with at least 12 weeks of Dupixent (dupilumab)
Exclusion Criteria:	• Concurrent use with another therapeutic immunomodulator agent
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a dermatologist, allergist, or immunologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
NEONATAL FC RECEPTOR ANTAGONISTS
Affected Medications: VYVGART (efgartigimod alfa), VYVGART HYTRULO (efgartigimod alfa and hyaluronidase), RYSTIGGO (rozanolixizumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design **Vyvgart** o Generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive **Rystiggo** o Generalized myasthenia gravis (gMG) in adult patients who are AChR or anti-muscle-specific tyrosine kinase (MuSK) antibody positive **Vyvgart Hytrulo** o Generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive o Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)
Required Medical Information:	**<u>Myasthenia Gravis</u>** • Diagnosis of generalized Myasthenia Gravis (gMG) confirmed by one of the following: o A history of abnormal neuromuscular transmission test o A positive edrophonium chloride test o Improvement in gMG signs or symptoms with an acetylcholinesterase inhibitor • Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV • Positive serologic test for AChR or MuSK antibodies (for Rystiggo) • Documentation of **ONE** of the following: o MG-Activities of Daily Living (MG-ADL) total score of 6 or greater o Quantitative Myasthenia Gravis (QMG) total score of 12 or greater

	<u>CIDP (Vyvgart Hytrulo only)</u>
	• Documented baseline in strength/weakness using an objective clinical measuring tool (INCAT, Medical Research Council (MRC) muscle strength, 6 Minute Walk Test, Rankin, Modified Rankin)
	• Documented disease course is progressive or relapsing and remitting for 2 months or longer
	• Abnormal or absent deep tendon reflexes in upper or lower limbs
	• Electrodiagnostic evidence of demyelination indicated by one of the following:
	  ○ Motor distal latency prolongation in 2 nerves
	  ○ Reduction of motor conduction velocity in 2 nerves
	  ○ Prolongation of F-wave latency in 2 nerves
	  ○ Absence of F-waves in at least 1 nerve
	  ○ Partial motor conduction block of at least 1 motor nerve
	  ○ Abnormal temporal dispersion in at least 2 nerves
	  ○ Distal CMAP duration increase in at least 1 nerve
	• Cerebrospinal fluid (CSF) analysis indicates all of the following (if electrophysiologic findings are non-diagnostic):
	  ○ CSF white cell count of less than 10 cells/mm^3
	  ○ CSF protein is elevated (greater than or equal to 45mg/dL)

Appropriate Treatment Regimen & Other Criteria:	• Currently on a stable dose of at least one gMG therapy (acetylcholinesterase inhibitor, corticosteroid, or non-steroidal immunosuppressive therapy (NSIST)) that will be continued during initial treatment with Vyvgart, Vyvgart Hytrulo, or Rystiggo
	
	• Documentation of **ONE** of the following:
	  ○ Treatment failure with an adequate trial (one year or more) of at least 2 immunosuppressive therapies (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)
	  ○ Need for ongoing rescue therapy (at least 3 courses in the past 12 months) with plasmapheresis, plasma exchange, or intravenous immunoglobulin (IVIG) while consistently taking immunosuppressive therapy (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)

	Coverage for **Rystiggo** is provided when one of the following is met:
	• Currently receiving treatment with Rystiggo, excluding when the product is obtained as samples or via manufacturer's patient assistance programs
	• Documented treatment failure or intolerable adverse event with Vyvgart for AChR antibody positive gMG
	• Documented treatment failure with rituximab for MuSK antibody positive gMG (preferred products: Riabni, Ruxience)
	
	Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced
	
	Reauthorization:
	• Documentation of treatment success and clinically significant response to therapy defined as:
	  ○ A minimum 2-point reduction in MG-ADL score from baseline or improvement in QMG total score
	  ○ Absent or reduced need for rescue therapy compared to baseline
	• Documentation that the patient requires continuous treatment, after an initial beneficial response, due to new or worsening disease activity
	
	Note: a minimum of 50 days for Vyvgart/Vyvgart Hytrulo or 63 days for Rystiggo must have elapsed from the start of the previous treatment cycle
	
	CIDP (Vyvgart Hytrulo only)
	• Documented trial and failure of at least 3 months of intravenous or subcutaneous immune globulin
	
	Reauthorization:
	• Documentation of a clinical response to therapy based on an objective clinical measuring tool (e.g., INCAT, Medical Research Council (MRC) muscle strength, 6-Minute walk test, Rankin, Modified Rankin)

Exclusion Criteria:	• Immunoglobulin G (IgG) levels less than 600 mg/dL at baseline • Concurrent use with other disease-modifying biologics for the treatment of gMG
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
NILOTINIB
Affected Medications: TASIGNA (nilotinib)

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 • Documentation Philadelphia chromosome or BCR::ABL1-positive mutation status
Appropriate Treatment Regimen & Other Criteria:	• For patients with Chronic Myeloid Leukemia (CML) and low-risk score, documented clinical failure with imatinib **Reauthorization** requires documentation of disease responsiveness to therapy (as applicable, BCR-ABL1 transcript levels, cytogenetic response)
Exclusion Criteria:	• Karnofsky Performance Status 50% or less, ECOG performance score 3 or greater
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
NIROGACESTAT
Affected Medications: OGSIVEO (nirogacestat)

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

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

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

Renewal for hyaluronic acid (HA) after previous administration of HA product		
1. Is there documentation of treatment success that lasted at least 6 months from date of previous HA administration AND documented intolerable adverse event to Synvisc, Synvisc-One, and Orthovisc with date and description of reactions?	Yes – Go to #2	No – Criteria not met
2. Is the requested dose within the Food and Drug Administration (FDA)-approved label and PacificSource quantity limitations?	Yes – Approve up to 6 months	No – Criteria not met
Quantity Limitations		
• Durolane: 1 injection per course • Euflexxa: 3 injections per course • Gel-One: 1 injection per course • Gelsyn-3: 3 injections per course • GenVisc 850: 3 to 5 injections per course • Hyalgan: 5 injections per course • Hymovis: 2 injections per course • Monovisc: 1 injection per course • Supartz: 3 to 5 injections per course • Synojoynt: 3 injections per course • Triluron: 3 injections per course • Trivisc: 3 injections per course • Visco-3: 3 injections per course		

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

<table>
<tr><td>Covered Uses:</td><td>• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
• For oncology indications: National Comprehensive Cancer Network (NCCN) indications with evidence level of 2A or higher</td></tr>
<tr><td>Required Medical Information:</td><td></td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td>• Approval of a non-preferred medical drug listed below requires documentation of an intolerable adverse event to all the preferred alternatives, and the adverse event was not an expected adverse event attributed to the active ingredient

<table>
<tr><td>Drug</td><td>Non-Preferred code (Manufacturer)</td><td>Preferred Alternatives</td></tr>
<tr><td>Bortezomib (Velcade)</td><td>J9046 (Dr. Reddys)</td><td>J9041, J9048, J9049</td></tr>
<tr><td>Pemetrexed (Pemfexy, Alimta, Pemrydi RTU)</td><td>J9304 (Apotex)</td><td>J9294, J9296, J9297, J9305, J9314, J9324</td></tr>
</table>

<u>Reauthorization</u>: documentation of disease responsiveness to therapy</td></tr>
<tr><td>Exclusion Criteria:</td><td></td></tr>
<tr><td>Age Restriction:</td><td></td></tr>
<tr><td>Prescriber/Site of Care Restrictions:</td><td>• All approvals are subject to utilization of the most cost-effective site of care</td></tr>
<tr><td>Coverage Duration:</td><td>• Authorization: 12 months, unless otherwise specified</td></tr>
</table>

POLICY NAME:
NUEDEXTA
Affected Medications: NUEDEXTA (dextromethorphan hydrobromide/quinidine sulfate)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. ○ 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
	  ○ stroke.
	• Baseline Center for Neurologic Study-Lability Scale (CNS-LS) score of 13 or greater
	• Documentation of treatment failure to a 30-day trial of each of the following:
	  ○ serotonin reuptake inhibitor (SSRI)
	  ○ tricyclic antidepressant (TCA)

Appropriate Treatment Regimen & Other Criteria:	• Reauthorization requires documentation of treatment success defined as decreased frequency of pseudobulbar affect (PBA) episodes.
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist • 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)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ 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 Molybdenum cofactor deficiency (MoCD) Type A based on the following</u>**:
	• Family history
	  ○ Affected siblings with confirmed MoCD Type A; or a history of deceased sibling(s) with classic MoCD presentation
	  ○ One or both parents are known to carry a copy of the mutated gene [Molybdenum Cofactor Synthesis 1 (MOCS1)]
	  ○ Child has consanguineous parents with a family history of MoCD
	• Onset of clinical and/or laboratory signs and symptoms consistent with MoCD Type A:
	  ○ Clinical presentation: intractable seizures, exaggerated startle response, high-pitched cry, axial hypotonia, limb hypertonia, feeding difficulties
	  ○ 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
	
	<u>Confirmed diagnosis of MoCD Type A</u>:
	• Genetic confirmation of the presence of mutation in molybdenum cofactor synthesis gene 1 (MOSC1) to confirm MoCD Type A
	• In patients with a presumptive diagnosis of MoCD Type A, the diagnosis must be confirmed immediately using genetic testing
	
	<u>Reauthorization</u>:

	• Documentation of clinically significant response to therapy as determined by prescribing provider • 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) • MoCD Type C (gephyrin or GPHN mutation)
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neonatologist, pediatrician, pediatric neurologist, neonatal neurologist, or geneticist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 1 month, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

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

POLICY NAME:
NUSINERSEN
Affected Medications: SPINRAZA (nusinersen)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Spinal muscular atrophy (SMA)

Required Medical Information:	• Diagnosis of SMA type 1, 2, or 3 confirmed by genetic testing of chromosome 5q13.2 demonstrating ONE of the following:
	  ○ Homozygous gene deletion of SMN1 (survival motor neuron 1)
	  ○ Homozygous gene mutation of SMN1
	  ○ Compound heterozygous gene mutation of SMN1
	• Documentation of 2 or more copies of the SMN2 (survival motor neuron 2) gene
	• Documentation of previous treatment history
	• Documentation of one of the following baseline motor assessments appropriate for patient age and motor function:
	  ○ Hammersmith Infant Neurological Examination (HINE-2)
	  ○ Hammersmith Functional Motor Scale (HFSME)
	  ○ Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)
	  ○ Upper Limb Module (ULM) test
	  ○ 6-Minute Walk Test (6MWT)
	• Documentation of ventilator use status
	  ○ Patient is NOT ventilator-dependent (defined as using a ventilator at least 16 hours per day on at least 21 of the last 30 days)
	  ○ This does not apply to patients who require non-invasive ventilator assistance
	• Planned treatment regimen

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure with or intolerable adverse event on Evrysdi **Reauthorization** requires documentation of improvement in baseline motor assessment score, clinically meaningful stabilization, or delayed progression of SMA-associated signs and symptoms

Exclusion Criteria:	• SMA type 4 • Advanced SMA at baseline (complete paralysis of limbs, permanent ventilation support) • Prior treatment with SMA gene therapy (i.e., onasemnogene abeparvovec-xioi) • Will not use in combination with other agents for SMA (e.g., onasemnogene abeparvovec-xioi, risdiplam, etc.)
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or provider who is experienced in treatment of spinal muscular atrophy • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 8 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
OCRELIZUMAB
Affected Medications: OCREVUS (ocrelizumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.
	  ○ Primary progressive multiple sclerosis (PPMS)
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive disease (SPMS)

Required Medical Information:	**<u>All Indications:</u>** • Diagnosis confirmed with magnetic resonance imaging (MRI) per revised McDonald diagnostic criteria for MS ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS **<u>Primary Progressive MS:</u>** • Documentation of at least one year of disease progression and baseline Expanded Disability Status Scale (EDSS) of 3.0 to 6.5

Appropriate Treatment Regimen & Other Criteria:	**<u>Relapsing forms of MS:</u>**
	• Coverage of Ocrevus (ocrelizumab) requires documentation of one of the following:
	  ○ Documented disease progression or intolerable adverse event with rituximab (biosimilar products, Riabni and Ruxience, preferred)
	  ○ Currently receiving treatment with Ocrevus (ocrelizumab), excluding via samples or manufacturer's patient assistance program
	
	<u>Reauthorization</u> requires documentation of treatment success

Exclusion Criteria:	• Active hepatitis B infection • Concurrent use of other disease-modifying medications indicated for the treatment of MS

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

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Idiopathic pulmonary fibrosis
	  ○ Chronic fibrosing interstitial lung diseases with a progressive phenotype
	  ○ Systemic sclerosis-associated interstitial lung disease (SSc-ILD)

Required Medical Information:	• Documentation of baseline liver function tests in all patients, at regular intervals during the first three months, then periodically thereafter or as clinically indicated
	
	<u>Idiopathic Pulmonary Fibrosis (IPF)</u>
	• Documentation of diagnosis of idiopathic pulmonary fibrosis supported by one of the following:
	  ○ Presence of usual interstitial pneumonia (UIP)
	  ○ High resolution computed tomography (HRCT)
	  ○ Surgical lung biopsy
	• Documentation of baseline forced vital capacity (FVC) greater than or equal to 50% of the predicted value
	• Documentation of predicted diffuse capacity for carbon monoxide (DLCO) greater than or equal to 30%
	
	<u>Systemic Sclerosis-Associated Interstitial Lung Disease (SSc-ILD)</u>
	• Documentation of diagnosis of Systemic Sclerosis-Associated Interstitial Lung Disease from the American College of Rheumatology / European League Against Rheumatism classification criteria
	• Documentation of onset of disease (first non-Raynaud symptom) of less than 7 years
	• Documentation of greater than or equal to 10% fibrosis on a chest high resolution computed tomography (HRCT) scan conducted within the previous 12 months
	• Documentation of baseline FVC greater than or equal to 40% of predicted
	• Documentation of predicted DLCO 30-89% of predicted

	<u>Chronic Fibrosing Interstitial Lung Diseases with a Progressive Phenotype</u> • Documentation of a diagnosis of chronic fibrosing interstitial lung diseases with a progressive phenotype • Documentation of relevant fibrosis (greater than 10% fibrotic features) on chest high resolution computed tomography (HRCT) scan with clinical signs of progression (defined as FVC decline at least 10%, FVC decline at least 5% with worsening symptoms, and/or imaging in the previous 24 months) • FVC greater than or equal to 45% of predicted • DLCO 30% to less than 80% of predicted
Appropriate Treatment Regimen & Other Criteria:	**<u>IPF:</u>** • Documented treatment failure, contraindication, or intolerance to pirfenidone **<u>SSc-ILD:</u>** • Documented treatment failure with mycophenolate (MMF) **<u>Reauthorization</u>** requires documentation of treatment success
Exclusion Criteria:	• Documentation of airway obstruction (such as pre-bronchodilator FEV/FVC less than 0.7) • Transaminases more than 5 times the upper limit of normal or elevated transaminases accompanied by symptoms (jaundice, hyperbilirubinemia). • 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 • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of acid sphingomyelinase deficiency (ASMD) in adult and pediatric patients

Required Medical Information:	• Documentation of acid sphingomyelinase deficiency as evidenced by one of the following:
	  ○ Enzyme assay showing diminished (less than 10% of controls) or absent acid sphingomyelinase activity (ASM)
	  ○ Gene sequencing showing biallelic pathogenic SMPD1 mutation
	• Documentation of clinical presentation (e.g., hepatosplenomegaly, interstitial lung disease, liver fibrosis, growth restriction of childhood) outside the central nervous system
	• Documentation of current body mass index (BMI), weight, and height
	• For adults 18 years of age and older, documentation of both of the following:
	  ○ Diffusion capacity of lungs (DLCO) is less than or equal to 70% of the predicted normal value
	  ○ Spleen volume greater than or equal to 6 multiples of normal (MN) measured by magnetic resonance imaging (MRI)
	• For pediatrics 18 years of age and younger, documentation of both of the following:
	  ○ Spleen volume greater than or equal to 5 MN measured by MRI
	  ○ Height of -1 Z-score or lower

Appropriate Treatment	**Dosing:** Dosed every two weeks based on FDA label • Body mass index (BMI) less than or equal 30, the dosage is based on actual body weight (kg)

Regimen & Other Criteria:	• BMI of greater than 30 is dosed based on adjusted body weight • Adjusted body weight = (height in m^2) x 30 • Availability: 20 mg single-dose vials • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced **<u>Reauthorization</u>** requires documentation of improvement in patient specific disease presentation such as: • Improvement in PFT or DLCO • Improvement in spleen and/or liver volume or function • Improvement/stability in platelet counts • 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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
OMALIZUMAB
Affected Medications: XOLAIR (omalizumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of moderate to severe allergic asthma in adults and pediatric patients 6 years of age and older
	  ○ Add-on maintenance treatment of chronic rhinosinusitis with nasal polyps (CRSwNP) in adult patients
	  ○ Treatment of symptomatic chronic spontaneous urticaria (CSU) in patients 12 years of age and older
	  ○ Reduction of allergic reactions (Type I), including anaphylaxis, that may occur with accidental exposure to one or more foods in adults and pediatric patients aged 1 year and older with an IgE-mediated food allergy

Required Medical Information:	**<u>Allergic Asthma</u>**
	• Documentation of moderate to severe allergic asthma defined by all of the following:
	  ○ A positive skin test or in vitro reactivity to a perennial aeroallergen (e.g., house dust mite, animal dander [dog, cat], cockroach, feathers, mold spores)
	  ○ A serum total IgE level at baseline of:
	  ▪ At least 30 IU/mL and less than 700 IU/mL in patients 12 years of age and older OR
	  ▪ At least 30 IU/mL and less than 1,300 IU/mL in patients 6 to 11 years of age
	  ○ FEV1 less than 80% at baseline or FEV1/FVC reduced by at least 5% from normal
	
	<u>CRSwNP</u>
	• Documentation of both of the following:
	  ○ Diagnosis of chronic rhinosinusitis and has undergone prior bilateral total ethmoidectomy

	○ Indicated for revision sinus endoscopic sinus surgery due to recurrent symptoms of nasal polyps (such as nasal obstruction/congestion, bilateral sinus obstruction) **<u>CSU</u>** • Documentation of active CSU where the underlying cause is not considered to be any other allergic condition or other form of urticaria • Documentation of presence of recurrent urticaria, angioedema, or both, for a period of six weeks or longer • Documented avoidance of triggers (such as nonsteroidal anti-inflammatory drugs [NSAIDs]) • Documented baseline score from an objective clinical evaluation tool, such as: ○ Urticaria Activity Score (UAS7) (Score of 28 or higher) ○ Urticaria Control Test (UCT)) (Score under 12) ○ Dermatology Life Quality Index (DLQI) (Score of 21 or higher) ○ Chronic Urticaria Quality of Life Questionnaire (CU-QoL) (Score of 75 or higher) **<u>IgE-Mediated Food Allergy</u>** • Serum total IgE level between 30 and 1850 IU/mL • Body weight between 10 and 150 kg • Diagnosis of IgE-mediated food anaphylactic allergy to three or more foods with documented positive skin prick test and positive serum IgE • Documentation of past IgE-mediated food anaphylactic reactions requiring use of epinephrine despite avoidance of food allergen and modifications to diet • Documentation that avoidance of food allergen alone is not feasible based on the number of allergens, malnutrition due to nutritional restrictions, and impaired quality of life causing food allergy-related anxiety
Appropriate Treatment	**<u>Allergic Asthma</u>** • Documented use of high-dose inhaled corticosteroid (ICS) plus a

Regimen & Other Criteria:	long-acting beta agonist (LABA) for at least three months with continued symptoms
	• Documentation of one of the following:
	  ○ 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 with at least 80% adherence.
	  ○ Documentation that chronic daily oral corticosteroids are required
	
	<u>CRSwNP</u>
	• Documented treatment failure with at least 1 intranasal corticosteroid (such as fluticasone) after ethmoidectomy
	• Documented treatment failure with Sinuva implant
	
	<u>CSU</u>
	• Documented treatment failure with up to 4-fold standard dosing (must be scheduled) of one of the following second generation H1-antihistamine products for at least one month: cetirizine, fexofenadine, loratadine, desloratadine, or levocetirizine
	• Documented treatment failure with scheduled dosing of ALL of the following for at least one month each:
	  ○ Add-on therapy with a leukotriene antagonist (montelukast or zafirlukast)
	  ○ Add-on therapy with a H2-antagonist (famotidine or cimetidine)
	  ○ Add-on therapy with cyclosporine A
	
	<u>IgE-Mediated Food Allergy</u>
	• Trial and failure of oral immunotherapy (OIT)
	
	<u>Reauthorization</u>: documentation of treatment success and a clinically significant response to therapy

Exclusion Criteria:	• Use in combination with another monoclonal antibody (e.g., Fasenra, Nucala, Tezspire, Dupixent, Cinqair)

Age Restriction:	• **Allergic Asthma**: 6 years of age and older • **CRSwNP**: 18 years of age and older • **CSU**: 12 years of age and older
Prescriber/Site of Care Restrictions:	• **Allergic Asthma**: prescribed by, or in consultation with, an allergist, immunologist, or pulmonologist • **CRSwNP**: prescribed by, or in consultation with, an otolaryngologist • **CSU/IgE-Mediated Food Allergy**: prescribed by, or in consultation with, an allergist or immunologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
OMAVELOXOLONE
Affected Medications: Skyclarys (omaveloxolone)

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

POLICY NAME:
OMIDUBICEL
Affected Medications: OMISIRGE (Omidubicel)

Covered Uses:	• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better • All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
Required Medical Information:	• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course • Documented diagnosis of a hematologic malignancy • Clinically stable and eligible for umbilical cord blood transplantation (UCBT) following myeloablative conditioning

Appropriate Treatment Regimen & Other Criteria:	• Must **NOT** have a matched related donor (MRD), matched unrelated donor (MUD), mismatched unrelated donor (MMUD), or haploidentical donor readily available.
	• Documentation that **NONE** of the following are present:
	  ○ Other active malignancy
	  ○ Active or uncontrolled infection
	  ○ Active central nervous system (CNS) disease
	
	Reauthorization: None - Omisirge will be used as a one-time treatment

Exclusion Criteria:	• Karnofsky Performance Status (KPS) of 50% or less or Eastern Cooperative Oncology Group (ECOG) score of 3 or greater • HLA (human leukocyte antigen)-matched donor able to donate • Prior allo-HSCT (hematopoietic stem cell transplantation) • Pregnancy or lactation
Age Restriction:	• 12 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 2 months for 1 time administration, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Spinal muscular atrophy (SMA)

Required Medical Information:	• Diagnosis of SMA type 1 confirmed by genetic testing of chromosome 5q13.2 demonstrating ONE of the following:
	  ○ Homozygous gene deletion of SMN1 (survival motor neuron 1)
	  ○ Homozygous gene mutation of SMN1
	  ○ Compound heterozygous gene mutation of SMN1
	• Documentation of 2 or fewer copies of the SMN2 (survival motor neuron 2) gene
	• Documentation of previous treatment history
	• Documentation of ventilator use status:
	  ○ Patient is NOT ventilator-dependent (defined as using a ventilator at least 16 hours per day on at least 21 of the last 30 days)
	  ○ This does not apply to patients who require non-invasive ventilator assistance
	• Documentation of anti-adeno-associated virus (AAV) serotype 9 antibody titer less than or equal 1:50
	• Patient weight and planned treatment regimen

Appropriate Treatment Regimen & Other Criteria:	
Exclusion Criteria:	• Prior treatment with SMA gene therapy (i.e., onasemnogene abeparvovec-xioi) • Will not use in combination with other agents for SMA (e.g., nusinersen, risdiplam, etc.) • Advanced SMA at baseline (complete paralysis of limbs, permanent ventilation support)

Age Restriction:	• Children less than 2 years of age
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 • 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: ABECMA, ABRAXANE, ADCETRIS, ADSTILADRIN, AKEEGA, ALECENSA, ALIQOPA, ALKERAN, ALUNBRIG 180mg ORAL TABLET, ANKTIVA, ARZERRA, ASPARLAS, AUGTYRO, AYVAKIT, AZEDRA, BALVERSA, BAVENCIO, BELEODAQ, BELRAPZO, BENDAMUSTINE, BENDEKA, BESPONSA, BLENREP, BLINCYTO, BOSULIF, BRAFTOVI, BREYANZI, BRUKINSA, CABOMETYX, CALQUENCE, CAPRELSA, CARVYKTI, CLOFARABINE, CLOLAR, COLUMVI, COMETRIQ, COPIKTRA, COSELA, COTELLIC, CYRAMZA, DACOGEN, DARZALEX, DARZALEX FASPRO, DAURISMO, DOXIL, DOXORUBICIN LIPOSOMAL, ELAHERE, ELREXFIO, EMPLICITI, ENHERTU, EPKINLY, ERBITUX, ERIVEDGE, ERLEADA, ERLOTINIB, ERWINAZE, EVOMELA, FOTIVDA, FRUZAQLA, GAZYVA, GAVRETO, GEFITINIB, GILOTRIF, HEPZATO, HYCAMTIN, IBRANCE, IBRUTINIB, ICLUSIG, IDHIFA, IMBRUVICA, IMDELLTRA, IMFINZI, IMJUDO, IMLYGIC IRESSA, INLYTA, INQOVI, INREBIC, IOBENGUANE I-131, ISTODAX, IXEMPRA, JAKAFI, JAYPIRCA, JELMYTO, JEMPERLI, JEVTANA, KADCYLA, KEYTRUDA, KIMMTRAK, KRAZATI, KYMRIAH, KYPROLIS, LAPATINIB, LARTRUVO, LENALIDOMIDE, LENVIMA, LIBTAYO, LONSURF, LOQTORZI, LORBRENA, LUMAKRAS, LUMOXITI, LUNSUMIO, LUTATHERA, LYNPARZA, LYTGOBI, MARGENZA, MARQIBO, MATULANE, MEKINIST, MEKTOVI, MELPHALAN, MONJUVI, MYLOTARG, NAB-PACLITAXEL, NEXAVAR, NERLYNX, NILANDRON, NINLARO, NIVOLUMAB, NUBEQA, ODOMZO, OJEMDA, OJJAARA, ONCASPAR, ONIVYDE, ONUREG, OPDIVO, OPDUALAG, ORSERDU, PADCEV, PAZOPANIB, PEMAZYRE, PEPAXTO, PERJETA, PHOTOFRIN, PIQRAY, PLUVICTO, POLIVY, POMALYST, POTELIGEO, PROLEUKIN, PROVENGE, QINLOCK, RETEVMO, REVLIMID, REZLIDHIA, REZUROCK, ROMIDEPSIN, ROZLYTREK, RUBRACA, RYBREVANT, RYDAPT, RYLAZE, RYTELO, SARCLISA, SORAFENIB, STIVARGA, SUNITINIB, SUTENT, SYNRIBO, TABRECTA, TAFINLAR, TAGRISSO, TALVEY, TALZENNA, TARCEVA, TAZVERIK, TECARTUS, TECENTRIQ, TECVAYLI, TEMODAR, TEMOZOLOMIDE, TEPADINA, TEPMETKO, TIBSOVO, TIVDAK, TORISEL, TREANDA, TRODELVY, TRUQAP, TURALIO, TYKERB, VANFLYTA, VECTIBIX, VENCLEXTA, VERZENIO, VIDAZA, VIVIMUSTA, VIZIMPRO, VONJO, VORANIGO, VOTRIENT, VYXEOS, XALKORI, XOFIGO, XOSPATA, XPOVIO, XTANDI, YERVOY, YESCARTA, YONDELIS, ZALTRAP, ZEJULA TABLETS, ZELBORAF, ZEPZELCA, ZOLINZA, ZYDELIG, ZYKADIA, ZYNLONTA, ZYNYZ

