

ACUTE HEREDITARY ANGIOEDEMA THERAPY

MEDICATION(S)

BERINERT, FIRAZYR, ICATIBANT, KALBITOR, RUCONEST

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For initial authorization: All of the following must be met:

1. Diagnosis of Hereditary Angioedema Types (HAE) I, II or III and one of the following clinical criteria:
 - a. Self-limiting, non-inflammatory subcutaneous angioedema without urticaria, recurrent, and lasting more than 12 hours, or
 - b. Self-remitting abdominal pain without clear organic etiology, recurrent, and lasting more than six (6) hours, or
 - c. Recurrent laryngeal edema.

AND

2. One of the following:
 - a. For HAE Type I and Type II, documentation of at least two (2) complement studies taken at least one (1) month apart with the patient in their basal condition and after the first year of life that show:
 - i. C4 is less than 50 percent of the lower limit of normal

AND

- ii. One of the following:
 1. C1-Inhibitor (C1-INH) protein is less than 50 percent of the lower limit of normal, or
 2. C1-INH function is less than 50 percent of the lower limit of normal
 - b. For HAE with normal C1-INH or HAE Type III:
 - i. Confirmed Factor 12 (FXII), ANGPT1, PLG, KNG1 gene mutation

OR

- ii. Positive family history for HAE AND attacks lack response with high dose antihistamines or corticosteroids.

AND

3. For coverage of Berinert®, Kalbitor®, Firazyr®, or Ruconest®: For patients not established on therapy only: Documentation of trial and failure or contraindication to generic icatibant

For reauthorization:

Documentation must be provided showing benefit of therapy with reduction of length and severity of HAE

attack episodes.

For quantities exceeding the formulary quantity limit:

1. Documentation of frequent HAE attacks defined as greater than or equal to two (2) attacks per month on average.

QUANTITY LIMIT (subject to audit):

Beriner[®] - 2 injections per 30 days

Ruconest[®] - 2 injections per 30 days

Kalbitor[®] - 2 boxes (6 vials) per 30 days

Firazyr[®] - 3 injections (3 boxes, total of 9ml) per 30 days

AGE RESTRICTION

Kalbitor[®] - 12 years and older

Firazyr[®] - 18 years and older

Ruconest[®] - 13 years and older

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with an Immunologist or an Allergist.

COVERAGE DURATION

Initial authorization will be approved for up to 6 months. Reauthorization will be approved for up to one (1) year.

OTHER CRITERIA

N/A

ADAKVEO

MEDICATION(S)

ADAKVEO

COVERED USES

N/A

EXCLUSION CRITERIA

Used in combination with voxelotor (Oxbryta®)

REQUIRED MEDICAL INFORMATION

Initial authorization:

1. Confirmed medical history or diagnosis of sickle cell disease
2. Patient has experienced at least two (2) sickle cell-related pain crises in the prior year
3. Documentation that patient meets one of the following:
 - a. Patient will continue taking hydroxyurea with the requested therapy and patient has been on a maximally tolerated dose of hydroxyurea for at least six (6) months
 - b. Patient has had a therapeutic failure of hydroxyurea despite use of a maximally tolerated dose for at least six (6) months
 - c. Patient has had an intolerance or contraindication to hydroxyurea (For many patients myelosuppression is dose-dependent and reversible, intolerance due to myelosuppression will only be considered if patient continues to experience myelosuppression despite dose adjustments)

Reauthorization: Documentation that the number or severity of sickle cell-related pain crises has decreased from baseline

AGE RESTRICTION

May be approved for patients 16 years of age and older.

PRESCRIBER RESTRICTION

Must be prescribed by, or in consultation with, a hematologist or a provider experienced with the treatment of sickle cell disease.

COVERAGE DURATION

Initial authorization and reauthorization will be approved for 1 year

OTHER CRITERIA

N/A

ALPHA-1 PROTEINASE INHIBITORS

MEDICATION(S)

ARALAST NP, GLASSIA, PROLASTIN C, ZEMAIRA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

Documentation of:

1. One of the following:

- a. Serum alpha-1 antitrypsin (AAT) concentrations less than 11 micromol/L (approximately 50 mg/dL by nephelometry or 80mg/dL by immunodiffusion)
- b. Patient has one of the following high-risk phenotypes by protease inhibitor (PI) typing: PI*ZZ, PI*Z(null), PI*(null,null)

AND

2. Diagnosis of emphysema with one of the following:

- a. Forced expiratory volume per one second (FEV-1) of 35 to 65% of predicted volume
- b. Rapid lung function decline as evidence by reduction of FEV-1 of 100 mL/year or greater

AND

3. Documentation that the patient has never smoked or has abstained from smoking for at least the previous six months

Reauthorization requires documentation of positive clinical response to therapy (e.g., reduction in exacerbations, reduced progression of emphysema as assessed by computed tomography (CT) densitometry, slowing of FEV-1 decline)

QUANTITY LIMIT:

60 mg/kg infused every seven days, subject to audit.

Note: Dose may be rounded down to the nearest gram (500 mg for Aralast®) within 10% of calculated dose.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Initial authorization will be approved for six months and reauthorization will be approved for one year.

OTHER CRITERIA

N/A

BENLYSTA

MEDICATION(S)

BENLYSTA

COVERED USES

N/A

EXCLUSION CRITERIA

Belimumab will not be approved if any of the following are present:

1. Severe active central nervous system lupus
2. Current use of other biologic immunomodulator
3. Documentation of no previous use of dialysis in the past 12 months or currently using dialysis
4. Concurrent use of Lupkynis®

REQUIRED MEDICAL INFORMATION

For Systemic Lupus Erythematosus (SLE) and active lupus nephritis:

All of the following must be met:

1. Documented diagnosis of Systemic Lupus Erythematosus (SLE) or active lupus nephritis by a rheumatologist or nephrologist

AND

2. Documentation of laboratory test results indicating that patient has presence of auto-antibodies, defined as one of the following:

- a. Positive Antinuclear antibody (ANA)
- b. Positive anti-double-stranded DNA (anti-dsDNA) on two or more occasions, OR if tested by ELISA, an antibody level above laboratory reference range
- c. Positive anti-Smith (Anti-Sm)
- d. Positive anti-Ro/SSA and anti-La/SSB antibodies

AND

3. Documented failure of an adequate trial (such as inadequate control with ongoing disease activity and/or frequent flares), contraindication, or intolerance to at least one of the following:

- a. For SLE without Active Lupus Nephritis:

- i. Oral corticosteroid(s)
- ii. Azathioprine
- iii. Methotrexate
- iv. Mycophenolate mofetil
- v. Hydroxychloroquine
- vi. Chloroquine
- vii. Cyclophosphamide

b. For SLE with Active Lupus Nephritis:

i. mycophenolate for induction followed by mycophenolate for maintenance, OR

ii. cyclophosphamide for induction followed by azathioprine for maintenance.

4. Documentation that patient will continue to receive standard therapy (e.g., corticosteroids, hydroxychloroquine, mycophenolate, azathioprine, methotrexate)

Reauthorization:

1. Documentation of positive clinical response to belimumab (e.g. improvement in functional impairment, decrease of corticosteroid dose, decrease in pain medications, decrease in the number of exacerbations since prior to start of belimumab, reduction of renal related events)

Patient currently receiving standard therapy for SLE and active lupus nephritis

QUANTITY LIMIT:

- Belimumab 200 mg/mL single-dose prefilled auto injector and glass syringe for subcutaneous injection: 4 mL per 28 days

- o Adults with SLE without active lupus nephritis allowed loading dose: none

- o Adults with SLE with active lupus nephritis allowed loading dose: 400-mg dose (two 200-mg injections) once weekly for four doses, then 200 mg once weekly thereafter

- Belimumab powder for solution for IV use only (subject to audit): Initial dose of 10 mg/kg IV every two weeks for three doses and then continue every four weeks thereafter as maintenance

- o Applicable to adults with SLE or active lupus nephritis and pediatric patients with SLE

- Belimumab IV is available as:

- o 120 mg in a 5-mL single-dose vial

- o 400 mg in a 20-mL single-dose vial for injection

- Correct vial combination for each patient should be calculated to minimize waste

AGE RESTRICTION

For SLE without active lupus nephritis:

Age five years and older for IV infusion

Age 18 years and older for subcutaneous injection

For SLE with Active Lupus Nephritis:

Age 18 years and older for IV infusion or subcutaneous injection

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with a rheumatologist or nephrologist

COVERAGE DURATION

Initial authorization will be approved for six months. Reauthorization will be approved for 12 months.

OTHER CRITERIA

N/A

BOTULINUM TOXIN

MEDICATION(S)

BOTOX, BOTOX COSMETIC, DYSPORT, MYOBLOC, XEOMIN

COVERED USES

N/A

EXCLUSION CRITERIA

- When the above criteria are not met, botulinum toxin is considered investigational and not covered.
- Botulinum toxin is considered cosmetic and is not covered for the treatment of glabellar lines and/or fine wrinkles on the face.
 - o PrabotulinumtoxinA (Jeuveau®) will not be covered as it is only FDA approved for the treatment of glabellar lines and/or fine wrinkles on the face.

REQUIRED MEDICAL INFORMATION

OnabotulinumtoxinA (Botox®) may be covered for the following indications when criteria are met:

1. Chronic migraine headaches in adults when all of the following is met:
 - a. Documentation of at least 15 headache days per month with headaches lasting four hours or longer
 - b. Documentation of trial and failure, intolerance, or contraindication to at least TWO of the following classes used for migraine prevention. Trial and failure is defined as inadequate response following a minimum three months of consistent use.
 - i. Antidepressants (e.g., amitriptyline, venlafaxine)
 - ii. Beta-blockers (e.g., metoprolol, propranolol, timolol)
 - iii. Antiepileptics (e.g., divalproex, valproate, topiramate)
 - c. For patients established on a Calcitonin Gene Related Peptide (CGRP) receptor antagonist for migraine prophylaxis, combination therapy with Botox® may be considered medically necessary if the following criteria are met:
 - i. The patient has been established on, and adherent to, CGRP prophylaxis therapy (i.e., Aimovig®, Emgality®, Ajoovy®) for at least six months and has a documented improvement in frequency and/or severity of migraine headaches
 - ii. Patient continues to have at least 15 headache days per month with headaches lasting four hours or longer, despite use of CGRP prophylaxis monotherapy
 - d. Reauthorization for Botox® monotherapy or combination therapy with CGRP for prophylaxis will require documentation of a 30% reduction in headache days from baseline.
2. Spasticity in patients at least two years of age
3. Cervical dystonia in adults
4. Strabismus and blepharospasm associated with dystonia in patients at least 12 years of age
5. Severe axillary hyperhidrosis in adults after documented trial and failure, intolerance or contraindication

to topical agents

- a. Note: The safety and effectiveness of onabotulinumtoxinA for hyperhidrosis in other body areas have not been established.
6. Overactive bladder in adults with:
 - a. Symptoms of urge urinary incontinence, urgency, and frequency
 - b. Documented trial and failure, intolerance, or contraindication to at least one month of anticholinergic medication (e.g., oxybutynin, tolterodine)
7. Urinary incontinence in patients at least five years of age:
 - a. Due to detrusor over activity related to a neurologic condition (e.g., spinal cord injury, multiple sclerosis)
 - b. Documented trial and failure, intolerance, or contraindication at least one month of anticholinergic medication (e.g., oxybutynin, tolterodine)
8. Excessive salivation due to advanced Parkinson's disease
9. Hemifacial spasm
10. Chronic anal fissure when all of the following is met:
 - a. Prescribed by, or in consultation with, a gastroenterologist or colorectal surgeon
 - b. Documentation of trial and failure, intolerance, or contraindication to at least six weeks of therapy with either topical nitrates or topical calcium channel blockers
 - c. One of the following:
 - i. Documentation that the patient is not a good candidate for surgery or appropriate medical rationale is provided for avoiding surgery
 - ii. Botox® is to be used in conjunction with fissurotomy
 - d. The use of Botox® in combination with sphincterotomy or anal advancement flap is considered experimental and investigational and will not be covered
11. Spastic dysphonia (laryngeal dystonia) for adductor type when prescribed by, or in consultation with, a specialist in laryngology
12. Achalasia in patients ineligible for definitive treatments, such as pneumatic dilation, surgical myotomy or peroral endoscopic myotomy (POEM)
 - a. The use of Botox® in combination with pneumatic dilation is considered experimental and investigational and will not be covered

AbobotulinumtoxinA (Dysport®) may covered for the following indications:

1. Spasticity in patients two years of age and older
2. Cervical dystonia in adults
3. Blepharospasm in adults

IncobotulinumtoxinA (Xeomin®) may covered for the following indications:

1. Chronic sialorrhea in patients two years and older
2. Upper limb spasticity in patients at least two years of age
3. Cervical dystonia in adults
4. Blepharospasm in adults

RimabotulinumtoxinB (Myobloc®) may covered for the following indications:

1. Cervical dystonia in adults
2. Chronic sialorrhea in adult patients

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Initial authorization and reauthorization will be approved for one year

OTHER CRITERIA

N/A

BRINEURA - MEDICAL BENEFIT

MEDICATION(S)

BRINEURA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For initial authorization all the following criteria must be met:

1. Diagnosis of neuronal ceroid lipofuscinosis type 2 (CLN2) confirmed by both of the following:
 - a. Deficiency of tripeptidyl peptidase 1 (TPP1) enzyme activity (in a sample of leukocytes, fibroblasts, dried blood spot or saliva)
 - b. Genetic testing revealing one pathogenic mutation on each parental allele of TPP1/CLN2 gene
2. Documentation of symptomatic disease (e.g., seizures, changes in gait, falls, difficulty in ambulating, loss of language/delay in language development, visual failures)
3. Baseline Motor Domain of the CLN2 Clinical Rating Scale score of at least one (1)

Reauthorization requires documentation of response to therapy, defined as both of the following:

1. No more than a 1-point decline in the Motor Domain of the CLN2 Clinical Rating Scale
2. Motor Domain of the CLN2 Clinical Rating Scale score remains above zero

AGE RESTRICTION

May be covered for ages 3-17 years

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with a neurologist or medical geneticist

COVERAGE DURATION

Initial approval and reauthorization will be for 1 year

OTHER CRITERIA

N/A

CABENUVA

MEDICATION(S)

CABENUVA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For new starts:

1. Patient must have a confirmed diagnosis of human immunodeficiency virus type -1 (HIV-1)
2. Patient has been stable and adherent with their current antiviral regimen for a minimum of six (6) months (adherence may be confirmed by pharmacy claims)
3. Patient has a recent viral HIV-1 RNA of less than 50 copies/mL on current oral antiviral regimen
4. Documentation that patient does not have a history of treatment failure

For continuation of therapy:

1. Documentation that patient has been adherent with therapy
2. Documentation that patient has maintained a viral HIV-1 RNA of less than 50 copies/mL

AGE RESTRICTION

May be approved for patients aged 18 years and older

PRESCRIBER RESTRICTION

Must be prescribed by, or in consultation with, an infectious disease specialist

COVERAGE DURATION

Initial authorization for 1 year. Reauthorization will be approved until no longer eligible with the plan, subject to formulary and/or benefit changes.

OTHER CRITERIA

N/A

CABLIVI

MEDICATION(S)

CABLIVI

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

Initial Criteria:

1. Diagnosis of acquired thrombotic thrombocytopenic purpura
2. Documentation that therapy will be given in combination with plasma exchange therapy
3. Documentation that therapy will be given in combination with immunosuppressive therapy (i.e., glucocorticoids, rituximab)

Reauthorization criteria:

If the request is for a new treatment cycle:

1. Documentation of previous positive response to therapy (such as an improvement in platelet counts, reduction in neurological symptoms, or improvements in organ-damage markers)
2. Documentation that therapy will be given in combination with plasma exchange therapy and immunosuppressive therapy (i.e., glucocorticoids, rituximab)
3. Documentation that length of therapy post plasma exchange will not exceed 58 days
4. Documentation that patient has not had more than two (2) recurrences of acquired thrombotic thrombocytopenic purpura while on therapy with caplacizumab. Recurrence is defined as initial platelet normalization followed by a reduction in platelet count that necessitates re-initiation of plasma exchange.

If request is for treatment extension:

1. Documentation of positive response to therapy (such as an improvement in platelet counts, reduction in neurological symptoms, or improvements in organ-damage markers)
2. Documentation that patient has signs of persistent underlying disease such as persistent severe ADAMTS13 deficiency
3. Documentation that length of therapy post plasma exchange will not exceed 58 days

QUANTITY LIMIT:

One (1) vial per day

AGE RESTRICTION

Approved for patients 18 years of age and older

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with an oncologist or hematologist

COVERAGE DURATION

Initial authorization will be approved for 30 days. Reauthorization will be approved up to a total duration of 58 days post-plasma-exchange.

OTHER CRITERIA

N/A

CAR-T (KYMRIAH, YESCARTA) - MEDICAL BENEFIT

MEDICATION(S)

ABECMA, BREYANZI, KYMRIAH, TECARTUS, YESCARTA

COVERED USES

N/A

EXCLUSION CRITERIA

Previous treatment with chimeric antigen receptor therapy or other genetically modified T-cell therapy. Repeat administration of CAR-T therapy is considered experimental and investigational because the effectiveness of this approach has not been established.