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 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
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial approval: 4 months, unless otherwise specified • 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>• Short term use of opioids with an MME per day greater than 90 MME requires one of the following:
○ Recent surgery
○ Acute injury

• Chronic use of opioids with a Morphine Milligram Equivalents (MME) per day greater than 90 MME requires:
○ A comprehensive individual treatment plan including attestation of a pain management agreement between the prescriber and patient
○ Continued assessment and documentation of risk of abuse
○ Documentation that previous tapers have been attempted or documentation of a taper plan or rationale for avoidance of taper initiation</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td><u>Calculating morphine milligram equivalents (MME)</u>

<table><tr><th>Opioid</th><th>Factor</th></tr><tr><td>Methadone</td><td>4.7</td></tr><tr><td>Codeine</td><td>0.15</td></tr><tr><td>Fentanyl transdermal (mcg/hr)</td><td>2.4</td></tr><tr><td>Hydrocodone</td><td>1</td></tr><tr><td>Hydromorphone</td><td>5</td></tr><tr><td>Morphine</td><td>1</td></tr><tr><td>Oxycodone (Roxicodone, Oxycontin)</td><td>1.5</td></tr><tr><td>Oxymorphone</td><td>3</td></tr><tr><td>Tramadol</td><td>0.2</td></tr><tr><td>Buprenorphine patch</td><td>**</td></tr></table></td></tr>
</table>

	<table><tr><td>Tapentadol</td><td>0.4</td></tr><tr><td>Oxycodone myristate</td><td>1.67</td></tr></table> ** The MME conversion factor for buprenorphine patches is based on the assumption that: • One milligram of parenteral buprenorphine is equivalent to 75 milligrams of oral morphine and • One patch delivers the dispensed micrograms (mcg) per hour over a 24-hour day. **Example**: 5 mcg/hr buprenorphine patch X 24 hrs = 120 mcg/day buprenorphine = 0.12 mg/day 0.12 mg per day X 75 (1 mg buprenorphine=75 mg morphine) = 9 mg/day oral MME. In other words, the conversion factor not accounting for days of use would be 9/5 or 1.8. • Since the buprenorphine patch remains in place for 7 days, we have multiplied the conversion factor by 7 (1.8 X 7 = 12.6). In this example, MME/day for four 5 mcg/hr buprenorphine patches dispensed for use over 28 days would work out as follows: **Example**: 5 mcg/hr buprenorphine patch X (4 patches/28 days) X 12.6 = 9 MME/day. 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 • Chronic pain related to sickle cell disease • Pain related to hospice care • Surgery or documented acute injury – 1 month approval
Age Restriction:	

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

POLICY NAME:
OPZELURA
Affected Medications: OPZELURA CREAM (1.5%)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. ○ Atopic dermatitis
Required Medical Information:	**<u>Severe Atopic Dermatitis</u>** • 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 • Body Surface Area (BSA) of at least 10% OR • 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). <u>Reauthorization</u> • 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) • Previous 8-week treatment course • 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) • 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-INTRANASAL FENTANYL
Affected Medications: ABSTRAL, ACTIQ, FENTORA, FENTANYL CITRATE BUCCAL TABLET, LAZANDA, SUBSYS, FENTANYL CITRATE LOZENGE ON A HANDLE

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Management of breakthrough pain in cancer patients who are already receiving and who are tolerant to around-the-clock opioid therapy for their underlying persistent cancer pain

Required Medical Information:	• Documentation of ALL of the following:
	  ○ This drug is being prescribed for breakthrough cancer-related pain
	  ○ The patient is currently receiving, and will continue to receive, around-the-clock opioid therapy for underlying persistent cancer pain
	  ○ The patient is opioid tolerant, defined as taking one of the following for one week or longer:
	  ▪ At least 60 mg of oral morphine per day
	  ▪ At least 25 mcg of transdermal fentanyl per hour
	  ▪ At least 30 mg of oral oxycodone per day
	  ▪ At least 8 mg of oral hydromorphone per day
	  ▪ At least 25 mg of oral oxymorphone per day
	  ▪ At least 60 mg of oral hydrocodone per day
	  ▪ An equianalgesic dose of another opioid

Appropriate Treatment Regimen & Other Criteria:	• Documentation of **ONE** of the following:
	  ○ The patient is unable to swallow, or has dysphagia, esophagitis, mucositis, or uncontrollable nausea/vomiting
	  ○ The patient has documented intolerance or allergies to two other short-acting narcotics (such as oxycodone, morphine sulfate, hydromorphone, etc.)
	
	PDL only: Actiq requests will require documentation of clinical trial and failure with fentanyl citrate lozenge on a handle

	<u>Reauthorization</u> requires documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist or specialist in the treatment of cancer-related pain • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise exclude by plan design ○ Testosterone replacement therapy in adult males for conditions associated with a deficiency or absence of endogenous testosterone: primary hypogonadism or hypogonadotropic hypogonadism • Gender Dysphoria

Required Medical Information:	**<u>Hypogonadism in Adults</u>**
	• 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
	
	<u>For members 65 years and above</u>
	• Yearly evaluation of need is completed discussing need for hormone replacement therapy
	• Yearly documentation that provider has educated patient on risks of hormone replacement (heart attack, stroke)
	• Yearly documentation that provider has discussed limited efficacy and safety for hormone replacement in patients experiencing age related decrease in testosterone levels
	
	<u>Gender Dysphoria</u>
	• Documented diagnosis of gender dysphoria
	• If under 18 years of age, documentation of all of the following:
	  ○ Current Tanner stage 2 or greater OR baseline and current estradiol and testosterone levels to confirm onset of puberty
	  ○ Confirmed diagnosis of gender dysphoria that is persistent
	  ○ The patient has the capacity to make a fully informed decision and to give consent for treatment
	  ○ Any significant medical or mental health concerns are reasonably well controlled
	  ○ A comprehensive mental health evaluation has been completed by a licensed mental health professional (LMHP) and provided in accordance with the most current version

	of the World Professional Association for Transgender Health (WPATH) Standards of Care • **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:	**All Indications:** • Documented failure with transdermal testosterone **Reauthorization**: documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Treatment of sexual dysfunction • Treatment of symptoms of menopause
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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 24 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1

Required Medical Information:	**<u>Pulmonary Arterial Hypertension (PAH) WHO Group 1</u>**
	• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:
	  ○ Mean pulmonary artery pressure of at least 20 mm Hg
	  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg
	  ○ Pulmonary vascular resistance of at least 2.0 Wood units
	• Etiology of PAH: idiopathic, heritable, or associated with connective tissue disease
	• PAH secondary to one of the following conditions:
	  ○ Connective tissue disease
	  ○ Human immunodeficiency virus (HIV) infection
	  ○ Cirrhosis
	  ○ Anorexigens
	  ○ Congenital left to right shunts
	  ○ Schistosomiasis
	  ○ Drugs and toxins
	  ○ Portal hypertension
	• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class II or higher symptoms
	• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blockers) unless there are contraindications:
	  ○ 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 • The pulmonary hypertension has progressed despite maximal medical and/or surgical treatment of the identified condition

	• Documentation that treprostinil is used as a single route of administration (Remodulin, Tyvaso, Orenitram should not be used in combination) • 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.) **<u>Reauthorization</u>** requires documentation of treatment success defined as one or more of the following: • Improvement in walking distance • Improvement in exercise ability • Improvement in pulmonary function • Improvement or stability in WHO functional class
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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified.

POLICY NAME:
ORGOVYX
Affected Medications: ORGOVYX (relugolix)

Covered Uses:	• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher
Required Medical Information:	
Appropriate Treatment Regimen & Other Criteria:	**Prostate Cancer** • Documented treatment failure or intolerable adverse event with leuprolide or degarelix **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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
OSILODROSTAT
Affected Medications: ISTURISA (osilodrostat)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Cushing's disease

Required Medical Information:	• Documented diagnosis of Cushing's disease
	• Documentation of at least **TWO** of the following:
	  ○ Mean (at least two measurements) 24-hour urine free cortisol (mUFC) greater than 1.5 times the upper limit of normal (ULN) for the assay
	  ○ Bedtime salivary cortisol (at least two measurements) greater than 145 ng/dL
	  ○ Overnight dexamethasone suppression test (DST) with a serum cortisol greater than 1.8 mcg/dL

Appropriate Treatment Regimen & Other Criteria:	• Documentation confirming pituitary surgery is not an option **OR** previous surgery has not been curative **Reauthorization** Reauthorization requires documentation of treatment success defined as mUFC normalization (i.e., less than or equal to the ULN)
Exclusion Criteria:	
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist, neurologist, or adrenal surgeon • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
OTESECONAZOLE
Affected Medications: VIVJOA (oteseconazole)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ To reduce the incidence of recurrent vulvovaginal candidiasis (RVVC) in females with a history of RVVC who are **not** of reproductive potential, alone or in combination with fluconazole
Required Medical Information:	• Diagnosis of RVVC defined as three or more episodes of symptomatic vulvovaginal candidiasis infection within the past 12 months • Documented presence of signs/symptoms of current acute vulvovaginal candidiasis with a positive potassium hydroxide (KOH) test • 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 • **Not to exceed one treatment course per year** **<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 or who are pregnant or breastfeeding
Age Restriction:	• 18 years of age and older

Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 3 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of neurotrophic keratitis

Required Medical Information:	• Documentation of decreased corneal sensitivity (≤ 4 cm using the Cochet-Bonnet aesthesiometer) within the area of the recurrent/persistent epithelial defect or corneal ulcer **AND** outside of the area of the defect, in at least one corneal quadrant
	• Documentation of one of the following:
	  ○ Stage 2 neurotrophic keratitis, confirmed by presence of recurrent or persistent corneal epithelial defect
	  ○ Stage 3 neurotrophic keratitis, confirmed by presence of corneal ulceration (with or without stromal melting and perforation)

Appropriate Treatment Regimen & Other Criteria:	• Documented progression in disease severity with all of the following treatments:
	  ○ Preservative-free artificial tears, gel, or ointments
	  ○ Therapeutic corneal or scleral contact lenses
	  ○ Amniotic membrane transplantation and conjunctival flap surgery OR tarsorrhaphy OR cyanoacrylate glue OR soft-bandage contact lens
	• Dose may not exceed more than 1 vial per eye per day
	<u>Reauthorization</u> requires documentation of treatment response, as shown by a reduction in corneal staining with fluorescein

Exclusion Criteria:	• Active or suspected ocular or periocular infections
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an ophthalmologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 8 weeks, unless otherwise specified • Reauthorization: 8 weeks, unless otherwise specified • Lifetime Limit: 16 weeks (per affected eye)

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Narcolepsy with cataplexy
	  ○ Narcolepsy with excessive daytime sleepiness (EDS)
	  ○ Idiopathic Hypersomnia (IH) (Xywav only)

Required Medical Information:	• Diagnosis confirmed by polysomnography and multiple sleep latency test • Other causes of sleepiness have been ruled out or treated (including but not limited to obstructive sleep apnea, insufficient sleep syndrome, shift work, the effects of substances or medications, or other sleep disorders) **<u>Narcolepsy with cataplexy</u>**: • Documentation of cataplexy episodes defined as more than one episode of sudden loss of muscle tone with retained consciousness **<u>Narcolepsy with EDS or IH:</u>** • Current evaluation of symptoms and Epworth Sleepiness Scale (ESS) score of more than 10 despite treatment

Appropriate Treatment Regimen & Other Criteria:	**<u>Narcolepsy with cataplexy:</u>**
	• Documented treatment failure with TWO of the following for at least 1 month each:
	  ○ Venlafaxine
	  ○ Fluoxetine
	  ○ Duloxetine
	  ○ Tricyclic antidepressant (such as clomipramine, protriptyline)
	
	<u>Narcolepsy or IH, with EDS:</u>
	• Documented treatment failure to all of the following (1 in each category required) for at least 1 month each:
	  ○ Modafinil or armodafinil

	○ Methylphenidate, or dextroamphetamine, or lisdexamfetamine ○ Sunosi (Narcolepsy with EDS only) **<u>Reauthorization:</u>** • Narcolepsy with cataplexy: requires clinically significant reduction in cataplexy episodes • Narcolepsy or IH, with EDS: requires clinically significant improvement in activities of daily living and in Epworth Sleepiness Scale (ESS) score
Exclusion Criteria:	• Concurrent use of alcohol, sedative/hypnotic drugs, or other central nervous system depressants. • Use for other untreated causes of sleepiness
Age Restriction:	• 7 years of age and older for cataplexy or EDS due to narcolepsy • 18 years of age and older for EDS due to IH
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a sleep specialist or neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
OZANIMOD
Affected Medications: ZEPOSIA (ozanimod)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design:
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive disease (SPMS)
	  ○ Ulcerative Colitis

Required Medical Information:	**<u>Multiple Sclerosis</u>** • Diagnosis confirmed with magnetic resonance imaging (MRI), per revised McDonald diagnostic criteria for MS ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS **<u>Ulcerative Colitis</u>** • Diagnosis supported by endoscopy/colonoscopy/sigmoidoscopy or biopsy with moderate to severely active disease despite current treatment

Appropriate Treatment Regimen & Other Criteria:	**<u>Relapsing forms of MS</u>**
	• Coverage of Zeposia (ozanimod) requires documentation of one of the following:
	  ○ Documented disease progression or intolerable adverse event with one of the following: teriflunomide, dimethyl fumarate or fingolimod
	  ○ Currently receiving treatment with Zeposia (ozanimod), excluding via samples or manufacturer's patient assistance program
	
	<u>Ulcerative Colitis</u>
	• Documented failure with at least two oral treatments for a minimum of 12 weeks each: corticosteroids, sulfasalazine, azathioprine, mesalamine, balsalazide, cyclosporine, 6-mercaptopurine

	AND • Documented treatment failure with or intolerable adverse event with all preferred pharmacy drugs (Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Xeljanz, Stelara, Rinvoq) **<u>Reauthorization</u>** requires provider attestation of treatment success
Exclusion Criteria:	• MS: concurrent use of other disease-modifying medications indicated for the treatment of multiple sclerosis • UC: concurrent use with a JAK inhibitor or biologic medication for the treatment of ulcerative colitis
Age Restriction:	
Prescriber/Site of Care Restrictions:	• MS: prescribed by, or in consultation with, a neurologist or a multiple sclerosis specialist • UC: prescribed by, or in consultation with, a gastroenterologist • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization:
	  ○ UC: 6 months, unless otherwise specified
	  ○ MS: 12 months, unless otherwise specified
	• Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Mitigation of allergic reactions, including anaphylaxis, that may occur with accidental exposure to peanut

Required Medical Information:	• Documented treatment plan, including dose and frequency
	• Diagnosis of peanut allergy confirmed by one of the following:
	  ○ A positive skin prick test (SPT) response to peanut with a wheal diameter at least 3 mm larger than the control
	  ○ Serum peanut-specific IgE level greater than or equal to 0.35 kUA/L
	• Documented history of an allergic reaction to peanut with all of the following:
	  ○ Signs and symptoms of a significant systemic allergic reaction to peanut (e.g., hives, swelling, wheezing, hypotension, gastrointestinal symptoms)
	  ○ The reaction occurred within a short period of time following a known ingestion of peanut or peanut-containing food
	  ○ The reaction was severe enough to warrant a prescription for an epinephrine injection
	• Documentation indicating a significant impact on quality of life due to peanut allergies

Appropriate Treatment Regimen & Other Criteria:	**<u>Dosing:</u>** • Requests for initial dose escalation: must be between 1 and 17 years of age • Requests for up-dosing and maintenance phase: 1 year of age and older **<u>Reauthorization</u>** requires documentation of completion of the appropriate initial dose escalation and up-dosing phases prior to moving on to the maintenance phase AND documentation of treatment success and a clinically significant response to therapy, defined by one or more of the following:

	• Improvement in quality of life • Reduction in severe allergic reactions • Reduction in epinephrine use • Reduction in physician office visits, ER visits, or hospitalizations due to peanut allergy
Exclusion Criteria:	• Use for the emergency treatment of allergic reactions, including anaphylaxis • Uncontrolled asthma • History of eosinophilic esophagitis (EoE) and other eosinophilic gastrointestinal disease • History of cardiovascular disease, including uncontrolled or inadequately controlled hypertension • History of a mast cell disorder, including mastocytosis, urticarial pigmentosa, and hereditary or idiopathic angioedema
Age Restriction:	• 1 year of age and older (see Appropriate Treatment Regimen & Other Criteria for specific age-related dosing requirements)
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an allergist or immunologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
PALIVIZUMAB
Affected Medications: SYNAGIS (palivizumab)

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:** 1. Congenital heart disease (CHD): a. With cardiac transplantation, cardiac bypass, or extra-corporeal membrane oxygenation b. That is hemodynamically significant (e.g., acyanotic heart disease, congestive heart failure, or moderate to severe pulmonary hypertension) 2. Chronic lung disease (CLD) of prematurity: 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 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) 3. Cystic Fibrosis **and**: a. Clinical evidence of CLD and/or nutritional compromise 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) c. A weight for length less than the 10th percentile 4. Congenital airway abnormality or neuromuscular condition (not cystic fibrosis) that impairs the ability to clear airway secretions 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>** • The first dose of Synagis should be administered prior to commencement of the RSV season • 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)

	• No more than 5 monthly doses During the RSV season, November 1 through March 31 • Discontinue prophylaxis therapy if hospitalized for RSV
Exclusion Criteria:	• For use in the treatment of RSV disease • Received Beyfortus during the current RSV season
Age Restriction:	**Refer to numbered conditions above in "Required Medical Information":** • 1a. Less than 2 years of age • 1b. Less than 1 year of age • 2a. Less than 1 year of age; Gestational Age less than 32 weeks • 2b. Less than 2 years of age; Gestational Age less than 32 weeks • 3a. Less than 1 year of age • 3b. Less than 2 years of age • 3c. Less than 2 years of age • 4. Less than 1 year of age • 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:	Authorization: • 5 months (November 1 through March 31) [5 monthly doses] , unless otherwise specified • 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:
PALOVAROTENE
Affected Medications: SOHONOS (palovarotene)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Fibrodysplasia ossificans progressiva (FOP)
Required Medical Information:	• Documented diagnosis of FOP confirmed by *ACVR1* R206H mutation by molecular genetic testing • Radiographic features of FOP including joint malformations (such as hallux valgus deformity, malformed first metatarsal, absent or fused interphalangeal joint), and progressive heterotopic ossification (HO) • Documentation of experiencing at least two flare-ups in the past 12 months requiring prescription non-steroidal anti-inflammatory drugs (NSAIDs) and oral glucocorticoids such as prednisone
Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** requires documentation of treatment success defined as a decrease in HO volume or number of flare-ups compared to baseline
Exclusion Criteria:	• Patients weighing less than 10 kg • Pregnancy
Age Restriction:	• Females 8 years of age and older • Males 10 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a physician who specializes in rare connective tissue diseases • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Reduce phenylalanine (Phe) blood concentrations in adults with phenylketonuria (PKU) who have uncontrolled blood Phe greater than 600 micromol/L on existing management
Required Medical Information:	• Documentation of a diagnosis of PKU • Documentation of treatment failure with dual therapy of sapropterin and a Phe restricted diet as shown by a blood Phe level greater than 600 micromol/L (10 mg/dL) despite treatment
Appropriate Treatment Regimen & Other Criteria:	• Documentation that Palynziq will not be used in combination with sapropterin **Reauthorization** requires documentation of one of the following: • Reduction in baseline Phe levels by 20 percent • Increase in dietary Phe tolerance • Improvement in clinical symptoms
Exclusion Criteria:	
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist in metabolic disorders or an endocrinologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Adjunct to calcium and vitamin D to control hypocalcemia in hypoparathyroidism

Required Medical Information:	• Documentation of the following lab values:
	  ○ 25-hydroxyvitamin D levels within normal limits (approximately 30-74 ng/mL) while on standard of care (such as calcitriol)
	  ○ 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:
	  ○ Calcium 2000 mg daily
	  ○ Vitamin D (metabolite or analog)
	• 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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial approval: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of osteoporosis in men and postmenopausal women at high risk for fracture (teriparatide, Tymlos, and Forteo)
	  ○ Treatment of glucocorticoid-induced osteoporosis in men and women at high risk for fracture (teriparatide and Forteo only)

Required Medical Information:	• Diagnosis of osteoporosis as defined by at least **one** of the following:
	  ○ T-score –2.5 or lower (current or past) at the lumbar spine, femoral neck, total hip, or 1/3 radius site
	  ○ 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:
	  ▪ FRAX 10-year probability of major osteoporotic fracture is 20% or greater
	  ▪ FRAX 10-year probability of hip fracture is 3% or greater
	  ○ History of non-traumatic fractures in the absence of other metabolic bone disorders (postmenopausal women with osteoporosis only)
	• For glucocorticoid-induced osteoporosis, in addition to the above, must also provide documentation of the following:
	  ○ Treatment with 5 mg or higher of prednisone (or equivalent) per day for at least 3 months

Appropriate Treatment Regimen & Other Criteria:	Documentation of **one** of the following:
	• Treatment failure (new fracture or worsening T-score despite adherence to an adequate trial of therapy), contraindication, or intolerance to **BOTH** of the following:
	  ○ Oral or intravenous bisphosphonate (such as, alendronate, risedronate, zoledronic acid or ibandronate)
	  ○ Prolia (denosumab)

	• High risk of fracture defined as T-score -3.5 or lower, **OR** T-score -2.5 or lower with a history of fragility fractures
	
	For **Forteo** requests: Documented treatment failure with Tymlos and teriparatide
	
	<u>Total duration of therapy with parathyroid hormone analogs should not exceed 2 years in a lifetime</u>
	• Forteo or teriparatide may be **reauthorized** for up to one additional year beyond two years of parathyroid hormone analog use (maximum of 3 total years) if meeting the following criteria:
	  ○ Documentation of treatment success with parathyroid hormone use, defined as reduced frequency of fragility fractures or stable T-score while on Forteo or teriparatide
	  ○ Documentation that after 24 months of parathyroid hormone analog use, the patient remains at or has returned to having a high risk for fracture as evidenced by new fragility fracture or decline in T-score

Exclusion Criteria:	• Paget's Disease • Open epiphyses (such as, pediatric or young adult patient) • Bone metastases or skeletal malignancies • Hereditary disorders predisposing to osteosarcoma • Prior external beam or implant radiation therapy involving the skeleton • Concurrent use of bisphosphonates, other parathyroid hormone analogs, or RANK ligand inhibitors • Preexisting hypercalcemia • Pregnancy
Age Restriction:	
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 24 months (no reauthorization), unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ To reduce the risk of ototoxicity associated with cisplatin in pediatric patients 1 month of age and older with localized, non-metastatic solid tumors.
Required Medical Information:	• Documentation of a treatment plan that is a cisplatin-based regimen treating a localized, non-metastatic solid tumor
Appropriate Treatment Regimen & Other Criteria:	
Exclusion Criteria:	• Metastatic disease
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • 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

<table>
<tr><td>Covered Uses:</td><td>• All Food and Drug Administration (FDA)-approved and compendia-supported indications not otherwise excluded by plan design</td></tr>
<tr><td>Required Medical Information:</td><td>• Documentation of anticipated treatment course, to include full antiviral regimen, and duration of therapy

<u>Chronic Hepatitis C (CHC):</u>
• Documentation chronic hepatitis C virus (HCV) genotype by liver biopsy or by Food and Drug Administration (FDA)-approved serum test
• Baseline HCV RNA level

<u>Chronic Hepatitis B (CHB):</u>
• Documentation of HBeAg-positive or HBeAg-negative chronic hepatitis B virus (HBV) infection
• Baseline HBV DNA level
• Current (within 12 weeks) alanine transaminase (ALT) level

<u>Chronic Hepatitis C and B:</u>
• 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
• Documentation if HIV/HCV/HBV coinfection</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td><u>Chronic Hepatitis C:</u>
• 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

<u>Chronic Hepatitis B:</u>
• Documentation of ONE of the following scenarios:
<table>
<tr><th>HBeAg</th><th>HBV DNA</th><th>ALT</th></tr>
<tr><td colspan="3">Without cirrhosis</td></tr>
<tr><td>Positive</td><td>Greater than 20,000</td><td>Greater than 2 times the</td></tr>
</table>
</td></tr>
</table>

		copies/mL	upper limit of normal (ULN)
	Negative	Greater than 2,000 copies/mL	Greater than 2 times the ULN
	Negative	Greater than 2,000 copies/mL	1-2 times the ULN and moderate/severe liver inflammation/fibrosis
	With compensated cirrhosis		
	Either	Greater than 2,000 copies/mL	Any ALT
Exclusion Criteria:	• Treatment of patients with CHC who have had solid organ transplantation • Autoimmune hepatitis • Hepatic decompensation (Child-Pugh score greater than 6)		
Age Restriction:	• CHC: 5 years of age and older • CHB: 18 years of age and older		
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a gastroenterologist, hepatologist, or infectious disease specialist • All approvals are subject to utilization of the most cost-effective site of care		
Coverage Duration:	• CHC: 12 weeks, unless otherwise specified (depends on regimen and diagnosis) • CHB: 12 months, unless otherwise specified		

POLICY NAME:
PEGLOTICASE
Affected Medications: KRYSTEXXA (pegloticase)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Chronic gout in adults refractory to conventional therapy

Required Medical Information:	• Baseline serum uric acid (SUA) level greater than 8 mg/dL
	• Documentation of **ONE** of the following:
	  ○ 2 or more gout flares per year that were inadequately controlled by colchicine and/or nonsteroidal anti-inflammatory drugs (NSAIDS) or oral/injectable corticosteroids
	  ○ At least 1 non-resolving subcutaneous gouty tophus
	  ○ Chronic gouty arthritis (defined clinically or radiographically as joint damage due to gout)

Appropriate Treatment Regimen & Other Criteria:	• Documented contraindication, intolerance or clinical failure (defined as inability to reduce SUA level to less than 6 mg/dL) following a 12-week trial at maximum tolerated dose to BOTH:
	  ○ Xanthine oxidase inhibitor (allopurinol or febuxostat)
	  ○ Combination of a xanthine oxidase inhibitor AND a uricosuric agent (such as probenecid). If xanthine oxidase inhibitor is contraindicated, trial with uricosuric agent required.
	• Documentation Krystexxa will be used in combination with oral methotrexate 15 mg weekly unless contraindicated
	
	<u>Reauthorization</u> will require **ALL** of the following:
	• Documentation of SUA less than 6 mg/dL prior to next scheduled Krystexxa dose
	• Documentation of response to treatment such as reduced size of tophi or number of flares or affected joints
	• Rationale to continue treatment after resolution of tophi or reduction in symptoms

Exclusion Criteria:	• Concurrent use with oral urate-lowering therapies

Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a nephrologist or rheumatologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 6 months, unless otherwise specified

POLICY NAME:
PEMIVIBART
Affected Medications: PEMGARDA (pemivibart)

Covered Uses:	• All Food and Drug Administration (FDA) or compendia supported indications not otherwise excluded by plan design ○ Pre-exposure prophylaxis (PrEP) of coronavirus disease 2019 (COVID-19) in moderate-to-severe immune compromised individuals 12 years of age and older weighing at least 40 kg

Required Medical Information:	• Documentation of moderate-to-severe immune compromise due to a medical condition or receipt of immunosuppressive medications or treatments, and are unlikely to mount an adequate response to COVID-19 vaccination, meeting one of the following:
	  ○ Active treatment for solid tumor and hematologic malignancies
	  ○ Hematologic malignancies associated with poor responses to COVID-19 vaccines regardless of current treatment status (e.g., chronic lymphocytic leukemia, non-Hodgkin lymphoma, multiple myeloma, acute leukemia)
	  ○ Receipt of solid-organ transplant or an islet transplant and taking immunosuppressive therapy
	  ○ Receipt of chimeric antigen receptor (CAR)-T-cell or hematopoietic stem cell transplant (within 2 years of transplantation or taking immunosuppressive therapy)
	  ○ Moderate or severe primary immunodeficiency (e.g., common variable immunodeficiency disease, severe combined immunodeficiency, DiGeorge syndrome, Wiskott-Aldrich syndrome)
	  ○ Advanced or untreated human immunodeficiency viruses (HIV) infection (people with HIV and CD4 cell counts less than 200/mm^3, history of an AIDS-defining illness without immune reconstitution, or clinical manifestations of symptomatic HIV)
	  ○ Active treatment with high-dose corticosteroids (at least 20 mg prednisone or equivalent per day when administered for 2 or more weeks), alkylating agents, antimetabolites, transplant-related immunosuppressive drugs, cancer chemotherapeutic agents classified as

	severely immunosuppressive, and biologic agents that are immunosuppressive or immunomodulatory (such as B-cell depleting agents) • Documentation of prophylactic use • Baseline SARS-CoV-2 titers that show undetectable antibodies • Weight of 40 kg or more
Appropriate Treatment Regimen & Other Criteria:	• **Dosing** is in accordance with FDA labeling and does not exceed 4500 mg once every 3 months **Reauthorization** requires documentation of continued immune compromise and low SARS-CoV-2 titers
Exclusion Criteria:	• Positive SARS-CoV-2 antigen test or PCR test within the last 3 months • Received COVID-19 vaccine within the last 3 months
Age Restriction:	• 12 years of age and older
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 3 months, unless otherwise specified

POLICY NAME:
PENICILLAMINE
Affected Medications: PENICILLAMINE CAPSULE

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Cystinuria
	  ○ Wilson's Disease
	  ○ Rheumatoid arthritis
	  ○ Copper measurement in urine

Required Medical Information:	• Documented treatment plan including routine urinalysis, WBCs, hemoglobin, platelet count, liver function tests, renal function tests due to risk of fatalities due to aplastic anemia, agranulocytosis, thrombocytopenia, myasthenia gravis, and Goodpasture's Syndrome
	
	<u>Wilson's Disease</u>
	• Diagnosis confirmed by **ONE** of the following:
	  ○ Genetic testing results confirming biallelic pathogenic *ATP7B* mutations (in either symptomatic or asymptomatic individuals)
	  ○ Liver biopsy findings consistent with Wilson's disease
	  ○ Presence of Kayser-Fleischer (KF) rings **AND** serum ceruloplasmin level less than 20 mg/dL **AND** 24-hour urinary copper excretion greater than 40 mcg
	  ○ Presence of Kayser-Fleischer (KF) rings **AND** 24-hour urinary copper excretion greater than 100 mcg
	  ○ Absence of KF rings with serum ceruloplasmin level less than 10 mg/dL **AND** 24-hour urinary copper excretion greater than 100 mcg
	
	<u>Rheumatoid arthritis</u>
	• Documentation of severe, active disease defined by one of the following:
	  ○ The Disease Activity Score derivative for 28 joints (DAS-28) greater than 3.2
	  ○ The Simplified Disease Activity Index (SDAI) greater than 11

	○ The Clinical Disease Activity Index (CDAI) greater than 10 ○ Weighted Routine Assessment of Patient Index Data 3 (RAPID3) of at least 2.3
Appropriate Treatment Regimen & Other Criteria:	**<u>Rheumatoid arthritis</u>** • Has failed to respond to an adequate trial of conventional therapies (such as methotrexate, sulfasalazine, hydroxychloroquine, leflunomide, Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Xeljanz, Rinvoq, and Inflectra) **<u>Reauthorization</u>** requires documentation of disease responsiveness to therapy • For Wilson's disease, must have normalization of free serum copper (non-ceruloplasmin bound copper) to less than 15 mcg/dL and 24-hour urinary copper in the range of 200 to 500 mcg
Exclusion Criteria:	• Use of penicillamine during pregnancy (except for treatment of Wilson's disease or cystinuria)
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist familiar with the toxicity and dosage considerations (such as a hepatologist, gastroenterologist, or liver transplant physician for Wilson's Disease) • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of sweating and hypertension associated with pheochromocytoma

Required Medical Information:	• Documented diagnosis of pheochromocytoma that requires treatment to control episodes of hypertension and sweating
	• This drug will be used for one of the following:
	  ○ Preoperative preparation for a scheduled surgical resection
	  ○ Chronic treatment of pheochromocytoma that is not amenable to surgery

Appropriate Treatment Regimen & Other Criteria:	• Documentation of treatment failure, intolerance, or contraindication to a selective alpha-1 adrenergic receptor blocker (e.g., doxazosin, terazosin, prazosin) **Reauthorization** will require documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist or a specialist with experience in the management of pheochromocytoma • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Preoperative preparation: 1 month, unless otherwise specified • Chronic treatment: 12 months, unless otherwise specified

POLICY NAME:
PHESGO
Affected Medications: PHESGO (pertuzumab-trastuzumab-hyaluronidase-zzxf)

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
	• 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:	• Documentation of an intolerable adverse event to **all** of the preferred products (Perjeta in combination with Kanjinti, Perjeta in combination with Ogivri) and the adverse event was not an expected adverse event attributed to the active ingredients **Reauthorization** requires documentation of disease responsiveness to therapy
Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1

Required Medical Information:	• Diagnosis of World Health Organization (WHO) Group 1 PAH confirmed by right heart catheterization meeting the following criteria:
	  ○ Mean pulmonary artery pressure of at least 20 mm Hg
	  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg
	  ○ Pulmonary vascular resistance of at least 2.0 Wood units
	• New York Heart Association (NYHA)/WHO Functional Class II or higher symptoms
	• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blocker) unless there are contraindications:
	  ○ Low systemic blood pressure (systolic blood pressure less than 90)
	  ○ Low cardiac index
	  ○ Presence of severe symptoms (functional class IV)

Appropriate Treatment Regimen & Other Criteria:	• For all brand requests: Documented inadequate response or intolerance to sildenafil citrate 20 mg tablets and tadalafil 20 mg tablets • Requests for oral suspension must have documented inability to swallow tablets **<u>Reauthorization</u>** requires documentation of treatment success defined as one or more of the following: • Improvement in walking distance • Improvement in exercise ability • Improvement in pulmonary function

	• Improvement or stability in WHO functional class
Exclusion Criteria:	• Concomitant nitrate therapy on a regular or intermittent basis • Concomitant use of riociguat, a guanylate cyclase stimulator • Use for erectile dysfunction
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a cardiologist or pulmonologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Idiopathic Pulmonary Fibrosis

Required Medical Information:	• Documentation of ALL of the following:
	  ○ Presence of usual interstitial pneumonia (UIP) on high resolution computed tomography (HRCT), and/or surgical lung biopsy
	  ○ Baseline forced vital capacity (FVC) greater than or equal to 50 percent of the predicted value
	  ○ 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 • 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 consultation with, a pulmonologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
PLEGRIDY
Affected Medications: PLEGRIDY (peglyated interferon beta-1a)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive disease (SPMS)

Required Medical Information:	• Diagnosis confirmed with magnetic resonance imaging (MRI), per revised McDonald diagnostic criteria for MS ○ 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 indicated for the treatment of multiple sclerosis
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or multiple sclerosis specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Approval: 24 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Late-onset Pompe disease for patients weighing 40 kg or more and who are not improving on their current enzyme replacement therapy (ERT)

Required Medical Information:	• Diagnosis of late-onset Pompe disease confirmed by one of the following:
	  ○ Enzyme assay demonstrating a deficiency of acid alpha-glucosidase (GAA) enzyme activity
	  ○ DNA testing that identifies mutations in the GAA gene
	• One or more clinical signs or symptoms of late-onset Pompe disease:
	  ○ Progressive proximal weakness in a limb-girdle distribution
	  ○ Delayed gross-motor development in childhood
	  ○ Involvement of respiratory muscles causing respiratory difficulty (such as reduced forced vital capacity [FVC] or sleep disordered breathing)
	  ○ Skeletal abnormalities (such as scoliosis or scapula alata)
	  ○ Low/absent reflexes
	• Documentation that patient has a 6-minute walk test (6MWT) of 75 meters or more
	• Documentation of a sitting percent predicted forced vital capacity (FVC) of 30% or more
	• Patient weight

Appropriate Treatment Regimen & Other Criteria:	• Documentation of planned treatment regimen for both Pombiliti and Opfolda which are within FDA-labeling • Documentation that patient is no longer improving after at least one year of current enzyme replacement therapy (ERT) with

	Lumizyme (alglucosidase alfa) or Nexviazyme (avalglucosidase alfa-ngpt) **<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy as evidenced by an improvement, stabilization, or slowing of progression in percent-predicted FVC and/or 6MWT
Exclusion Criteria:	• Pregnancy or, if female of reproductive potential, not using effective contraception during treatment • Use of invasive or noninvasive ventilation support for more than 6 hours a day while awake • Diagnosis of infantile-onset Pompe disease • Concurrent treatment with Lumizyme or Nexviazyme • Pombiliti or Opfolda as monotherapy • Use of Opfolda for Gaucher disease
Age Restriction:	• 18 years of age or older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a metabolic specialist, endocrinologist, biochemical geneticist, or provider experienced in the management of Pompe disease • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
PONVORY
Affected Medications: Ponvory (ponesimod)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive disease (SPMS)

Required Medical Information:	• Diagnosis confirmed with magnetic resonance imaging (MRI), per revised McDonald diagnostic criteria for MS ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS
Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure with TWO of the following (minimum 12-week trial each): fingolimod, teriflunomide, Mayzent **Reauthorization:** provider attestation of treatment success
Exclusion Criteria:	• Concurrent use of other disease-modifying medications indicated for the treatment of multiple sclerosis
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or a multiple sclerosis specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
Required Medical Information:	• Susceptibility cultures matching posaconazole activity • Current body weight (for pediatric patients)

Appropriate Treatment Regimen & Other Criteria:	**<u>Treatment of invasive aspergillosis</u>**
	• Documentation of resistance (or intolerable adverse event) to voriconazole
	
	<u>Prophylaxis of invasive Aspergillus and Candida infections</u>
	• 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
	• Documentation of resistance (or intolerable adverse event) to one other compendia-supported systemic agent (e.g. fluconazole, itraconazole, voriconazole)
	
	<u>Treatment of oropharyngeal candidiasis (OPC):</u>
	• Documented failure (or intolerable adverse event) to 10 days or more of treatment with all of the following:
	  ○ Fluconazole
	  ○ Itraconazole

Exclusion Criteria:	
Age Restriction:	• Posaconazole delayed release tablets – 2 years of age and older, who weigh greater than 40 kg • Noxafil oral suspension – 13 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an infectious disease specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Approval: 6 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of CD55-deficient protein-losing enteropathy (PLE) or CHAPLE disease
Required Medical Information:	• Diagnosis of CD-55-deficient PLE confirmed by biallelic CD55 loss-of-function mutation using molecular genetic testing • Documentation of hypoalbuminemia (serum albumin of 3.2 g/dL or less) • Clinical signs and features of active PLE including abdominal pain, diarrhea, peripheral edema, or facial edema • Documentation of at least two albumin transfusions or hospitalizations in the past year

Appropriate Treatment Regimen & Other Criteria:	• **Dosing** is in accordance with FDA labeling and does not exceed the following:
	  ○ Loading Dose: 30 mg/kg by intravenous infusion for 1 dose
	  ○ Maintenance Dose: Starting on day 8;
	  10 mg/kg as a subcutaneous injection once weekly
	  May be increased to 12 mg/kg starting week 4
	  ○ Maximum maintenance dosage of 800 mg once weekly
	
	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced
	
	Reauthorization requires documentation of positive clinical response with all the following:
	• Improvement or stabilization of clinical symptoms
	• Improvement or normalization of serum albumin concentrations
	• Reduction in albumin transfusion requirements and/or hospitalizations

Exclusion Criteria:	• Receiving concurrent therapy with Soliris (eculizumab) • Unresolved Neisseria meningitidis, Streptococcus pneumoniae, or Haemophilus influenzae type b (Hib) infection

Age Restriction:	• 1 year of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist, gastroenterologist, or provider that specializes in rare genetic hematologic diseases • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
PRAMLINTIDE
Affected Medications: SYMLINPEN (pramlintide)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Type 1 diabetes mellitus
	  ○ Type 2 diabetes mellitus

Required Medical Information:	• Documentation of inadequate glycemic control (HbA1c greater than 7 percent) on optimized insulin therapy AND • Patient will take SymlinPen in addition to mealtime insulin therapy
Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** will require documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• HbA1c level greater than 9 percent • 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:
PROLIA
Affected Medications: PROLIA (denosumab)

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

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

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

Coverage Duration:	• Authorization: 1 month (one implant per impacted eye), unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of adults with paroxysmal nocturnal hemoglobinuria (PNH)

Required Medical Information:	• Patients must be administered a meningococcal vaccine at least two weeks prior to initiation of the requested therapy and revaccinated according to current Advisory Committee on Immunization Practices (ACIP) guidelines
	• Detection of PNH clones of at least 5% by flow cytometry diagnostic testing
	  ○ Presence of at least 2 different glycosylphosphatidylinositol (GPI) protein deficiencies (e.g., CD55, CD59, etc.) within at least 2 different cell lines (e.g., granulocytes, monocytes, erythrocytes)
	• Baseline lactate dehydrogenase (LDH) levels greater than or equal to 1.5 times the upper limit of normal range.
	• One of the following PNH-associated clinical findings:
	  ○ Presence of a thrombotic event
	  ○ Presence of organ damage secondary to chronic hemolysis
	  ○ History of 4 or more blood transfusions required in the previous 12 months

Appropriate Treatment Regimen & Other Criteria:	• For Empaveli: documented inadequate response, contraindication, or intolerance to ravulizumab (Ultomiris) • For Fabhalta: documented inadequate response, contraindication, or intolerance to another complement inhibitor such as ravulizumab (Ultomiris) or Empaveli **<u>Reauthorization</u>** requires documentation of treatment success defined as a decrease in serum LDH, stabilized/improved hemoglobin, decreased transfusion requirement, and reduction in thromboembolic events compared to baseline
Exclusion Criteria:	• Concurrent use with other biologics for PNH (Soliris, Ultomiris, Empaveli, or Fabhalta) except when cross tapering according to FDA approved dosing • Current meningitis infection or other unresolved serious infection caused by encapsulated bacteria

Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Primary biliary cholangitis (PBC)
Required Medical Information:	• Liver function tests (including alkaline phosphatase and bilirubin) • Child-Pugh score

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
	  ○ ULN ALP defined as 118 U/L for females or 124 U/L for males
	  ○ ULN of total bilirubin defined as 1.1 mg/dL for females or 1.5 mg/dL for males
	
	<u>Reauthorization</u> will require documentation of treatment success defined as a significant reduction in Alkaline phosphatase (ALP) and/or bilirubin levels

Exclusion Criteria:	• Complete biliary obstruction • Decompensated cirrhosis (e.g., Child-Pugh Class B or C) or a prior decompensation event • Compensated cirrhosis with evidence of portal hypertension (e.g., ascites, gastroesophageal varices, persistent thrombocytopenia) • Use in combination with another drug on this policy (Ocaliva, Iqirvo)
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hepatologist • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
PYRIMETHAMINE
Affected Medications: Daraprim, pyrimethamine

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Toxoplasmosis

Required Medical Information:	• Documentation of recent *Toxoplasma* infection
	• Documentation of one of the following:
	  ○ Severe symptoms (pneumonitis, myocarditis, etc) or prolonged symptoms greater than 4 weeks with significant impact on quality of life
	  ○ Immunocompromised status

Appropriate Treatment Regimen & Other Criteria:	• Dosing Regimen (adult):
	  ○ Day 1: Pyrimethamine 100 mg, sulfadiazine 2-4 gm divided four times daily, leucovorin 5-25 mg
	  ○ Day 2: Pyrimethamine 25-50 mg, sulfadiazine 2-4 gm divided four times daily, leucovorin 5-25 mg
	  ○ Day 3 and beyond: Pyrimethamine 25-50 mg, sulfadiazine 500 mg-1 gm divided four times daily, leucovorin 5-25 mg

Exclusion Criteria:	• Treatment regimen does not contain leucovorin and a sulfonamide (or alternative if allergic to sulfa)
Age Restriction:	
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: Up to 6 weeks, with no reauthorization unless otherwise specified

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

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

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis
	  ○ Atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy
	  ○ Generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive
	  ○ Neuromyelitis optica spectrum disorder (NMOSD) who are anti-aquaporin-4 (AQP4) antibody positive for adult patients

Required Medical Information:	**<u>PNH</u>**
	• Detection of PNH clones of at least 5% by flow cytometry diagnostic testing
	  ○ Presence of at least 2 different glycosylphosphatidylinositol (GPI) protein deficiencies (e.g., CD55, CD59, etc.) within at least 2 different cell lines (e.g., granulocytes, monocytes, erythrocytes)
	• Baseline lactate dehydrogenase (LDH) levels greater than or equal to 1.5 times the upper limit of normal range.
	• One of the following PNH-associated clinical findings:
	  ○ Presence of a thrombotic event
	  ○ Presence of organ damage secondary to chronic hemolysis
	  ○ History of 4 or more blood transfusions required in the previous 12 months
	
	<u>aHUS</u>
	• Clinical presentation of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury
	• Patient shows signs of thrombotic microangiopathy (TMA) (e.g., changes in mental status, seizures, angina, dyspnea, thrombosis, increasing blood pressure, decreased platelet count, increased serum creatinine, increased LDH, etc.)
	• ADAMTS13 activity level greater than or equal to 10%
	• Shiga toxin E. coli related hemolytic uremic syndrome (ST-HUS) has been ruled out

	• History of 4 or more blood transfusions required in the previous 12 months
	
	<u>gMG</u>
	• Diagnosis of gMG confirmed by **ONE** of the following:
	  ○ A history of abnormal neuromuscular transmission test
	  ○ A positive edrophonium chloride test
	  ○ Improvement in gMG signs or symptoms with an acetylcholinesterase inhibitor
	• Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV
	• Positive serologic test for AChR antibodies
	• Documentation of **ONE** of the following:
	  ○ MG-Activities of Daily Living (MG-ADL) total score of 6 or greater
	  ○ Quantitative Myasthenia Gravis (QMG) total score of 12 or greater
	
	<u>NMOSD</u>
	• Diagnosis of NMOSD with aquaporin-4 immunoglobulin G (AQP4-IgG) antibody positive disease confirmed by all of the following:
	  ○ Documentation of positive test for AQP4-IgG antibodies via cell-based assay
	  ○ Exclusion of alternative diagnoses (such as multiple sclerosis)
	  ○ At least **ONE** core clinical characteristic:
	  ▪ Acute optic neuritis
	  ▪ Acute myelitis
	  ▪ Area postrema syndrome (episode of otherwise unexplained hiccups or nausea/vomiting)
	  ▪ Acute brainstem syndrome
	  ▪ Symptomatic narcolepsy **OR** acute diencephalic clinical syndrome with NMSOD-typical diencephalic MRI lesions
	  ▪ Symptomatic cerebral syndrome with NMOSD-typical lesion on magnetic resonance imaging (MRI) [see table below]

<table>
<tr><td></td><td>
Acute cerebral syndrome with NMOSD-typical brain lesion on MRI [see table below]
<table>
<tr><th>Clinical presentation</th><th>Possible MRI findings</th></tr>
<tr><td>Diencephalic syndrome</td><td>• Periependymal lesion
• Hypothalamic/thalamic lesion</td></tr>
<tr><td>Acute cerebral syndrome</td><td>• Extensive periependymal lesion
• Long, diffuse, heterogenous, or edematous corpus callosum lesion
• Long corticospinal tract lesion
• Large, confluent subcortical or deep whitematter lesion</td></tr>
</table>
</td></tr>
<tr><td>Appropriate Treatment Regimen & Other Criteria:</td><td>
<u>aHUS</u>
• Failure to respond to plasma therapy within 10 days
  ○ Trial of plasma therapy not required if one of the following is present:
    ▪ Life-threatening complications of HUS such as seizures, coma, or heart failure
    ▪ Confirmed presence of a high-risk complement genetic variant (e.g., CFH or CFI)

<u>gMG</u>
• Documentation of one of the following:
  ○ Treatment failure with an adequate trial (one year or more) of at least 2 immunosuppressive therapies (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)
  ○ Has required three or more courses of rescue therapy (plasmapheresis/plasma exchange and/or intravenous immunoglobulin), while on at least one immunosuppressive therapy, over the last 12 months
• Documented inadequate response, contraindication, or intolerance to efgartigimod-alfa (Vyvgart)

<u>NMOSD</u>
• Documented inadequate response, contraindication, or
</td></tr>
</table>

	intolerance to **ALL** of the following: ○ Rituximab (preferred products: Riabni, Ruxience) ○ Satralizumab-mwge (Enspryng) ○ Inebilizumab-cdon (Uplizna) **<u>Reauthorization</u>** requires: • gMG: documentation of treatment success defined as an improvement in MG-ADL and QMG scores from baseline • PNH: documentation of treatment success defined as a decrease in serum LDH, stabilized/improved hemoglobin, decreased transfusion requirement, and reduction in thromboembolic events compared to baseline • aHUS: documentation of treatment success defined as a decrease in serum LDH, stabilized/improved serum creatinine, increased platelet count, and decreased plasma exchange/infusion requirement compared to baseline • NMOSD: documentation of treatment success defined as the stabilization or improvement in neurological symptoms as evidenced by a decrease in acute relapses, Expanded Disability Status Scale (EDSS) score, hospitalizations, or plasma exchange treatments
Exclusion Criteria:	• Current meningitis infection • Concurrent use with other disease-modifying biologics for requested indication, unless otherwise specified
Age Restriction:	• PNH, aHUS: 1 month of age and older • gMG: 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist ○ PNH: hematologist ○ aHUS: hematologist or nephrologist ○ gMG: neurologist ○ NMOSD: neurologist or neuro-ophthalmologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
REBLOZYL
Affected Medications: REBLOZYL (luspatercept-aamt)

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** • 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) • Documentation of anemia in adults without previous erythropoiesis-stimulating agent (ESA) use (ESA-naive) with very low- to intermediate-risk myelodysplastic syndromes (MDS) who may require RBC transfusions • Baseline complete blood count (CBC) within 2 months and then prior to each administration, or more frequently as indicated • Documentation of current RBC transfusion regimen
Appropriate Treatment Regimen & Other Criteria:	• Documentation of serum EPO over 500 mU/mL with a need for RBC transfusions (very low- to intermediate-risk MDS) **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 • Use as immediate correction as a substitute for RBC transfusions • Diagnosis of alpha thalassemia • Known pregnancy
Age Restriction:	• 18 years of age and older

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

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive disease (SPMS)

Required Medical Information:	• Diagnosis confirmed with magnetic resonance imaging (MRI), per revised McDonald diagnostic criteria for MS ○ 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 • 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)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Amyotrophic lateral sclerosis (ALS)
Required Medical Information:	• Definite or probable Amyotrophic lateral sclerosis (ALS) based on El Escorial revised (Airlie House) criteria • Symptom onset within 18 months • Slow vital capacity (SVC) of at least 60 percent • 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:
	  ○ Member is stable on riluzole
	  ○ Prescriber has indicated clinical inappropriateness of riluzole
	Reauthorization: Documentation of treatment success as determined by prescriber including retaining most activities of daily living

Exclusion Criteria:	• Presence of a tracheostomy • Use of permanent assisted ventilation
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1
	  ○ Pulmonary Arterial Hypertension in patients requiring transition from epoprostenol