REQUIRED MEDICAL INFORMATION

For all indications, the following criteria must be met:

1. Documentation of adequate bone marrow, cardiac, pulmonary and organ function (e.g., kidney) to minimize risks of serious adverse reactions (e.g., cytokine release syndrome)

For B-cell precursor acute lymphoblastic leukemia (ALL), Kymriah® may be approved when all of the following criteria are met:

1. Documentation of cluster of differentiation 19 (CD19) positive, B-cell precursor acute lymphoblastic leukemia (ALL), and
2. Disease is considered refractory, or in second or later relapse, as defined by any one of the following scenarios:
 - a. Second or later bone marrow relapse, or
 - b. Bone marrow relapse after allogenic stem cell transplant, or
 - c. Primary refractory (not achieving a complete response after two cycles of standard chemotherapy), or
 - d. Chemorefractory (not achieving a complete response after one cycle of standard chemotherapy for relapsed disease), and
3. Member is not eligible for allogenic stem cell transplant, and
4. For Philadelphia chromosome (Ph)-positive disease only: Have failed adequate trials of, contraindication, or intolerance to two prior lines of tyrosine kinase inhibitor (TKI) therapy (e.g., imatinib, dasatinib, nilotinib, ponatinib)
5. Performance score on Karnofsky or Lansky Scale is greater than or equal to 50% or Eastern Cooperative Oncology Group (ECOG) performance score is 0-3
6. No evidence of active infection or inflammatory disorder (including hepatitis B or C, active graft vs. host disease)

For relapsed or refractory large B-cell lymphoma, Breyanzi®, Yescarta® or Kymriah® may be approved

when all of the following criteria are met:

1. Confirmed diagnosis of relapsed or refractory FDA approved large B-cell lymphomas
2. Refractory or relapse to two or more prior treatment regimens. Prior therapy must have included the following unless otherwise not indicated/tolerated: a. An anthracycline containing chemotherapy regimen (e.g. doxorubicin), and b. Anti-CD20 monoclonal antibody (e.g. rituximab)
3. Asymptomatic or minimally symptomatic with Eastern cooperative oncology group (ECOG) performance status 0-1
4. Member does not have any of the following:
 - a. Primary central nervous system (CNS) lymphoma
 - b. Evidence of active infection or inflammatory disorder (including hepatitis B or C, active graft vs. host disease)

For relapsed or refractory mantle cell lymphoma (MCL), Tecartus™ may be approved when all of the following criteria are met:

1. Histologically confirmed mantle-cell lymphoma [i.e. cyclin D1 overexpression or chromosomal translocation]
2. Disease is considered relapsed or refractory
3. Previous use to the following therapy: anthracycline or bendamustine containing chemotherapy, an anti-CD20 monoclonal antibody, and BTK inhibitor therapy
4. Asymptomatic or minimally symptomatic with Eastern cooperative oncology group (ECOG) performance status 0-1
5. No evidence of active infection or inflammatory disorder (including hepatitis B or C, active graft vs. host disease)

For multiple myeloma, Abecma® may be approved when all of the following criteria are met:

1. Confirmed diagnosis of multiple myeloma
2. Refractory or relapsed disease to four or more prior lines of therapy. Prior therapy must have included an immunomodulatory agent, a proteasome inhibitor, and an anti-CD38 monoclonal antibody
3. Asymptomatic or minimally symptomatic with Eastern Cooperative Oncology Group (ECOG) performance status 0-1
4. No evidence of active infection (including hepatitis B or C, active graft vs. host disease)

AGE RESTRICTION

Abecma®:

1. Approved for 18 years of age and older

Breyanzi®:

2. Approved for 18 years of age and older

Kymriah®:

1. Approved for 25 years of age or younger for acute lymphoblastic leukemia (ALL)
2. Approved for 18 years of age and older for relapsed or refractory large B-cell lymphoma

Tecartus™:

1. Approved for 18 years of age and older

Yescarta®:

1. Approved for 18 years of age and older

PRESCRIBER RESTRICTION

Must be prescribed by, or in consultation with, an oncologist

COVERAGE DURATION

Two months (limited to one treatment course per lifetime, with four doses of tocilizumab [Actemra®] at up to 800mg per dose).

OTHER CRITERIA

N/A

CONSTIPATION AGENTS

MEDICATION(S)

RELISTOR 12 MG/0.6 ML SYRINGE, RELISTOR 12 MG/0.6 ML VIAL, RELISTOR 8 MG/0.4 ML SYRINGE

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

1. For all requests, the patient must have an FDA labeled indication for the requested agent.
2. For all requests, medication will not be used concomitantly with other intestinal secretagogues, selective 5-HT agonists or peripherally acting mu-opioid receptor antagonists covered by this policy
3. For patients already established on the requested product (starting on samples will not be considered as established on therapy):
 - i. Documentation of response to therapy (e.g., less straining, less pain on defecation, improved stool consistency, increased number of stools per week or reduction in the number of days between stools)
4. For patients not established on the requested product must meet ALL of the following indication-specific criteria:
 - i. For chronic idiopathic constipation (CIC):
 - a. Documentation of weekly constipation (less than three (3) spontaneous bowel movements) for at least three (3) months
 - b. Screen for constipation-inducing medications and medical rationale provided for continuing these medications, if applicable
 - c. Inadequate response or contraindication to a reasonable trial (at least two (2) weeks treatment) to ALL of the following:
 - 1) Regular use of dietary fiber supplementation (e.g. cereal, citrus, fruits or legumes) or use of bulking agents (e.g., psyllium or methylcellulose taken with adequate fluids),
 - 2) A stimulant laxative (e.g. senna, bisacodyl)
 - 3) Routine laxative therapy, with a different mechanism of action than the laxative(s) listed above (e.g., lactulose, Miralax®)
 - 4) Lubiprostone (Amitiza®)
 - ii. For irritable bowel syndrome with constipation (IBS-C):

a. Documentation of recurrent abdominal pain occurring, on average, at least one (1) day per week during the previous three (3) months with two (2) or more of the following criteria:

- 1) Related to defecation (either increased or improved pain)
- 2) Associated with a change in stool frequency
- 3) Associated with a change in stool form (appearance)

b. Inadequate response or contraindication to a reasonable trial (at least two (2) weeks treatment) to ALL of the following:

- 1) Regular use of dietary fiber supplementation (e.g. cereal, citrus, fruits or legumes) or use of bulking agents (e.g., psyllium or methylcellulose taken with adequate fluids)
- 2) Routine laxative therapy with polyethylene glycol (Miralax®)

c. For Zelnorm®: patient is a woman aged 65 years or younger without contraindication to therapy.

Contraindications include:

- 1) History of myocardial infarction (MI), stroke, transient ischemic attack (TIA), or angina
- 2) History of ischemic colitis or other forms of intestinal ischemia, bowel obstruction, symptomatic gallbladder disease, suspected sphincter of Oddi dysfunction, or abdominal adhesion
- 3) Moderate or severe hepatic impairment
- 4) Severe renal disease or end-stage renal disease

iii. For opioid-induced constipation (OIC):

a. Patient is on chronic opioid therapy

b. Documentation of less than three (3) spontaneous bowel movements per week

c. Inadequate response or contraindication to a reasonable trial (at least two (2) weeks treatment) of ALL of the following:

- 1) A stimulant laxative (e.g. senna, bisacodyl)
- 2) Routine laxative therapy, with a different mechanism of action than the laxative above (e.g. lactulose, Miralax®)
- 3) For Relistor®: Failure, contraindication, or intolerance to one (1) of the following medications:

i. Naloxegol (Movantik®)

ii. Lubiprostone (Amitiza®)

iii. Naldemedine (Symproic®)

QUANTITY LIMIT:

Relistor:

- 8-mg syringe: one (1) single use syringe per day (12 ml per 30 days)
- 12-mg syringe or vial: one (1) single use syringe or vial per day (18 ml per 30 days)
- 150-mg tablet: three (3) tablets per day

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

For OIC: Initial authorization will be approved for six (6) months. Reauthorization will be approved for one (1) year

For CIC or IBS: Authorization will be approved until no longer eligible with the plan, subject to formulary or benefit changes

OTHER CRITERIA

N/A

CONTINUOUS GLUCOSE MONITORS FOR PERSONAL USE

MEDICATION(S)

DEXCOM, DEXCOM G4, DEXCOM G5, DEXCOM G5-G4 SENSOR, DEXCOM G6, FREESTYLE LIBRE 10 DAY READER, FREESTYLE LIBRE 10 DAY SENSOR, FREESTYLE LIBRE 14 DAY READER, FREESTYLE LIBRE 14 DAY SENSOR, FREESTYLE LIBRE 2 READER, FREESTYLE LIBRE 2 SENSOR

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

I. Continuous glucose monitors may be considered medically necessary and covered for the treatment of insulin-dependent diabetes when all of the following criteria are met:

A. The requested device is FDA-approved and is being used in accordance with the approved indications of use, and

B. The patient has been on a program of multiple daily injections of insulin (i.e., at least two injections per day), and

C. Documented history of inadequate glycemic control despite compliance with frequent self-monitoring (four or more finger sticks per day) and patient has any of the following problems controlling blood glucose level:

1. Documented hypoglycemia unawareness, or
2. Documented recurring episodes (two or more events) of clinically significant hypoglycemia (less than 54 mg/dl) or fasting hyperglycemia (greater than 150 mg/dl), or
3. Glycosylated hemoglobin level (HbA1C) greater than 7%, or
4. History of recurring, symptomatic hypoglycemia, or
5. Fasting blood sugars frequently exceeding 200 mg/dL, or
6. History of severe glycemic fluctuations, or
7. Documented need for more than five daily injections of insulin or patient is using an insulin pump.

Replacement of Continuous Glucose Monitors

II. Upgrade or replacement of continuous glucose monitor systems may be considered medically necessary and covered when there is documentation that one or more of the device components meet all of the following criteria (A.-C.):

- A. Are no longer functional, and
- B. Are not under warranty, and
- C. Cannot be repaired.

III. Upgrade or replacement of continuous glucose monitor systems is considered not medically necessary and not covered when criterion II above is not met.

Upon approval, concurrent use of test strips will be limited to:

- Dexcom G6/Freestyle Libre: 50 test strips per 90-day supply
- o An additional 50 test strips per 90 days may be approved with documentation that the patient has low blood glucose levels requiring verification at least two times per week (See Diabetic DME policy).

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Authorization will be approved until no longer eligible with the plan, subject to formulary or benefit changes

OTHER CRITERIA

N/A

CRYSVITA - MEDICAL BENEFIT

MEDICATION(S)

CRYSVITA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

Initial authorization for new starts:

1. One of the following diagnoses:
 - a. Diagnosis of X-linked hypophosphatemia (XLH) supported by ONE or more of the following:
 - i. Confirmed PHEX mutation in the patient or a directly related family member with appropriate X-linked inheritance
 - ii. Elevated Serum fibroblast growth factor 23 (FGF23) level greater than 30 pg/mL
 - b. Clinical diagnosis of tumor-induced osteomalacia (TIO) and all of the following:
 - i. Associated with tumors that cannot be identified or curatively resected
 - ii. FGF23 level of at least 100 pg/mL, and
2. Documentation that serum phosphorus level is below the normal range for age, (use laboratory-specific reference ranges if available), and
3. One of the following:
 - a. Patient's epiphyseal plate has NOT fused, or
 - b. Patient meets all of the following:
 - i. Patient's epiphyseal plate has fused, and
 - ii. Patient is experiencing clinical signs and symptoms of disease (e.g., limited mobility, musculoskeletal pain, bone fractures), and
4. Failure of calcitriol with an oral phosphate agent, unless contraindicated or clinically significant adverse effects are experienced, and
5. Documentation of patient's current weight and that dosing is in accordance with the United States Food and Drug Administration approved labeling

For patients established on therapy with burosumab for X-linked hypophosphatemia all of the following criteria must be met:

1. Documentation of recent serum phosphorus level and levels have normalized while on therapy, and
2. Documentation of at least one of the following responses to therapy:
 - a. Improvement in skeletal deformities

- b. Healing of fracture or pseudofractures
 - c. Reduction in number of fractures/pseudofractures
 - d. Increase in growth velocity, and
3. Documentation of patient's current weight and that dosing continues to be in accordance with the United States Food and Drug Administration approved labeling

For patients established on therapy with burosumab for hypophosphatemia in tumor induced osteomalacia (TIO) all of the following criteria must be met:

- 1. Documentation that tumor continues to be unidentifiable or unresectable
- 2. Documentation of recent serum phosphorus level and levels have normalized while on therapy, and
- 3. Documentation of at least one of the following responses to therapy:
 - a. Improvement in skeletal deformities
 - b. Healing of fracture or pseudofractures
 - c. Reduction in number of fractures/pseudofractures
 - d. Increase in growth velocity, and
- 4. Documentation of patient's current weight and that dosing continues to be in accordance with the United States Food and Drug Administration approved labeling

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Prescribed by, or in consultation with, an endocrinologist or specialist experienced in the treatment of metabolic bone disorders.

COVERAGE DURATION

Initial authorization will be approved for six (6) months and reauthorization will be approved for one (1) year.

OTHER CRITERIA

N/A

DISPOSABLE INSULIN PUMPS

MEDICATION(S)

OMNIPOD 5 PACK POD, OMNIPOD DASH 5 PACK POD

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

I. Disposable insulin pumps may be considered medically necessary and covered for the treatment of insulin-dependent diabetes when one of the following criteria are met:

A. The request is for an insulin pump without a continuous glucose monitor for Type 1 diabetes, or

B. All of the following:

1. The requested device is FDA-approved and is being used in accordance with the approved indications of use, and
2. The patient has been on a program of multiple daily injections of insulin (i.e., at least 2 injections per day), and
3. Documented history of inadequate glycemic control despite compliance with frequent self-monitoring (4 or more finger sticks per day) and patient has any of the following problems controlling blood glucose level:
 - i. Documented hypoglycemia unawareness, or
 - ii. Documented recurring episodes (two or more events) of clinically significant hypoglycemia (less than 54 mg/dl) or fasting hyperglycemia (greater than 150 mg/dl), or
 - iii. Glycosylated hemoglobin level (HbA1C) greater than 7%, or
 - iv. History of recurring, symptomatic hypoglycemia, or
 - v. Fasting blood sugars frequently exceeding 200 mg/dL, or
 - vi. History of severe glycemic fluctuations, or
 - vii. Documented need for more than 5 daily injections of insulin.

Replacement of Disposable Insulin Pumps

II. Upgrade or replacement of existing advanced diabetes management technology may be considered medically necessary and covered when there is documentation that one or more of the device components meet all of the following criteria (A.-C.):

- A. Are no longer functional, and
- B. Are not under warranty, and
- C. Cannot be repaired.

III. Upgrade or replacement of existing advanced diabetes management technology is considered not

medically necessary and not covered when criterion II above is not met.

QUANTITY LIMIT:

Omnipod Dash pods: 10 pods per 30 days

- Requests for additional pods may be covered when the patients' total daily dose of insulin is more than 65 units per day

Note: The Omnipod pump Personal Diabetes Manager (PDM) is supplied by the manufacturer for most Commercial patients, so patients should be referred to Insulet Corporation for free starter kit or for replacement at 1-800-591-3455.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Authorization will be approved until no longer eligible with the plan, subject to formulary or benefit changes

OTHER CRITERIA

N/A

DURYSTA

MEDICATION(S)

DURYSTA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

The following criteria must be met:

1. The patient is not receiving re-treatment of eye(s) previously treated with bimatoprost intracameral implant (Durysta®)
2. Trial and failure, intolerance or contraindication to at least two ophthalmic products (either as monotherapy or as concomitant therapy) from two different pharmacological classes, one of which is an ophthalmic prostaglandin

AGE RESTRICTION

Approved for 18 years and older

PRESCRIBER RESTRICTION

Must be prescribed by an ophthalmologist

COVERAGE DURATION

Authorization will be approved for six months. Approval will be for a one-time use in each treated eye (one implant per treated eye, a total of two implants per patient)

OTHER CRITERIA

N/A

EGRIFTA

MEDICATION(S)

EGRIFTA, EGRIFTA SV

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

1. Diagnosis of HIV-associated lipodystrophy

AND

2. Documentation of patient's waist circumference

a. Waist circumference greater than or equal to 37.4 inches (95 cm) for males

b. Waist circumference greater than or equal to 37 inches (94 cm) for females

AND

3. Documentation of waist-to-hip ratio

a. Waist-to-hip ratio greater than or equal to 0.94 for males

b. Waist-to-hip ratio greater than or equal to 0.88 for females

AND

4. Documentation of a body mass index (BMI) greater than 20 kilograms per meter squared

AND

5. Documentation of fasting blood glucose (FBG) of less than or equal to 150 mg/dL (8.33 mmol/L)

AND

6. Documentation that patient has been on a stable regimen of antiretrovirals for at least eight (8) weeks

Reauthorization will require documentation of clinical improvement (e.g., decrease in waist circumference, improvement in visceral adipose tissue).

AGE RESTRICTION

Adults 18 years of age and older

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Initial authorization and reauthorization will be approved for 6 months.

OTHER CRITERIA

N/A

ELZONRIS

MEDICATION(S)

ELZONRIS

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For initial authorization all of the following criteria must be met:

1. Diagnosis of blastic plasmacytoid dendritic cell neoplasm (BPDCN)
2. Documentation that patient has a current Eastern Cooperative Oncology Group (ECOG) status of 0-1
3. Documentation that patient has a baseline serum albumin level of at least 3.2 g/dL
4. Documentation that patient has adequate cardiac function, defined as LVEF of at least 50% and none of the following:
 - a. Uncontrolled or any NYHA Class 3 or 4 congestive heart failure
 - b. Uncontrolled angina
 - c. History of myocardial infarction or stroke within 6 months of initiating therapy
 - d. Uncontrolled hypertension
 - e. Clinically significant arrhythmias not controlled by medication

Reauthorization requires documentation of positive response to therapy, such as a lack of disease progression.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with an oncologist or hematologist

COVERAGE DURATION

Initial and reauthorization will be approved for 6 months

OTHER CRITERIA

N/A

ENZYME REPLACEMENT THERAPY

MEDICATION(S)

ALDURAZYME, CERAZYME, ELAPRASE, ELELYSO, FABRAZYME, KANUMA, LUMIZYME, MEPSEVII, NAGLAZYME, VIMIZIM, VPRIV

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For initial authorization both of the following must be met:

1. Documentation of FDA-labeled indication for the requested product

AND

2. Dosing is within FDA-labeled guidelines.

Note: If request is for a non-FDA approved dose, medical rational must be submitted in support of therapy with a higher dose for the intended diagnosis (i.e., high-quality peer reviewed literature, accepted compendia or evidence-based practice guidelines) and exceptions will be considered on a case by case basis.

REAUTHORIZATION:

Both of the following must be met:

1. Documentation of successful response to therapy (e.g., disease stability or improvement in symptoms).

2. Dosing is within FDA-labeled guidelines

Note: If request is for a non-FDA approved dose, medical rational must be submitted in support of therapy with a higher dose for the intended diagnosis (i.e., high-quality peer reviewed literature, accepted compendia or evidence-based practice guidelines) and exceptions will be considered on a case by case basis.