Required Medical Information:	**<u>Pulmonary Arterial Hypertension (PAH) WHO Group 1</u>**
	• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:
	  ○ Mean pulmonary artery pressure of at least 20 mm Hg
	  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg
	  ○ Pulmonary vascular resistance of at least 2.0 Wood units
	• Etiology of PAH: idiopathic PAH, hereditary PAH, OR
	• PAH secondary to one of the following conditions:
	  ○ Connective tissue disease
	  ○ Human immunodeficiency virus (HIV) infection
	  ○ Cirrhosis
	  ○ Anorexigens
	  ○ Congenital left to right shunts
	  ○ Schistosomiasis
	  ○ Drugs and toxins
	  ○ Portal hypertension
	• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class II or higher symptoms
	• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blockers) unless there are contraindications:
	  ○ Low systemic blood pressure (systolic blood pressure less than 90)
	  ○ Low cardiac index
	  OR
	  ○ Presence of severe symptoms (functional class IV)

Appropriate Treatment Regimen &	• The pulmonary hypertension has progressed despite maximal medical and/or surgical treatment of the identified condition

Other Criteria:	• Documentation that treprostinil is used as a single route of administration (Remodulin, Tyvaso, Orenitram should not be used in combination) • Treatment with oral calcium channel blocking agents has been tried and failed, or has been considered and ruled out • 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 **Reauthorization** requires documentation of treatment success defined as one or more of the following: • Improvement in walking distance • Improvement in exercise ability • Improvement in pulmonary function • Improvement or stability in WHO 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:	• Prescribed by, or in consultation with, a cardiologist or pulmonologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
RESLIZUMAB
Affected Medications: CINQAIR (reslizumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Add-on maintenance treatment of adult patients with severe asthma with an eosinophilic phenotype

Required Medical Information:	• Diagnosis of severe asthma with an eosinophilic phenotype, defined by both of the following:
	  ○ Baseline eosinophil count of at least 400 cells/μL
	  ○ FEV1 less than 80% at baseline or FEV1/FVC reduced by at least 5% from normal

Appropriate Treatment Regimen & Other Criteria:	• Documented use of high-dose inhaled corticosteroid (ICS) plus a long-acting beta agonist (LABA) for at least three months with continued symptoms
	• Documentation of one of the following:
	  ○ 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
	  ○ Documentation that chronic daily oral corticosteroids are required
	• Documented treatment failure or intolerable adverse event with all of the preferred products (Dupixent, Fasenra, Nucala, and Xolair)
	
	• Availability: 100 mg/10 mL vials
	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced
	
	<u>Reauthorization</u>: documentation of treatment success and a clinically significant response to therapy

Exclusion Criteria:	• Use in combination with another monoclonal antibody (e.g., Dupixent, Nucala, Xolair, Fasenra, Tezspire)

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

POLICY NAME:
RESMETIROM
Affected Medications: REZDIFFRA (resmetirom)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of adults with noncirrhotic nonalcoholic steatohepatitis (NASH) with moderate to advanced liver fibrosis (consistent with stages F2 to F3 fibrosis), in conjunction with diet and exercise

Required Medical Information:	• Diagnosis of NASH or metabolic dysfunction–associated steatohepatitis (MASH) with moderate to advanced (F2 to F3) liver fibrosis confirmed by **ONE** of the following:
	  ○ Conclusive result from a well-validated non-invasive test such as:
	  ▪ Fibroscan-AST (FAST) score
	  ▪ MAST (score from MRI–proton density fat fraction, Magnetic resonance elastography [MRE], and serum AST)
	  ▪ MEFIB (Fibrosis-4 Index ≥1.6 and MRE ≥3.3 kPa)
	  ○ Liver biopsy (also required if non-invasive testing is inconclusive or other causes for liver disease have not been ruled out)
	• Other causes for liver steatosis have been ruled out (such as alcohol-associated liver disease, chronic hepatitis C, Wilson disease, drug-induced liver disease)
	• Baseline lab values for AST and ALT

Appropriate Treatment Regimen & Other Criteria:	• Documentation of abstinence from alcohol consumption
	• Documentation of comprehensive comorbidity management being undertaken, including all of the following:
	  ○ Use of diet and exercise for weight management
	  ○ Medications to manage associated comorbid conditions, such as thyroid disease (must not have active disease), diabetes, dyslipidemia, hypertension, or cardiovascular

	conditions **<u>Reauthorization</u>** requires documentation of disease responsiveness to therapy based on improvements or stability in laboratory results, such as ALT and AST, or fibrosis as evaluated by a non-invasive test
Exclusion Criteria:	• History of excessive alcohol use or alcohol-associated liver disease • Current excessive alcohol use • Continued use of medications associated with liver steatosis • Stage 4 liver disease or cirrhosis • Use for other liver disease • Active or untreated thyroid disease
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hepatologist or gastroenterologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Immune reconstitution in pediatric patients with congenital athymia

Required Medical Information:	• Documentation of congenital athymia associated with one of the following:
	  ○ Complete DiGeorge Syndrome (cDGS)
	  ○ Forkhead Box N1 (*FOXN1*) deficiency
	  ○ 22q11.2 deletion
	  ○ CHARGE Syndrome (Coloboma, Heart defects, Atresia of the nasal choanae, Retardation of growth and development, Genitourinary anomalies, Ear anomalies)
	  ○ CHD7 mutation
	  ○ 10p13-p14 deletion

Appropriate Treatment Regimen & Other Criteria:	• Congenital athymia confirmed by flow cytometry that demonstrates:
	  ○ Fewer than 50 naïve T cells/mm3 in the peripheral blood OR
	  ○ Less than 5% of total T cells being naïve T cells

Exclusion Criteria:	• Treatment of patients with severe combined immunodeficiency (SCID) • Prior thymus transplant
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a pediatric immunologist or prescriber experienced in the treatment of congenital athymia • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 1 month (1 treatment only), unless otherwise specified

POLICY NAME:
RILONACEPT
Affected Medications: ARCALYST (rilonacept)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ 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
	  ○ The maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in adults and pediatric patients weighing at least 10 kg
	  ○ 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:
	• Diagnosis of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome (FCAS), and Muckle-Wells Syndrome (MWS)
	• Diagnosis of Deficiency of Interleukin-1 Receptor Antagonist (DIRA)
	  ○ Must include genetic testing results which confirm the presence of homozygous mutations in the interleukin-1 receptor antagonist (IL1RN) gene
	  ○ Disease must currently be in remission
	• Diagnosis of Recurrent Pericarditis with an inflammatory phenotype shown by one of the following:
	  ○ Fever, elevated C-Reactive protein (CRP), elevated white blood cell count, elevated erythrocyte sedimentation rate (ESR), pericardial late gadolinium enhancement (LGE) on cardiac magnetic resonance (CMR), or pericardial contrast enhancement on computed tomography (CT) scan

Appropriate Treatment Regimen & Other Criteria:	**<u>All Indications:</u>** • Documented treatment failure or intolerable adverse event with trial of Kineret (anakinra)

	<u>Recurrent Pericarditis:</u>
	• Documented treatment failure or intolerable adverse event to triple therapy with all of the following:
	  ○ Colchicine
	  ○ Non-steroidal anti-inflammatory (NSAID) or aspirin
	  ○ Glucocorticoid
	
	<u>Reauthorization:</u>
	• **All indications:** documentation of treatment success and a clinically significant response to therapy
	• **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 • Concurrent therapy with anakinra, tumor necrosis factor (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 rheumatologist, immunologist, cardiologist, or dermatologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
RIOCIGUAT
Affected Medications: ADEMPAS (riociguat)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1
	  ○ Chronic-Thromboembolic Pulmonary Hypertension (WHO Group 4)

Required Medical Information:	**<u>Chronic Thromboembolic Pulmonary Hypertension (CTEPH)</u>**
	• Documentation of CTEPH (WHO Group 4) meeting the following criteria:
	  ○ Evidence of thromboembolic occlusion of proximal or distal pulmonary vasculature on CT/MRI or V/Q scan
	  ○ Mean pulmonary arterial pressure greater than 20 mm Hg
	  ○ PAWP less than 15 mm Hg
	  ○ Elevated pulmonary vascular resistance over 2 Wood units
	
	<u>Pulmonary Arterial Hypertension (PAH)</u>
	• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:
	  ○ Mean pulmonary artery pressure of at least 20 mm Hg
	  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg
	  ○ Pulmonary vascular resistance of at least 2.0 Wood units
	• Etiology of PAH (idiopathic, heritable, or associated with connective tissue disease)
	• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class II or higher symptoms
	• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blocker) unless there are contraindications:
	  ○ 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:	**<u>CTEPH</u>** • Documentation of failure of or inability to receive pulmonary endarterectomy surgery • Current therapy with anticoagulants **<u>PAH</u>** • Documented failure to the following therapy classes: Phosphodiesterase type 5 (PDE5) inhibitors **AND** endothelin receptor antagonists **<u>Reauthorization</u>** requires documentation of treatment success defined as one or more of the following: • Improvement in walking distance • Improvement in exercise ability • Improvement in pulmonary function • Improvement or stability in WHO functional class
Exclusion Criteria:	• Concomitant use with nitrates or nitric oxide donors (such as amyl nitrite) • Concomitant use with specific PDE-5 inhibitors (such as sildenafil, tadalafil, or vardenafil) or non-specific PDE inhibitors (such as dipyridamole or theophylline)
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a cardiologist or a pulmonologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
RISDIPLAM
Affected Medications: EVRYSDI (risdiplam)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Spinal muscular atrophy (SMA)

Required Medical Information:	• Diagnosis of SMA type 1, 2, or 3 confirmed by genetic testing of chromosome 5q13.2 demonstrating ONE of the following:
	  ○ Homozygous gene deletion of SMN1 (survival motor neuron 1)
	  ○ Homozygous gene mutation of SMN1
	  ○ Compound heterozygous gene mutation of SMN1
	• Documentation of 4 or fewer copies of the SMN2 (survival motor neuron 2) gene
	• Documentation of one of the following baseline motor assessments appropriate for patient age and motor function:
	  ○ Hammersmith Infant Neurological Examination (HINE-2)
	  ○ Hammersmith Functional Motor Scale (HFSME)
	  ○ Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)
	  ○ Upper Limb Module (ULM) test
	  ○ 6-Minute Walk Test (6MWT)
	• Documentation of previous treatment history
	• Documentation of ventilator use status:
	  ○ Patient is NOT ventilator-dependent (defined as using a ventilator at least 16 hours per day on at least 21 of the last 30 days)
	  ○ This does not apply to patients who require non-invasive ventilator assistance
	• Patient weight and planned treatment regimen

Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** requires documentation of improvement in baseline motor assessment score, clinically meaningful stabilization, or delayed progression of SMA-associated signs and symptoms
Exclusion Criteria:	• SMA type 4 • Advanced SMA at baseline (complete paralysis of limbs, permanent ventilation support)

	• Prior treatment with SMA gene therapy (i.e., onasemnogene abeparvovec-xioi) • Will not use in combination with other agents for SMA (e.g., onasemnogene abeparvovec-xioi, nusinersen, etc.)
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or provider who is experienced in treatment of spinal muscular atrophy • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
RITUXIMAB
Affected Medications: RITUXAN (rituximab), RITUXAN HYCELA (rituximab and hyaluronidase human), TRUXIMA (rituximab-abbs), RUXIENCE (rituximab-pvvr), RIABNI (rituximab-arrx)

Covered Uses:	• All Food and Drug Administration (FDA)-approved and compendia supported indications not otherwise excluded by plan design
	  ○ Rheumatoid arthritis (RA)
	  ○ Microscopic Polyangiitis (MPA)
	  ○ Granulomatosis with Polyangiitis (GPA)
	  ○ Eosinophilic granulomatosis with polyangiitis (EGPA)
	  ○ Relapsing forms of multiple sclerosis (MS)
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive disease (SPMS)
	  ○ Neuromyelitis Optica Spectrum Disorder (NMOSD)
	  ○ Pemphigus Vulgaris (PV) and other autoimmune blistering skin diseases
	  ○ Thrombocytopenia in patients with immune thrombocytopenia (ITP)
	• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher

Required Medical Information:	• Documentation of disease staging, all prior therapies used, and anticipated treatment course
	
	<u>RA</u>
	• Documentation of moderate to severe disease despite current treatment
	• Documented current level of disease activity with one of the following (or equivalent objective scale):
	  ○ Disease Activity Score derivative for 28 joints (DAS-28) greater than 3.2
	  ○ Simplified Disease Activity Index (SDAI) greater than 11
	  ○ Clinical Disease Activity Index (CDAI) greater than 10
	  ○ Weighted RAPID3 of at least 2.3

	MPA or GPA
	• Documentation of active GPA or MPA
	
	EGPA
	• Non-severe disease: documentation of active EGPA
	OR
	• Severe disease: documentation of organ or life-threatening manifestations as defined by the American College of Rheumatology/Vasculitis Foundation (ACR/VF)
	
	Relapsing Forms of MS
	• Diagnosis confirmed with magnetic resonance imaging (MRI) per revised McDonald diagnostic criteria for multiple sclerosis (MS)
	  ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS
	
	NMOSD
	• Diagnosis of seropositive aquaporin-4 immunoglobulin G (AQP4-IgG) NMOSD confirmed by all of the following:
	  ○ Documentation of AQP4-IgG-specific antibodies on cell-based assay
	  ○ Exclusion of alternative diagnoses (such as multiple sclerosis)
	  ○ At least **one** core clinical characteristic:
	  ▪ Acute optic neuritis
	  ▪ Acute myelitis
	  ▪ Acute area postrema syndrome (episode of otherwise unexplained hiccups or nausea/vomiting)
	  ▪ Acute brainstem syndrome
	  ▪ Symptomatic narcolepsy **OR** acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesion on magnetic resonance imaging (MRI) [*see table below*]
	  ▪ Acute cerebral syndrome with NMOSD-typical brain lesion on MRI [*see table below*]

	<table><tr><th>Clinical presentation</th><th>Possible MRI findings</th></tr><tr><td>Diencephalic syndrome</td><td>• Periependymal lesion
	• Hypothalamic/thalamic lesion</td></tr><tr><td>Acute cerebral syndrome</td><td>• Extensive periependymal lesion
	• Long, diffuse, heterogenous, or edematous corpus callosum lesion
	• Long corticospinal tract lesion
	• Large, confluent subcortical or deep white matter lesion</td></tr></table>
	
	<u>PV and other autoimmune blistering skin diseases (such as but not limited to pemphigus foliaceus, bullous pemphigoid, cicatricial pemphigoid, epidermolysis bullosa acquisita, and paraneoplastic pemphigus)</u>
	• Diagnosis confirmed by biopsy
	• Documented severe or refractory disease with failure to conventional topical and oral systemic therapies
	
	<u>Thrombocytopenia in patients with ITP</u>
	• Platelet count less than 20,000/mcL **AND**
	• One of the following:
	  ○ Documented steroid dependence to maintain platelets/prevent bleeding for at least 3 months
	  ○ Lack of clinically meaningful response to corticosteroids (defined as inability to increase platelets to at least 50,000/mcL)

Appropriate Treatment Regimen & Other Criteria:	**<u>All Uses</u>** • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced

	• Coverage of Truxima, Rituxan, or Rituxan Hycela requires documentation of one of the following:
	  ○ 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
	
	Oncology Uses
	• Documentation of ECOG performance status of 1 or 2 OR Karnofsky performance score greater than 50%
	
	RA
	• Initial Course: Documented failure with two of the preferred pharmacy drugs (Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Xeljanz, Rinvoq)
	  ○ Dose is approved for up to 2 doses of 1,000 mg given 2 weeks apart
	• 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.
	
	Relapsing Forms of MS
	• Initial: May include one-time induction dose (e.g., 1,000 mg once every 2 weeks for 2 doses)
	• Maintenance: Approvable up to 2,000 mg annually. Higher doses will require documentation to support
	
	NMOSD
	• Initial: May include one-time induction dose (e.g., 1,000 mg once every 2 weeks for 2 doses)
	• Maintenance: Approvable up to 2,000 mg annually. Higher doses will require documentation to support (e.g., detection of CD19+ lymphocytes)
	
	MPA and GPA
	• Initial: May include one-time induction dose (e.g., 1,000 mg

	once every 2 weeks for 2 doses **or** 375 mg/m^2 once weekly for 4 doses), to be used in combination with a systemic glucocorticoid
	• Maintenance: Approvable for up to 1,000 mg annually. Higher doses will require documentation to support (e.g., positive ANCA titers, detection of CD19+ lymphocytes)
	
	<u>EGPA</u>
	• Non-severe disease:
	  ○ Documented treatment failure with a corticosteroid
	  ○ Documented treatment failure to an adequate trial (at least 12 weeks) with an oral immunosuppressive therapy: azathioprine, methotrexate, mycophenolate, leflunomide
	• Severe disease:
	  ○ Documentation that rituximab will be administered in combination with a systemic glucocorticoid
	
	<u>PV and other autoimmune blistering skin diseases</u>
	• Documentation that rituximab will be administered in combination with a systemic glucocorticoid (if appropriate)
	• Documented treatment failure with 12 weeks of a corticosteroid **AND**
	• Documented treatment failure with 12 weeks of an immunosuppressant at an adequate dose (e.g., azathioprine, mycophenolate, methotrexate, etc.) or other appropriate corticosteroid-sparing therapy
	
	<u>All other indications</u>
	• A Food and Drug Administration (FDA)-approved or compendia supported dose, frequency, and duration of therapy
	• Documented treatment failure of first-line recommended and conventional therapies
	
	<u>Reauthorization</u>: documentation of disease responsiveness to therapy

Exclusion	• MS: Concurrent anti-CD20-directed therapy or other disease-modifying medications indicated for the treatment of MS

Criteria:	• Other non-oncology indications: Concurrent use with targeted immune modulators
Age Restriction:	
Prescriber/Site of Care Restrictions:	• For RA, MPA, GPA, EGPA: Prescribed by, or in consultation with, a rheumatologist • For CLL, NHL: Prescribed by, or in consultation with, an oncologist • For MS, NMOSD: Prescribed by, or in consultation with, a neurologist or MS specialist • For PV: Prescribed by, or in consultation with, a dermatologist • All approvals are subjects to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: ○ PV, MPA, GPA, EGPA – 3 months, unless otherwise specified ○ Oncology – 4 months, unless otherwise specified ○ RA, MS, NMOSD – 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Primary hyperoxaluria type 1 (PH1)

Required Medical Information:	• A diagnosis of primary hyperoxaluria type 1 (PH1) confirmed by genetic testing confirming presence of AGXT gene mutation
	• Metabolic testing demonstrating elevated urinary **oxalate** excretion
	• Presence of clinical manifestations diagnostic of PH1 such as:
	  ○ Metabolic testing demonstrating elevated urinary **glycolate** excretion
	  ○ Normal levels of L-glyceric acid (elevation indicates PH type 2)
	  ○ Normal levels of hydroxy-oxo-glutarate (elevation indicates PH type 3)
	• For Rivfloza: eGFR of 30 or more

Appropriate Treatment Regimen & Other Criteria:	• For Rivfloza: Trial and failure or contraindication with Oxlumo **Reauthorization** requires documentation of the following criteria related to treatment success: • Reduction from baseline in urine or plasma oxalate levels • Improvement, stabilization, or slowed worsening of one or more clinical manifestation of PH1 (i.e., nephrocalcinosis, renal stone events, renal impairment, systemic oxalosis)
Exclusion Criteria:	• Diagnosis of primary hyperoxaluria type 2 or type 3 • Secondary hyperoxaluria • Concurrent use of another RNA interference drug for PH1
Age Restriction	• For Rivfloza: age in accordance with FDA labeling
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a nephrologist, urologist, geneticist, or specialist in the treatment of PH1 • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
ROMIPLOSTIM
Affected Medications: NPLATE (romiplostim)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Adult patients with immune thrombocytopenia (ITP) who have had an insufficient response to corticosteroids, immunoglobulins, or splenectomy
	  ○ 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
	  ○ Adult and pediatric patients (including term neonates) with acute exposure to myelosuppressive radiation doses

Required Medical Information:	**<u>Thrombocytopenia in patients with ITP</u>**
	• Documentation of **ONE** of the following:
	  ○ Platelet count less than 20,000/microliter
	  ○ Platelet count less than 30,000/microliter AND symptomatic bleeding
	  ○ Platelet count less than 50,000/microliter AND increased risk for bleeding (such as peptic ulcer disease, use of antiplatelets or anticoagulants, history of bleeding at higher platelet count, need for surgery or invasive procedure)
	
	<u>Hematopoietic syndrome of acute radiation syndrome</u>
	• Suspected or confirmed exposure to radiation levels greater than 2 gray (Gy)

Appropriate Treatment Regimen & Other Criteria:	• Current weight
	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced
	
	<u>Thrombocytopenia in patients with ITP</u>
	• Documentation of inadequate response, defined as platelets did not increase to at least 50,000/microliter, to the following therapies:
	  ○ **ONE** of the following:
	  ▪ Inadequate response with at least 2 therapies for immune thrombocytopenia, including corticosteroids, rituximab, or immunoglobulin

	▪ Splenectomy ○ Promacta **<u>Reauthorization (ITP only):</u>** • Response to treatment with platelet count of at least 50,000/microliter (not to exceed 400,000/microliter) **OR** • The platelet counts have not increased to a level of at least 50,000/microliter and member has NOT been on the maximum dose for at least 4 weeks **<u>Hematopoietic syndrome of acute radiation syndrome</u>** • Approved for one-time single subcutaneous injection of 10 mcg/kg
Exclusion Criteria:	• Treatment of thrombocytopenia due to myelodysplastic syndrome (MDS) • Use in combination with another thrombopoietin receptor agonist, spleen tyrosine kinase inhibitor, or similar treatments (Promacta, Nplate, Tavalisse)
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hematologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	**Thrombocytopenia in patients with ITP** • Initial Approval: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified **Hematopoietic syndrome of acute radiation syndrome** • Approval: 1 month, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of osteoporosis in postmenopausal women at high risk for fracture, defined as one of the following:
	  • History of osteoporotic fracture
	  • Multiple risk factors for fracture
	  • 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:
	  ○ 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
	  ○ 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:
	  ▪ FRAX 10-year probability of major osteoporotic fracture is 20% or greater
	  ▪ FRAX 10-year probability of hip fracture is 3% or greater
	  ○ 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:
	  ○ Intravenous bisphosphonate (zoledronic acid or ibandronate)
	  ○ Prolia (denosumab)
	
	<u>Total duration of therapy with Evenity should not exceed 12 months in a lifetime</u>

Exclusion Criteria:	• Heart attack or stroke event within the preceding year • Concurrent use of bisphosphonates, parathyroid hormone

	analogs, or RANK ligand inhibitors • Hypocalcemia that is uncorrected prior to initiating Evenity
Age Restriction:	
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months (no reauthorization), unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Lennox-Gastaut Syndrome(LGS)

Required Medical Information:	**<u>All Indications</u>**
	• Patient weight
	• Documentation that rufinamide will be used as adjunctive therapy
	
	<u>Lennox-Gastaut Syndrome (LGS)</u>
	• Documentation of at least 8 drop seizures per month while on stable antiepileptic drug therapy
	• Documented treatment and inadequate seizure control with at least three guideline directed therapies including:
	  ○ Valproate **and**
	  ○ Lamotrigine **and**
	  ○ Topiramate, felbamate, **or** clobazam

Appropriate Treatment Regimen & Other Criteria:	• Dosing: not to exceed 3200 mg daily **<u>Reauthorization</u>** requires documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Familial Short QT syndrome • Use as monotherapy for seizure control
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
RYPLAZIM
Affected Medications: RYPLAZIM

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Plasminogen Deficiency Type 1

Required Medical Information:	**<u>Plasminogen Deficiency type 1 (must meet all of the following):</u>**
	• Diagnosis of symptomatic congenital plasminogen deficiency (C-PLGD) as evidenced by documentation of all of the following:
	  ○ Genetic testing showing presence of biallelic pathogenic variants in plasminogen (PLG)
	  ○ Baseline plasminogen activity level less than or equal to 45% of laboratory standard
	  ○ Presence of (ligneous) pseudomembranous lesions with documented size, location and number of total lesions
	• Abnormal plasminogen antigen plasma level less than 9 mg/dL as confirmed by an enzyme-linked immunosorbent assay
	• 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 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) • If plasminogen activity is less than 10% above baseline level then increase to every 2 day dosing • If between 10-20% of baseline then maintain every 3 day dosing • If above 20% of baseline then change dosing to every 4 days. **<u>Maintain dosing frequency as determined above for 12 weeks while treating active lesions</u>** • If lesions do not resolve by 12 weeks, or there are new or recurrent lesions, increase dosing frequency in one-day

	increments every 4-8 weeks up to Q2D dosing while reassessing clinical improvement until lesion resolution or until the lesions stabilize without further worsening.
	• If desired clinical change does not occur by 12 weeks, check trough plasminogen activity level.
	  ○ 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.
	  ○ 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
	
	***If lesions resolve by 12 weeks, continue at same dosing frequency and monitor for new or recurrent lesions every 12 weeks.
	
	• Dosing may not exceed 6.6 mg/kg every 2 days.
	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced.
	
	<u>Reauthorization (must meet all of the following):</u>
	• Trough plasminogen activity level (taken 72 hours after dose) greater than 10% above baseline level
	• Documented improvement (reduction) in lesion size and number
	• Dosing may not exceed 6.6 mg/kg every 2 days.

Exclusion Criteria:	• Prior treatment failure with Ryplazim • 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 • Prescribed by, or in consultation with, a hematologist in coordination with Hemophilia Treatment Center (HTC) or other

	specialized center of excellence
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
SACROSIDASE
Affected Medications: SUCRAID (sacrosidase)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Oral replacement therapy for congenital sucrase-isomaltase deficiency (CSID)

Required Medical Information:	• Documentation of confirmed congenital sucrose-isomaltase deficiency, diagnosed by one of the following:
	  ○ Small bowel biopsy
	  ○ Sucrose breath test
	  ○ Genetic test
	• Documentation of current symptoms (e.g., diarrhea, abdominal pain or cramping, bloating, gas, loose stools, nausea, vomiting)
	
	Reauthorization: requires documentation of treatment success and a clinically significant response to therapy (fewer stools, lower number of symptoms)

Appropriate Treatment Regimen & Other Criteria:	
Exclusion Criteria:	
Age Restriction:	• 5 months of age or older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a gastroenterologist or genetic specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

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

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

<table>
<tr><td>Covered Uses:</td><td>• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
○ Neuromyelitis optica spectrum disorder (NMOSD) in adults who are anti-aquaporin-4 (AQP4) antibody positive</td></tr>
<tr><td>Required Medical Information:</td><td><u>NMOSD</u>
• Diagnosis of seropositive aquaporin-4 immunoglobulin G (AQP4-IgG) NMOSD confirmed by all the following:
○ Documentation of AQP4-IgG-specific antibodies on cell-based assay
○ Exclusion of alternative diagnoses (such as multiple sclerosis)
○ At least one core clinical characteristic:
▪ Acute optic neuritis
▪ Acute myelitis
▪ Acute area postrema syndrome (episode of otherwise unexplained hiccups or nausea/vomiting)
▪ Acute brainstem syndrome
▪ Symptomatic narcolepsy OR acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesion on magnetic resonance imaging (MRI) [see table below]
▪ Acute cerebral syndrome with NMOSD-typical brain lesion on MRI [see table below]
<table><tr><th>Clinical presentation</th><th>Possible MRI findings</th></tr><tr><td>Diencephalic syndrome</td><td>• Periependymal lesion
• Hypothalamic/thalamic lesion</td></tr><tr><td>Acute cerebral syndrome</td><td>• Extensive periependymal lesion
• Long, diffuse, heterogenous, or</td></tr></table></td></tr>
</table>

		edematous corpus callosum lesion • Long corticospinal tract lesion • Large, confluent subcortical or deep white matter lesion
	• History of at least 1 attack in the past year, or at least 2 attacks in the past 2 years, requiring rescue therapy	
Appropriate Treatment Regimen & Other Criteria:	• Documented inadequate response, contraindication, or intolerance to rituximab (preferred agents Riabni and Ruxience) **<u>Reauthorization</u>** requires documentation of treatment success	
Exclusion Criteria:	• Active Hepatitis B Virus (HBV) infection • Active or untreated latent tuberculosis • Concurrent with other disease-modifying biologics for requested indication	
Age Restriction:	• 18 years of age or older	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or neuro-ophthalmologist • All approvals are subject to utilization of the most cost-effective site of care	
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified	

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of Lysosomal Acid Lipase (LAL) deficiency

Required Medical Information:	• Diagnosis of LAL deficiency or Rapidly Progressive LAL deficiency within the first 6 months of life confirmed by one of the following:
	  ○ Absence or deficiency in lysosomal acid lipase activity
	  ○ Mutation in the lipase A, lysosomal acid type (*LIPA*) gene
	• Documentation of patient weight
	• Documentation of prescribed treatment regimen (dose and frequency)
	• Baseline fasting lipid panel including LDL-c prior to initiating therapy (not required for Rapidly Progressive LAL deficiency)

Appropriate Treatment Regimen & Other Criteria:	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced **<u>Reauthorization</u>**: • Rapidly Progressive LAL deficiency: documentation of improvement in weight-for-age Z-score • LAL deficiency: documentation of improvement in LDL-c
Exclusion Criteria:	
Age Restriction:	• 1 month of age or older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist or metabolic specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

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

POLICY NAME:
SELUMETINIB
Affected Medications: KOSELUGO (selumetinib)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Neurofibromatosis type 1 with symptomatic, inoperable plexiform neurofibromas in pediatric patients 2 years of age and older • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better

Required Medical Information:	• Documented body surface area (BSA) and requested dose
	
	<u>Neurofibromatosis type 1 (NF1) with inoperable plexiform neurofibromas</u>
	• Documentation of diagnosis of symptomatic and/or progressive, inoperable NF1, defined as one or more plexiform neurofibromas that cannot be completely removed without risk for substantial morbidity due to encasement of, or close proximity to, vital structures, invasiveness, or high vascularity
	• Documentation of 2 or more of the following clinical diagnostic criteria as evaluated by a multidisciplinary specialist care team (A child of a parent with NF1 can be diagnosed if one or more of these criteria are met):
	  ○ 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
	  ○ Freckling in the axillary or inguinal region
	  ○ Two or more neurofibromas of any type or one plexiform neurofibroma
	  ○ Optic pathway glioma
	  ○ Two or more iris Lisch nodules identified by slit lamp examination or two or more choroidal abnormalities

	○ A distinctive osseous lesion such as sphenoid dysplasia, anterolateral bowing of the tibia, or pseudarthrosis of a long bone ○ A heterozygous pathogenic NF1 variant with a variant allele fraction of 50% in apparently normal tissue such as white blood cells **NCCN Indications** • 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 • For NF1: defined as a decrease in tumor volume from baseline and improvement in symptoms, such as pain
Exclusion Criteria:	**NCCN Indications** • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater
Age Restriction:	**Neurofibromatosis type 1 (NF1) with inoperable plexiform neurofibromas** • 2 to 18 years of age
Prescriber/Site of Care Restrictions:	**Neurofibromatosis type 1 (NF1) with inoperable plexiform neurofibromas** • Prescribed by, or in consultation with, a pediatric oncologist or specialist with experience in the treatment of neurofibromatosis **NCCN Indications** • Prescribed by, or in consultation with, an oncologist
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
SEROSTIM
Affected Medications: SEROSTIM (somatropin)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ HIV (human immunodeficiency virus)-associated wasting, cachexia

Required Medical Information:	• Documentation of current body mass index (BMI), actual body weight, and ideal body weight (IBW)
	• Serostim is used in combination with antiretroviral therapy to which the patient has documented compliance
	• Alternative causes of wasting (e.g., inadequate nutrition intake, malabsorption, opportunistic infections, hypogonadism) have been ruled out or treated appropriately
	• 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
	• Diagnosis of HIV-association wasting syndrome or cachexia confirmed by **one** of the following:
	  ○ Unintentional weight loss greater than or equal to 10% of body weight over prior 12 months
	  ○ Unintentional weight loss greater than or equal to 5% of body weight over prior 6 months
	  ○ BMI less than 20 kg/m^2
	  ○ Weight is less than 90% of IBW

Appropriate Treatment Regimen & Other Criteria:	**<u>Reauthorization</u>:** • Documentation of treatment success and clinically significant response to therapy (e.g., improved or stabilized BMI, increased physical endurance compared to baseline, etc.) • Documentation of continued compliance to antiretroviral regimen

Exclusion Criteria:	• Acute critical illness due to complications following open heart or abdominal surgery, multiple accidental trauma or acute respiratory failure • Active malignancy • Acute respiratory failure • Active proliferative or severe non-proliferative diabetic retinopathy
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an infectious disease specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 8 months (maximum duration of therapy 48 weeks total)

POLICY NAME:
SIGNIFOR
Affected Medications: SIGNIFOR (pasireotide)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Cushing's disease

Required Medical Information:	• Documented diagnosis of Cushing's disease
	• Documentation of at least **TWO** of the following:
	  ○ Mean 24-hour urine free cortisol (mUFC) greater than 1.5 times the upper limit of normal (ULN) for the assay (at least two measurements)
	  ○ Bedtime salivary cortisol greater than 145 ng/dL (at least two measurements)
	  ○ Overnight dexamethasone suppression test (DST) with a serum cortisol greater than 1.8 mcg/dL

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure or intolerable adverse event to ketoconazole and cabergoline • Documentation confirming pituitary surgery is not an option **OR** previous surgery has not been curative **Reauthorization** requires documentation of treatment success defined as mUFC normalization (i.e., less than or equal to the ULN)
Exclusion Criteria:	• Severe hepatic impairment (Child Pugh C)
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Acromegaly
	  ○ Cushing's disease

Required Medical Information:	**<u>Acromegaly</u>**
	• Documentation confirming clinical manifestations of disease
	• Diagnosis of acromegaly confirmed by **ONE** of the following:
	  ○ Elevated pre-treatment serum insulin-like growth factor-1 (IGF-1) level for age/gender
	  ○ Serum growth hormone (GH) level of 1 microgram/mL or greater after an oral glucose tolerance test (OGTT)
	
	<u>Cushing's Disease</u>
	• Documented diagnosis of Cushing's disease
	• Documentation of at least **TWO** of the following:
	  ○ Mean 24-hour urine free cortisol (mUFC) greater than 1.5 times the upper limit of normal (ULN) for the assay (at least two measurements)
	  ○ Bedtime salivary cortisol greater than 145 ng/dL (at least two measurements)
	  ○ Overnight dexamethasone suppression test (DST) with a serum cortisol greater than 1.8 mcg/dL

Appropriate Treatment Regimen & Other Criteria:	**<u>Acromegaly</u>**
	• Documented treatment failure or intolerance to **ONE** of the following: lanreotide (Somatuline Depot), Sandostatin LAR, or pegvisomant (Somavert)
	• Documentation confirming **ONE** of the following:
	  ○ Inadequate response to surgery or radiotherapy
	  ○ Not a candidate for surgical management or radiotherapy (e.g., medically unstable, high risk for complications under anesthesia, major systemic complications of acromegaly, severe hypertension, uncontrolled diabetes, etc.)

	• **Dosing:** Not to exceed 60 mg every 4 weeks (after 3 months of 40 mg) **<u>Reauthorization</u>** requires documentation of treatment success shown by decreased/normalized IGF-1 or GH levels **<u>Cushing's Disease</u>** • Documentation confirming pituitary surgery is not an option **OR** previous surgery has not been curative • Documented treatment failure or intolerance to ketoconazole and cabergoline • **Dosing:** Not to exceed 40 mg every 4 weeks (after 4 months of 10 mg) **<u>Reauthorization</u>** requires documentation of treatment success defined as mUFC normalization (i.e., less than or equal to the ULN)
Exclusion Criteria:	• Severe hepatic impairment (Child Pugh C)
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
SILTUXIMAB
Affected Medications: SYLVANT (siltuximab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of patients with multicentric Castleman's disease (MCD) who are human immunodeficiency virus (HIV) negative and human herpesvirus-8 (HHV-8) negative • 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 • The diagnosis was confirmed by biopsy of lymph gland • Documented negative tests for HIV and HHV-8 • Patient weight
Appropriate Treatment Regimen & Other Criteria:	**Dosing** • **MCD:** 11 mg/kg intravenous (IV) infusion once every 3 weeks until treatment failure • **Cytokine release syndrome (CRS):** 11 mg/kg IV one time only • Availability: 100 mg and 400 mg vials • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced **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 • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• **MCD:**
	  ○ Initial Authorization: 4 months, unless otherwise specified
	  ○ Reauthorization: 12 months, unless otherwise specified
	• **CRS:** 1 month (1 dose only), unless otherwise specified

POLICY NAME:
SIPONIMOD
Affected Medications: MAYZENT (siponimod)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive multiple sclerosis (SPMS)

Required Medical Information:	• Diagnosis confirmed with magnetic resonance imaging (MRI), per revised McDonald diagnostic criteria for MS ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS

Appropriate Treatment Regimen & Other Criteria:	• Coverage of Mayzent (siponimod) requires documentation of one of the following:
	  ○ Documented disease progression or intolerable adverse event with one of the following: teriflunomide, dimethyl fumarate or fingolimod
	  ○ Currently receiving treatment with Mayzent (siponimod), excluding via samples or manufacturer's patient assistance program
	
	<u>Reauthorization</u> requires provider attestation of treatment success

Exclusion Criteria:	• Presence of CYP2C9*3/*3 genotype • Concurrent use of other disease-modifying medications indicated for the treatment of MS
Age Restriction:	

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or a MS specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
SODIUM PHENYLBUTYRATE
Affected Medications: SODIUM PHENYLBUTYRATE

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Adjunctive therapy in the chronic management of patients with urea cycle disorders (UCDs) involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC), or argininosuccinic acid synthetase (AS)
	  ○ Neonatal-onset deficiency (complete enzymatic deficiency, presenting within the first 28 days of life)
	  ○ Late-onset disease (partial enzymatic deficiency, presenting after the first month of life) with history of hyperammonemic encephalopathy

Required Medical Information:	• Diagnosis confirmed by blood, enzymatic, biochemical, or genetic testing
Appropriate Treatment Regimen & Other Criteria:	• Oral tablets require documented inability to use sodium phenylbutyrate powder • Documented treatment failure with dietary protein restriction and/or amino acid supplementation alone • Must be used in combination with dietary protein restriction **<u>Reauthorization</u>** will require **BOTH** of the following: • Documentation of treatment success defined as ammonia levels maintained within normal limits • That this drug continues to be used in combination with dietary protein restriction
Exclusion Criteria:	• Use for management of acute hyperammonemia
Age Restriction:	

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist experienced in the treatment of metabolic diseases • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
SOLRIAMFETOL
Affected Medications: SUNOSI (solriamfetol)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Excessive daytime sleepiness associated with narcolepsy
	  ○ Excessive daytime sleepiness associated with obstructive sleep apnea

Required Medical Information:	**<u>Narcolepsy</u>** • Diagnosis confirmed by polysomnography and multiple sleep latency test • Symptoms of excessive daytime sleepiness consistent with narcolepsy have been present for at least 3 months • An Epworth Sleepiness Scale score of more than 10 despite treatment **<u>Obstructive Sleep Apnea (OSA)</u>** • Diagnosis confirmed by sleep study • An Epworth Sleepiness Scale score of more than 10 despite drug treatment and current use of continuous positive airway pressure (CPAP) for at least 3 months • Documentation that CPAP use will be continued during treatment with solriamfetol **<u>All indications:</u>** • Documented trial and failure or contraindication to modafinil OR armodafinil • For narcolepsy only, documented trial and failure or contraindication to ONE of the following: methylphenidate, dextroamphetamine, lisdexamfetamine, amphetamine-dextroamphetamine **<u>Reauthorization</u>** requires clinically significant improvement in activities of daily living and in Epworth Sleepiness Scale score
Appropriate Treatment Regimen & Other Criteria:	

Exclusion Criteria:	• Use for other untreated causes of sleepiness • Concurrent use of sedative/hypnotic drugs or other central nervous system depressants
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a sleep specialist or neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design **<u>Octreotide, Sandostatin LAR:</u>** ○ Acromegaly ○ Symptomatic treatment of metastatic carcinoid tumors (carcinoid syndrome) ○ Symptomatic treatment of vasoactive intestinal peptide tumors (VIPomas) **<u>Lanreotide, Somatuline Depot:</u>** ○ Acromegaly ○ Carcinoid syndrome (to reduce the frequency of short-acting somatostatin analog rescue therapy) ○ Unresectable, well- or moderately-differentiated, locally advanced or metastatic gastroenteropancreatic neuroendocrine tumors (GEP-NETs) • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher
Required Medical Information:	**<u>Acromegaly</u>** • Initiation of therapy, patient meets the following: ○ Clinical evidence of acromegaly ○ Pre-treatment high insulin-like growth factor (IGF-1) level for age/gender ○ Patient had an inadequate or partial response to surgery and/or radiotherapy OR there is a clinical reason for avoidance of surgery or radiotherapy ○ Clinical reasons for avoidance of surgery or radiotherapy include: ▪ 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 ▪ Uncontrolled diabetes **<u>All other indications</u>** Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course
Appropriate Treatment Regimen & Other Criteria:	**<u>All indications</u>** • 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 • For patients experiencing breakthrough symptoms while taking the long-acting depot, supplementary doses of SQ octreotide may be necessary **<u>Sandostatin LAR</u>** • Coverage for the non-preferred product **Sandostatin LAR** is provided when **ONE** of the following criteria is met: o Currently receiving treatment with Sandostatin LAR, excluding when the product is obtained as samples or via manufacturer's patient assistance programs o Documented inadequate response or intolerable adverse event with Lanreotide, Somatuline Depot, OR Somavert (pegvisomant; acromegaly only) **<u>Lanreotide, Somatuline Depot</u>** • GEP-NETs must use 120 mg injection **<u>Reauthorization:</u>** • **Acromegaly:** requires that the IGF-1 level is decreased or normalized

	• **All other indications:** requires documentation of disease responsiveness to therapy
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist, endocrinologist, or gastroenterologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Approval: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
SOMAVERT
Affected Medications: SOMAVERT (pegvisomant)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Acromegaly

Required Medical Information:	• Documentation confirming clinical manifestations of disease
	• Diagnosis of acromegaly confirmed by **ONE** of the following:
	  ○ Elevated pre-treatment serum insulin-like growth factor-1 (IGF-1) level for age/gender
	  ○ Serum growth hormone (GH) level of 1 microgram/mL or greater after an oral glucose tolerance test (OGTT)

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure or intolerance to octreotide or lanreotide (Somatuline Depot)
	• Documentation confirming one of the following:
	  ○ Inadequate response to surgery or radiotherapy
	  ○ Not a candidate for surgical management or radiotherapy (e.g., medically unstable, high risk for complications under anesthesia, major systemic complications of acromegaly, severe hypertension, uncontrolled diabetes, etc.)
	• **Dosing:** Not to exceed 30 mg daily
	
	<u>Reauthorization</u> requires documentation of treatment success shown by decreased/normalized IGF-1 or GH levels

Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA) approved indications not otherwise excluded by plan design ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1

Required Medical Information:	• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:
	  ○ Mean pulmonary artery pressure of at least 20 mm Hg
	  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg
	  ○ Pulmonary vascular resistance of at least 5 Wood units
	  ○
	• Etiology of PAH: idiopathic PAH, hereditary PAH
	OR
	• PAH secondary to one of the following conditions:
	  ○ Connective tissue disease
	  ○ Simple, congenital systemic to pulmonary shunts at least 1 year following repair
	  ○ Drugs and toxins
	• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class II or III symptoms
	• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blockers) unless there are contraindications:
	  ○ Low systemic blood pressure (systolic blood pressure less than 90)
	  ○ Low cardiac index (cardiac index less than 2 L/min/m^2)
	OR
	  ○ Presence of severe symptoms (functional class IV)
	• Baseline 6-minute walk test (6MWD)

Appropriate Treatment Regimen & Other Criteria:	• Documentation that drug will be used as an add-on treatment with all the following (one from each category) at therapeutic doses for at least 90 days:
	  ○ Phosphodiesterase-5 (PDE-5) inhibitor: sildenafil, tadalafil
	  ○ Endothelin Receptor Antagonist: ambrisentan, bosentan, Opsumit
	  ○ Prostacyclin: treprostinil, epoprostenol, Ventavis

	• Documentation of inadequate response or intolerance to oral calcium channel blocking agents (nifedipine, diltiazem) if positive Acute Vasoreactivity Test • Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced **<u>Reauthorization</u>** requires documentation of treatment success defined as one or more of the following: • Improvement in walking distance (6MWD) • Improvement or stability in WHO functional class
Exclusion Criteria:	• Human immunodeficiency virus (HIV)-associated PAH • PAH associated with portal hypertension • Schistosomiasis-associated PAH • Pulmonary veno occlusive disease • Platelet count less than 50,000/mm^3 (50 x 10^9/L) • Hemoglobin (Hgb) at screening above gender-specific upper limit of normal (ULN)
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a cardiologist or pulmonologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
SPARSENTAN
Affected Medications: FILSPARI (sparsentan)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Reduce proteinuria in adults with primary immunoglobulin A nephropathy (IgAN) at risk of rapid disease progression

Required Medical Information:	• Diagnosis of primary immunoglobulin A nephropathy (IgAN) confirmed with biopsy
	• Documentation of **ONE** of the following (with labs current within 30 days of request):
	  ○ Proteinuria defined as equal to or greater than 1 g/day
	  ○ Urine protein-to-creatinine ratio (UPCR) greater than 1.5 g/g

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure (defined as proteinuria equal to or greater than 1 g/day **OR** UPCR greater than 1.5 g/g) with a minimum of 12 weeks of each of the following:
	  ○ Maximum tolerated dose of an angiotensin-converting enzyme (ACE) inhibitor or angiotensin receptor blocker (ARB)
	  ○ High dose glucocorticoid therapy such as oral prednisone or methylprednisolone (or an adverse effect to two or more glucocorticoid therapies that is not associated with the corticosteroid class)

Exclusion Criteria:	• Hepatic impairment (Child-Pugh class A-C)
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a nephrologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 9 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Generalized pustular psoriasis flares (GPP, also called von Zumbusch psoriasis)

Required Medical Information:	• Diagnosis of generalized pustular psoriasis as confirmed by the following:
	  ○ The presence of widespread sterile pustules arising on erythematous skin
	  ○ Pustulation is not restricted to psoriatic plaques
	• Signs and symptoms of an acute GPP flare of moderate-to-severe intensity as follows:
	  ○ A Generalized Pustular Psoriasis Physician Global Assessment (GPPGA) total score of greater than or equal to 3
	  ○ A GPPGA pustulation category subscore of greater than or equal to 2
	  ○ Greater than or equal to 5% body surface area (BSA) covered with erythema and the presence of pustules

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure of acute disease flare (or documented intolerable adverse event) with:
	  ○ A one-week trial of cyclosporine
	  AND
	  ○ Infliximab (preferred biosimilars Inflectra, Renflexis)
	• Treatment for each flare is limited to two 900 mg infusions of Spevigo separated by 1 week

Exclusion Criteria:	• Previous use of Spevigo • Erythrodermic plaque psoriasis without pustules or with pustules restricted to psoriatic plaques • Synovitis-acne-pustulosis-hyperostosis-osteitis syndrome • Drug-induced acute generalized exanthematous pustulosis

Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a dermatologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 1 month with no reauthorization, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Indicated, in conjunction with an oral antidepressant, for the treatment of:
	  ▪ Treatment-resistant depression (TRD) in adults
	  ▪ 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>** • Assessment of patient's risk for abuse or misuse • Baseline Patient Health Questionnaire-9 (PHQ-9) score (or other standard rating scale) **<u>Diagnosis of Major Depressive Disorder (MDD) with acute suicidal ideation or behavior:</u>** • Assessment of patient's risk for abuse or misuse • 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>**
	• Documented treatment failure (defined by less than 50% improvement in depression symptom severity using a standard rating scale such as a PHQ-9) to an adequate trial (at least 6 weeks each), or intolerance, of at least three antidepressants from at least two different classes, during the current depressive episode
	• Failure to respond to augmentation therapy such as:
	  ○ Two antidepressants with different mechanisms of action used concurrently
	  ○ An antidepressant and a second-generation antipsychotic used concurrently
	  ○ An antidepressant and lithium used concurrently
	  ○ An antidepressant and buspirone used concurrently
	  ○ An antidepressant and thyroid hormone used concurrently

- 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
- Spravato will be used in combination with an oral antidepressant (at a therapeutic dose)
- Dosing according to the approved label:

		Adults
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
	Weeks 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

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 measures depressive symptoms
- Spravato continues to be used in combination with an oral antidepressant

Major depressive disorder (MDD) with acute suicidal ideation or behavior:
- Documentation of current inpatient psychiatric hospitalization OR adequate documentation of why patient is not currently at inpatient level of care
- Spravato will be used in combination with an oral antidepressant
- Dosing: 84 mg twice weekly for 4 weeks maximum (No reauthorization unless requirements for TRD met)

Exclusion Criteria:	• Concomitant psychotic disorder • Bipolar or related disorders • History of substance use disorder • Use as an anesthetic agent • Pregnancy • Aneurysmal vascular disease (including thoracic and abdominal aorta, intracranial, and peripheral arterial vessels) or arteriovenous malformation • History of intracerebral hemorrhage • Hypersensitivity to esketamine, ketamine, or any of the excipients
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a psychiatrist who is REMS certified • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	Initial authorization: • Major depressive disorder (MDD) with acute suicidal ideation or behavior: 1 month (limit #24 nasal spray devices in 28 days of treatment only), unless otherwise specified • TRD: 2 months (Induction phase – maximum of 23 nasal spray devices in first 28 days followed by once weekly maintenance phase), unless otherwise specified Reauthorization: (TRD indication only): 6 months, unless otherwise specified

POLICY NAME:
STIRIPENTOL
Affected Medications: DIACOMIT (stiripentol)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of seizures associated with Dravet syndrome (DS)
Required Medical Information:	• Current Weight • Documentation that therapy is being used as adjunct to clobazam for seizures • Documentation of at least 4 generalized clonic or tonic-clonic seizures in the last month while on stable antiepileptic drug therapy

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment and inadequate control of seizures with at least four guideline directed therapies including:
	  ○ Valproate
	  ○ Clobazam
	  ○ Topiramate
	  ○ Clonazepam, levetiracetam, or zonisamide
	
	Reauthorization will require documentation of treatment success and a reduction in seizure severity, frequency, or duration

Exclusion Criteria:	
Age Restriction:	• 6 months of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Perinatal/infantile or Juvenile onset hypophosphatasia (HPP)

Required Medical Information:	**<u>Diagnosis of Perinatal/Infantile or Juvenile onset hypophosphatasia (HPP) with ALL of the following:</u>**
	• Age of onset less than 18 years
	• One of the following:
	  ○ Clinical manifestations consistent with hypophosphatasia at onset prior to age 18, such as: vitamin B6 dependent seizures, respiratory insufficiency, failure to thrive, non-traumatic fracture, dental abnormalities, low score on 6-minute walk test, low bone density score
	  ○ Skeletal abnormalities confirmed with radiographic imaging (such as flared and frayed metaphyses, widened growth plate, bowed arms or legs, rachitic chest deformity, craniosynostosis)
	• Genetic test confirming mutation of tissue-non-specific alkaline phosphatase (TNSALP) gene
	• Low level of serum alkaline phosphatase (ALP) evidenced by lab result below reference range for patient's age and gender
	• Elevated levels of one of the following:
	  ○ Urine or serum concentration of phosphoethanolamine (PEA)
	  ○ Serum concentration of pyridoxal 5'-phosphate (PLP) in the absence of vitamin supplements within one week prior to the test
	  ○ Urinary inorganic pyrophosphate (PPi)

Appropriate Treatment Regimen & Other Criteria:	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced ○ Please note: the 80 mg/0.8 mL vial is for patients weighing greater than 40 kilograms only

	<u>Reauthorization</u> requires documentation of:
	• Laboratory results confirming a decrease in urine concentration of urine or serum phosphoethanolamine (PEA), serum concentration of pyridoxal 5'-phosphate (PLP), or urinary inorganic pyrophosphate (PPi)
	• Improvement or stabilization in the clinical signs and symptoms of hypophosphatasia, such as:
	  ○ Radiographic evidence of improvement in skeletal deformities or growth
	  ○ Improvement in 6-minute walk test
	  ○ Improved bone density
	  ○ Reduction in fractures
	  ○ Respiratory function/breathing
	  ○ Improvement in developmental milestones

Exclusion Criteria:	• Other types of osteomalacia or hypophosphatasia, including adult onset hypophosphatasia
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an endocrinologist or specialist experienced in the treatment of metabolic bone disorders • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Primary immunodeficiency (PID)/Wiskott-Aldrich syndrome
	  ▪ Such as: x-linked agammaglobulinemia, common variable immunodeficiency (CVID), transient hypogammaglobulinemia of infancy, immunoglobulin G (IgG) subclass deficiency with or without immunoglobulin A (IgA) deficiency, antibody deficiency with near normal immunoglobulin levels) and combined deficiencies (severe combined immunodeficiencies, ataxia-telangiectasia, x-linked lymphoproliferative syndrome) [list not all inclusive]
	  ○ Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