QUANTITY LIMIT:

Initial dose approval will be based on patient's current weight. Increases in dose will require new authorization with patient's weight and relevant chart notes

AGE RESTRICTION

• Aldurazyme®: N/A

• Cerezyme®: N/A

- Elaprase®: The safety and efficacy have not been established in patients less than 16 months of age
- Elelyso®: The safety and efficacy have not been established in patients less than four years of age
- Fabrazyme®: Safety and efficacy not established in patients under two years of age
- Kanuma®: N/A
- Lumizyme®: N/A
- Mepsevii®: N/A
- Naglazyme®: N/A
- Vimizim®: The safety and effectiveness have not been established in patients less than five years of age
- Vpriv®: The safety and efficacy have not been established in patients less than four years of age

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with a Hepatologist, Endocrinologist, Medical Geneticist, Cardiologist, Pulmonologist, or Bone and Mineral specialist.

COVERAGE DURATION

Initial authorization will be approved for six months. Reauthorization will be approved for one year.

OTHER CRITERIA

N/A

ERYTHROPOIESIS STIMULATING AGENTS (ESAS)

MEDICATION(S)

ARANESP, EPOGEN, PROCRIT, RETACRIT

COVERED USES

N/A

EXCLUSION CRITERIA

- Patients with uncontrolled hypertension
- Anemia induced from hepatitis C therapy

REQUIRED MEDICAL INFORMATION

1. All diagnoses with the exception of 2e (preoperative use in patients scheduled for elective non-cardiac, nonvascular surgery), must have documented Hemoglobin (HGB) levels of less than or equal to 10g/dl within the 30 days prior to initiation of therapy

AND

2. Must meet all of the listed criteria below for each specific diagnosis:

a. Treatment of Anemia in Chronic Kidney Disease (CKD)

i. Adequate iron stores as indicated by current (within the last 3 months) serum ferritin level greater than or equal to 100 mcg/L or serum transferrin saturation greater than or equal to 20%

b. Treatment of anemia in patients with cancer:

i. Adequate iron stores as indicated by current (within the last 3 months) serum ferritin level more than or equal to 100 mcg/L or serum transferrin saturation more than or equal to 20%

AND

ii. One of the following clinical scenarios:

1. Patient has comorbid chronic kidney disease

2. Patient undergoing palliative treatment

3. Patient is currently on myelosuppressive chemotherapy and anemia is not able to be managed by transfusion therapy

c. Treatment of Anemia in Myelodysplastic Syndromes (MDS) or with myelofibrosis

i. Adequate iron stores as indicated by current (within the last 3 months) serum ferritin level more than or equal to 100 mcg/L or serum transferrin saturation more than or equal to 20%

ii. Must have documented current (within last 3 months) endogenous serum erythropoietin levels less than or equal to 500 mU/mL

d. Anemia associated with zidovudine-treated HIV-infection patients

i. Documented current (within last 3 months) endogenous serum erythropoietin level is less than or equal to 500 mU/ml

ii. Zidovudine dose is less than or equal to 4200mg/week

- e. Preoperative use in patients scheduled for elective noncardiac and nonvascular surgery, all of the following criteria must be met:
- i. Member has preoperative HGB between 10 and 13 g/dL
 - ii. The surgery has a high-risk for perioperative blood loss (e.g., expected to lose more than 2 units of blood)
 - iii. Patient is unwilling to donate autologous blood pre-operatively

Reauthorization:

1. Documentation of continued medical necessity (e.g., ongoing chronic kidney disease)
2. Documented HGB levels of less than or equal to 12 g/dl within previous 30 days

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Initial authorization and reauthorization will be for one (1) year

OTHER CRITERIA

N/A

EVENITY

MEDICATION(S)

EVENITY, EVENITY (2 SYRINGES)

COVERED USES

N/A

EXCLUSION CRITERIA

Myocardial infarction or stroke within the preceding year, hypocalcemia

REQUIRED MEDICAL INFORMATION

For the treatment or prevention of osteoporosis, must meet ONE of the following criteria:

1. Patient has a history of multiple or severe vertebral fractures, or history of fragility fractures
2. Patient has a spine or hip bone mineral density (BMD) T-score less than or equal to -2.5 and high risk for fracture, defined as one of the following:
 - a. Age more than 80 years
 - b. Chronic glucocorticoid use
 - c. Documented increased fall risk
3. Patient has a spine or hip BMD T-score less than or equal to -2.5 and one of the following:
 - a. Documented failure to anti-resorptive therapy (e.g., denosumab, bisphosphonates). Failure is defined as a new fracture or worsening BMD while adherent to therapy
 - b. Documented contraindication or intolerance to therapy with all of the following: 1. denosumab, 2. oral bisphosphonate (e.g., alendronate), and 3. IV bisphosphonate therapy (i.e., zoledronic acid)
4. Patient has a spine or hip BMD T-score between -1.0 and -2.5 and BOTH of the following:
 - a. Fracture Risk Assessment (FRAX) probability score for hip fracture of at least 3% or, for other major osteoporosis fracture, of at least 20%
 - b. One of the following:
 - i. Documented failure to anti-resorptive therapy (e.g., denosumab, bisphosphonates). Failure is defined as a new fracture or worsening BMD while adherent to therapy
 - ii. Documented contraindication or intolerance to therapy with all of the following:
 1. Denosumab
 2. Oral bisphosphonate (e.g., alendronate)
 3. IV bisphosphonate therapy (i.e., zoledronic acid)

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with an endocrinologist or rheumatologist.

COVERAGE DURATION

May be approved for up to one (1) year, ensuring the total duration of Evenity® therapy does not exceed one (1) year of total therapy duration.

OTHER CRITERIA

N/A

EXON-SKIPPING THERAPIES FOR DUCHENNE MUSCULAR DYSTROPHY

MEDICATION(S)

AMONDYS-45, EXONDYS-51, VILTEPSO, VYONDYS-53

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

N/A

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

N/A

OTHER CRITERIA

Exon-skipping therapies for Duchene Muscular Dystrophy are not considered medically necessary and will not be covered due to the lack of clinical evidence of improved outcomes and safety.

EXTAVIA

MEDICATION(S)

EXTAVIA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

Initial Authorization for multiple sclerosis (MS), all of the following criteria must be met:

1. Must have one of the following confirmed diagnoses:
 - a. Relapsing-remitting disease (RRMS)
 - b. Secondary progressive multiple sclerosis (SPMS)
 - c. Clinically isolated syndrome (CIS)
2. Documented inadequate response or intolerance to generic dimethyl fumarate or glatiramer, or contraindication to BOTH dimethyl fumarate and glatiramer
3. Documentation of trial and failure, contraindication, or intolerance to one additional of the following preferred agents OR medical rationale why therapies cannot be tried:
 - a. Interferon-beta 1a (Avonex®, Rebif® or Plegridy®)
 - b. Interferon-beta 1b (Betaseron®)
 - c. Teriflunomide (Aubagio®)
 - d. Fingolimod (Gilenya®)
 - e. Ozanimod hydrochloride (Zeposia®)
 - f. Siponimod (Mayzent®)

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with a neurologist.

COVERAGE DURATION

Authorization will be approved until no longer eligible with the plan, subject to formulary and/or benefit changes.

OTHER CRITERIA

N/A

FERTILITY AND RELATED MEDICATIONS

MEDICATION(S)

CETROTIDE, CHORIONIC GONAD 10,000 UNIT VL, CHORIONIC GONAD 12,000 UNIT VL, CHORIONIC GONAD 50,000 UNIT VL, CHORIONIC GONAD 6,000 UNIT VL, FOLLISTIM AQ, GANIRELIX ACETATE, GONAL-F, GONAL-F RFF, GONAL-F RFF REDI-JECT, MENOPUR, NOVAREL, OVIDREL, PREGNYL

COVERED USES

N/A

EXCLUSION CRITERIA

1. Hypogonadism, unrelated to infertility
2. Cryptorchidism

REQUIRED MEDICAL INFORMATION

- I. For treatment of infertility, preferred gonadotropins may be covered if the patient's benefit covers the planned infertility treatment [e.g. intrauterine insemination (IUI) vs. in vitro fertilization (IVF)] and
 - a. For non-preferred gonadotropins: documented intolerance or contraindication to at least two of the preferred agents
- II. For fertility preservation, preferred gonadotropins may be covered if the patient's benefit covers fertility preservation and the following criteria are met:
 - a. The gonadotropin will be used for retrieval and storage of eggs and/or sperm
 - b. The patient will be undergoing treatment for cancer that is expected to cause irreversible infertility as recommended by evidence-based guidelines such as the National Comprehensive Cancer Network (NCCN),
 - c. For non-preferred gonadotropins: documented intolerance or contraindication to at least two of the preferred agents

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

- For infertility: Must be prescribed by, or in consultation with, a reproductive endocrinologist.
- For fertility preservation: Must be prescribed by, or in consultation with, an oncologist or reproductive endocrinologist.

COVERAGE DURATION

Authorization will be approved for one year

OTHER CRITERIA

N/A

FORTEO

MEDICATION(S)

FORTEO, TERIPARATIDE

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For the treatment or prevention of osteoporosis

1. Must meet ONE of the following criteria:

- a. Patient has a history of multiple or severe vertebral fractures, or history of fragility fractures, or
- b. Patient has a spine or hip bone mineral density (BMD) T-score less than or equal to -2.5 and high risk for fracture, defined as one of the following:

- i. Age more than 80 years
- ii. Chronic glucocorticoid use
- iii. Documented increased fall risk, or

c. Patient has a spine or hip BMD T-score less than or equal to -2.5 and one of the following:

- i. Documented failure to anti-resorptive therapy (e.g., denosumab, bisphosphonates). Failure is defined as a new fracture or worsening BMD while adherent to therapy
- ii. Documented contraindication or intolerance to therapy with all of the following: 1. denosumab, 2. oral bisphosphonate (e.g., alendronate), and 3. IV bisphosphonate therapy (i.e., zoledronic acid), or

d. Patient has a spine or hip BMD T-score between -1.0 and -2.5 and BOTH of the following:

- i. Fracture Risk Assessment (FRAX) probability score for hip fracture of at least 3% or, for other major osteoporosis fracture, of at least 20%
- ii. One of the following:

1. Documented failure to anti-resorptive therapy (e.g., denosumab, bisphosphonates). Failure is defined as a new fracture or worsening BMD while adherent to therapy

2. Documented contraindication or intolerance to therapy with all of the following:

- a. Denosumab
- b. Oral bisphosphonate (e.g., alendronate)
- c. IV bisphosphonate therapy (i.e., zoledronic acid).

2. For female patients only:

- a. Documentation of trial and failure to Tymlos® (abaloparatide). Failure is defined as a new fracture or worsening bone mineral density while adherent to Tymlos® (abaloparatide).

AND

b.Total duration of treatment with Tymlos® (abaloparatide) has not exceeded two years.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with an endocrinologist or rheumatologist

COVERAGE DURATION

May be approved for up to 2 years, ensuring the cumulative duration of osteoanabolic therapy does not exceed 2 years in a lifetime. Duration of osteoanabolic therapy is defined as cumulative duration spent on any of the three therapies: abaloparatide, teriparatide, or romosozumab.

OTHER CRITERIA

N/A

GAMIFANT

MEDICATION(S)

GAMIFANT

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

Initiation Criteria:

1. Diagnosis of primary HLH based on a molecular diagnosis OR family history consistent with primary HLH OR 5 out of the following 8 criteria fulfilled:

- a. Fever
- b. Splenomegaly
- c. Cytopenias affecting 2 of 3 lineages in the peripheral blood: hemoglobin less than 9 g/dL, platelets less than $100 \times 10^9/L$, neutrophils less than $1 \times 10^9/L$
- d. Hypertriglyceridemia (fasting triglycerides greater than 3 mmol/L or equal or greater than 265 mg/dL) and/or hypofibrinogenemia (equal or less than 1.5 g/L)
- e. Hemophagocytosis in bone marrow, spleen, or lymph nodes with no evidence of malignancy
- f. Low or absent NK-cell activity
- g. Ferritin equal or greater than 500 mcg/L
- h. Soluble CD 25 equal or greater than 2400 U/mL

2. Refractory, recurrent, or progressive disease or intolerance with conventional HLH therapy (corticosteroids, methotrexate, cyclosporine A, etoposide, anti-thymocyte globulin) based on one of the following criteria:

- a. Having not responded or not achieved a satisfactory response
- b. Having not maintained a satisfactory response to conventional HLH therapy
- c. Intolerance to conventional HLH treatments

3. Patient is a candidate for stem cell transplant and emapalumab is being used as part of the induction or maintenance phase for stem cell transplant and will be discontinued at the initiation of conditioning for stem cell transplant

4. Dosing is in accordance with the United States Food and Drug Administration approved labeling

5. Documentation that patient currently has no active infection (e.g. mycobacteria and Histoplasma Capsulatum)

Reauthorization Criteria:

1. Patient continues to be a candidate for stem cell transplant
2. Documentation of disease improvement such as:
 - a. Complete response defined as normalization of all HLH abnormalities (i.e. no fever, no splenomegaly, neutrophils more than $1 \times 10^9/L$, platelets more than $100 \times 10^9/L$, ferritin less than $2,000 \mu g/L$, fibrinogen more than $1.50 g/L$, D-dimer less than $500 \mu g/L$, normal CNS symptoms, no worsening of sCD25 more than 2-fold baseline)
 - b. Partial response defined as normalization of more than or equal to 3 HLH abnormalities
 - c. HLH improvement defined as more than or equal 3 HLH abnormalities improved by at least 50% from baseline
3. Documentation that patient is being monitored for serious infections (such as tuberculosis, adenovirus, EBV, and CMV)
4. Documentation that dose does not exceed max FDA approved dosing of 10 mg/kg per dose for two doses per week

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with a hematologist or oncologist

COVERAGE DURATION

Initial authorization approved for 3 months, reauthorization for 1 month

OTHER CRITERIA

N/A

GIVLAARI

MEDICATION(S)

GIVLAARI

COVERED USES

N/A

EXCLUSION CRITERIA

Use post liver transplant

REQUIRED MEDICAL INFORMATION

For initial authorization, all of the following criteria must be met:

1. Confirmed diagnosis of acute hepatic porphyria [i.e., acute intermittent porphyria, hereditary corproporhyria, variegate porphyria, aminolevulinic acid (ALA) dehydratase deficient porphyria]

AND

2. One of the following:

a. Active disease defined as two (2) documented porphyria attacks within the past six (6) months which required either hospitalization, urgent care visit, or intravenous hemin administration, or

b. Patient is currently receiving treatment with prophylactic hemin to prevent porphyria attacks

3. Documentation that patient will not receive concomitant prophylactic hemin treatment while on therapy with givosiran therapy

4. Documentation that patient's dosing is in accordance with FDA labeling (patient's current weight must be included in documentation) and is subject to audit

Reauthorization requires documentation of one of the following:

1. Reduction in the number or severity of porphyria attacks

2. Reduction in number of hospitalizations due to acute porphyria attacks

3. Decreased hemin administration from baseline

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with a hepatologist, gastroenterologist, or hematologist

COVERAGE DURATION

Initial authorization will be approved for 6 months.

Reauthorization will be approved for 1 year.

OTHER CRITERIA

N/A

GONADOTROPIN RELEASING HORMONE AGONISTS

MEDICATION(S)

FENSOLVI, LUPANETA PACK, LUPRON DEPOT, LUPRON DEPOT-PED, SUPPRELIN LA, TRELSTAR, TRIPTODUR, VANTAS, ZOLADEX

COVERED USES

N/A

EXCLUSION CRITERIA

Treatment of male infertility

REQUIRED MEDICAL INFORMATION

For oncological indications: Use must be for a FDA approved indication or indication supported by National Comprehensive Cancer Network guidelines with recommendation 2A or higher

For anemia associated with uterine leiomyomata (fibroids)

1. Documented trial, failure, intolerance or contraindication to at least 30 days of therapy with iron supplementation alone

AND

2. Documentation that Lupron® will be used in combination with iron supplementation

For uterine leiomyomata (fibroids)

1. Documentation that surgical removal of fibroids is planned within four months

AND

2. And one of the following, less invasive surgical methods will be employed:

a. Documentation of an enlarged uterus that will require a midline rather than transverse incision.

b. Documentation that shrinking the uterus or fibroids will allow for a vaginal hysterectomy rather than an abdominal procedure.

For endometriosis:

1. Documentation that other causes of gynecologic pain have been ruled out (e.g., irritable bowel syndrome, interstitial cystitis, urinary tract disorders)

2. For Synarel®: documented trial and failure to Lupron® with add-back progesterone therapy (such as norethindrone acetate) or Lupaneta® Pack.

Reauthorization

For Lupron® requires documentation that it will be used in combination with “add-back” progesterone therapy (e.g. norethindrone) to help prevent bone mineral density loss.

Reauthorization

For Synarel® and Zoladex® is not recommended. Treatment is only recommended for up to six months with these agents for endometriosis

For central precocious puberty

Note, a one-time dose may be approved for diagnostic purposes

For Initial Authorization:

1. Documentation of a history of early onset of secondary sexual characteristics (age eight years and under for females or nine years and under for males)

AND

2. Confirmation of diagnosis by one of the following:

a. Pubertal response to a GnRH or GnRH analog (such as leuprolide) stimulation test [e.g., stimulated peak luteinizing hormone (LH) of approximately 4.0 to 6.0 IU/L and/or elevated ratio of LH/follicle-stimulating hormone at 0.66 or greater (reference range may vary depending on assay)]

b. Pubertal level of basal LH levels (0.3 IU/L or greater)

c. Bone age advanced one year beyond the chronological age

AND

3. For Synarel®: documented trial and failure or contraindication/intolerance to Lupron® and, either Triptodur® or Supprelin LA®

For Reauthorization:

1. Clinical response to treatment (i.e., pubertal slowing or decline, height velocity, bone age, LH, or estradiol and testosterone level), and

2. Documentation that hormonal and clinical parameters are being monitored periodically during treatment to ensure adequate hormone suppression.

Discontinuation of leuprolide should be considered before age 11 years for females and age 12 years for males. However, treatment discontinued at the appropriate age of onset of puberty should be at discretion of the treating provider.

For gender-affirming services:

1. Prescribed by or in consultation with an endocrinologist

2. Demonstration that puberty has progressed to a minimum of Tanner Stage 2

3. For Medicaid: request is for Vantas® (histrelin acetate 50 mg implant)

For Endometrial thinning/dysfunctional uterine bleeding:

1. Documentation for use prior to endometrial ablation

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Anemia from fibroids: Authorization will be approved for up to three months (NO reauthorization)

Uterine leiomyomata (fibroids): Authorization will be approved for four months. No reauthorization

Endometriosis: For Lupron® and Lupaneta® Pack – authorization/reauthorization will be approved for up to six months (total of 12 months). For Synarel®/Zoladex® - initial authorization for up to six months and no reauthorization

CPP: Authorization/reauthorization will be approved for up to one year

GID: Authorization/reauthorization will be approved for up to one year

Endometrial Thinning/Dysfunctional Uterine Bleeding: Initial authorization for two months. No reauthorization.