Required Medical Information:	• Monthly intravenous immune globulin (IVIG) dose for those transitioning
	• Patient weight
	
	<u>Primary Immunodeficiency (PID)</u>
	• Type of immunodeficiency
	• Documentation of one of the following:
	  ○ Recent IgG level less than 200
	  ○ 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:
	  ▪ Four or more ear infections within 1 year
	  ▪ Two or more serious sinus infections within 1 year
	  ▪ Two or more months of antibiotics with little effect
	  ▪ Two or more pneumonias within 1 year
	  ▪ Recurrent or deep skin abscesses
	  ▪ Need for intravenous antibiotics to clear infections

	▪ Two or more deep-seated infections including septicemia • Documentation showing a deficiency in producing antibodies in response to vaccination including all of the following: o Titers that were drawn before challenging with vaccination o Titers that were drawn between 4 and 8 weeks after vaccination **<u>Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)</u>** • Documented baseline in strength/weakness has been documented using an objective clinical measuring tool (INCAT, Medical Research Council (MRC) muscle strength, 6 Minute Walk Test, Rankin, Modified Rankin) • Documented disease course is progressive or relapsing and remitting for 2 months or longer • Abnormal or absent deep tendon reflexes in upper or lower limbs • Electrodiagnostic evidence of demyelination indicated by one of the following: o Motor distal latency prolongation in 2 nerves o Reduction of motor conduction velocity in 2 nerves o Prolongation of F-wave latency in 2 nerves o Absence of F-waves in at least 1 nerve o Partial motor conduction block of at least 1 motor nerve o Abnormal temporal dispersion in at least 2 nerves o Distal CMAP duration increase in at least 1 nerve • Cerebrospinal fluid (CSF) analysis indicates all of the following (if electrophysiologic findings are non-diagnostic): o CSF white cell count of less than 10 cells/mm^3 o CSF protein is elevated (greater than or equal to 45mg/dL)
Appropriate Treatment	• Meets all criteria for IVIG approval • Exceptions may be given for patients without prior intravenous (IV) or subcutaneous (SC) immune globulin use

Regimen & Other Criteria:	**PID** • Documentation of at least 3 months of IVIG therapy **CIDP** • HyQvia, Hizentra and Gamunex-c only • Refractory to or intolerant of corticosteroids (prednisolone, prednisone) given in therapeutic doses over at least three months **Reauthorization:** • PID: Documented disease response defined as a decrease in the frequency or severity of infections • CIDP: ○ Documentation of a beneficial clinical response to maintenance therapy, without relapses, based on an objective clinical measuring tool OR ○ 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 • History of hypersensitivity to immune globulin or product components • Hyperprolinemia type I or II
Age Restriction:	• PID: 2 years of age and older • CIDP: 18 years of age and older
Prescriber/Site of Care Restrictions:	• PID: prescribed by, or in consultation with, an immunologist • CIDP: prescribed by, or in consultation with, a neurologist or rheumatologist with CIDP expertise
Coverage Duration:	Initial Authorization: • CIDP: 3 months, unless otherwise specified • PID: 12 months, unless otherwise specified Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of hemolysis in adults with cold agglutinin disease (CAD)

Required Medical Information:	**<u>Cold Agglutinin Disease (CAD)</u>**
	• Documentation of current weight
	• Diagnosis of CAD as confirmed by all of the following:
	  ○ Chronic hemolysis as confirmed by hemoglobin level of 10 g/dL or less AND elevated indirect bilirubin level
	  ○ Positive monospecific direct antiglobulin test (DAT) or Coombs test for C3d
	  ○ A positive DAT or Coombs test for IgG of 1+ or less
	  ○ Cold agglutinin titer of greater than or equal to 64 at 4°C

Appropriate Treatment Regimen & Other Criteria:	**<u>Cold Agglutinin Disease (CAD)</u>**
	• Dosing:
	  ○ 39 kg to less than 75 kg: 6,500 mg/dose
	  ○ 75 kg or greater: 7,500 mg/dose
	  ○ Administered weekly for the first two weeks, then every two weeks thereafter
	
	<u>Reauthorization:</u> documentation of disease responsiveness to therapy (e.g., increased hemoglobin, normalized markers of hemolysis [bilirubin, lactate dehydrogenase, reticulocyte count], reduced blood transfusion requirements)

Exclusion Criteria:	• Disease secondary to infection, rheumatologic disease, systemic lupus erythematosus, or overt hematologic malignancy • 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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of wild type or hereditary transthyretin amyloid cardiomyopathy (ATTR-CM) to reduce cardiovascular mortality and cardiovascular-related hospitalizations in adults

Required Medical Information:	• Diagnosis of ATTR-CM supported by **ONE** of the following (a, b, or c):
	  a. Cardiac tissue biopsy confirms presence of ATTR amyloid deposits by immunohistochemistry (IHC) or mass spectrometry
	  b. Documentation of **BOTH** of the following (i and ii):
	  i. Noncardiac tissue biopsy confirms presence of ATTR amyloid deposits by IHC or mass spectrometry
	  ii. Imaging consistent with cardiac amyloidosis (echocardiogram [ECG], cardiac magnetic resonance [CMR], or positron emission tomography [PET])
	  c. Documentation of **ALL** the following (i, ii, and iii):
	  i. Grade 2 to 3 uptake on cardiac scintigraphy (utilizing Tc-PYP, Tc-DPD, or Tc-HMDP radiotracers)
	  ii. Normal serum kappa/lambda free light chain (sFLC) ratio, serum protein immunofixation, **AND** urine protein immunofixation
	  iii. Imaging consistent with cardiac amyloidosis (ECG, CMR, or PET)
	• Documentation of New York Heart Association (NYHA) Functional Class I to III

Appropriate Treatment	**<u>Reauthorization</u>** requires documentation of disease responsiveness (improvement in symptoms, quality of life, or 6-

Regimen & Other Criteria:	Minute Walk Test; slowing or stabilization of disease progression; reduced cardiovascular-related hospitalizations, etc.)
Exclusion Criteria:	• NYHA Functional Class IV heart failure • Presence of light-chain (primary) amyloidosis • Prior liver or heart transplant • Implanted cardiac mechanical assist device • Combined use with transthyretin-lowering therapy
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a cardiologist or specialist experienced in the treatment of amyloidosis • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of blastic plasmacytoid dendritic cell neoplasm (BPDCN) in adults and in pediatric patients at least 2 years of age • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or better

Required Medical Information:	• Diagnosis of BPDCN is confirmed by ALL of the following:
	  ○ A biopsy showing the morphology of plasmacytoid dendritic blast cells
	  ○ At least 3 of the following plasmacytoid dendritic cell (pDC) markers are expressed by immunohistochemistry (IHC) or flow cytometry:
	  ▪ CD123
	  ▪ CD4
	  ▪ CD56
	  ▪ TCF4
	  ▪ TCL1
	  ▪ CD303
	  ▪ CD304
	  ○ The following pDC markers are negative: CD3, CD14, CD19, CD34, lysozyme, myeloperoxidase
	  ○ Diagnosis is made by a board-certified hematopathologist or dermatopathologist
	• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course

Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** requires documentation of disease responsiveness to therapy
Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater • Pregnancy

Age Restriction:	• 2 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a prescriber experienced in the treatment of BPDCN • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

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

1. Is there documented current disease activity with one of the following (or equivalent objective scale)? ○ Disease Activity Score derivative for 28 joints (DAS-28) greater than 3.2 ○ Clinical Disease Activity Index (CDAI) greater than 10 ○ Weighted Routine Assessment of Patient Index Data 3 (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 treatment with methotrexate, or if unable to tolerate methotrexate or contraindications apply, another disease modifying antirheumatic drug (sulfasalazine, hydroxychloroquine, leflunomide)?	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 (Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, 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) **Preferred Pharmacy Drugs –Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Cosentyx, Otezla, Stelara, Skyrizi, Tremfya** **Preferred Medical Drugs – Inflectra, Renflexis, Stelara** **Non-Preferred Medical Drugs – Remicade, Infliximab (J1745), Avsola**		
1. Is there documentation that the skin disease meets one of the following: ○ At least 10% body surface area involvement despite current treatment ○ Hand, foot, or mucous membrane involvement	Yes – Document and go to #2	No – Criteria not met
2. 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 #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 (Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Cosentyx, Otezla, Stelara, Skyrizi, Tremfya) 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 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) **Preferred Pharmacy Drugs – Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Otezla, Cosentyx, Xeljanz, Stelara, Tremfya, Rinvoq, Skyrizi** **Preferred Medical Drugs – Inflectra, Renflexis, Stelara, Simponi Aria** **Non-Preferred Medical Drugs – Remicade, Orencia IV, Infliximab (J1745), Avsola, Cosentyx Intravenous**		
1. Is there documentation of Classification for Psoriatic Arthritis (CASPAR) criteria score 3 or greater based on chart notes: ○ 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 ○ Nail lesions (onycholysis, pitting): one point ○ Dactylitis (present or past, documented by a rheumatologist): one point ○ Negative rheumatoid factor (RF): one point ○ Juxta-articular 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 (sulfasalazine, cyclosporine, leflunomide)?	Yes – Document and 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 (Hadlima,	Yes – Go to #5	No – Criteria not met

Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Cosentyx, Otezla, Stelara, Xeljanz, Tremfya, Rinvoq, Skyrizi) AND one of the preferred medical drugs (Inflectra, Renflexis, Simponi Aria)?		
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 **Preferred Pharmacy Drugs – Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Cosentyx, Xeljanz, Rinvoq** **Preferred Medical Drugs –Inflectra, Renflexis, Simponi Aria** **Non-preferred Medical Drugs –Remicade, Infliximab (J1745), Avsola, Cosentyx Intravenous**		
1. Is there a diagnosis of axial spondyloarthritis (SpA) confirmed by sacroiliitis on imaging AND at least 1 Spondyloarthritis (SpA) feature: ○ Inflammatory back pain (4 of 5 features met): ▪ Onset of back discomfort before the age of 40 years ▪ Insidious onset ▪ Improvement with exercise ▪ No improvement with rest ▪ Pain at night (with improvement upon arising) ○ Arthritis ○ Enthesitis	Yes – Go to #2	No – Criteria not met

○ Uveitis ○ Dactylitis (inflammation of entire digit) ○ Psoriasis ○ Crohn's disease/ulcerative colitis ○ Good response to NSAIDs ○ Family history of SpA ○ Elevated CRP **OR** ○ 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? OR 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
5. Is there documented failure with one of the preferred pharmacy drugs (Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, 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 (CD) **Preferred Pharmacy Drugs – Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Stelara, Skyrizi, Rinvoq** **Preferred Medical Drugs – Inflectra, Renflexis, Skyrizi Intravenous, Stelara** **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 documented failure with at least two oral treatments for a minimum of 12 weeks: corticosteroids, azathioprine, 6-mercaptopurine, methotrexate, sulfasalazine, balsalazide? **OR** 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: ○ Fistulizing disease ○ Stricture ○ Presence of abscess/phlegmon ○ Deep ulcerations	Yes – Document and go to #4	No – Criteria not met

○ 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 (Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Stelara, Skyrizi, 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
Ulcerative Colitis (UC) **Preferred Pharmacy Drugs – Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Rinvoq, Xeljanz, Stelara, Skyrizi** **Preferred Medical Drugs –Inflectra, Renflexis, Stelara, Skyrizi Intravenous** **Non-Preferred Medical Drugs –Remicade, Entyvio, Omvoh, 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	Yes – Document and got to #4	No – Go to #3

with severe cramps and evidence of systemic toxicity (fever, tachycardia, 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 (Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, 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) **Preferred Pharmacy Drugs – Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Enbrel, Xeljanz, Rinvoq** **Preferred Medical Drug – Simponi Aria** **Non-Preferred Medical Drugs – Orencia IV, Actemra IV, Tofidence IV, Tyenne 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 corticosteroids AND At least one of methotrexate or leflunomide for a minimum of 12 weeks?	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 (Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, 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 – Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz		
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	
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) **Preferred Pharmacy Drugs – Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Cosentyx** **Preferred Medical Drugs –Inflectra, Renflexis** **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	Yes – Document and go to #2	No – Criteria not met

Documentation of baseline count of abscess and inflammatory nodules?		
2. Is there documented failure with at least a 90-day trial of oral antibiotics for treatment of HS (Doxycycline/tetracycline/minocycline or clindamycin plus rifampin) AND 8 weeks on a retinoid (Isotretinoin, Acitretin)?	Yes – Document and 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 drug (Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz, Cosentyx) 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, Tofidence IV, Tyenne IV		
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 angiography (MRA), positron emission tomography (PET) or PET with CT?	Yes – Go to #3	No – Criteria not met
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

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

	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) Preferred Drugs - Cosentyx Juvenile Psoriatic Arthritis (JPsA) Preferred Drugs – Cosentyx, Enbrel		
1. Is there diagnosis of ERA confirmed by presence of the following: • Arthritis persisting at least 6 weeks AND enthesitis present **OR** • Arthritis or enthesitis with two of the following features: ○ Sacroiliac tenderness or inflammatory lumbosacral pain ○ Positive HLA-B27 ○ Onset of arthritis in males greater than 6 years of age ○ Acute symptomatic anterior uveitis ○ 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: • Arthritis and psoriasis **OR** • Arthritis and at least 2 of the following: ○ Dactylitis ○ Nail pitting or onycholysis ○ 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

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 (GPP) Flare **Preferred Drugs – Inflectra, Renflexis** **Non-Preferred Medical Drugs – Remicade, Avsola, Infliximab (J1745)**		
1. Is there documentation of a diagnosis of generalized pustular psoriasis (GPP) confirmed by the following: a. The presence of widespread sterile pustules arising on erythematous skin 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: a. A Generalized Pustular Psoriasis Physician Global Assessment (GPPGA) total score of greater than or equal to 3 b. A GPPGA pustulation category subscore of greater than or equal to 2	Yes – Document and go to #3	No – Criteria not met

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 – Document and go to #4	No – Criteria not met
4. Is the request for Remicade, Avsola, or Infliximab (J1745)?	Yes – Go to #5	No – Go to #6
5. Is there documented failure with 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 dermatology 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 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., Hadlima 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

PacificSource HEALTH PLANS

Quantity Limitations

- **Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz**
  - Induction
    - **PP/Uveitis:** 80 mg as a single dose, followed by 40 mg every other week beginning 1 week after initial dose (160 mg total in first 28 days)
    - **CD/UC/HS:** 160 mg on day 1, followed by 80 mg on day 15, then maintenance dosing beginning on day 29
  - Maintenance
    - **RA/PP/PsA/CD/UC/AS/nr-axSpA/Uveitis/JIA:** 40 mg every other week
    - **HS:** 40 mg every week **OR** 80 mg every other week
  - Dose escalation (40 mg every week **OR** 80 mg every other week)
    - **RA/PP/CD/UC:** Approval will require documentation of lost or inadequate response after a minimum of 16 weeks with standard maintenance dosing
- **Enbrel**
  - 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
    - Hidradenitis Suppurativa: 4 two-packs (300 mg) in 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
- Hidradenitis Suppurativa: 1 two-pack (300 mg) per 28 days

- **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
  - Ulcerative Colitis:
    - Induction: 1200 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)**
  - CD: 45 mg once daily for 12 weeks, then 15 mg once daily. May increase to 30 mg once daily for patients with refractory, severe or extensive disease.
    ****45mg limited to 84 tablets (first 12 weeks of treatment)**
  - Polyarticular JIA/Pediatric Psoriatic Arthritis: 10 kg to <20 kg: 3 mg (3 mL solution) twice daily; 20 kg to <30 kg: 4 mg (4 mL solution) twice daily; 30 kg and greater: 6 mg (6 mL solution) twice daily or 15 mg tablet once daily
- **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, Tofidence Intravenous, Tyenne 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)
  - GCA: 6mg/kg every 4 weeks
  - 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
Abatacept (Orencia SQ & Orencia IV)			≥2 yo		≥2 yo	≥18 yo		Acute GVHD prophylaxis: IV: ≥2 yo
Adalimumab (Hadlima, Hyrimoz (Cordavis), Adalimumab-adaz)	≥18 yo	≥6 yo ≥18 yo (biosimilars)	≥2 yo ≥4 yo (biosimilars)	≥18 yo	≥18 yo	≥18 yo	≥5 yo	Uveitis (noninfectious) ≥2 yo HS ≥12 yo
Anakinra (Kineret)						≥18 yo		NOMID
Apremilast (Otezla)				≥6 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 ≥2 yo HIDS ≥2 yo MKD ≥2 yo FMF ≥2 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		JPsA ≥2 yo
Golimumab (Simponi & Simponi Aria)	≥18 yo		≥2 yo (Simponi Aria)		≥18 yo (Simponi) ≥2 yo (Simponi Aria)	≥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	GPP≥18 yo
Ixekizumab (Taltz)	≥18 yo			≥6 yo	≥18 yo			Nr-axSpA ≥18 yo
Rituximab (Rituxan) [See standalone policy]						≥18 yo		CLL ≥18 yo NHL ≥18 yo; ≥6 yo (Rituxan) GPA ≥18 yo; ≥2 yo (Rituxan) Pemphigus Vulgaris ≥18 yo RRMS ≥18 yo
Risankizumab-rzaa (Skyrizi)		≥18 yo		≥18 yo	≥18 yo		≥18 yo	
Sarilumab (Kevzara)						≥18 yo		
Secukinumab (Cosentyx)	≥18 yo			≥6 yo	≥2 yo			Nr-axSpA ≥18 yo ERA ≥ 4 yo JPsA ≥ 2 yo HS ≥18 yo
Tildrakizumab-asmn (Ilumya)				≥18 yo				
Tocilizumab (Actemra SQ & Actemra IV)			≥2 yo			≥18 yo		CRS >2 yo GCA >18 yo

Tofacitinib (Xeljanz)	≥18 yo		≥2 yo		≥18 yo	≥18 yo	≥18 yo	
Upadacitinib (Rinvoq)	≥18 yo	≥18 yo			≥18 yo	≥18 yo	≥18 yo	AD ≥12 yo Nr-axSpA ≥18 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)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Reduce the loss of kidney function in adults with primary immunoglobulin A nephropathy (IgAN) who are at risk for disease progression
Required Medical Information:	• Diagnosis of primary immunoglobulin A nephropathy (IgAN) confirmed with biopsy • 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** • 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 with a minimum of 12 weeks of an angiotensin-converting enzyme (ACE) inhibitor or angiotensin receptor blocker (ARB) **AND** • Documentation of treatment failure with a minimum of 12 weeks of glucocorticoid therapy such as oral prednisone or methylprednisolone (treatment failure defined as proteinuria equal to or greater than 1 g/day or an adverse effect to two glucocorticoid therapies that is not associated with the corticosteroid class) **AND** • Documentation of treatment failure with a minimum of 12 weeks of Filspari (treatment failure defined as proteinuria equal to or greater than 1 g/day or an adverse effect to Filspari) **<u>No reauthorization</u>** – Recommended duration of therapy is 9 months followed by a 2-week dose taper prior to discontinuation
Exclusion Criteria:	• Other glomerulopathies or nephrotic syndrome
Age Restriction:	• 18 years of age and older

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a nephrologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 10 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of Non-24-Hour Sleep-Wake Disorder (Non-24)
	  ○ Treatment of nighttime sleep disturbances in Smith-Magenis Syndrome (SMS)

Required Medical Information:	**<u>Non-24</u>**
	• Documentation of being totally blind with no light perception
	• Diagnosis of Non-24 hour sleep wake disorder meeting ALL of the following:
	  ○ Documented history of insomnia, excessive daytime sleepiness, or both, that alternates with asymptomatic periods
	  ○ Symptoms have been present for at least three months
	  ○ Drift in rest-activity patterns demonstrated by at least 4 weeks of data from daily sleep logs and actigraphy
	  ○ Documentation that other sleep disorders were treated or ruled out using a sleep study
	
	<u>Smith-Magenis Syndrome (SMS)</u>
	• Diagnosis of Smith-Magenis Syndrome (SMS) confirmed by both of the following:
	  ○ Genetic test showing mutation or deletion of the retinoic acid-induced 1 (RAI1) gene
	  ○ Documentation of significant nighttime sleep disturbances

Appropriate Treatment Regimen & Other Criteria:	**<u>Non-24</u>** • Documentation of treatment failure with at least 12 weeks of melatonin **<u>Smith-Magenis Syndrome (SMS)</u>** • Documented trial and failure with treatment regimen that includes both melatonin taken at bedtime AND acebutolol taken during daytime for at least 12 weeks

	<u>Reauthorization</u> requires documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Sleep disorders other than Non-24 and SMS such as insomnia, shift work disorder, jet lag disorder, irregular sleep-wake rhythm disorder, delayed sleepwake phase disorder, advanced sleep-wake rhythm disorder • Sleep disturbances caused by taking sedative or stimulant central nervous system-active drugs • Sleep disturbances caused by other conditions

Age Restriction:	• Non-24: 18 years of age and older
	• SMS:
	  ○ Capsules: 16 years of age and older
	  ○ Suspension: 3 to 15 years of age

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

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Acute bacterial skin and skin structure infections (ABSSSI) caused by susceptible isolates of the following Gram-positive microorganisms:
	  ▪ Staphylococcus aureus (including methicillin-resistant [MRSA] and methicillin-susceptible [MSSA] isolates)
	  ▪ Streptococcus pyogenes
	  ▪ Streptococcus agalactiae
	  ▪ Streptococcus anginosus Group (including Streptococcus anginosus, Streptococcus intermedius, and Streptococcus constellatus)
	  ▪ Enterococcus faecalis

Required Medical Information:	• Documentation of confirmed or suspected diagnosis • Documentation of treatment history and current treatment regimen • Documentation of culture and sensitivity data • Documentation of planned treatment duration

Appropriate Treatment Regimen & Other Criteria:	• Dosing: 200 mg once daily for 6 days
	
	Requests for the intravenous formulation will require both of the following:
	• Documentation of treatment failure, contraindication, or intolerable adverse event with intravenous linezolid **AND**
	• Documentation of treatment failure, contraindication, or intolerable adverse event with at least 2 of the following drugs/drug classes:
	  ○ Vancomycin
	  ▪ Avoidance of vancomycin due to nephrotoxicity will require documentation of multiple (at least 2 consecutive) increased serum creatinine concentrations (increase of 0.5 mg/dL [44 mcmol/L] or at least 50 percent increase from baseline, whichever is greater), without an alternative explanation

	○ Daptomycin ○ Cephalosporin (cefazolin) Requests for the oral tablet formulation will require both of the following: • Documentation of treatment failure, contraindication, or intolerable adverse event with oral linezolid **AND** • Documentation of treatment failure, contraindication, or intolerable adverse event with at least 2 of the following drugs/drug classes: ○ Trimethoprim-sulfamethoxazole ○ Tetracycline (doxycycline, minocycline) ○ Clindamycin
Exclusion Criteria:	
Age Restriction:	• 12 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a infectious disease specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 1 month, unless otherwise specified.

POLICY NAME:
TEDUGLUTIDE
Affected Medications: GATTEX (teduglutide)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of Short Bowel Syndrome (SBS)
Required Medical Information:	• Documentation of confirmed SBS diagnosis • Dependence on parenteral nutrition (PN) and/or intravenous (IV) fluids at least 12 consecutive months continuously • Receiving three or more days per week of PN support such as fluids, electrolytes, and/or nutrients

Appropriate Treatment Regimen & Other Criteria:	• Documentation of inability to be weaned from PN despite use of the following conventional measures:
	  ○ Dietary manipulations, oral rehydration solutions
	  ○ Antidiarrheal/motility agents: loperamide or diphenoxylate
	  ○ Antisecretory agents: H2 receptor antagonists or proton pump inhibitors
	OR
	• Developed significant complications or severe impairment in quality of life related to parenteral nutrition use (such as loss of vascular access sites, recurrent catheter-related bloodstream infections, and liver disease)
	
	• Dose does not exceed 0.05 mg/kg daily
	
	<u>Reauthorization</u> requires documentation of clinically significant benefit defined by parenteral support reduction of 1 day or greater a week

Exclusion Criteria:	• Weight of less than 10 kg • Onset or worsening of gallbladder/biliary disease • Onset or worsening of pancreatic disease • Presence of any gastrointestinal malignancy • Presence of intestinal or stomal obstruction

Age Restriction:	• 1 year of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a gastroenterologist or SBS specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 6 months, unless otherwise specified

POLICY NAME:
TENAPANOR
Affected Medications: XPHOZAH (tenapanor)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of hyperphosphatemia associated with chronic kidney disease (CKD)
Required Medical Information:	• Diagnosis of hyperphosphatemia associated with CKD and currently on dialysis treatment • Documentation of progressively or persistently elevated serum phosphate that is greater than 5.5 mg/dL over the past 6 months despite adherence to phosphate binders and dietary restrictions • Documentation that Xphozah will be used as add-on therapy to phosphate binder therapy unless contraindicated or clinically significant adverse effects were experienced

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure with at least an 8-week trial, at maximally indicated doses, of two or more of the following:
	  ○ calcium acetate
	  ○ lanthanum carbonate
	  ○ sevelamer
	  ○ Velphoro
	  ○ Auryxia
	
	Reauthorization requires documentation of treatment success defined as reduction in serum phosphorus from pretreatment level and maintenance of serum phosphorus level at 5.5 mg/dL or lower

Exclusion Criteria:	• Known or suspected mechanical gastrointestinal obstruction
Age Restriction:	• 18 years of age and older

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

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ For the treatment of chronic hepatitis B virus (HBV) infection in adults and pediatric patients 6 years of age and older with compensated liver disease
Required Medical Information:	• Documentation confirming diagnosis of chronic hepatitis B infection • 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:
	  ○ Inadequate virologic response or intolerable adverse event to tenofovir disoproxil fumarate
	  ○ 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)
	  ○ 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)
	
	Reauthorization: documentation of treatment success and a clinically significant response to therapy

Exclusion Criteria:	• Decompensated hepatic impairment (Child-Pugh B or C)
Age Restriction:	• 6 years of age and older
Prescriber Restrictions:	• Prescribed by, or in consultation with, a hepatologist, gastroenterologist, or infectious disease specialist • All approvals are subject to utilization of the most cost-effective site of care

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

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

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

Documentation of the patient’s current body surface area (BSA) or height and weight to calculate BSA
Treatment plan, including planned dose and frequency

</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>

Availability: 2 mg/2 mL (1 mg/mL) single-dose vials
Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced

</td></tr>
<tr><td>Exclusion Criteria:</td><td>

Prior treatment with Tzield
Diagnosis of Stage 3 type 1 diabetes (clinical type 1 diabetes)
Diagnosis of Type 2 diabetes
Current active serious infection or chronic infection
Pregnant or lactating

</td></tr>
<tr><td>Age Restriction:</td><td>

8 to 45 years of age
See Required Medical Information for age requirements based on first-degree or second-degree relative

</td></tr>
<tr><td>Prescriber/Site of Care Restrictions:</td><td>

Prescribed by, or in consultation with, an endocrinologist
All approvals are subject to utilization of the most cost-effective site of care

</td></tr>
<tr><td>Coverage Duration:</td><td>

Authorization: 3 months, unless otherwise specified (one 14-day infusion only)

</td></tr>
</table>

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Thyroid Eye Disease (TED) regardless of TED activity or duration

Required Medical Information:	• Documentation that baseline disease is under control prior to starting therapy, as defined by one of the following:
	  ○ Patient is euthyroid (thyroid function tests are within normal limits)
	  ○ Patient has recent and mild hypo- or hyperthyroidism (thyroid function tests show free thyroxine (T4) and free triiodothyronine (T3) levels less than 50% above or below normal limits) and will undergo treatment to maintain euthyroid state
	• TED has an appreciable impact on daily life, defined as:
	  ○ Proptosis greater than or equal to 3 mm increase from baseline (prior to diagnosis of TED) and/or proptosis greater than or equal to 3 mm above normal for race and gender
	  OR
	  ○ Current moderate-to-severe active TED with a Clinical Activity Score (CAS) greater than or equal to 4 (on the 7-item scale) for the most severely affected eye and symptoms such as: lid retraction greater than or equal to 3 mm, moderate or severe soft tissue involvement, diplopia, and/or proptosis greater than or equal to 3 mm above normal for race and gender

Appropriate Treatment Regimen & Other Criteria:	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced • Evidence of stable, well-controlled disease if comorbid inflammatory bowel disease (IBD) or diabetes • Documented failure to intravenous or oral steroid at appropriate dose over 12 weeks

Exclusion Criteria:	• Use of more than one course of Tepezza treatment • Prior orbital irradiation, orbital decompression, or strabismus surgery • Decreasing visual acuity, new defect in visual field, color vision defect from optic nerve involvement within the previous 6 months • Corneal decompensation that is unresponsive to medical management
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an ophthalmologist • 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:
TESTOPEL
Affected Medications: TESTOPEL (testosterone pellets)

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

	○ The clinical rationale for supporting the client's request for cross-hormone therapy and statement that the client meets eligibility criteria; ○ Informed consent required from both patient and guardian documented by prescribing provider ○ Permission to contact the licensed mental health professional for coordination of care • 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 • **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 • Reauthorization: documentation of recent testosterone levels within normal limits **Gender Dysphoria:** • 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 • 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:
TEZEPELUMAB-EKKO
Affected Medications: TEZSPIRE (tezepelumab-ekko)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Add-on maintenance treatment of patients aged 12 years and older with severe asthma

Required Medical Information:	• Diagnosis of severe asthma defined by the following:
	  ○ For adults: FEV1 less than 80% at baseline or FEV1/FVC reduced by at least 5% from normal
	  ○ For adolescents aged 12 to 17: FEV1 less than 90% at baseline or FEV1/FVC reduced by at least 5% from normal

Appropriate Treatment Regimen & Other Criteria:	• Documented use of high-dose inhaled corticosteroid (ICS) plus a long-acting beta agonist (LABA) for at least three months with continued symptoms • 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 with at least 80% adherence **Reauthorization:** documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Use in combination with another monoclonal antibody (e.g., Fasenra, Nucala, Xolair, Dupixent, Cinqair)
Age Restriction:	• 12 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an allergist, immunologist, or pulmonologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
THALIDOMIDE
Affected Medications: THALOMID (thalidomide)

Covered Uses:	• All Food and Drug Administration (FDA)-approved or compendia-supported indications not otherwise excluded by plan design
	  ○ Multiple Myeloma (MM)
	  ○ Erythema Nodosum (ENL)
	  ○ Systemic light chain amyloidosis
	  ○ AIDS-related aphthous stomatitis
	  ○ Waldenström macroglobulinemia
	  ○ Graft-versus-host disease, chronic (refractory)
	• 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>** • NCCN (National Comprehensive Cancer Network) regimen with evidence level of 2A or higher **<u>Systemic light chain amyloidosis</u>** • NCCN (National Comprehensive Cancer Network) regimen with evidence level of 2A or higher **<u>Waldenström Macroglobulinemia</u>** • NCCN (National Comprehensive Cancer Network) regimen with evidence level of 2A or higher **<u>AIDS-related or Severe recurrent aphthous stomatitis</u>** • Documented trial and failure with BOTH topical and systemic corticosteroids **<u>Erythema Nodosum Leprosum (ENL)</u>** • Acute treatment of the cutaneous manifestations of moderate to severe ENL (Type 2 reaction) • Maintenance therapy for prevention and suppression of the cutaneous manifestations of ENL recurrence

	<u>Reauthorization</u>: Documentation of disease responsiveness to therapy
Exclusion Criteria:	• Pregnancy • Karnofsky Performance Status less than or equal to 50% or ECOG performance score greater than or equal to 3
Age Restriction:	• 12 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist or infectious disease specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
THYMOGLOBULIN
Affected Medications: THYMOGLOBULIN

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.
	• Renal transplant acute rejection treatment and induction therapy
	• Off-label uses:
	  ○ Heart transplant
	  ○ Intestinal and multivisceral transplantation
	  ○ Lung transplant
	  ○ 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 • Prophylaxis: 1.5mg/kg of body weight administered daily for 4-7 days • Clinical rationale for avoiding Simulect (basiliximab) in prophylaxes
Exclusion Criteria:	• Active acute or chronic infections that contraindicates any additional immunosuppression
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Physicians experienced in immunosuppressive therapy for the management of renal transplant patients. • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial approval: 1 Month, unless otherwise specified • Reauthorization: 1 Month, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Management of Cystic Fibrosis (CF) patients with Pseudomonas aeruginosa

Required Medical Information:	• Diagnosis of Cystic Fibrosis (phenotyping not required).
	• Culture and sensitivity report confirming presence of pseudomonas aeruginosa in the lungs
	• Baseline forced expiratory volume in 1 second (FEV1)
	  ○ Tobi Podhaler: FEV1 equal to or between 25% and 80%
	  ○ Bethkis: FEV1 equal to or between 40% and 80%
	  ○ Kitabis Pak: FEV1 equal to or between 25% and 75%

Appropriate Treatment Regimen & Other Criteria:	• For Tobi Podhaler, Kitabis Pak, Bethkis, and Tobi: Documentation of failure with nebulized tobramycin or clinical rationale for avoidance • Use is limited to a 28 days on and 28 days off regimen **Reauthorization** requires documentation of improved respiratory symptoms and need for long-term use
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a pulmonologist, or provider who specializes in CF • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
TOFERSEN
Affected Medications: QALSODY (tofersen)

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

Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ **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)
	  ○ **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>** • Serum sodium less than 125 mEq/L at baseline **OR** • Serum sodium less than 135 mEq/L at baseline and symptomatic (nausea, vomiting, headache, lethargy, confusion) **<u>ADPKD</u>** • Diagnosis of typical ADPKD confirmed by family history, imaging, and if applicable, genetic testing • Estimated glomerular filtration rate (eGFR) greater than or equal to 25 mL/min/1.73m^2 • High risk for rapid progression determined by Mayo imaging class 1C, 1D, or 1E
Appropriate Treatment Regimen & Other Criteria:	**<u>Hyponatremia</u>** • Treatment is initiated or re-initiated in a hospital setting prior to discharge **<u>ADPKD</u>** • Documentation of intensive blood pressure control with an angiotensin-converting enzyme (ACE) inhibitor or angiotensin

	receptor blocker (ARB), unless contraindicated **<u>Reauthorization (for ADPKD)</u>** requires documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Patients requiring intervention to raise serum sodium urgently to prevent or treat serious neurological symptoms • Patients who are unable to sense or respond to thirst • Hypovolemic hyponatremia • Anuria • 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 • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	**<u>Hyponatremia</u>** • Authorization: 1 month (no reauthorization), unless otherwise specified **<u>ADPKD</u>** • Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

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

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design • 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 • Documentation of cutaneous T-cell lymphoma (CTCL), stage and type confirmed by biopsy. • Extent of skin involvement (limited/localized or generalized)

Appropriate Treatment Regimen & Other Criteria:	**<u>Limited/localized skin involvement (topical bexarotene and mechlorethamine)</u>**
	• Documented clinical failure to ALL of the following:
	  ○ Topical corticosteroids (high or super-high potency) such as clobetasol, betamethasone, fluocinonide, halobetasol
	  ○ Topical imiquimod
	  ○ Phototherapy
	
	<u>Generalized skin involvement (topical mechlorethamine only)</u>
	• Documentation of failure or contraindication to at least 1 skin-directed therapy
	
	<u>Reauthorization</u>: documentation of disease responsiveness to therapy

Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater • Pregnancy
Age Restriction:	• 18 years of age and older

Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
TOPICAL DERMATITIS AND PSORIATIC AGENTS
Affected Medications: VTAMA (tapinarof 1% cream), ZORYVE (roflumilast 0.3% cream), ZORYVE (roflumilast 0.3% foam), ZORYVE (roflumilast 0.15% cream)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Plaque psoriasis (Vtama and Zoryve 0.3% cream)
	  ○ Seborrheic dermatitis (Zoryve 0.3% foam)
	  ○ Atopic dermatitis (Zoryve 0.15% cream)

Required Medical Information:	**<u>Plaque Psoriasis</u>**
	• Diagnosis of chronic plaque psoriasis
	• Documentation that the skin disease meets **<u>ONE</u>** of the following:
	  ○ At least 10% body surface area (BSA) involvement despite current treatment
	  ○ Hand, foot, or mucous membrane involvement
	
	<u>Seborrheic Dermatitis</u>
	• Diagnosis of moderate to severe seborrheic dermatitis with presence of lesions that are characteristic of the condition (such as erythematous plaques and yellowish scales distributed on areas with sebaceous glands)
	• Documentation of persistent itching, scaling, and erythema despite current therapy
	
	<u>Atopic Dermatitis</u>
	• Documentation of atopic dermatitis that meets **<u>ONE</u>** of the following:
	  ○ At least 10% body surface area (BSA) involvement despite current treatment
	  ○ Hand, foot, face, or mucous membrane involvement

Appropriate Treatment Regimen & Other Criteria:	• For all indications, documented failure with a high or super-high potency topical corticosteroid (such as betamethasone dipropionate, clobetasol, fluocinonide, or halobetasol)

	<u>Plaque Psoriasis</u>
	• Documented failure with ALL the following:
	  ○ Calcipotriene cream or calcitriol ointment
	  ○ Tazarotene cream
	  ○ Vtama also requires documented treatment failure with 8 weeks of Zoryve
	
	<u>Seborrheic Dermatitis</u>
	• Documented failure with ALL of the following:
	  ○ Topical calcineurin inhibitor (such as tacrolimus or pimecrolimus)
	  ○ Topical antifungal (such as ketoconazole, ciclopirox, or selenium sulfide)
	
	<u>Atopic Dermatitis</u>
	• Documented failure with one of the following:
	  ○ Topical calcineurin-inhibitor (such as tacrolimus or pimecrolimus)
	  ○ Eucrisa
	
	<u>Reauthorization</u> will require documentation of disease responsiveness to therapy defined as:
	• For plaque psoriasis and atopic dermatitis, BSA reduction when compared to baseline
	• For seborrheic dermatitis, reduction in itching, scaling, and erythema when compared to baseline

Exclusion Criteria:	
Age Restriction:	• Vtama: 18 years of age and older • Zoryve cream: 6 years of age and older • Zoryve foam: 9 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a dermatologist, allergist, or immunologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
TRALOKINUMAB
Affected Medications: ADBRY (tralokinumab)

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? - 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

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**
  - Availability: 150 mg/mL prefilled syringes, 300 mg/2 mL autoinjectors
  - Dosing:
    - Adults 18 years and older: 600 mg as single dose, then 300 mg every 2 weeks.
      - If less than 100 kg and clear/almost clear is achieved, dosing may be reduced to 300 mg every 4 weeks
    - Pediatric patients 12 to 17 years old: 300 mg as a single dose, then 150 mg every 2 weeks.

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

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 • 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:	• Maximum duration for adjuvant breast cancer therapy is 12 months **<u>All Indications</u>** • Coverage for a non-preferred product (Trazimera, Herzuma, Ontruzant, Herceptin, or Herceptin Hylecta) requires documentation of the following: ○ 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 **<u>Reauthorization</u>** will require 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 • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• For new starts to adjuvant breast cancer therapy – approve for 12 months with no reauthorization
	• For all other clinical scenarios:
	  ○ Initial Authorization: 4 months, unless otherwise specified
	  ○ Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
TRIENTINE
Affected Medications: TRIENTINE HYDROCHLORIDE, CUVRIOR (trientine tetrahydrochloride)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. ○ Wilson's disease

Required Medical Information:	• Diagnosis of Wilson's disease confirmed by **ONE** of the following:
	  ○ Genetic testing results confirming biallelic pathogenic *ATP7B* mutations (in either symptomatic or asymptomatic individuals)
	  ○ Liver biopsy findings consistent with Wilson's disease
	  ○ Presence of Kayser-Fleischer (KF) rings **AND** serum ceruloplasmin level less than 20 mg/dL **AND** 24-hour urinary copper excretion greater than 40 mcg
	  ○ Presence of Kayser-Fleischer (KF) rings **AND** 24-hour urinary copper excretion greater than 100 mcg
	  ○ Absence of KF rings with serum ceruloplasmin level less than 10 mg/dL **AND** 24-hour urinary copper excretion greater than 100 mcg

Appropriate Treatment Regimen & Other Criteria:	• For Cuvrior, must meet **BOTH** of the following:
	  ○ Documented treatment failure with a minimum 6-month trial of penicillamine that was not due to tolerability
	  ○ Documented intolerable adverse event to a maximally tolerated dosage of generic trientine hydrochloride and the adverse event was not an expected adverse event attributed to the active ingredient
	
	Reauthorization: Documentation of treatment success and a clinically significant response to therapy as shown by normalization of free serum copper (non-ceruloplasmin bound copper) to less than 15 mcg/dL and 24-hour urinary copper in the range of 200 to 500 mcg

Exclusion Criteria:	For trientine hydrochloride: • Treatment of rheumatoid arthritis • Treatment of cystinuria • Treatment of biliary cirrhosis
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a hepatologist, gastroenterologist, or liver transplant provider • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization:12 months, unless otherwise specified

POLICY NAME:
TRIPTORELIN
Affected Medications: TRELSTAR, TRIPTODUR (triptorelin)

Covered Uses:	• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher
	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Prostate Cancer (Trelstar)
	  ○ Central Precocious Puberty (Triptodur)
	• Gender Dysphoria

Required Medical Information:	**<u>Central Precocious Puberty (CPP)</u>**
	• Documentation of CPP confirmed by one of the following labs:
	  ○ Elevated basal luteinizing hormone (LH) level greater than 0.2 - 0.3 mIU/L
	  ○ Elevated leuprolide-stimulated LH level greater than 3.3 - 5 IU/L (dependent on type of assay used)
	• Bone age greater than 2 standard deviations (SD) beyond chronological age
	
	<u>Gender Dysphoria</u>
	• Documentation of all the following:
	  ○ Current Tanner stage 2 or greater OR baseline and current estradiol and testosterone levels to confirm onset of puberty
	  ○ Confirmed diagnosis of gender dysphoria that is persistent
	  ○ The patient has the capacity to make a fully informed decision and to give consent for treatment
	  ○ Any significant medical or mental health concerns are reasonably well controlled
	  ○ A comprehensive mental health evaluation has been completed by a licensed mental health professional (LMHP) and provided in accordance with the most current version of the World Professional Association for Transgender Health (WPATH) Standards of Care

Appropriate Treatment	For all Triptodur requests: • Documentation of treatment failure with leuprolide

Regimen & Other Criteria:	**<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Use as neoadjuvant androgen deprivation therapy (ADT) for radical prostatectomy
Age Restriction:	• CPP: 2 years of age through 11 years for females, 2 years of age through 12 years for males
Prescriber/Site of Care Restrictions:	• Oncology: prescribed by, or in consultation with, an oncologist • CPP: prescribed by, or in consultation with, a pediatric endocrinologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Oncology Initial Authorization: 4 months, unless otherwise specified • CPP Approval/Oncology Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
TROFINETIDE
Affected Medications: DAYBUE (trofinetide)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of Rett syndrome (RTT)

Required Medical Information:	• Documented diagnosis of typical RTT (per the revised diagnostic criteria for Rett Syndrome) AND a period of regression followed by recovery or stabilization
	• Documented presence of mutation in the *MECP2* gene
	• Documentation of all the following:
	  ○ Partial or complete loss of acquired purposeful hand skills
	  ○ Partial or complete loss of acquired spoken language
	  ○ Gait abnormalities: Impaired (dyspraxic) or absence of ability
	  ○ Stereotypic hand movements such as hand wringing/squeezing, clapping/tapping, mouthing, and washing/rubbing automatisms
	• Current weight (within past 30 days)
	  ○ Must weigh minimum of 9 kilograms

Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** requires documentation of treatment success determined by treating provider
Exclusion Criteria:	• Brain injury secondary to trauma or severe infection • Grossly abnormal psychomotor development in first 6 months of life
Age Restriction:	• 2 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or provider experienced in the management of Rett syndrome • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
TROGARZO
Affected Medications: TROGARZO (ibalizumab-uiyk/IV infusion)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of human immunodeficiency virus type 1 (HIV-1) infection, in combination with other antiretrovirals, in heavily treatment-experienced adults with multidrug resistant HIV-1 infection failing their current antiretroviral regimen

Required Medical Information:	• Documentation of all prior therapies used
	• Documentation of active antiretroviral therapy for at least 6 months
	• Documented resistance to at least one antiretroviral agent from three different classes:
	  ○ Nucleoside reverse-transcriptase inhibitors (NRTIs)
	  ○ Non-nucleoside reverse-transcriptase inhibitors (NNRTIs)
	  ○ Integrase strand transfer inhibitors (INSTIs)
	  ○ Protease inhibitors (PIs)
	• Documentation of current (within the past 30 days) HIV-1 RNA viral load of at least 200 copies/mL

Appropriate Treatment Regimen & Other Criteria:	• Prescribed in combination with an optimized background antiretroviral regimen
	
	<u>Reauthorization</u> requires all of the following:
	• Treatment plan includes continued use of optimized background antiretroviral regimen
	• Documentation of treatment success as evidenced by one of the following:
	  ○ Reduction in viral load from baseline or maintenance of undetectable viral load
	  ○ Absence of postbaseline emergence of ibalizumab resistance-associated mutations confirmed by resistance testing

Exclusion Criteria:	

Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an infectious disease or HIV specialist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization 12 months, unless otherwise specified

POLICY NAME:
TUCATINIB
Affected Medications: TUKYSA (tucatinib)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design • 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 • Documentation of RAS wild-type, human epidermal growth factor receptor-2 (HER2) positive, unresectable or metastatic colorectal cancer that has progressed following treatment with fluoropyrimidine, oxaliplatin, and irinotecan-based chemotherapy **OR** • Advanced, unresectable or metastatic, HER2-positive breast cancer with prior treatment of 1 or more anti-HER2-based regimens in the metastatic setting
Appropriate Treatment Regimen & Other Criteria:	**Colorectal cancer** • Documented intolerable adverse event to Lapatinib **Reauthorization:** documentation of disease responsiveness to therapy
Exclusion Criteria:	• Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater • Colorectal cancer ONLY: previous treatment with a HER2 inhibitor
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial approval: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
TYVASO
Affected Medications: TYVASO (treprostinil inhalation)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 1
	  ○ Pulmonary Arterial Hypertension (PAH) World Health Organization (WHO) Group 3

Required Medical Information:	**<u>Pulmonary Arterial Hypertension (PAH) WHO Group 1</u>**
	• Documentation of PAH confirmed by right-heart catheterization meeting the following criteria:
	  ○ Mean pulmonary artery pressure of at least 20 mm Hg
	  ○ Pulmonary capillary wedge pressure less than or equal to 15 mm Hg
	  ○ Pulmonary vascular resistance of at least 2.0 Wood units
	• Etiology of PAH: idiopathic PAH, hereditary PAH, OR
	• PAH secondary to one of the following conditions:
	  ○ Connective tissue disease
	  ○ Human immunodeficiency virus (HIV) infection
	  ○ Drugs
	  ○ Congenital left to right shunts
	  ○ Schistosomiasis
	  ○ Portal hypertension
	• New York Heart Association (NYHA)/World Health Organization (WHO) Functional Class III or higher symptoms
	• Documentation of Acute Vasoreactivity Testing (positive result requires trial/failure to calcium channel blockers) unless there are contraindications:
	  ○ Low systemic blood pressure (systolic blood pressure less than 90)
	  ○ Low cardiac index
	  OR
	  ○ Presence of severe symptoms (functional class IV)
	
	<u>Pulmonary Hypertension Associated with Interstitial Lung Disease WHO Group 3</u>

	• 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 OR • Pulmonary fibrosis and emphysema OR • Connective tissue disorder
Appropriate Treatment Regimen & Other Criteria:	• The pulmonary hypertension has progressed despite maximal medical and/or surgical treatment of the identified condition • Documentation that treprostinil is used as a single route of administration (Remodulin, Tyvaso, Orenitram should not be used in combination) **<u>WHO Group 1 only:</u>** • Treatment with oral calcium channel blocking agents has been tried and failed, or has been considered and ruled out • 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 o Ambrisentan and tadalafil o Bosentan and riociguat o Macitentan and sildenafil **<u>Reauthorization</u>** requires documentation of treatment success defined as one or more of the following: • Improvement in walking distance • Improvement in exercise ability • Improvement in pulmonary function • Improvement or stability in WHO functional class
Exclusion Criteria:	• PAH secondary to pulmonary venous hypertension such as left sided atrial or ventricular disease, left sided valvular heart disease, or disorders of the respiratory system such as chronic obstructive pulmonary disease, obstructive sleep apnea or other sleep disordered breathing, alveolar hypoventilation disorders, etc.
Age Restriction:	

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

POLICY NAME:
UBLITUXIMAB-XIIY
Affected Medications: BRIUMVI (ublituximab-xiiy)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of relapsing forms of multiple sclerosis (MS), including the following:
	  ▪ Clinically isolated syndrome (CIS)
	  ▪ Relapsing-remitting multiple sclerosis (RRMS)
	  ▪ Active secondary progressive disease (SPMS)

Required Medical Information:	• Diagnosis confirmed with magnetic resonance imaging (MRI), per revised McDonald diagnostic criteria for MS ○ Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS

Appropriate Treatment Regimen & Other Criteria:	<u>Relapsing forms of MS</u>:
	• Documentation of one of the following:
	  ○ Documented disease progression or intolerable adverse event with rituximab (biosimilar products, Riabni and Ruxience, preferred)
	  ○ Currently receiving treatment with Briumvi, excluding via samples or manufacturer's patient assistance program
	
	<u>Reauthorization</u> requires documentation of treatment success

Exclusion Criteria:	• Active Hepatitis B infection • Concurrent use of medications indicated for treatment of relapsing-remitting multiple sclerosis
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or a multiple sclerosis specialist • All approved are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Initial approval: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
UPNEEQ
Affected Medications: UPNEEQ (oxymetazoline opthalmic solution)

Covered Uses:	• Upneeq (oxymetazoline opthalmic solution) is not considered medically necessary due to insufficient evidence of therapeutic value.
Required Medical Information:	
Appropriate Treatment Regimen & Other Criteria:	
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	
Coverage Duration:	

POLICY NAME:
VAGINAL PROGESTERONE
Affected Medications: FIRST-PROGESTERONE VGS 100 MG, FIRST-PROGESTERONE VGS 200 MG

Covered Uses:	• Prevention of preterm birth in pregnancy
Required Medical Information:	• Pregnancy with maternal risk factor(s) for preterm birth (such as race, low maternal weight, smoking, substance use, or short interpregnancy interval) • Current week of gestation and estimated delivery date
Appropriate Treatment Regimen & Other Criteria:	• May continue until completion of 36 weeks gestation
Exclusion Criteria:	• Treatment of infertility
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a gynecologist or obstetrician • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: up to 6 months, unless otherwise specified

POLICY NAME:
VALOCTOCOGENE ROXAPARVOVEC-RVOX
Affected Medications: ROCTAVIAN (valoctocogene roxaparvovec-rvox) - Available on Medical Benefit only

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Hemophilia A (Factor VIII deficiency)

Required Medical Information:	• Documentation of diagnosis of Hemophilia A
	• Documentation of current testing with negative results for active factor VIII inhibitors on 2 consecutive occasions (at least one week apart within the past 12 months) and is not receiving a bypassing agent (e.g., Feiba)
	• Documentation of baseline circulating level of factor with Factor VIII activity level equal to or less than 1 IU/dL or 1% endogenous factor VIII
	• Evidence of any bleeding disorder NOT related to hemophilia A has been ruled out
	• No detectable antibodies to AAV5 as determined by an FDA-approved/CLIA-compliant test
	• Has received stable dosing of prophylactic Factor VIII replacement therapy on a regular basis for at least 1 year
	• Baseline lab values (must be less than 2 times upper limit of normal):
	  ○ ALT
	  ○ AST
	  ○ Total bilirubin
	  ○ Alkaline phosphatase (ALP)

Appropriate Treatment Regimen & Other Criteria:	**<u>Dosing</u>** • 6×10^{13} vector genomes/kg (which is 3 mL/kg) as a single one-time dose
Exclusion Criteria:	• History of or current presence of Factor VIII inhibitors • Prior gene therapy administration

	• Active Hepatitis B or C infection or other active acute or uncontrolled chronic infection • Cirrhosis • Female gender at birth • Allergy to mannitol
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation, with a hematologist or specialist with experience in treatment of hemophilia • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 2 months (one time infusion), unless otherwise specified

POLICY NAME:
VAMOROLONE
Affected Medications: AGAMREE (vamorolone)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Duchenne muscular dystrophy (DMD) in patients 2 years of age and older