Oncological Indications: Authorization/reauthorization will be approved for one year

In vitro fertilization: Authorization/reauthorization will be approved for one year

OTHER CRITERIA

N/A

HEMLIBRA - MEDICAL BENEFIT

MEDICATION(S)

HEMLIBRA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

1. Use is for routine prophylaxis to prevent or reduce the frequency of bleeding episodes
AND
2. Diagnosis of hemophilia A (congenital factor VIII deficiency) and documentation of ANY of the following:
 - a. Factor VIII inhibitors (defined as at least 5 Bethesda units per milliliter)
 - b. Severe hemophilia (defined as pre-treatment factor VIII level less than 1%)
 - c. Moderate hemophilia (defined as pre-treatment factor VIII level of 1% to less than 5%) or mild hemophilia (defined as pre-treatment factor VIII level of 5% to less than 40%) with:
 - i. One (1) or more spontaneous episodes of bleeding into the central nervous system, large joints (ankles, knees, hips, elbows, shoulders) or other serious, life-threatening bleed

When the above criteria are met, Hemlibra® (emicizumab-kxwh) will be approved for a loading dose of 3 mg/kg once weekly for four (4) weeks, followed by any of the three (3) maintenance dosing regimens below:

- 1.5 mg/kg once weekly
- 3 mg/kg every 2 weeks
- 6 mg/kg every 4 weeks

Reauthorization criteria: Documentation of positive clinical response to emicizumab therapy (e.g. reduction in the number/severity of bleeds)

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

To be prescribed by, or in consultation with a hematologist.

COVERAGE DURATION

Initial authorization: six (6) months

Reauthorization: 12 months

OTHER CRITERIA

N/A

HOMOZYGOUS FAMILIAL HYPERCHOLESTEROLEMIA (FH) AGENTS

MEDICATION(S)

EVKEEZA

COVERED USES

N/A

EXCLUSION CRITERIA

1. Concomitant use of therapies on this policy
2. Current pregnancy
3. Diagnosis of Heterozygous familial hypercholesterolemia or other hyperlipidemia disorders

REQUIRED MEDICAL INFORMATION

All of the following must be met:

1. Diagnosis of Homozygous Familial Hypercholesterolemia (HoFH) as evidenced by either genetic or clinical confirmation, as outlined below:
 - a. Genetic confirmation: biallelic functional mutations in the low density lipoprotein receptor (LDLR), apolipoprotein B (apo B), or proprotein convertase subtilisin/kexin type 9 (PCSK9) genes
 - b. Clinical confirmation defined as untreated total cholesterol greater than 500 mg/dL and one of the following:
 - i. Presence of xanthomas before the age of 10 years, or
 - ii. Untreated total cholesterol level greater than 250 mg/dL in both parents
2. Current use of all of the following therapies:
 - a. High-intensity statin therapy, defined as atorvastatin 80 mg daily or rosuvastatin 40 mg daily, unless contraindicated or documented statin intolerance
 - b. Ezetimibe, unless contraindicated or prior intolerance
 - c. PCSK-9 inhibitor (e.g., evolocumab), unless contraindicated or prior intolerance
3. Documentation of LDL cholesterol levels greater than 100 mg/dL despite at least six months of use of the therapies outlined above

Initial reauthorization requires documentation of at least a 30% reduction in LDL cholesterol levels from pre-treatment levels

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by, or in consultation with, a cardiologist, endocrinologist, or board certified lipidologist.

COVERAGE DURATION

Initial authorization will be approved for six months. Reauthorization will be approved until no longer eligible with the plan, subject to formulary and/or benefit changes.

OTHER CRITERIA

N/A

HP ACTHAR GEL

MEDICATION(S)

ACTHAR

COVERED USES

N/A

EXCLUSION CRITERIA

All other indications beside infantile spasms are not considered medically necessary and are excluded for coverage.

REQUIRED MEDICAL INFORMATION

For infantile spasm: H.P. Acthar Gel® will be approved for one month of therapy at the following dose: 75 units/m(2) injected intramuscularly twice daily

Reauthorization will require medical rationale for continuing treatment, as recommended treatment duration is for 2 weeks followed by two-week taper to avoid adrenal insufficiency.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Initial authorization/reauthorization will be approved for one month.

OTHER CRITERIA

N/A

IL-5 INHIBITORS - CINQAIR FASENRA, NUCALA - MEDICAL BENEFIT

MEDICATION(S)

CINQAIR, FASENRA, FASENRA PEN, NUCALA

COVERED USES

N/A

EXCLUSION CRITERIA

Concurrent use with another therapeutic immunomodulator agent utilized for the same indication.

REQUIRED MEDICAL INFORMATION

For initial authorization, must meet all of the following criteria:

For eosinophilic asthma:

1. Documentation of eosinophilic asthma by one of the following:
 - a. A blood eosinophil count of greater than 150 cells/microliter in the past 12 months
 - b. Past history of eosinophilic asthma if currently on daily maintenance treatment with oral glucocorticoids
2. Documentation of treatment with maximally tolerated dose of medium to high-dose inhaled corticosteroid plus an additional asthma controller (e.g., long-acting inhaled beta2-agonist, leukotriene receptor antagonist) and has been compliant to therapy in the past three months (this may be verified by pharmacy claims information)
3. Documentation of severe asthma with inadequate asthma control despite above therapy, defined as one of the following:
 - a. Asthma Control Test (ACT) score less than 20 or Asthma Control Questionnaire (ACQ) score greater than or equal to 1.5
 - b. At least two asthma exacerbations requiring oral systemic corticosteroids in the last 12 months
 - c. At least one asthma exacerbation requiring hospitalization, emergency room or urgent care visit

For Eosinophilic Granulomatosis with Polyangiitis (EGPA):

1. Request is for Nucala®
2. Confirmed diagnosis of eosinophilic granulomatosis with polyangiitis (EGPA)
3. History or presence of asthma
4. Blood eosinophil level of at least 10% or an absolute eosinophil count of more than 1000 cells/microliter
5. Documentation of one of the following
 - a. History of relapse requiring an increase in glucocorticoid dose, initiation or increase in other immunosuppressive therapy, or hospitalization in the previous two years while receiving at least 7.5 mg/day prednisone (or equivalent)

OR

- b. Failure to achieve remission following a standard induction regimen administered for at least three

months OR recurrence of symptoms of EGPA while tapering of glucocorticoids

i. Standard treatment regimens include: prednisone [or equivalent] dosed at least 7.5 mg/day in combination with an immunosuppressant such as cyclophosphamide, azathioprine, methotrexate, or mycophenolate mofetil

For Hyperesoinophilic Syndrome (HES)

1. Request is for Nucala®
2. Document of primary HES without an identifiable nonhematologic secondary cause such as parasitic infections, solid tumors, or T cell lymphoma
3. Blood eosinophil count of 1,000 cells/microliter or higher for at least six months
4. Documentation of use of HES therapy including one of the following in the past for the past 12 months:
 - a. chronic or episodic oral corticosteroids (OCS)
 - b. immunosuppressive therapy
 - c. cytotoxic therapy
5. Documentation of at least two HES flares within the past 12 months (defined as HES-related worsening of clinical symptoms or blood eosinophil counts requiring an escalation in therapy)

Reauthorization documentation of response to therapy, such as attainment and maintenance of remission or decrease in number of relapses

AGE RESTRICTION

Nucala®: Approved for six years of age or older for eosinophilic asthma, approved for 18 years of age and older for EGPA and approved for 12 years of age and older for HES

Cinqair®: Approved for 18 years of age or older

Fasenra®: Approved for 12 years of age or older

PRESCRIBER RESTRICTION

For eosinophilic asthma: must be prescribed by or in consultation with an asthma specialist (such as a pulmonologist, immunologist, or allergist)

For Eosinophilic Granulomatosis with Polyangiitis: must be prescribed by or in consultation with a pulmonologist, neurologist, or rheumatologist

For hypereosinophilic syndrome (HES): must be prescribed by or in consultation with hematologist, immunologist, pulmonologist, cardiologist, or neurologist.

COVERAGE DURATION

For EGPA and HES: Initial authorization and reauthorization will be approved for one year.

For asthma: Initial authorization will be approved for one year and reauthorization will be approved until no longer eligible with the plan, subject to formulary or benefit changes

OTHER CRITERIA

N/A

IMMUNE GAMMA GLOBULIN (IGG)

MEDICATION(S)

ASCENIV, BIVIGAM, CUTAQUIG, CUVITRU, FLEBOGAMMA DIF, GAMASTAN, GAMASTAN S-D, GAMMAGARD LIQUID, GAMMAGARD S-D, GAMMAKED, GAMMAPLEX, GAMUNEX-C, HIZENTRA, HYQVIA, OCTAGAM, PANZYGA, PRIVIGEN, XEMBIFY

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

Initial Authorization for ALL indications:

1. The medical diagnosis is an FDA approved indication or is listed as a covered medical condition below and any indication specific criteria in the policy is met

AND

2. Requested dosage, frequency and length of therapy are supported by FDA-approved labeling, accepted compendia and/ or evidence-based practice guidelines. If request is for a non-standard dose, frequency or length, medical rationale should be provided and exceptions will be considered on a case by case basis.

Dosing is subject to audit.

Re-Authorization for ALL indications:

1. Documentation of response to therapy and any indication specific re-authorization criteria listed below is met

Indication-Specific Requirements:

Primary immune deficiency disorders such as agammaglobulinemia, hypogammaglobulinemia (i.e., common variable immunodeficiency), Hyper-IgM (i.e., X-linked or autosomal recessive hypogammaglobulinemia), Wiskott-Aldrich syndrome

1. The patient has one of the following:

a. The patient has a total IgG less than 200 mg/dL at baseline prior to immune globulin therapy

b. The patient has abnormal Bruton tyrosine kinase (BTK) gene or absence of BTK protein

c. The patient has an absence of B lymphocytes

d. The patient meets all of the following:

i. One of the following:

1. The patient has selective IgG subclass deficiency [Defined as deficiency of one or more IgG subclasses

(e.g., IgG1, IgG2, IgG3, or IgG4) more than two standard deviations (SD) below age-specific mean, assessed on two separate occasions during infection free period

2. The patient has specific antibody deficiency (SAD) with normal levels of both immunoglobulin and total IgG subclasses

3. The patient has hypogammaglobulinemia (defined as total IgG less than 700 mg/dL OR more than two SDs below mean for the patient's age at baseline prior to immune globulin therapy)

ii. The patient has a lack of response or inability to mount an adequate response to protein and/or polysaccharide antigens (e.g., inability to make IgG antibody against either diphtheria and tetanus toxoids, or pneumococcal polysaccharide vaccine, or both)

iii. The patient has evidence of recurrent, persistent, severe, difficult-to-treat infections (e.g., recurring otitis media, bronchiectasis, recurrent infections requiring IV antibiotics) despite aggressive prophylactic management and treatment with antibiotics

Reauthorization:

1. Documentation that treatment has been effective in reducing the number or severity of clinical infections

Prevention of infections in patients with B-cell chronic lymphocytic leukemia (CLL):

1. Documented pre-treatment endogenous IgG less than 700 mg/dL OR more than two standard deviations below mean for the patient's age

OR

2. History of recurrent, severe bacterial infections requiring antibiotics and/or hospitalization

Kawasaki Disease:

1. Documentation that use is for acute treatment given in conjunction with aspirin and within 10 days of the onset of symptoms

Idiopathic or Immune Thrombocytopenic Purpura (ITP):

(Platelet counts expressed per microliter and should be obtained within the past 30 days)

For children with ITP:

1. Documentation of one of the following:

a. Platelet count less than 20,000 and significant mucous membrane bleeding

b. Platelet count less than 10,000 and minor purpura

c. Rapid increase in platelets required due to planned surgery, dental extractions, or other procedures likely to cause blood loss

Pregnant Women with ITP:

1. Documentation of one of the following:

a. Platelet count is less than 100,000

b. Past history of splenectomy

c. Past history of delivered infant with autoimmune thrombocytopenia

Adult Patients with ITP:

1. Documentation of one of the following:
 - a. Platelet count of less than 30,000
 - b. Platelet count less than 50,000 with acute bleeding or high-risk of bleeding
 - c. To defer or avoid splenectomy
 - d. Rapid increase in platelets required due to planned surgery, dental extractions, or other procedures likely to cause blood loss (platelet count goal is generally greater than 50,000)
2. Documentation that IGG product will be used in combination with corticosteroid therapy or corticosteroid therapy is contraindicated

Dermatomyositis and polymyositis:

1. Documented trial, failure, intolerance or contraindication to systemic corticosteroids (i.e. prednisone or methylprednisolone)
AND
2. Documented trial, failure, intolerance or contraindication to immunosuppressant therapy (e.g., methotrexate, azathioprine, cyclosporine, 6-mercaptopurine, chlorambucil, cyclophosphamide)
AND
3. Documentation of severe symptoms/disability despite previous therapy with above agents

Reauthorization: Documented response to therapy

Chronic inflammatory demyelinating polyneuropathy (CIDP):

1. Documentation of severe disability
AND
2. One of the following:
 - a. Documented trial, failure, intolerance or contraindication to systemic corticosteroids (i.e. prednisone or methylprednisolone)
 - b. Documentation of pure motor CIDP

Autoimmune Hemolytic Anemia:

1. Documented trial, failure, intolerance or contraindication to systemic corticosteroids (i.e. prednisone or methylprednisolone)
AND
2. Documented trial, failure, intolerance or contraindication to another conventional therapy for autoimmune hemolytic anemia (e.g., splenectomy, cyclophosphamide, azathioprine, cyclosporine)

Guillain-Barre Syndrome:

1. Documentation that symptom onset is within two weeks or symptoms are severe (e.g. unable to

ambulate independently)

AND

2. Documented trial, failure, intolerance or contraindication to plasma exchange

Multifocal motor neuropathy:

1. Confirmed diagnosis: motor involvement of at least two nerves (for more than one month) without symptoms of sensory abnormalities

AND

2. Documentation of severe disease/disability

Multiple Sclerosis:

1. Documentation of relapsing/remitting disease

AND

2. Documented trial, failure, intolerance or contraindication to at least two conventional therapies (e.g., glatiramer, interferon beta, dimethyl fumarate)

Myasthenia Gravis:

Myasthenic exacerbation:

1. Evidence of myasthenic exacerbation, defined by at least one of the following symptoms in the last month:

a. Difficulty swallowing

b. Acute respiratory failure

c. Major functional disability responsible for the discontinuation of physical activity

Refractory disease:

1. Documentation that patient has severely impaired function due to myasthenia gravis

AND

2. Documented trial, failure, intolerance or contraindication to at least two of the following conventional therapies:

a. Acetylcholinesterase inhibitors (e.g., pyridostigmine)

b. Corticosteroids (e.g., prednisone, methylprednisolone)

c. Immunosuppressive agents (e.g., azathioprine, cyclosporine, mycophenolate)

d. Plasma exchange

Allogenic Bone Marrow Transplantation or Hematopoietic Stem Cell Transplant (HSCT) Recipients:

1. Documentation of one of the following:

a. Therapy is requested for use within 100 days after transplantation (transplantation date must be documented)

OR

b. Documentation that patient has an IgG less than 400 mg/dL with a history of recurrent infections

Autoimmune mucocutaneous blistering disease: pemphigus vulgaris, pemphigus foliaceus, bullous pemphigoid, mucous membrane (cicatricial) pemphigoid, epidermolysis bullosa acquisita, pemphigoid gestationis, linear IgA bullous dermatosis

1. Documentation of biopsy proven disease

AND

2. Documented trial, failure, intolerance or contraindication to systemic corticosteroids with concurrent immunosuppressive treatment (e.g., azathioprine, cyclophosphamide, mycophenolate mofetil).

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with an appropriate specialist (e.g., a Neurologist for multiple sclerosis or an immunologist, hematologist or infections disease expert for primary immunodeficiency)

COVERAGE DURATION

Generally, initial authorization is up to six months subject to criteria and reauthorization is up to one year subject to criteria.

OTHER CRITERIA

N/A

INCRELEX

MEDICATION(S)

INCRELEX

COVERED USES

N/A

EXCLUSION CRITERIA

Subjects with secondary forms of Insulin-like growth factor (IGF)-1 deficiency:

- GH deficiency
- Malnutrition
- Hypothyroidism
- Chronic treatment with pharmacologic doses of anti-inflammatory steroids

Concurrent use of growth hormone therapy

Malignant neoplasia

REQUIRED MEDICAL INFORMATION

For Severe primary IGF-1 deficiency:

1. Height standard deviation score of less than or equal to -3.0

AND

2. Basal insulin-like growth factor (IGF)-1 standard deviation score of less than or equal to -3.0

AND

3. Normal or elevated growth hormone (GH) levels.

AND

4. Documentation of open epiphyses by bone radiograph

For Growth hormone (GH) gene deletion

1. Documentation of open epiphyses by bone radiograph

AND

2. Patient has developed neutralizing antibodies to growth hormone

Reauthorization will require evidence that the medication remains effective, growth velocity is above 2.0 cm/year, evidence of open epiphyses, and documentation of expected adult height goal that is not yet obtained.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Initial authorization and reauthorization will be approved for one year.

OTHER CRITERIA

For Severe primary IGF-1 deficiency:

1. Height standard deviation score of less than or equal to -3.0

AND

2. Basal insulin-like growth factor (IGF)-1 standard deviation score of less than or equal to -3.0

AND

3. Normal or elevated growth hormone (GH) levels.

AND

4. Documentation of open epiphyses by bone radiograph

For Growth hormone (GH) gene deletion

1. Documentation of open epiphyses by bone radiograph

AND

2. Patient has developed neutralizing antibodies to growth hormone

Reauthorization will require evidence that the medication remains effective, growth velocity is above 2.0 cm/year, evidence of open epiphyses, and documentation of expected adult height goal that is not yet obtained.

INJECTABLE ANTI-CANCER MEDICATIONS

MEDICATION(S)

ABRAXANE, ACTIMMUNE, ADCETRIS, ALIQOPA, ALKERAN 50 MG VIAL, ARRANON, ARZERRA, ASPARLAS, AZACITIDINE, AZEDRA DOSIMETRIC, AZEDRA THERAPEUTIC, BAVENCIO, BELEODAQ, BELRAPZO, BENDAMUSTINE HCL, BENDEKA, BESPONSA, BLENREP, BLINCYTO, BORTEZOMIB, COSELA, CYRAMZA, DACOGEN, DANYELZA, DARZALEX, DARZALEX FASPRO, DECITABINE, EMLICITI, ENHERTU, ERBITUX, FASLODEX, FOLOTYN, FULVESTRANT, HALAVEN, IMFINZI, IMLYGIC, ISTODAX, IXEMPRA, JELMYTO, JEMPERLI, JEVTANA, KADCYLA, KANJINTI, KEYTRUDA, KYPROLIS, LIBTAYO, LUMOXITI, LUTATHERA, MARGENZA, MELPHALAN HCL, MONJUVI, MVASI, OGIVRI, ONIVYDE, ONTRUZANT, OPDIVO 100 MG/10 ML VIAL, OPDIVO 240 MG/24 ML VIAL, OPDIVO 40 MG/4 ML VIAL, PADCEV, PEPAXTO, PERJETA, PHESGO, POLIVY, PORTRAZZA, POTELIGEO, ROMIDEPSIN, RYBREVAANT, SARCLISA, SYLATRON, SYNRIBO, TECENTRIQ, TEMODAR 100 MG VIAL, TEMSIROLIMUS, TORISEL, TRAZIMERA, TREANDA, TRODELVY, VECTIBIX, VELCADE, VIDAZA, VYXEOS, XOFIGO, YERVOY, YONDELIS, ZALTRAP, ZEPZELCA, ZIRABEV, ZYNLONTA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For initial authorization:

1. Use must be for a FDA approved indication or indication supported by National Comprehensive Cancer Network guidelines with recommendation 2A or higher
2. For requests for trastuzumab or bevacizumab: Documented trial and failure, intolerance, or contraindication to the use of both of the preferred biosimilar medications, as follows:
 - a. Trastuzumab preferred products: Ogivri® (trastuzumab-dkst) and Kanjinti® (trastuzumab-anns)
 - b. Bevacizumab preferred products: Mvasi® (bevacizumab-bvzr) and Zirabev® (bevacizumab-awwb)

For patients established on therapy: documentation of adequate response to the medication must be provided.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by, or in consultation with an oncologist

COVERAGE DURATION

Authorization will be approved until no longer eligible with the plan, subject to formulary and/or benefit changes.

OTHER CRITERIA

N/A

INTERLEUKIN – 1 INHIBITORS (ARCALYST, ILARIS)

MEDICATION(S)

ARCALYST, ILARIS

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For Cryopyrin-Associated Periodic Syndrome (CAPS) including Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS) confirmed by:

1. Laboratory evidence of genetic mutation NLRP-3 (Nucleotide-binding domain, leucine rich family (NLR) pyrin domain containing 3) or CIAS1 (Cold-Induced Auto-inflammatory Syndrome-1),

AND

2. Classic symptoms associated with Familial Cold Auto-Inflammatory Syndrome (FCAS) or Muckle-Wells Syndrome (MWS) – recurrent intermittent fever and rash typically associated with natural or artificial cold

For Arcalyst® only:

For maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA):

1. Diagnosis of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) confirmed by laboratory evidence of genetic mutation in IL1RN (encodes for interleukin-1 receptor antagonist)

2. Current inflammatory remission of DIRA

3. Weight of at least 10 kg

For recurrent pericarditis:

1. Diagnosis of recurrent pericarditis (RP) confirmed by an acute episode of pericarditis followed by a 4-6 week symptom free period prior to the next episode without an identified cause

2. Documentation trial and failure, contraindication or intolerance to NSAIDs or glucocorticoids

For Ilaris® only:

For Familial Mediterranean Fever (FMF), and all the following:

1. Documented trial and failure, contraindication or intolerance to colchicine,

AND

2. Classic symptoms associated with FMF (febrile episodes, pain in the abdomen, chest, or arthritis of large joints).

Diagnosis of Hyperimmunoglobulin D (Hyper-IgD) Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD) confirmed by:

1. Laboratory evidence of genetic mutation MVK (mevalonate kinase),

AND

2. Classic symptoms associated with HIDs (abdominal pain, lymphadenopathy, aphthous ulcers).

Diagnosis of Tumor Necrosis Factor (TNF) receptor Associated Periodic Syndrome (TRAPS) confirmed by:

1. Laboratory evidence of genetic mutation TNFRSF1A (tumor necrosis factor receptor super family),

AND

2. Classic symptoms associated with TRAPs (abdominal pain, skin rash, musculoskeletal pain, eye manifestations).

Diagnosis of Active Still's Disease including Systemic Juvenile Idiopathic Arthritis (SJIA) and Adult-Onset Still's Disease:

1. Documentation of trial and failure, intolerance, or contraindication to at least one conventional therapy (e.g., methotrexate, leflunomide, hydroxychloroquine, sulfasalazine)

AND

2. Documentation of trial, failure, intolerance, or contraindication to both etanercept (Enbrel®) and adalimumab (Humira®)

Reauthorization: Documentation submitted of improvement of symptoms (such as fever, urticaria-like rash, arthralgia, myalgia, fatigue, and conjunctivitis for CAPS)

AGE RESTRICTION

Arcalyst®: may be covered for patients aged 12 years and older with CAPS (which includes FCAS, MWS), treatment of recurrent pericarditis (RP), and reduction in risk of recurrence of pericarditis.

Ilaris® may be covered for patients aged four years of age and older in patients with CAPS (which includes FCAS, MWS), Periodic Fever Syndromes including TRAPS, HIDS/MKD, and FMF

Ilaris® may be covered for patients aged two years of age and older in patients with Active Systemic Juvenile Idiopathic Arthritis and Adult Onset Still's Disease (AOSD)

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Initial authorization will be approved for six months. Reauthorization will be approved for one year.

OTHER CRITERIA

N/A

KRYSTEXXA - MEDICAL BENEFIT

MEDICATION(S)

KRYSTEXXA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For initial therapy, all of the following criteria must be met:

- 1) Diagnosis of chronic gout
- 2) Documentation of inadequate response to both of the following medications, unless contraindication to both of the following: xanthine oxidase inhibitor (e.g., allopurinol) and uricosuric (e.g., probenecid). Inadequate response is defined as inability to achieve uric acid levels of less than 6 mg/dL after at least three months of continuous therapy.
- 3) Documentation of symptomatic gout, as defined by one or more of the following, despite therapies outlined in criterion 2 above:
 - a) At least two gout flares per year
 - b) Non-resolving tophi

Reauthorization requires documentation of a decreased uric acid level from baseline

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with a rheumatologist.

COVERAGE DURATION

Initial authorization and reauthorization will be approved for six months.

OTHER CRITERIA

N/A

LEMTRADA - MEDICAL BENEFIT

MEDICATION(S)

LEMTRADA

COVERED USES

N/A

EXCLUSION CRITERIA

- In combination with other disease modifying therapy indicated for the treatment of MS
- For treatment beyond two years or beyond two treatment courses

REQUIRED MEDICAL INFORMATION

1. Documentation of confirmed diagnosis of relapsing form of multiple sclerosis or active secondary progressive disease

AND

2. Documentation of active disease (e.g. patients with frequent attacks or who are rapidly progressing in disability) after an adequate trial to ocrelizumab (Ocrevus®)

AND

3. Documentation of active disease after at least one additional of the following disease modifying therapies, unless all are contraindicated.

- a. Interferon-beta 1a (Avonex®, Rebif® or Plegridy®) or interferon-beta 1b (Betaseron®)
- b. Generic dimethyl fumarate
- c. Glatiramer acetate (Copaxone®)
- d. Natalizumab (Tysabri®)
- e. Teriflunomide (Aubagio®)
- f. Fingolimod (Gilenya®)
- g. Diroximel fumarate (Vumerity®)
- h. Ozanimod hydrochloride (Zeposia®)
- i. Siponimod (Mayzent®)

*Adequate trial is defined as at least six months of continuous therapy. Discontinuation of therapy due to drug intolerance will not be considered as failure to therapy.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Prescribed by, or in consultation with, a neurologist

COVERAGE DURATION

May be approved for up to two years, ensuring the cumulative duration of therapy does not exceed two years in a lifetime.

OTHER CRITERIA

N/A

LUXTURNA - MEDICAL BENEFIT

MEDICATION(S)

LUXTURNA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

All of the following must be met:

1. Confirmed biallelic RPE65 gene mutation, and
2. Has not previously had the intended treatment eye treated with gene therapy for retinal dystrophy RPE65 mutations, and
3. Documentation by an ophthalmologist within the previous six months of BOTH of the following:
 - a. Presence of sufficient viable retinal cells in the intended treatment eye as evidenced by an area of retina within the posterior pole of more than 100 micrometer thickness shown on optical coherence tomography, and
 - b. The member has remaining light perception in the intended treatment eye

AGE RESTRICTION

Approved for 12 months of age and older

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with an ophthalmologist from a certified Luxturna® administration site

COVERAGE DURATION

Authorization is limited to one treatment course per eye per lifetime. Approval duration will be for 12 weeks.

OTHER CRITERIA

N/A

MEDICAL NUTRITION - COMMERCIAL

MEDICATION(S)

RELIZORB

COVERED USES

N/A

EXCLUSION CRITERIA

- L-methylfolate (such as Deplin®) in the treatment of depression
- Non- elemental enteral nutrition (such as Boost® and Ensure®) is considered a benefit exclusion when given orally based on plan limitations
- Use of Relizorb™ is considered experimental and investigational for use with enteral feedings due to the lack of evidence to assess safety and efficacy on health outcomes

REQUIRED MEDICAL INFORMATION

For Enteral Nutrition Via Feeding Tube

1. Member has a feeding tube and nutrition will be administered via feeding tube (e.g., nasogastric [NG], nasojejunal [NJ], gastrostomy [PEG], jejunostomy [J-tube, PEG-J, PEJ]

OR

2. ALL of the following:

a. Established or anticipated inadequate oral intake for adults of at least 7 days. For children and infants the length of time of inadequate oral intake will be considered on a case-by-case basis.

AND

b. Documentation of ONE of the following

i. A medical condition that prevents food from reaching the digestive tract (e.g. head and neck cancer with reconstructive surgery, central nervous system disease that interferes with neuromuscular mechanisms of ingestion).

ii. Enteral nutrition comprises the sole source or is an essential source of nutrition (at least 75 percent of estimated basal caloric requirements) and is used as a therapeutic regimen to prevent serious disability in the patient.

iii. Recent unplanned weight loss of at least 10% in the past 3 months or less due to: increased metabolic need resulting from severe trauma, malabsorption difficulties due to underlying medical condition/disease (e.g. cystic fibrosis) or severe anorexia nervosa.

iv. Documentation of failure to thrive in patients under the age of 17.

AND

c. Adequate nutrition is not possible by dietary adjustment and/or oral supplementation.

Elemental Formula

FOR INBORN ERRORS OF METABOLISM

ALL of the following must be met:

1. Member has a confirmed in-born error of metabolism (including, but not limited to phenylketonuria [PKU], maternal phenylketonuria, maple syrup urine disease, citrullinemia, homocystinuria, histidinemia, tyrosinemia)

AND

2. Failure to use medical food will predictably result in adverse medical outcomes

AND

3. Treatment of the condition cannot be met through normal dietary supplementation or modification

Oral Administration

ALL of the following must be met:

1. Member has a diagnosis of severe intestinal malabsorption (such as eosinophilic gastrointestinal disorder, severe Crohn's disease, short bowel)

AND

2. Documentation that enteral formula is the sole or essential source of nutrition (at least 75 percent of estimated basal caloric requirements), and is used as a therapeutic regimen to prevent serious disability in the patient

AND

3. Treatment of the condition cannot be met through normal dietary supplementation or modification

AND

4. A physician has issued a written order for the formula

For patients with milk allergy, nutrition supplementation may be covered if the following criteria are met:

1. Documentation that the allergy is severe and causing symptoms, such as eosinophilic esophagitis, food protein-induced enterocolitis syndrome, failure to thrive, or anaphylaxis reactions

AND

2. Documentation that the requested nutritional support is the sole or essential source of nutrition (at least 75 percent of estimated basal caloric requirements)

AND

3. Therapy has been recommended by a pediatric specialist and/or dietician

REAUTHORIZATION:

Continued coverage requires documentation that the requested therapeutic regimen is medically necessary to prevent serious disability in the patient

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Initial authorization and reauthorization will be approved for up to one year.

For permanent or progressive conditions, authorization will be approved until no longer eligible with the plan, subject to formulary or benefit changes.

OTHER CRITERIA

N/A

MEDICALLY INFUSED THERAPEUTIC IMMUNOMODULATORS (TIMS)

MEDICATION(S)

ACTEMRA 200 MG/10 ML VIAL, ACTEMRA 400 MG/20 ML VIAL, ACTEMRA 80 MG/4 ML VIAL, AVSOLA, CIMZIA 200 MG VIAL KIT, ENTYVIO, ILUMYA, INFLECTRA, ORENCIA 250 MG VIAL, REMICADE, RENFLEXIS, SIMPONI ARIA, STELARA 130 MG/26 ML VIAL, STELARA 45 MG/0.5 ML VIAL

COVERED USES

N/A

EXCLUSION CRITERIA

Combination therapy with another therapeutic immunomodulator (TIM) agent or apremilast (Otezla®).

REQUIRED MEDICAL INFORMATION

1. For all requests, the patient must have an FDA labeled indication for the requested agent, or use to treat the indication is supported in drug compendia (i.e., American Hospital Formulary Service-Drug Information (AHFS-DI) or Truven Health Analytics' DRUGDEX® System.)

AND

2. The requested agent will not be given concurrently with another therapeutic immunomodulator (TIMs) agent or apremilast (Otezla®)

AND

3. One of the following:

a. For patients already established on the requested TIMs agent:

i. Documentation of response to therapy (e.g., slowing of disease progression or decrease in symptom severity and/or frequency)

ii. Note: Medications obtained as samples, coupons, or any other method of obtaining medications outside of an established health plan benefit are NOT considered established on therapy.

b. Patients not established on the requested TIMs agent must meet ALL of the following criteria:

i. Requests for infliximab products Remicade® and Avsola® will require documentation of failure, intolerance, or contraindication to the preferred infliximab products Inflectra® and Renflexis® in addition the indication-specific criteria below.

ii. For moderate to severe Ulcerative Colitis:

1. For non-preferred agents: documentation of trial, failure, intolerance, or contraindication to a preferred infliximab product (Inflectra® or Renflexis®) or vedolizumab (Entyvio®)

iii. For moderate to severe Crohn's Disease:

1. For non-preferred agents: documentation of trial, failure, intolerance, or contraindication to a preferred infliximab (Inflectra® or Renflexis®)

iv. For Rheumatoid Arthritis:

1. Documentation of trial and failure, intolerance, or contraindication to at least one oral disease modifying

- anti-rheumatic agent (DMARD) (e.g., methotrexate, leflunomide, hydroxychloroquine, sulfasalazine)
- 2.For non-preferred agents: documentation of trial, failure, intolerance, or contraindication to a preferred infliximab product (Inflectra® or Renflexis®)
- v.For moderate to severe Plaque Psoriasis:
- 1.Documentation of trial and failure, intolerance, or contraindication to at least one conventional therapy (e.g., methotrexate, tazarotene, topical corticosteroids, calcitriol)
- 2.For non-preferred agents: documentation of trial, failure, intolerance, or contraindication to a preferred infliximab product (Inflectra® or Renflexis®)
- vi.For Psoriatic Arthritis:
- 1.Documentation of trial and failure, intolerance, or contraindication to at least one conventional therapy (e.g., methotrexate, leflunomide, hydroxychloroquine, sulfasalazine)
- 2.For non-preferred agents: documentation of trial, failure, intolerance, or contraindication to a preferred infliximab product (Inflectra® or Renflexis®)
- vii.For Ankylosing Spondylitis:
- 1.For non-preferred agents: documentation of trial, failure, intolerance, or contraindication to a preferred infliximab product (Inflectra® or Renflexis®)
- viii.For giant cell arteritis:
- 1.Documentation of trial and failure, intolerance, or contraindication to at least one conventional therapy (e.g., systemic corticosteroid therapy)
- ix.For immune checkpoint inhibitor related diarrhea/colitis, infliximab (Inflectra® or Renflexis®) may be covered if the following criteria are met:
- 1.Documentation of severe diarrhea/colitis (G3-4)
- 2.Documentation of inadequate response to a 1-2 day trial of intravenous prednisone or methylprednisolone
- x.For systemic sclerosis (SSc-ILD): tocilizumab (Actemra®) may be covered if the following criteria are met:
- 1.Patient has interstitial lung disease, as evidence by high-resolution computed tomography (HRCT)

Note:

- Conventional therapy requirements may be waived if the patient has previously used another therapeutic immunomodulator agent OR apremilast (Otezla®) for the same indication.
- Conventional therapy and preferred agent requirements may be waived with clinically appropriate medical rationale

AGE RESTRICTION

Age must be appropriate based on FDA-approved indication

PRESCRIBER RESTRICTION

- Rheumatoid arthritis, ankylosing spondylitis: must be prescribed by, or in consultation with, a

rheumatologist

- Psoriasis: must be prescribed by, or in consultation with, a dermatologist
- Psoriatic arthritis: must be prescribed by, or in consultation with, a dermatologist or rheumatologist
- Inflammatory Bowel Disease: must be prescribed by, or in consultation with, a gastroenterologist
- Systemic sclerosis-associated interstitial lung disease: must be prescribed by, or in consultation with, a pulmonologist or rheumatologist
- Giant cell arteritis: must be prescribed by, or in consultation with, a rheumatologist or neurologist
- Immune checkpoint inhibitor related diarrhea/colitis: must be prescribed by, or in consultation with, an oncologist or gastroenterologist

COVERAGE DURATION

For immune checkpoint inhibitor related diarrhea/colitis: Authorization will be approved for three months

For all other indications: Authorization will be approved until no longer eligible with the plan, subject to formulary or benefit changes

OTHER CRITERIA

N/A

MIRCERA - MEDICAL BENEFIT

MEDICATION(S)

MIRCERA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For the treatment of adults with anemia associated with chronic kidney disease:

1. Documented Hemoglobin (HGB) levels of less than or equal to 10g/dl or hematocrit (HCT) levels of less than or equal to 30% within 30 days prior to initiation of therapy
2. Adequate iron stores as indicated by current (within the last 3 months) serum ferritin level greater than or equal to 100 mcg/L or serum transferrin saturation greater than or equal to 20%

For the treatment of pediatric patients 5 to 17 years of age who are converting from another erythropoiesis-stimulating agent (ESA) after their hemoglobin level was stabilized with an ESA:

1. Documented hemodialysis for at least 8 weeks
2. Documented stable maintenance treatment with epoetin alfa, epoetin beta, or darbepoetin alfa for at least 8 weeks prior to initiation of therapy
3. Documented stable hemoglobin (HGB) levels for at least 8 weeks prior to initiation of therapy.

Reauthorization:

1. Documentation of continued medical necessity (e.g., ongoing chronic kidney disease)
2. Documented HGB levels of less than or equal to 12g/dl or HCT levels of less than or equal to 36% within previous 30 days

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Initial authorization and reauthorization will be approved for one year.

OTHER CRITERIA

N/A

NPLATE - MEDICAL BENEFIT

MEDICATION(S)

NPLATE

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For immune thrombocytopenia,

Initial authorization all of the following must be met:

1. A diagnosis of immune thrombocytopenia (ITP), AND
2. Patient is at risk for bleeding with a platelet count of less than or equal to 30,000 cells per microliter, AND
3. Treatment by at least one of the following was ineffective or not tolerated:
 - a. Systemic corticosteroids, OR
 - b. Immune globulin, OR
 - c. Splenectomy

Reauthorization will require submission of platelet values demonstrating a response to therapy and a weekly dose below 10 microgram/kg.

For Hematopoietic Syndrome of Acute Radiation Syndrome [HSARS], all of the following must be met:

1. Documentation of acute exposure to radiation, AND
2. Documentation of myelosuppression defined as leukopenia, thrombocytopenia, or anemia

QUANTITY LIMITS:

Nplate is available as 125-, 250-, and 500- mcg single-dose vials of lyophilized powder. Quantity approved may be rounded down to nearest available vial size within 10% of calculated dose.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Prescribed by or in consultation with an oncologist, hematologist, or hepatologist.

COVERAGE DURATION

Initial authorization will be approved for up to three (3) months. Reauthorization will be approved for up to

six (6) months.

OTHER CRITERIA

N/A

OPHTHALMIC VEGF INHIBITORS: EYLEA, LUCENTIS, MACUGEN

MEDICATION(S)

BEOVU, EYLEA, LUCENTIS, MACUGEN

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

Must have one of the following diagnoses and meet any required criteria:

1. Neovascular (wet) age-related macular degeneration (AMD)

a. For ranibizumab (Lucentis®), aflibercept (Eylea®), brolucizumab (Beovu®)

i. Documentation that bevacizumab has been ineffective, not tolerated, or contraindicated (examples of contradictions include but are not limited to: serous pigmented epithelial detachment (PED), hemorrhagic PED, subretinal hemorrhage, or posterior uveal bleeding syndrome)

OR

ii. Rationale is provided why therapy with bevacizumab is not appropriate for the patient

a. For pegaptanib (Macugen®):

i. Documentation that bevacizumab and either ranibizumab (Lucentis®) or aflibercept (Eylea®) has been ineffective, not tolerated, or contraindicated

OR

ii. Rationale is provided why therapy is not appropriate for the patient

2. Diabetic macular edema or Diabetic retinopathy

a. For ranibizumab (Lucentis®) or aflibercept (Eylea®):

i. Documentation that bevacizumab has been ineffective, not tolerated, or contraindicated (examples of contradictions include but are not limited to: serous pigmented epithelial detachment (PED), hemorrhagic PED, subretinal hemorrhage, or posterior uveal bleeding syndrome)

OR

ii. Request is for aflibercept (Eylea®) and patients baseline visual acuity is 20/50 or worse

OR

iii. Rationale is provided why therapy with bevacizumab is not appropriate for member

3. Macular edema following retinal vein occlusion

a. For ranibizumab (Lucentis®) or aflibercept (Eylea®):

i. Documentation that bevacizumab has been ineffective, not tolerated, or contraindicated (examples of contradictions include but are not limited to: serous pigmented epithelial detachment (PED), hemorrhagic PED, subretinal hemorrhage, or posterior uveal bleeding syndrome)

OR

ii. Rationale is provided why therapy with bevacizumab is not appropriate for the patient

4. Myopic Choroidal Neovascularization (mCNV)

a. Request is for ranibizumab (Lucentis®)

Reauthorization or continuation of therapy:

Documentation of positive response to therapy (such as stabilization or improvement in vision)

QUANTITY LIMITS:

Approval may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines and are subject to medical claims audits

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed and administered by an ophthalmologist or retinal specialist

COVERAGE DURATION

Authorization will be approved until no longer eligible with the plan, subject to formulary or benefit changes

OTHER CRITERIA

N/A

OXLUMO

MEDICATION(S)

OXLUMO

COVERED USES

N/A

EXCLUSION CRITERIA

1. Patients with a history of liver transplant
2. Patients with an estimated glomerular filtration rate (eGFR) less than 30 mL/min/1.73m²

REQUIRED MEDICAL INFORMATION

Initial authorization for new starts:

1. Patient has a diagnosis of primary hyperoxaluria type 1 (PH1)
2. Diagnosis of PH1 has been confirmed by one of the following:
 - a. Genetic testing demonstrating mutation in the alanine: glyoxylate aminotransferase (AGXT) gene
 - b. Liver biopsy demonstrating significantly decreased or absent alanine: glyoxylate aminotransferase (AGT) enzyme activity
3. Documentation of one of the following:
 - a. Elevated urine oxalate (UOx) excretion as measured by body surface area-normalized daily UOx output greater than upper limit of normal (ULN)
 - b. Elevated UOx excretion as measured by UOx: creatinine ratio above age-specific upper limit of normal (ULN) OR
 - c. Elevated plasma oxalate (POx) concentration (POx concentration greater than ULN)
4. Documentation of a trial of high fluid intake of at least 3 liters per meter-squared of Body Surface Area (BSA) per day and that high fluid intake will continue with therapy
5. Concurrent use of pyridoxine or previous trial of at least 3 months with no significant improvement in urine oxalate concentration

Reauthorization or continuation of therapy:

1. Documentation of a clinically significant reduction in urine or plasma oxalate levels relative to pre-treatment baseline
2. Patient continues with concurrent high fluid intake (at least 3 liters per meter-squared BSA per day) and pyridoxine (unless individual is a pyridoxine non-responder)

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Initial authorization will be approved for 6 months. Reauthorization will be approved for 12 months

OTHER CRITERIA

N/A

PREVYMIS

MEDICATION(S)

PREVYMIS 240 MG/12 ML VIAL, PREVYMIS 480 MG/24 ML VIAL

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

ALL of the following must be met:

1. Member is within 100 days post-allogeneic transplant, and
2. Cytomegalovirus (CMV) Recipient positive, and
3. Member has ONE of the following:
 - a. Graft Versus Host Disease (GVHD) requiring greater than or equal to 1 mg/kg/day use of prednisone [or equivalent]
 - b. Receipt of lymphocyte depleting therapy (e.g. antithymocyte globulin [ATG], antithymocyte globulin equine [ATGAM], antithymocyte globulin rabbit [thymoglobulin], alemtuzumab, fludarabine) within the previous 6 months
 - c. Transplant was a cord blood allograft
 - d. History of CMV drug resistance within the past 6 months
4. If IV letermovir is being requested, rationale for not using oral formulation must be provided (e.g. patient is unable to swallow)

AGE RESTRICTION

Approved for 18 years and older.

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with a hematologist, oncologist, or Infectious Disease specialist.

COVERAGE DURATION

3 months, up to 100 days post-transplant

OTHER CRITERIA

N/A

PROPHYLACTIC HEREDITARY ANGIOEDEMA THERAPY

MEDICATION(S)

CINRYZE, HAEGARDA, TAKHZYRO

COVERED USES

N/A

EXCLUSION CRITERIA

Combination prophylaxis therapy with Cinryze®, Haegarda®, Takhzyro®, or Orladeyo®

REQUIRED MEDICAL INFORMATION

All of the following must be met:

1. Documentation of one of the following clinical criteria:

- a. Self-limiting, non-inflammatory subcutaneous angioedema without urticaria, recurrent, and lasting more than 12 hours, or
- b. Self-remitting abdominal pain without clear organic etiology, recurrent, and lasting more than six hours, or
- c. Recurrent laryngeal edema

AND

2. Documentation of greater than or equal to 2 HAE attacks per month on average for the past 3 months despite removal of triggers (e.g., estrogen containing oral contraceptive, angiotensin converting enzyme inhibitors) unless medically necessary

AND

3. One of the following:

a. For HAE Type I and Type II, documentation of at least two (2) complement studies taken at least one month apart with the patient in their basal condition and after the first year of life that show:

i. C4 is less than 50 percent of the lower limit of normal

AND

ii. one of the following:

a. C1-inhibitor (C1-INH) protein is less than 50 percent of the lower limit of normal, or

b. C1-INH function is less than 50 percent of the lower limit of normal

b. For HAE with normal C1-INH or HAE Type III:

i. Confirmed Factor 12 (FXII) ANGPT1, PLG, KNG1 gene mutation

OR

ii. Positive family history for HAE AND attacks lack response with high dose antihistamines or corticosteroids.

For coverage of Cinryze®:

Documentation of trial and failure or contraindication to Haegarda®.

REAUTHORIZATION:

Documentation must be provided showing benefit of therapy with reduction of frequency and severity of HAE attack episodes by greater than or equal to 50% from baseline.

QUANTITY LIMITS:

Cinryze®: 16 vials (500 units each vial) for 28 days

Haegarda®: Weight based 60 units/kg twice weekly for a 28-day supply (see appendix 2)

Takhzyro®: 2 vials (300 mg each vial) per 28-day supply

Orladeyo®: 30 capsules (150mg each) per 30-day supply

Dosing regimens beyond quantity limits will only be approved if evidence-based-rationale is provided.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with an immunologist or an allergist.

COVERAGE DURATION

Initial prior authorization will be approved for 3 months. Reauthorization may be approved for one year.

OTHER CRITERIA

N/A

PROVENGE - MEDICAL BENEFIT

MEDICATION(S)

PROVENGE

COVERED USES

N/A

EXCLUSION CRITERIA

- Concomitant use of chemotherapy, systemic steroid medications at greater than physiologic replacement doses and/or other systemic immunosuppressive agents to treat autoimmune disease or prevent allogeneic transplant rejection
- Presence of hepatic or other visceral metastases

REQUIRED MEDICAL INFORMATION

All of the following criteria must be met:

1. Asymptomatic or minimally symptomatic metastatic disease (e.g. no opioid use for malignant cancer pain)
2. Castrate-resistant or castration-recurrent prostate cancer, defined as both of the following:
 - a. Radiographic, clinical or biochemical [i.e., prostate-specific antigen (PSA)] progression despite therapy with androgen ablation therapy (e.g. orchiectomy, GnRH agonists/antagonists)

AND

- b. Testosterone level less than 50 ng/dL
3. Eastern Cooperative Oncology Group (ECOG) performance status of 0-1
 4. Life expectancy more than six (6) months

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by, or in consultation with, an oncologist.

COVERAGE DURATION

Authorization will be approved for 3 complete doses administered at approximately 2 week intervals (6 weeks) for one course of therapy per lifetime.

OTHER CRITERIA

N/A

PULMONARY ARTERIAL HYPERTENSION

MEDICATION(S)

EPOPROSTENOL SODIUM, FLOLAN, REMODULIN, REVATIO 10 MG/12.5 ML VIAL, SILDENAFIL 10 MG/12.5 ML VIAL, TREPROSTINIL, TYVASO, TYVASO INSTITUTIONAL START KIT, TYVASO REFILL KIT, TYVASO STARTER KIT, UPTRAVI 1,800 MCG VIAL, VELETRI

COVERED USES

N/A

EXCLUSION CRITERIA

- Heart failure caused by reduced left ventricular ejection fraction for epoprostenol (Flolan®, Veletri®)
- Idiopathic interstitial pneumonia for riociguat (Adempas®) only

REQUIRED MEDICAL INFORMATION

For initial authorization the following criteria must be documented:

1. Diagnosis of Pulmonary Hypertension (PH) confirmed by right heart catheterization as defined by:

i. Mean pulmonary artery pressure (mPAP) greater than or equal to 25 mmHg at rest

AND

ii. Pulmonary capillary wedge pressure (PCWP) or left ventricular end diastolic pressure (LVEDP) less than or equal to 15 mmHg

AND

iii. Pulmonary vascular resistance (PVR) greater than 3 Wood units (WU)

AND

2. Patient has one of the following:

i. World Health Organization (WHO) Group 1 classification PAH with WHO/New York Heart Association (NYHA) functional class as outlined below:

a. Flolan®, Veletri®, and Ventavis: Class III or IV

b. Tyvaso®: Class III or IV

c. All other therapies: Class II, III, or IV

ii. For Adempas® only, WHO Group 4 classification CTEPH with WHO/New York Heart Association (NYHA) functional class II, III, or IV

iii. For Tyvaso® only, WHO Group 3 classification PH-ILD

AND

3. For sildenafil citrate oral suspension or parenteral injection (Revatio®): Documentation of intolerance or allergy to excipient ingredients of all available generic sildenafil citrate tablets (Revatio®) or other medical rationale provided for use of oral suspension/parenteral injection over tablets.

Reauthorization: Documentation of response to therapy such as lack of disease progression, improvement

in WHO functional class

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Prescribed by or in consultation with a pulmonologist or cardiologist

COVERAGE DURATION

Initial authorization for 12 months. Reauthorization may be reviewed annually to assess continued medical necessity and effectiveness of medication.

OTHER CRITERIA

N/A

RADICAVA - MEDICAL BENEFIT

MEDICATION(S)

RADICAVA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

1. For initiation of therapy, all of the following criteria must be met:
 - a. Documentation of definite or probable amyotrophic lateral sclerosis (ALS) within the previous two years per the El Escorial (Airlie House) Criteria
 - b. Documentation of baseline ALS Functional Rating Scale-Revised (ALSFRS-R) with at least 2 points in each individual item
 - c. Forced vital capacity (FVC) of at least 80% (taken within the past three months)
 - d. Dosing is in accordance with the FDA approved labeling
2. For patients established on therapy:
 - a. Documentation of a clinical benefit from therapy such as slowing of disease progression or stabilization of functional ability and maintenance of activities of daily living (ADLs)
 - b. Documentation that patient is not dependent on invasive ventilation or tracheostomy
 - c. Dosing is in accordance with the FDA approved labeling

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Prescribed by, or in consultation with, a neurologist with expertise in ALS.

COVERAGE DURATION

Initial authorization and reauthorization will be approved for six months.

OTHER CRITERIA

N/A

REBLOZYL

MEDICATION(S)

REBLOZYL

COVERED USES

N/A

EXCLUSION CRITERIA

1. Evidence of active pregnancy
2. History of thrombosis

REQUIRED MEDICAL INFORMATION

For initial authorization for beta-thalassemia, all of the following must be met:

1. Diagnosis of beta-thalassemia, which can be confirmed by one of the following:
 - a. Hemoglobin analysis or genetic testing
 - b. Complete blood count that showed reduced Hgb level (less than seven (7) g/dL), mean corpuscular volume (MCV) between 50 and 70 fL, and mean corpuscular hemoglobin (MCH) between 12 and 20 pg
 - c. Peripheral blood smear results that show red blood cell (RBC) morphologic changes including microcytosis, hypochromia, anisocytosis, poikilocytosis and nucleated RBC
2. Documentation that patient is transfusion-dependent, defined as receiving at least 6-20 units RBC transfusions every 24 weeks
3. Documented baseline Hgb level of at least 9 g/dL, drawn within the previous 30 days

For continuation of therapy for beta-thalassemia beyond 9 weeks, ongoing documentation of patient response to therapy must include maintenance of reduced transfusion levels

For initial authorization for myelodysplastic syndrome (MDS), all of the following must be met:

1. Diagnosis of MDS with ring sideroblasts (MDS-RS), or myelodysplastic/myeloproliferative neoplasm with ring sideroblasts and thrombocytosis (MDS/MPN-RS-T)
2. Score of very low to intermediate risk based on the Revised International Prognostic Scoring System
3. Documentation that patient requires RBC transfusions of at least two (2) units every eight (8) weeks
4. One of the following:
 - a. Documented trial and failure [of at least two (2) months], intolerance, or contraindication to erythropoiesis-stimulating agents (i.e., erythropoietin or darbepoetin)
 - b. Documentation of endogenous erythropoietin level greater than 500 mU/mL

For reauthorization for MDS-RS: Documentation that patient was able to achieve transfusion independence

for at least eight weeks during previous treatment period

AGE RESTRICTION

At least 18 years of age

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with a hematologist

COVERAGE DURATION

Beta-thalassemia: Initial authorization will be for 9 weeks. Reauthorization will be for one (1) year.

MDS-RS: Initial authorization will be for 6 months. Reauthorization will be for one (1) year

OTHER CRITERIA

N/A

RITUXIMAB

MEDICATION(S)

RIABNI, RUXIENCE, TRUXIMA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For all indications for non-preferred rituximab products: Documented trial and failure, intolerance, or contraindication to the use of both of the preferred biosimilar medications: Ruxience® (rituximab-pvvr) and Truxima® (rituximab-abbs)

For Oncologic diagnoses:

For initial authorization: use must be for a FDA approved indication or indication supported by National Comprehensive Cancer Network guidelines with recommendation 2A or higher

For Rheumatoid Arthritis:

1. Documentation of trial, failure, intolerance, or contraindication to at least one of the following targeted immune modulators: etanercept (Enbrel®), adalimumab (Humira®), a preferred infliximab product or intravenous golimumab (Simponi® Aria).

AND

2. Documentation that rituximab will be used concurrently with methotrexate. If intolerance or contraindication to methotrexate, then in combination with another disease-modifying antirheumatic drug (DMARD) (e.g., leflunomide, sulfasalazine, hydroxychloroquine), unless medical rationale is provided to support monotherapy.

For Vasculitis – including antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis [e.g., Granulomatosis with Polyangiitis (GPA) and Microscopic Polyangiitis (MPA)] and refractory polyarteritis nodosa (resistant to cyclophosphamide):

1. Documentation that rituximab will be given in combination with glucocorticoids

AND

2. Documentation of severe disease (e.g., critical organ system involvement)

For Immune Thrombocytopenia (ITP):

1. Documentation of trial, failure, intolerance, or contraindication to systemic corticosteroid therapy

AND

2. Documentation of active bleeding, or high-risk of bleeding, or a platelet count less than 30,000 cells per microliter

For Relapsing and Remitting Multiple Sclerosis (RRMS):

1. One of the following:

a. Documentation of trial, failure, or intolerance to at least two disease modifying therapies indicated for RRMS

OR

b. Documentation that patient has highly active or aggressive disease

For Refractory Myasthenia Gravis:

1. Documentation that patient has severely impaired function due to myasthenia gravis

AND

2. Documented trial, failure, intolerance or contraindication to at least two of the following conventional therapies:

a. Acetylcholinesterase inhibitors (e.g., pyridostigmine)

b. Corticosteroids (e.g., prednisone, methylprednisolone)

c. Immunosuppressive agents (e.g., azathioprine, cyclosporine, mycophenolate)

d. Plasma exchange

For Autoimmune Hemolytic Anemia (AIHA):

1. In patients diagnosed with warm AIHA

a. Documentation of trial, failure, intolerance, or contraindication to glucocorticoids

OR

2. In patients diagnosed with cold AIHA or cold agglutinin disease

For Reauthorization: Documentation of adequate response to the medication must be provided.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by, or in consultation with, an oncologist, rheumatologist, neurologist (in the case of RRMS, NMO), dermatologist (in the case of PV), or nephrologist (in the case of renal disease).

COVERAGE DURATION

Initial authorization will be approved for six months and reauthorization will be approved for one year.

OTHER CRITERIA

N/A

SCENESSE

MEDICATION(S)

SCENESSE

COVERED USES

N/A

EXCLUSION CRITERIA

1. Current Bowen's disease, basal cell carcinoma, or squamous cell carcinoma
2. Personal history of melanoma or dysplastic nevus syndrome
3. Erythropoietic protoporphyria (EPP) or X-linked protoporphyria (XLP) with significant hepatic involvement

REQUIRED MEDICAL INFORMATION

For initial authorization:

1. Confirmed diagnosis of erythropoietic protoporphyria (EPP) or X-linked protoporphyria (XLP) by one of the following:
 - a. Gene sequencing showing an FECH, CLPX, or ALAS2 mutation
 - b. Elevated total erythrocyte protoporphyrin greater than 80 mcg/dL
 - c. Erythrocyte fractionation shows more than 50% metal-free vs. zinc protoporphyrin
2. Documentation of characteristic symptoms of EPP/XLP phototoxicity (e.g., intolerance to light with symptoms including itching, burning, pain, erythema, or scarring of the skin on contact with sunlight)
3. Documentation that sun avoidance and use of sunscreen and protective clothing have proven inadequate in controlling EPP/ XLP -associated painful skin reactions
4. Documentation that the condition is having a significant impact on quality of life (QOL)

For reauthorization:

1. Documentation of a positive response to therapy by one of the following:
 - a. Decreased severity and number of phototoxic reactions
 - b. Increased duration of sun exposure
 - c. Increased quality of life
2. For request of more than three implants per year: medical justification must be provided addressing why member needs coverage for more than six months out of the year (afamelanotide is typically given during periods of high sunlight exposure, e.g. from spring to autumn)

AGE RESTRICTION

Approved for 18 years of age or older

PRESCRIBER RESTRICTION

Must be prescribed by, or in consultation with a dermatologist or porphyria specialist

COVERAGE DURATION

Initial and reauthorization will be approved for six months for three implants (Medical justification is required for requests beyond three implants for seasonal coverage)

OTHER CRITERIA

N/A

SIGNIFOR LAR - MEDICAL BENEFIT

MEDICATION(S)

SIGNIFOR LAR

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

Treatment of patients with acromegaly:

1. Documentation that the patient has persistent disease (e.g., biochemical or clinical) following surgical resection or patient is ineligible for surgery

AND

2. Documentation of trial and failure, intolerance or contraindication to octreotide injection therapy or lanreotide subcutaneous depot

Note: Mild symptoms of disease are typically treated with a dopamine agonist (e.g., cabergoline)

Patients with Cushing's disease:

1. Diagnosis of endogenous Cushing's Disease

AND

2. Documentation of one of the following:

a. Patient has failed pituitary surgery or

b. Patient is not a candidate for surgery

Reauthorization:

Acromegaly: documentation of response to therapy, as defined as normalization of insulin-like growth factor (IGF)-1 and reduction of symptoms

Cushing's disease: documentation of positive clinical response to therapy (e.g., a clinically meaningful reduction in 24-hour urinary free cortisol levels, improvement in signs or symptoms of the disease)

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with an endocrinologist

COVERAGE DURATION

Initial authorization and reauthorization will be approved for one year.

OTHER CRITERIA

N/A

SOLIRIS - MEDICAL BENEFIT

MEDICATION(S)

SOLIRIS

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For Paroxysmal Nocturnal Hemoglobinuria (PNH), all of the following must be met:

1. Documented, confirmed diagnosis of paroxysmal nocturnal hemoglobinuria (PNH) by Flow Cytometric Immunophenotyping (FCMI) using at least two independent flow cytometry reagents on at least two cell lineages (e.g., RBCs and WBCs) demonstrating that the patient's peripheral blood cells are deficient in glychophosphatidylinositol (GPI)-linked proteins (which may include CD59, CD55, CD14, CD15, CD16, CD24, CD45, and CD64)

AND

2. Severe disease as indicated by at least one of the following (a or b):

a. Documented history of thrombosis, OR

b. Documentation of at least 10% PNH type III red cells AND at least one (1) of the following:

i. Transfusion dependence (eg. hemoglobin less than 7 g/dL or symptomatic anemia with hemoglobin less than 9 g/dL)

ii. Disabling fatigue

iii. End-organ complications

iv. Frequent pain paroxysms (eg. dysphagia or abdominal pain)

v. Lactate dehydrogenase (LDH) levels greater than or equal to 1.5 times the upper limit of normal

AND

3. Dose and frequency is in accordance with FDA-approved labeling

Reauthorization for PNH:

1. Documentation of reduced LDH levels, reduced transfusion requirements, or improvement in PNH related symptoms

2. Dose and frequency is in accordance with FDA-approved labeling

For Complement-Mediated Hemolytic Uremic Syndrome (HUS), all of the following must be met:

1. Diagnosis of non-infectious HUS (i.e. HUS is not due to infection with Shiga toxin-producing Escherichia coli)

AND

2. Clinical presentation that includes: microangiopathic hemolytic anemia (hemoglobin less than 10 g/dL), thrombocytopenia (platelets less than 150), and acute kidney injury (elevations in serum creatinine)

AND

3. Dose and frequency is in accordance with FDA-approved labeling

Reauthorization for HUS:

1. Documentation of improvement in at least two thrombotic microangiopathy endpoints, such as:

- Maintenance of platelet counts (i.e. improvements or reductions less than 25%)
- Reductions in LDH
- Reduction in number of needed plasmaphoresis or plasma infusion events
- Improvement in kidney function and reduction of dialysis

2. Dose and frequency is in accordance with FDA-approved labeling

For Generalized Myasthenia Gravis (gMG), all of the following must be met:

1. Anti-acetylcholine receptor (anti-AChR) antibody positive

AND

2. Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV

AND

3. Myasthenia Gravis -Activities of Daily Living (MG-ADL) total score greater than five

AND

4. Failed treatment for at least one year with the following:

A. At least TWO immunosuppressive therapies ([ISTs] such as azathioprine, mycophenolate mofetil, cyclosporine and tacrolimus, corticosteroids)

OR

B. ONE immunosuppressive therapy and required at least four infusions/ year of either intravenous immunoglobulin (IVIg) OR plasma exchange (PE)

AND

5. Dose and frequency is in accordance with FDA-approved labeling

Reauthorization for Myasthenia Gravis (MG):

1. Initial reauthorization may require documentation of improvement in MG-ADL by at least two points from baseline.

2. Dose and frequency is in accordance with FDA-approved labeling

For Neuromyelitis Optica Spectrum Disorder (NMOSD), all of the following must be met:

1. Diagnosis of neuromyelitis optica spectrum disorder as defined as the following:

A. Presence of at least one core clinical characteristic (optic neuritis, acute myelitis, area postrema syndrome, acute brainstem syndrome, symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions, symptomatic cerebral syndrome with NMOSD-typical brain

lesions)

AND

B. Anti-AQP4 antibody positive

AND

2. Documentation that other alternative diagnoses have been excluded (i.e. Multiple Sclerosis)

AND

3. Trial and failure, intolerance or contraindication to rituximab

AND

4. Medication will not be used in combination with complement inhibitor (e.g.ravulizumab-cwvz), anti-CD20-directed (e.g., rituximab), anti-CD19 directed (e.g., inebilizumab) or IL-6 inhibition pathway therapies (e.g., satralizumab)

AND

5. Dose and frequency is in accordance with FDA-approved labeling

Reauthorization for Neuromyelitis Optica Spectrums Disorder (NMOSD):

1. Documentation of positive clinical response to therapy

2. Medication will not be used in combination with complement inhibitor (e.g.ravulizumab-cwvz), anti-CD20-directed (e.g., rituximab), anti-CD19 directed (e.g., inebilizumab) or IL-6 inhibition pathway therapies (e.g., satralizumab)

3. Dose and frequency is in accordance with FDA-approved labeling

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

PNH or aHUS: Prescribed by an hematologist/oncologist or nephrologist

MG or NMOSD: Prescribed by a neurologist

COVERAGE DURATION

Initial authorization for up to three months and reauthorization will be approved for up to one year.

OTHER CRITERIA

N/A

SOMATOSTATIN ANALOGS

MEDICATION(S)

SANDOSTATIN LAR DEPOT, SOMATULINE DEPOT

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

I. Acromegaly:

a. Initial authorization requires all of the following:

- i. Confirmed diagnosis of acromegaly,
- ii. Documentation of an inadequate response to surgery or pituitary irradiation, or patient is not a candidate for surgical resection and pituitary irradiation,
- iii. Documentation of good response and tolerability to short-acting octreotide

b. Reauthorization requires documentation of a positive clinical response to therapy (e.g., reduction or normalization of IGF-1/GH level for same age and sex, reduction in tumor size)

II. Carcinoid Tumors, for Symptomatic Treatment of Diarrhea or Flushing:

a. Initial authorization requires all of the following:

- i. Documentation that patient has severe diarrhea or flushing caused by a carcinoid tumor
- ii. Documentation of good response and tolerability to short-acting octreotide

b. Reauthorization requires documentation of an improvement in the number of diarrhea and flushing episodes

III. Vasoactive Intestinal Peptide Tumors, for Symptomatic Treatment of Diarrhea:

a. Initial authorization requires all of the following:

- i. Documentation that patient has severe diarrhea caused by vasoactive intestinal peptide tumors
- ii. Documentation of good response and tolerability to short-acting octreotide

b. Reauthorization requires documentation of an improvement in the number of diarrhea episodes

IV. Chemotherapy induced diarrhea (Sandostatin LAR® only):

a. Initial authorization requires all of the following:

- i. Documentation that patient has severe diarrhea caused by chemotherapy
- ii. Documentation of an inadequate response or contraindication to loperamide
- iii. Documentation of good response and tolerability to short-acting octreotide

b. Reauthorization requires documentation of an improvement in the number of diarrhea episodes

V. For AIDS-related diarrhea (Sandostatin LAR® only):

a. Initial authorization requires all of the following:

- i. Documentation that patient has severe diarrhea
 - ii. Documentation of an inadequate response or contraindication to loperamide and diphenoxylate (Lomotil®)
 - iii. Documentation of good response and tolerability to short-acting octreotide
- b. Reauthorization requires documentation of an improvement in the number of diarrhea episodes
- VI. For oncologic diagnoses:
- a. Initial authorization: use must be for a FDA approved indication or indication supported by National Comprehensive Cancer Network guidelines with recommendation 2A or higher

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Initial authorization and reauthorization will be approved for one year

OTHER CRITERIA

N/A

SPINRAZA - MEDICAL BENEFIT

MEDICATION(S)

SPINRAZA

COVERED USES

N/A

EXCLUSION CRITERIA

1. Concomitant use with, or following, gene therapy for SMA (e.g., onasemnogene abeparvovec)
2. Use in combination with risdiplam (Evrysdi®)
3. Advanced symptoms of SMA (e.g., complete paralysis of limbs, tracheostomy or ongoing invasive ventilator support in the absence of an acute reversible illness)

REQUIRED MEDICAL INFORMATION

For initial authorization, all of the following criteria must be met:

1. Confirmed genetic diagnosis of Spinal Muscular Atrophy (SMA) with documentation of bi-allelic mutations in the survival motor neuron 1 (SMN1) gene and less than or equal to three copies of SMN2, AND
2. Documentation that patient is presymptomatic or has symptoms with an onset at age less than 30 years, AND
3. Documentation of baseline motor function, with one of the following standardized test appropriate based on the patient's age and level of function:
 - a. CHOP-INTEND: Children's hospital of Philadelphia Infant Test of Neuromuscular Disorders
 - b. HINE: Hammersmith Infant Neurological Examination
 - c. HFSME: Hammersmith Functional Motor Scale Expanded
 - d. 6MWT: six-minute walk test
 - e. RULM: Revised Upper Limb Module

NOTE the following guidance on selecting an appropriate test:

- Non-sitters (infants and kids): CHOP-INTEND, HINE (may need HFSME as they transition to sitting).
- Sitters: HFSME, RULM
- Walkers (kids): 6MWT, HFSME
- Walkers (adults): 6MWT, RULM
- Non-walkers (adults): RULM

For reauthorization: Improvement or maintenance of motor function, evidenced by stabilization or improvement in motor function test scores performed at baseline

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by, or in consultation with, a neurologist

COVERAGE DURATION

Initial authorization and reauthorization will be approved for one year

OTHER CRITERIA

N/A

SPRAVATO

MEDICATION(S)

SPRAVATO

COVERED USES

N/A

EXCLUSION CRITERIA

- Concomitant use with another dissociative agent
- Aneurysmal vascular disease (including thoracic and abdominal aorta, intracranial, and peripheral arterial vessels) or arteriovenous malformation
- History of intracerebral hemorrhage
- Current or prior DSM-5 diagnosis of a psychotic disorder or MDD with psychosis, bipolar or related disorders, comorbid obsessive compulsive disorder, intellectual disability, autism spectrum disorder, borderline personality disorder, antisocial personality disorder, histrionic personality disorder, or narcissistic personality disorder
- Current or recent history (i.e. within the last six months) of moderate or severe substance or alcohol use disorder

REQUIRED MEDICAL INFORMATION

For initial authorization for treatment-resistant depression (TRD), all of the following criteria must be met:

1. Individual has been diagnosed with treatment-resistant depression (TRD) by a psychiatrist within the previous three months. Clinical documentation must be provided that outlines the patient evaluation, plan for on-going management, and treatment options reviewed.
2. Baseline score from one of the following standardized depression rating scales confirming severe depression:
 - a. Patient Health Questionnaire-9 (PHQ-9) score of at least 20
 - b. Hamilton Depression Scale (HAM-D17) score of at least 24
 - c. Quick Inventory of Depressive Symptomatology, Clinician-Rated (QIDS-C16) score of at least 16
 - d. Montgomery Asberg Depression Rating Scale (MADRS) total score of at least 28
3. Individual has tried and failed three oral antidepressants in at least two different therapeutic classes for at least eight weeks of treatment at the highest tolerable dose or the FDA-approved maximum dose for the medication. Trials should have occurred within the previous two years
4. Individual has tried and failed augmentation therapy (i.e., two antidepressants with different mechanisms of action used concomitantly or an antidepressant and a second-generation antipsychotic, lithium, thyroid hormone, or anticonvulsant used concomitantly). Trial should have occurred within the previous two years
5. Documentation that esketamine (Spravato®) will be used in combination with oral antidepressant therapy

6. Dosing is in accordance with the United States Food and Drug Administration approved labeling

For reauthorization, all of the following criteria must be met:

1. Documentation of clinical improvement in depression symptoms as measured by a clinically significant decrease in baseline depression rating scores
2. Documentation of on-going management with a psychiatrist at minimum of every three months
3. Documentation that esketamine (Spravato®) will continue to be used in combination with oral antidepressant therapy
4. Dosing is in accordance with the United States Food and Drug Administration approved labeling

For initial authorization for depressive symptoms in adults with major depressive disorder (MDD) with acute suicidal ideation or behavior all of the following criteria must be met:

1. Individual has been diagnosed with depressive symptoms in adults with major depressive disorder (MDD) with acute suicidal ideation or behavior by a psychiatrist
2. Baseline score from one of the following standardized depression rating scales confirming severe depression:
 - a. PHQ-9 score of at least 20
 - b. MADRS total score of at least 28
 - c. HAMD 17 score of at least 24
 - d. QIDS-C 16 score of at least 16
3. Individual received standard of care treatment including one of the following:
 - a. Initiation of an antidepressant, or
 - b. Optimized oral antidepressant, or
 - c. Added augmentation therapy to current antidepressant
4. Dosing is in accordance with the United States Food and Drug Administration approved labeling

For continuation of care post initiation in inpatient setting all of the following criteria must be met:

1. Documentation of the number of doses provided in the inpatient setting
2. Documentation of clinical improvement in depression symptoms as measured by a clinically significant decrease in baseline depression rating scores
3. Documentation that esketamine (Spravato®) will continue to be used in combination with oral antidepressant therapy
4. Dosing is in accordance with the United States Food and Drug Administration approved labeling

AGE RESTRICTION

Approved for 18 years and older

PRESCRIBER RESTRICTION

Prescribed by, or in consultation with, a psychiatrist. The administration/monitoring of this product may be completed by any mental health provider.

COVERAGE DURATION

For treatment resistant depression: Initial authorization will be approved for three months. Reauthorization will be approved for six months

For Depressive symptoms in adults with major depressive disorder (MDD) with acute suicidal ideation or behavior: Initial authorization will be approved for one month or the remainder of weeks to one month of treatment post inpatient initiation. Reauthorization will only be approved for treatment resistant depression criteria.

OTHER CRITERIA

N/A

SYLVANT - MEDICAL BENEFIT

MEDICATION(S)

SYLVANT

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

Initial Authorization:

1. Confirmed diagnosis of Multicentric Castleman Disease (MCD)
- AND
2. Documentation of negative human immunodeficiency virus (HIV) status
- AND
3. Documentation of negative human herpes-virus 8 (HHV-8) status

Reauthorization will require positive response to therapy as well as documentation that patient remains HIV and HHV-8 negative

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with an oncologist, hematologist, or rheumatologist.

COVERAGE DURATION

Initial authorization and reauthorization will be approved for 1 year.

OTHER CRITERIA

N/A

TEPEZZA

MEDICATION(S)

TEPEZZA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

All of the following criteria must be met:

1. Confirmed diagnosis of moderate-to-severe thyroid eye disease/Grave's Orbitopathy, as defined by one (1) of the following:
 - a. Sight-threatening disease (e.g., dysthyroid optic neuropathy, corneal breakdown)
 - b. Eye disease significantly impacts quality of life and at least two (2) of the following:
 - i. Lid retraction of at least 2 mm, marginal reflex distance-1 (MRD1) greater than four (4), or presence of lagophthalmos
 - ii. Moderate or severe soft-tissue involvement (e.g. swelling or redness of the eyes)
 - iii. Inconstant diplopia (i.e., diplopia at extremes of gaze) or constant diplopia (i.e., continuous diplopia in primary or reading position)
2. Documentation of active disease, defined as a Clinical Activity Score of at least three (3)
3. Laboratory evidence of euthyroid state
4. Inadequate response to at least two (2) weeks of therapy with high-dose intravenous (IV) glucocorticoid therapy (equivalent to methylprednisolone 0.5 g once weekly) or inability to use this therapy (e.g., evidence of recent viral hepatitis, significant hepatic dysfunction, severe cardiovascular morbidity or psychiatric disorders) or documentation that patient has sight threatening disease (defined as presence of direct optic neuropathy or corneal breakdown)

Reauthorization is not considered medically necessary and will not be covered.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by, or in consultation with, an ophthalmologist

COVERAGE DURATION

Authorization will be approved for six (6) months for a total of eight (8) infusions

OTHER CRITERIA

N/A

TESTOSTERONE REPLACEMENT THERAPY (TRT)

MEDICATION(S)

AVEED, TESTOPEL, TESTOSTERONE 50 MG PELLETT

COVERED USES

N/A

EXCLUSION CRITERIA

Use for improvement of sexual signs and symptoms (e.g., decreased libido, sexual dysfunction)

REQUIRED MEDICAL INFORMATION

1. One of the following:

a. Diagnosis of gender dysphoria or gender identity disorder

OR

b. Diagnosis of primary or secondary (hypogonadatropic) hypogonadism

2. Documented trial and failure (defined as inability to reach therapeutic levels or fluctuations in levels resulting in symptoms) of the following:

a. For topical products: Generic topical testosterone 1%

b. For injectable products: Generic injectable testosterone cypionate

c. For all other products including pellets, oral and nasal products: Both generic topical testosterone 1% and generic injectable testosterone cypionate

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Authorization will be approved until no longer eligible with the plan, subject to formulary or benefit changes

OTHER CRITERIA

N/A

TOTAL PARENTERAL NUTRITION (TPN)

MEDICATION(S)

AMINOSYN, AMINOSYN II, AMINOSYN M, AMINOSYN 7%-ELECTROLYTE SOL, AMINOSYN-HBC, AMINOSYN-PF, AMINOSYN-RF, CLINIMIX, CLINIMIX E, CLINIMIX N14G30E, CLINISOL, CLINOLIPID, FREAMINE HBC, FREAMINE III, HEPATAMINE, INTRALIPID, NEPHRAMINE, NUTRILIPID, OMEGAVEN, PLENAMINE, PREMASOL, PROCALAMINE, PROSOL, SMOFLIPID, SYNTHAMIN 17 WITHOUT ELTYE, TRAVASOL, TROPHAMINE

COVERED USES

N/A

EXCLUSION CRITERIA

Coverage for intradialytic parenteral nutrition (IDPN) when offered in addition to regularly scheduled TPN infusions

REQUIRED MEDICAL INFORMATION

One of the following criteria must be met:

1. Member has a central or peripheral line and nutrition will be administered via this line.

OR

2. Documentation of a failure to enteral nutrition (either oral or via tube), defined as either a or b:

a. A documented loss of at least 10% of body weight over a three-month period

b. Member is unable to reach nutritional needs from combined oral and enteral intake (less than 75 percent of estimated basal caloric requirements)

OR

3. Evidence of structural or functional bowel disease (e.g. massive small bowel resection, short bowel syndrome) that makes oral and tube feedings not possible

OR

4. A condition in which it is necessary for the gastrointestinal tract to be totally non-functioning for a period of time (i.e. bowel rest)

Medically necessary intradialytic parenteral nutrition (IDPN) may be covered for members on chronic dialysis who meet criteria 2, 3 or 4 AND cannot tolerate daily TPN.

For continued coverage, annual assessment that documents the ongoing medical necessity of PN as per the above criteria will be required.

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

Initial authorization will be approved for 3 months and reauthorization will be approved for up to one year

OTHER CRITERIA

N/A

TRANSTHYRETIN (TTR) LOWERING AGENTS

MEDICATION(S)

ONPATTRO

COVERED USES

N/A

EXCLUSION CRITERIA

- New York Heart Association (NYHA) Heart Functional class III or IV
- Hereditary transthyretin-mediated amyloidosis with cardiomyopathy
- Others forms of amyloidosis that is not due to a genetic mutation in the TTR gene
- Patients without the presence of polyneuropathy symptoms associated with hATTR amyloidosis
- Patients with type I or type II diabetes
- Previous organ transplant(s) requiring immunosuppression
- Malignancy within the past five years
- Uncontrolled cardiac arrhythmia or unstable angina

REQUIRED MEDICAL INFORMATION

1. Diagnosis of hereditary transthyretin-mediated amyloidosis (hATTR) with polyneuropathy

AND

2. Documentation of a pathogenic TTR mutation

AND

3. Patient has a baseline polyneuropathy disability (PND) score of less than or equal to IIIB OR has a baseline familial amyloid polyneuropathy (FAP) stage of I or II

AND

4. Baseline neuropathy impairment score (NIS) between 5 and 130

AND

5. Baseline Norfolk Quality of Life-Diabetic Neuropathy Questionnaire (Norfolk-QOL-DN) score

AND

6. Demonstrate symptoms consistent with polyneuropathy of hATTR amyloidosis including at least two of the following:

- Peripheral sensorimotor polyneuropathy (e.g., tingling or increased pain in the hands, feet, hands and/or arms, loss of feeling in the hands and/or feet, numbness or tingling in the wrists, carpal tunnel syndrome, loss of ability to sense temperature, difficulty with fine motor skills, weakness in the legs, difficulty walking)
- Autonomic neuropathy symptoms (e.g., orthostasis, abnormal sweating, sexual dysfunction, recurrent urinary tract infection, dysautonomia [constipation and/or diarrhea, nausea, vomiting, anorexia, early satiety])

AND

7. For patisiran (Onpattro®): Not taking in combination with inotersen (Tegsedi®) or tafamidis
OR

For inotersen (Tegsedi®): Not taking in combination with patisiran (Onpattro®) or tafamidis

Reauthorization:

1. Documentation that patient is tolerating applicable gene therapy (i.e. inotersen (Tegsedi®) or patisiran (Onpattro®))

AND

2. Documented improvement or stabilization in polyneuropathy symptoms, defined as improvement or stabilization from baseline in the Neuropathy impairment score (NIS) AND at least one of the following measures:

3. Baseline polyneuropathy disability (PND) score

4. Familial amyloid polyneuropathy (FAP) stage

5. Norfolk Quality of Life-Diabetic Neuropathy Questionnaire (Norfolk-QOL-DN) score

QUANTITY LIMIT:

For inotersen (Tegsedi®): 4 syringes per 28 days

For patisiran (Onpattro®): See Appendix B

AGE RESTRICTION

Approved for patients 18 years of age and older

PRESCRIBER RESTRICTION

Prescribed by or in consultation with a neurologist or a physician who specializes in the treatment of amyloidosis

COVERAGE DURATION

Initial authorization will be approved for 6 months

Reauthorization will be approved for 12 months

OTHER CRITERIA

N/A

TROGARZO/ROKUBIA- MEDICAL BENEFIT

MEDICATION(S)

TROGARZO

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

Initial Authorization:

1. Inadequate response to six (6) months of treatment with anti-retroviral therapy (ART) and have failed therapy within the last eight (8) weeks
 - a. Defined as persistent viremic failure
 - b. Failure must not be due to non-adherence (adherence may be verified by pharmacy claims)
2. Documentation of multi-drug resistant human immunodeficiency virus (HIV)-1 infection with viral resistance to at least one antiretroviral medication from each of the three (3) following classes:
 - a. Non-nucleoside reverse transcriptase inhibitor
 - b. Nucleoside reverse transcriptase inhibitor
 - c. Protease inhibitor
3. Documentation of baseline viral load
4. Confirmation that patient will take an optimized background regimen of anti-retroviral therapy (ART) along with Trogarzo™ or Rukobia™ therapy

Re-authorization or continuation of therapy:

1. Patient has previously received treatment with Trogarzo™.
2. Documentation of a clinically significant decrease in viral load from baseline (prior to starting therapy)
3. Confirmation that patient will continue to take an optimized background regimen of anti-retroviral therapy (ART) with Trogarzo™ or Rukobia™ therapy

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with an infectious disease specialist.

COVERAGE DURATION

Initial authorization will be approved for 6 months and reauthorization will be approved for one (1) year.

OTHER CRITERIA

N/A

TYSABRI - MEDICAL BENEFIT

MEDICATION(S)

TYSABRI

COVERED USES

N/A

EXCLUSION CRITERIA

1. Use of natalizumab in combination with other disease modifying therapy (DMT) to treat patients with multiple sclerosis (e.g., dimethyl fumarate, glatiramer)
2. Use of natalizumab in combination with immunosuppressants or TNF inhibitors (e.g., adalimumab).

REQUIRED MEDICAL INFORMATION

1. For initiation of therapy for Multiple Sclerosis, all of the following criteria (a-c) must be met:
 - a. Must have one of the following confirmed diagnoses:
 - i. Relapsing-remitting disease (RRMS)
 - ii. Secondary progressive multiple sclerosis (SPMS)
 - iii. Clinically isolated syndrome (CIS)
 - b. One of the following:
 - i. Documentation of trial, failure, or intolerance to at least one of the following disease modifying therapies:
 1. Interferon therapy (Avonex®, Rebif®, Plegridy®, or Betaseron®)
 2. Generic dimethyl fumarate
 3. glatiramer acetate (Copaxone®)
 4. teriflunomide (Aubagio®)
 5. fingolimod (Gilenya®)
 6. ocrelizumab (Ocrevus®)
 7. ozanimod hydrochloride (Zeposia®)
 8. siponimod (Mayzent®)

OR

- ii. Documentation that patient has highly active or aggressive disease defined as one of the following:
 1. Relapse leading to deterioration in physical functioning or disabilities
 2. Magnetic resonance imaging (MRI) findings of new or worsening lesions
 3. Manifestations of multiple sclerosis-related cognitive impairment

AND

- c. Negative anti-JCV antibody status OR if anti-JCV antibody positive, the patient must meet the following criteria:
 - i. Confirmation patient has not used any of the following immunosuppressants agents: mitoxantrone, azathioprine, methotrexate, cyclophosphamide, or mycophenolate mofetil

AND

- ii. Medical rationale is provided for continued use despite increased risk of developing progressive multifocal leukoencephalopathy (PML)
- 2. For initiation of therapy for Crohn's disease, all of the following criteria (a-c) must be met:
 - a. Diagnosis of moderate to severe Crohn's disease, AND
 - b. Documentation of trial, failure, intolerance, or lack of response to a formulary TNF inhibitor (Remicade® and/or Humira®) indicated for Crohn's, AND
 - c. Negative anti-JCV antibody status OR if anti-JCV antibody positive, the patient must meet the following criteria:
 - i. Confirmation patient has not used any of the following immunosuppressants agents: mitoxantrone, azathioprine, methotrexate, cyclophosphamide, and mycophenolate mofetil, AND
 - ii. Medical rationale is provided for continued use despite increased risk of developing progressive multifocal leukoencephalopathy (PML)
- 3. For patients established on therapy: Documentation of positive clinical response to therapy must be provided

AGE RESTRICTION

N/A

PRESCRIBER RESTRICTION

Prescribed by, or in consultation with, either a neurologist (for multiple sclerosis) or gastroenterologist (for Crohn's disease).

COVERAGE DURATION

Authorization will be approved until no longer eligible with the plan, subject to formulary or benefit changes.

OTHER CRITERIA

N/A

ULTOMIRIS

MEDICATION(S)

ULTOMIRIS

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

Paroxysmal Nocturnal Hemoglobinuria (PNH):

Initial authorization all of the following must be met:

1. Confirmed diagnosis of paroxysmal nocturnal hemoglobinuria (PNH) by Flow Cytometric Immunophenotyping (FCMI) using at least two (2) independent flow cytometry reagents on at least two (2) cell lineages (e.g., RBCs and WBCs) demonstrating that the patient's peripheral blood cells are deficient in glychosphatidylinositol (GPI)-linked proteins (which may include CD59, CD55, CD14, CD15, CD16, CD24, CD45, and CD64)

AND

2. Severe disease as indicated by at least one of the following (a or b):

a. Documented history of thrombosis, OR

b. Documentation of at least 10% PNH type III red cells AND at least one of the following:

i. Transfusion dependence (e.g., hemoglobin less than 7 g/dL or symptomatic anemia with hemoglobin less than 9 g/dL)

ii. Disabling fatigue

iii. End-organ complications

iv. Frequent pain paroxysms (e.g., dysphagia or abdominal pain)

v. Lactate dehydrogenase (LDH) levels greater than or equal to 1.5 times the upper limit of normal

AND

3. Dose and frequency is in accordance with FDA-approved labeling

For patients currently on eculizumab (Soliris®) switching to ravulizumab (Ultomiris®) for PNH:

1. Confirmed documentation of paroxysmal nocturnal hemoglobinuria (criteria 1 above) and severe disease (criteria 2 above). However, this can be based on patient's history prior to starting eculizumab.

AND

2. Dose and frequency is in accordance with FDA-approved labeling

Reauthorization:

1. Documentation of reduced LDH levels, reduced transfusion requirements, or improvement in PNH related symptoms
2. Dose and frequency is in accordance with FDA-approved labeling

Complement-Mediated Hemolytic Uremic Syndrome (HUS)

Initial authorization all of the following must be met:

1. Diagnosis of non-infectious HUS (i.e. HUS is not due to infection with Shiga toxin-producing *Escherichia coli*)
AND
2. Clinical presentation that includes: microangiopathic hemolytic anemia (hemoglobin less than 10 g/dL), thrombocytopenia (platelets less than 150), and acute kidney injury (elevations in serum creatinine)
AND
3. Complement dysregulation confirmed by genetic testing (e.g., mutations in complement regulatory genes: factor H (CFH), membrane cofactor protein (CD46), factor I (CFI), thrombomodulin (THBD), the activator genes: factor B (CFB) and C3 and autoantibodies to CFH)
AND
4. Prior or current treatment with plasma therapy (plasmapheresis or plasma infusions) OR medical rationale of why plasma therapy is not appropriate for member
AND
5. Dose and frequency is in accordance with FDA-approved labeling

For patients currently on eculizumab (Soliris®) switching to ravulizumab (Ultomiris®)

1. Confirmed documentation of Complement-Mediated Hemolytic Uremic Syndrome (criteria 1, 2 and 3 above). However, this can be based on patient's history prior to starting eculizumab.
AND
2. Dose and frequency is in accordance with FDA-approved labeling

Reauthorization for HUS:

1. Documentation of improvement in at least two thrombotic microangiopathy endpoints, such as:
 - Maintenance of platelet counts (i.e. improvements or reductions less than 25%)
 - Reductions in LDH
 - Reduction in number of needed plasmapheresis or plasma infusion events
 - Improvement in kidney function and reduction of dialysis
2. Dose and frequency is in accordance with FDA-approved labeling

AGE RESTRICTION

For PNH: Approved for 18 years of age and older

For aHUS: No age restriction

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with a nephrologist, hematologist or an oncologist

COVERAGE DURATION

Initial authorization for up to 3 months and reauthorization will be approved for up to one year.

OTHER CRITERIA

N/A

UPLIZNA

MEDICATION(S)

UPLIZNA

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For Neuromyelitis Optica Spectrum Disorder (NMOSD), all of the following must be met:

1. Diagnosis of neuromyelitis optica spectrum disorder as defined as the following:
 - a. Presence of at least one core clinical characteristic (optic neuritis, acute myelitis, area postrema syndrome, acute brainstem syndrome, symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions, symptomatic cerebral syndrome with NMOSD-typical brain lesions)

AND

- b. Anti-AQP4 antibody positive
2. Documentation that other alternative diagnoses have been excluded (i.e. Multiple Sclerosis)
3. Trial and failure, intolerance or contraindication to rituximab
4. Medication will not be used in combination with complement-inhibitor, anti-CD20-directed, anti-CD19 directed, or IL-6 inhibition pathway therapies
5. Dose and frequency is in accordance with FDA-approved labeling

Reauthorization for Neuromyelitis Optica Spectrum Disorder (NMOSD):

1. Documentation of positive clinical response to therapy
2. Medication will not be used in combination with complement-inhibitor, anti-CD20-directed, anti-CD19 directed, or IL-6 inhibition pathway therapies
3. Dose and frequency is in accordance with FDA-approved labeling

AGE RESTRICTION

May be approved for patients aged 18 years and older

PRESCRIBER RESTRICTION

Must be prescribed by a neurologist

COVERAGE DURATION

Initial authorization will be approved for 6 months. Reauthorization will be approved for one year.

OTHER CRITERIA

N/A

XIAFLEX

MEDICATION(S)

XIAFLEX

COVERED USES

N/A

EXCLUSION CRITERIA

N/A

REQUIRED MEDICAL INFORMATION

For Dupuytren's contracture:

1. Both of the following diagnostic criteria:
 - a. Finger flexion contracture with a palpable cord of at least one finger (other than the thumb) of 20° to 100° in a metacarpophalangeal (MP) joint or 20° to 80° in a proximal interphalangeal (PIP) joint
 - b. Documentation of a positive "table top test," defined as the inability to simultaneously place the affected finger(s) and palm flat against a table top
2. Documentation that affected joint has not had surgical intervention within the previous 90 days

For Peyronie's disease:