Required Medical Information:	• Confirmation of Duchenne muscular dystrophy (DMD) diagnosis by genetic testing or biopsy showing lack of muscle dystrophin
	• Documentation of being ambulatory without needing an assistive device such as a wheelchair, walker, or cane
	• Baseline motor function assessment from one of the following:
	  ○ Time to Stand Test (TTSTAND)
	  ○ 6-minute walk test
	  ○ North Star Ambulatory Assessment (NSAA)
	  ○ Motor Function Measure (MFM)
	  ○ Hammersmith Functional Motor Scale (HFMS)
	• Patient weight and planned treatment regimen

Appropriate Treatment Regimen & Other Criteria:	• Documented treatment failure with a 6-month trial of prednisone, or intolerable adverse event causing one of the following:
	  ○ Clinically significant weight gain defined as greater than or equal to 10% of body weight gain over a 6-month period
	  ○ Psychiatric/behavioral issues (e.g., abnormal behavior, aggression, irritability) that persists beyond the first six weeks of prednisone treatment
	• **<u>Reauthorization</u>** requires a documented improvement from baseline or stabilization of motor function demonstrated by a motor function assessment tool

Exclusion Criteria:	
Age Restriction:	• 2 years of age and older

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

POLICY NAME:
VARIZIG
Affected Medications: VARIZIG (varicella zoster immune globulin (human) IM injection)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ For post exposure prophylaxis of varicella in high-risk individuals
Required Medical Information:	Documentation of immunocompromised patient, defined as: • Newborns of mothers with signs and symptoms of varicella shortly before or after delivery (five days before to two days after delivery) • 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 • 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 • Immunocompromised children and adults who lack evidence of immunity to varicella • Pregnant women who lack evidence of immunity to varicella ○ Lack evidence of immunity to varicella is defined as: those who are seronegative for varicella zoster antibodies OR those with unknown history of varicella
Appropriate Treatment Regimen & Other Criteria:	• If repeat dose is necessary due to re-exposure, use more than 3 weeks after initial administration.
Exclusion Criteria:	• Coagulation disorders
Age Restriction:	

Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Approval: 6 months, unless otherwise specified

POLICY NAME:
VELMANASE ALFA-TYCV
Affected Medications: LAMZEDE (velmanase alfa-tycv)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ The treatment of non-central nervous system manifestations of alpha-mannosidosis
Required Medical Information:	• Diagnosis of alpha-mannosidosis (AM) confirmed by enzyme assay demonstrating alpha-mannosidase activity less than 10% of normal activity • Documentation of symptoms consistent with AM such as hearing impairment, difficulty walking, skeletal abnormalities, or intellectual disabilities
Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** will require documentation of treatment success such as improvement in motor function, forced vital capacity (FVC), or reduction in frequency of infections
Exclusion Criteria:	• AM with only central nervous system manifestations and no other symptoms
Age Restriction:	
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care • Prescribed by, or in consultation with, a specialist familiar with the treatment of lysosomal storage disorders
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
VERTEPORFIN
Affected Medications: VISUDYNE (verteporfin)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Treatment of predominantly classic subfoveal choroidal neovascularization (CNV) due to one of the following:
	  ▪ Age-related macular degeneration (AMD)
	  ▪ Pathologic myopia
	  ▪ Presumed ocular histoplasmosis

Required Medical Information:	• Subfoveal choroidal neovascularization (CNV) lesions caused by age-related macular degeneration (AMD); or • Ocular histoplasmosis; or • Pathologic myopia 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 fluorescein 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:
	  ○ Currently receiving treatment with Visudyne, excluding when the product is obtained as samples or via manufacturer's patient assistance programs
	  ○ A documented inadequate response or intolerable adverse event with all of the preferred products (Avastin AND Byooviz or Cimerli)
	• Dosing: 6 mg/m2 body surface area given intravenously; may repeat at 3-month intervals (if evidence of choroidal neovascular leakage)
	• Dose-rounding to the nearest vial size within 10% of the prescribed dose will be enforced
	
	Reauthorization requires documented treatment success and a continued need for treatment with the non-preferred product

Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an ophthalmologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
VIGABATRIN
Affected Medications: VIGABATRIN, VIGADRONE (vigabatrin)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Refractory complex partial seizures (focal seizures with impaired awareness)
	  ○ Infantile spasms

Required Medical Information:	**<u>Infantile Spasms</u>** • Used as monotherapy for pediatric patients (1 month to 2 years of age) **<u>Refractory Complex Partial Seizures (focal seizures with impaired awareness)</u>** • Used as adjunctive therapy only
Appropriate Treatment Regimen & Other Criteria:	**<u>Refractory complex partial seizures (focal seizures with impaired awareness)</u>** • Documentation of treatment failure with at least 2 alternative therapies: carbamazepine, phenytoin, levetiracetam, topiramate, oxcarbazepine, or lamotrigine **<u>Reauthorization</u>** will require documentation of treatment success and a reduction in seizure severity, frequency, and/or duration
Exclusion Criteria:	• Use as a first line agent for complex partial seizures (focal seizures with impaired awareness)
Age Restriction:	• Infantile Spasms: 1 month to 2 years of age • Refractory complex partial seizures (focal seizures with impaired awareness): greater than 2 years of age
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	**<u>Infantile Spasms</u>**

	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months (or up to 2 years of age), unless otherwise specified **<u>Refractory Complex Partial Seizures (focal seizures with impaired awareness)</u>** • Authorization: 12 months, unless otherwise specified

POLICY NAME:
VIJOICE
Affected Medications: VIJOICE (alpelisib)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Treatment of severe manifestations of PIK3CA-related overgrowth spectrum (PROS) in patients who require systemic therapy

Required Medical Information:	• Documented diagnosis of PROS, to include any of the following:
	  ○ CLAPOS syndrome
	  ○ CLOVES syndrome
	  ○ Diffuse capillary malformation with overgrowth (DCMO)
	  ○ Dysplastic megalencephaly (DMEG)
	  ○ Facial infiltrating lipomatosis (FIL)
	  ○ Fibroadipose hyperplasia (FAH)/fibroadipose overgrowth (FAO)/hemihyperplasia multiple lipomatosis (HHML) syndrome
	  ○ Fibroadipose vascular anomaly (FAVA)
	  ○ Hemimegalencephaly (HMEG)
	  ○ Klippel-Trenaunay syndrome (KTS)
	  ○ Lipomatosis of nerve (LON)
	  ○ Megalencephaly-capillary malformation (MCAP) syndrome
	  ○ Muscular hemihyperplasia (HH)
	• Documentation of PIK3CA gene mutation
	• Documentation of clinical manifestations that were assessed by the treating provider as severe or life-threatening and necessitating systemic treatment
	• Documentation that clinical manifestations are a direct result of a lesion that is both of the following:
	  ○ Inoperable, as defined by the treating provider
	  ○ Causing functional impairment
	• Documentation of one or more target lesion(s) identified on imaging within 6 months prior to request, including location(s) and volume of lesion(s)

Appropriate Treatment Regimen & Other Criteria:	• Treatment failure (or intolerable adverse event) with sirolimus for at least 6 months at a dose of at least 2 mg daily in patients with lymphatic, venous, or combined manifestations of disease **<u>Reauthorization</u>** will require documentation of both of the following: ○ 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 ○ Absence of greater than or equal to a 20% increase from baseline in any target lesion, progression of non-target lesions, or appearance of a new lesion
Exclusion Criteria:	• Treatment of PIK3CA-mutated conditions other than PROS
Age Restriction:	• 2 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a specialist with experience in the treatment of PROS • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 6 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
VISTOGARD
Affected Medications: VISTOGARD (uridine triacetate)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ For the emergency treatment of adult and pediatric patients:
	  ▪ Following a fluorouracil or capecitabine overdose regardless of the presence of symptoms, **OR**
	  ▪ Who exhibit early-onset, severe, or life-threatening toxicity affecting the cardiac or central nervous system, and/or early-onset, unusually severe adverse reactions (e.g., gastrointestinal toxicity and/or neutropenia) within 96 hours following the end of fluorouracil or capecitabine administration

Required Medical Information:	• Documentation of fluorouracil or capecitabine administration • Documentation of overdose **OR** early-onset, severe adverse reaction, or life-threatening toxicity
Appropriate Treatment Regimen & Other Criteria:	• Dosing is in accordance with FDA labeling
Exclusion Criteria:	• Non-emergent treatment of adverse events associated with fluorouracil or capecitabine • Use more than 96 hours following the end of fluorouracil or capecitabine administration
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care

Coverage Duration:	• Authorization: 7 days, unless otherwise specified

POLICY NAME:
VMAT2 INHIBITORS
Affected Medications: TETRABENAZINE, AUSTEDO (deutetrabenazine), AUSTEDO XR (deutetrabenazine)

Covered Uses:	• All Food and Drug Administration (FDA)-approved and compendia supported indications not otherwise excluded by plan design
	  ○ Chorea associated with Huntington's disease
	  ○ Tardive dyskinesia

Required Medical Information:	**<u>Chorea related to Huntington's Disease</u>**
	• Diagnosis of Huntington's Disease with Chorea requiring treatment
	
	<u>Tardive Dyskinesia</u>
	• Diagnosis of moderate to severe tardive dyskinesia including all of the following:
	  ○ A history of at least one month of ongoing or previous dopamine receptor-blocking agent exposure
	  ○ Presence of dyskinetic or dystonic involuntary movements that developed either while exposed to a dopamine receptor-blocking agent, or within 4 weeks of discontinuation from an oral agent (8 weeks from a depot formulation)
	  ○ Other causes of abnormal movements have been excluded
	• Baseline evaluation of the condition using one of the following:
	  ○ Abnormal Involuntary Movement Scale (AIMS)
	  ○ Extrapyramidal Symptom Rating Scale (ESRS)

Appropriate Treatment Regimen & Other Criteria:	**<u>Tardive Dyskinesia</u>** • Persistent dyskinesia despite dose reduction or discontinuation of the offending agent **OR** • Documented clinical inability to reduce dose or discontinue the offending agent **<u>Reauthorization</u>** requires documentation of treatment success and a clinically significant response to therapy

	• Tardive Dyskinesia: must include an improvement in AIMS or ESRS score from baseline
Exclusion Criteria:	• Use for Huntington's comorbid with untreated or inadequately treated depression or actively suicidal • Concomitant use with another VMAT2 inhibitor or reserpine • Hepatic impairment
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist or psychiatrist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 3 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
VOCLOSPORIN
Affected Medications: LUPKYNIS CAPSULE 7.9 MG ORAL

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? 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? a. Estimated glomerular filtration rate (eGFR) b. Urine protein to creatinine ratio (uPCR) c. Blood pressure	Yes – Document and go to #3	No – Criteria not met
3. Is there documented treatment failure with at least 12 weeks of standard therapy with both mycophenolate mofetil (MMF) AND cyclophosphamide?	Yes – Document and go to #4	No – Criteria not met

4. Is there documented treatment failure with at least 12 weeks of subcutaneous Benlysta?	Yes – Document and go to #5	No – Criteria not met
5. Will Lupkynis be used in combination with MMF and corticosteroids or other background immunosuppressive therapy, other than cyclophosphamide?	Yes – Document and go to #6	No – Criteria not met
6. Is the drug prescribed by, or in consultation with, a rheumatologist, immunologist, nephrologist or kidney specialist?	Yes – Go to #7	No – Criteria not met
7. Is the requested dose within the Food and Drug Administration (FDA)-approved label and PacificSource quantity limitations?	Yes – Approve up to 12 months	No – Criteria not met
Renewal Criteria		
1. Is there documentation of treatment success defined as an increase in eGFR, decrease in uPCR, or decrease in flares and corticosteroid use?	Yes – Go to #2	No – Criteria not met
2. Is the requested dose within the Food and Drug Administration (FDA)-approved label and PacificSource quantity limitations?	Yes – Approve up to 12 months	No – Criteria not met
Quantity Limitations		

• **Lupkynis**
  ○ 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 m^2 or less at initiation: 15.8 mg BID
  ▪ Mild-to-moderate hepatic impairment (Child-Pugh A or B): 15.8 mg BID		

- Concomitant use with moderate CYP3A4 inhibitors: 15.8 mg in morning and 7.9 mg in afternoon.

POLICY NAME:
VORETIGENE NEPARVOVEC
Affected Medications: LUXTURNA (Voretigene neparvovec-rzyl intraocular suspension for subretinal injection)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design. ○ 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.) • Genetic testing documenting biallelic mutations of the RPE65 gene • Visual acuity of at least 20/800 OR have remaining light perception in the eye(s) receiving treatment • Visual acuity of less than 20/60 OR a visual field of less than 20 degrees • 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 • Patient has other pre-existing eye conditions or complicating systemic diseases that would eventually lead to irreversible vision loss and prevent the patient from receiving full benefit from treatment (e.g., severe diabetic retinopathy)
Age Restriction:	• 12 months of age and older

Prescriber/Site of Care Restrictions:	• Ophthalmologist or retinal surgeon with experience providing sub-retinal injections
Coverage Duration:	• Approval: 1 month - 1 injection per eye per lifetime, unless otherwise specified

POLICY NAME:
VOSORITIDE
Affected Medications: VOXZOGO (vosoritide)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ To increase linear growth in pediatric patients with achondroplasia with open epiphyses
Required Medical Information:	• Diagnosis of achondroplasia confirmed by molecular genetic testing showing a mutation in the fibroblast growth factor receptor type 3 (FGFR3) gene • Baseline height, growth velocity, and patient weight

Appropriate Treatment Regimen & Other Criteria:	• Documentation of all the following:
	  ○ Evaluation of epiphyses (growth plates) documenting they are open
	  ○ Growth velocity greater than or equal to 1.5 cm/yr
	
	<u>Reauthorization:</u>
	• Evaluation of epiphyses (growth plates) documenting they remain open
	• Growth velocity greater than or equal to 1.5 cm/yr

Exclusion Criteria:	• Hypochondroplasia • Other short stature condition other than achondroplasia • Evidence of growth plate closure
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a pediatric orthopedist, endocrinologist, or a provider with experience in treating skeletal dysplasia • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 12 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME:
XEOMIN, DYSPORT, MYOBLOC, and DAXXIFY
Affected Medications: XEOMIN (incobotulinum toxin A), DYSPORT (abobotulinumtoxinA), MYOBLOC (rimabotulinumtoxinB), JEUVEAU (prbotulinumtoxinA-xvfs), DAXXIFY (daxibotulinumtoxinA-lanm)

Covered Uses:	• All Food and Drug Administration (FDA)-approved and compendia-supported indications not otherwise excluded by plan design
	  ○ Dysport
	  ▪ Focal dystonia (cervical dystonia, blepharospasm, laryngeal spasm, oromandibular dystonia, severe writer's cramp)
	  ▪ Hemifacial spasm
	  ▪ Upper/lower limb spasticity
	  ○ Xeomin
	  ▪ Cervical dystonia
	  ▪ Blepharospasm
	  ▪ Upper limb spasticity
	  ▪ Chronic sialorrhea
	  ○ Myobloc, Daxxify
	  ▪ Cervical dystonia

Required Medical Information:	• Pertinent medical records and diagnostic testing • Complete description of the site(s) of injection • Strength and dosage of botulinum toxin used
Appropriate Treatment Regimen & Other Criteria:	**<u>Dysport</u>** • Approved first-line for focal dystonia, hemifacial spasm, drug-induced orofacial dyskinesia, upper or lower limb spasticity **<u>Xeomin</u>** • Approved first-line for cervical dystonia, blepharospasm, upper limb spasticity, chronic sialorrhea **<u>Myobloc</u>** • **Cervical dystonia:** Documentation of treatment failure with Botox, Dysport, and Xeomin

	• **Axillary hyperhidrosis:** Documentation of treatment failure with Botox • **Chronic sialorrhea:** Documentation of treatment failure with glycopyrrolate oral tablets **<u>Daxxify</u>** • **Cervical dystonia:** Documentation of treatment failure with Botox, Dysport, and Xeomin **<u>Quantity limitations</u>** • Maximum of 4 treatments per 12 months **<u>Reauthorization</u>** requires documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	• Cosmetic procedures (including glabellar lines, horizontal forehead lines, lateral canthal lines) • Migraine headache use (Botox is preferred product)
Age Restriction:	• Myobloc, Daxxify: 18 years of age and older
Prescriber/Site of Care Restrictions:	• **Blepharospasm:** Prescribed by, or in consultation with, a neurologist, ophthalmologist, or optometrist • **Other indications:** Prescribed by, or in consultation with, a neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
XGEVA
Affected Medications: XGEVA (denosumab)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design.
	  ○ One of these diagnoses:
	  ▪ Giant cell tumor
	  ▪ Bone metastases from solid tumors
	  ▪ Hypercalcemia of malignancy
	  ▪ Multiple myeloma
	• NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher

Required Medical Information:	• **Giant cell tumor**
	  ○ Unresectable disease or surgical resection would likely result in severe morbidity.
	• **Bone metastases from solid tumors**
	• **Hypercalcemia of malignancy**
	  ○ Refractory to bisphosphonate therapy or contraindication
	• **Multiple myeloma**
	  ○ Requires failure of zoledronic acid or pamidronate OR creatinine clearance less than 30 mL/min

Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** requires documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	
Age Restriction:	• **Giant cell tumor**: Adults and adolescents at least 12 years of age and skeletally mature weighing at least 45 kg • **All other indications**: 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care

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

POLICY NAME:
XIAFLEX
Affected Medications: XIAFLEX (collagenase clostridium histolyticum)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Dupuytren's contracture with a palpable cord
	  ○ Peyronie's disease

Required Medical Information:	**<u>Peyronie's disease:</u>** • Documented diagnosis of Peyronie's disease with a palpable plaque • Curvature deformity is at least 30 degrees at the start of therapy • Documentation of stable disease defined as symptoms that have remained unchanged for at least 3 months
Appropriate Treatment Regimen & Other Criteria:	**<u>Dupuytren's:</u>** • Authorization will be limited per joint as follows: One injection per month for a maximum of three injections per cord **<u>Reauthorization</u>** will require documentation of treatment success and a clinically significant response to therapy **<u>Peyronie's disease:</u>** • One treatment cycle consists of two Xiaflex injection procedures **<u>Reauthorization</u>** 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 • Maximum of 4 treatment cycles per plaque, administered at 6-week intervals
Exclusion Criteria:	• Peyronie's plaques that involve the penile urethra

Age Restriction:	
Prescriber/Site of Care Restrictions:	• **Peyronie's**: prescribed by, or in consultation with, a urologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• **Dupuytren's**: 12 weeks, unless otherwise specified • **Peyronie's**: 6 weeks, unless otherwise specified

POLICY NAME:
XIFAXAN
Affected Medications: XIFAXAN (rifaximin)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design
	  ○ Prevention of hepatic encephalopathy (HE)
	  ○ Treatment of Travelers' Diarrhea caused by noninvasive strains of *Escherichia coli (E. coli)*
	  ○ Treatment of Irritable Bowel Syndrome with Diarrhea (IBS-D)
	• Compendia-supported uses that will be covered (if applicable)
	  ○ Treatment of HE
	  ○ Treatment of recurrent Clostridium difficile (C. diff)-associated diarrhea
	  ○ Treatment of Small Intestinal Bacterial Overgrowth (SIBO)

Required Medical Information:	• Documentation of complete & current treatment course required • Documentation of E-coli bacterial cultures for travelers' diarrhea • Previous antibiotic history and documented allergies/hypersensitivity

Appropriate Treatment Regimen & Other Criteria:	**<u>Recurrent C. diff</u>**
	• Documentation confirming a current diagnosis of recurrent C. diff infection (CDI) with **ALL** of the following:
	  ○ CDI symptoms resolved on prior appropriate therapy and have reappeared within 8 weeks of completing prior therapy
	  ○ Presence of at least 3 unformed stools in 24 hours
	  ○ Positive stool test for toxigenic Clostridium difficile
	• Documented treatment failure with oral vancomycin
	
	<u>HE</u>
	• Documented treatment failure with at least 1 month of lactulose therapy defined as continued altered mental status or elevated ammonium levels despite adequate upward titration
	
	<u>Travelers' Diarrhea</u>

	• Documentation of **ALL** of the following:
	  ○ Travelers' diarrhea is caused by noninvasive strains of E. coli
	  ○ Systemic signs of infection (fever or blood in stool) are not present
	  ○ Member is returning from an area of high fluoroquinolone resistance
	• Documented treatment failure with a fluoroquinolone (e.g., ciprofloxacin, levofloxacin) and azithromycin
	
	SIBO
	• Documented diagnosis confirmed by a carbohydrate breath test
	• Documented treatment failure with trial of at least one of the following antibiotics: amoxicillin/clavulanic acid, ciprofloxacin, metronidazole
	
	IBS-D
	• Documentation confirming a Rome IV diagnosis with recurrent abdominal pain, on average, at least one day per week in the last 3 months, associated with two or more of the following:
	  ○ Related to defecation
	  ○ Associated with a change in stool frequency
	  ○ Associated with a change in stool form (appearance)
	• Symptom onset at least six months prior to diagnosis
	• Documented treatment failure with **ALL** of the following:
	  ○ Loperamide
	  ○ Dicyclomine or hyoscyamine
	  ○ Tricyclic antidepressant (e.g., amitriptyline, nortriptyline)
	
	• **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.

	<u>Reauthorization</u> will require documentation of treatment success and a clinically significant response to therapy
Exclusion Criteria:	**<u>Recurrent C. diff</u>** • Xifaxan exceeding 400 mg three times per day for 20 days **<u>HE</u>** • Xifaxan exceeding the recommended dose of 550 mg twice daily or 400 mg 3 times daily for the treatment or prevention of hepatic encephalopathy **<u>Travelers' Diarrhea</u>** • Xifaxan exceeding 200 mg three times per day for total of 3 days • Diarrhea complicated by fever or bloody stool, or caused by bacteria other than noninvasive strains of E. coli **<u>SIBO</u>** • Xifaxan exceeding 550 mg three times per day for 14 days **<u>IBS-D</u>** • Mild cases of irritable bowel syndrome or diagnosis of irritable bowel syndrome with constipation • Xifaxan exceeding 550 mg three times per day for 14 days
Age Restriction:	• 12 years of age and older
Prescriber/Site of Care Restrictions:	• All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	**<u>Recurrent C. diff</u>** • Authorization: 20 days, unless otherwise specified **<u>HE</u>** • Authorization: 12 months, unless otherwise specified **<u>Travelers' Diarrhea</u>** • Authorization: 7 days, unless otherwise specified **<u>SIBO</u>**

	• Authorization: 14 days, unless otherwise specified (one treatment per lifetime) **IBS-D** • Authorization: 14 days, unless otherwise specified (maximum of 3 treatment courses per lifetime)

POLICY NAME:
XURIDEN
Affected Medications: XURIDEN (uridine triacetate)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Hereditary orotic aciduria

Required Medical Information:	• Diagnosis of hereditary orotic aciduria confirmed by **ONE** of the following:
	  ○ Molecular genetic testing confirming biallelic pathogenic mutation in the UMPS gene
	  ○ Urinary orotic acid level above the normal reference range
	  ○ Clinical manifestations consistent with disease such as:
	  ▪ Megaloblastic anemia
	  ▪ Leukopenia
	  ▪ Developmental delays
	  ▪ Failure to thrive

Appropriate Treatment Regimen & Other Criteria:	**Reauthorization** requires documentation of treatment success based on **ONE** of the following: • Improvement of hematologic abnormalities such as megaloblastic anemia and leukopenia • Reduction of urinary orotic acid levels
Exclusion Criteria:	
Age Restriction:	
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a metabolic specialist or geneticist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Authorization: 12 months, unless otherwise specified

POLICY NAME:
YONSA
Affected Medications: YONSA (abiraterone)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design • NCCN (National Comprehensive Cancer Network) indications with evidence level of 2A or higher
Required Medical Information:	• Documentation of performance status, disease staging, all prior therapies used, and anticipated treatment course
Appropriate Treatment Regimen & Other Criteria:	• Documented inadequate response or intolerable adverse event with the preferred product abiraterone acetate **Reauthorization** requires documentation of disease responsiveness to therapy
Exclusion Criteria:	• Child-Pugh Class C • Karnofsky Performance Status 50% or less or ECOG performance score 3 or greater
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, an oncologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified.

POLICY NAME:
ZILUCOPLAN
Affected Medications: ZILBRYSQ (zilucoplan)

Covered Uses:	• All Food and Drug Administration (FDA)-approved indications not otherwise excluded by plan design ○ Generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive

Required Medical Information:	• Diagnosis of generalized myasthenia gravis (gMG) confirmed by one of the following:
	  ○ A history of abnormal neuromuscular transmission test
	  ○ A positive edrophonium chloride test
	  ○ Improvement in gMG signs or symptoms with an acetylcholinesterase inhibitor
	• Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV
	• Positive serologic test for AChR antibodies
	• MG-Activities of Daily Living (MG-ADL) total score of 6 or greater **OR** Quantitative Myasthenia Gravis (QMG) total score of 12 or greater

Appropriate Treatment Regimen & Other Criteria:	• Currently on a stable dose of at least one gMG therapy (acetylcholinesterase inhibitor, corticosteroid, or non-steroidal immunosuppressive therapy (NSIST)) that will be continued during initial treatment with Zilbrysq
	• Documentation of one of the following:
	  ○ Treatment failure with an adequate trial (one year or more) of at least two immunosuppressive therapies (azathioprine, mycophenolate, tacrolimus, cyclosporine, methotrexate)
	  ○ Has required three or more courses of rescue therapy (plasmapheresis/plasma exchange and/or intravenous immunoglobulin), while on at least one immunosuppressive therapy, over the last 12 months
	
	<u>Reauthorization</u>:
	• Documentation of treatment success and clinically significant response to therapy defined as:
	  ○ A minimum 2-point reduction in MG-ADL score from baseline AND

	○ Absent or reduced need for rescue therapy compared to baseline • That the patient requires continuous treatment, after an initial beneficial response, due to new or worsening disease activity
Exclusion Criteria:	• Current or recent systemic infection within 2 weeks • Concurrent use with other biologics (rituximab, eculizumab, IVIG, etc)
Age Restriction:	• 18 years of age and older
Prescriber/Site of Care Restrictions:	• Prescribed by, or in consultation with, a neurologist • All approvals are subject to utilization of the most cost-effective site of care
Coverage Duration:	• Initial Authorization: 4 months, unless otherwise specified • Reauthorization: 12 months, unless otherwise specified

POLICY NAME: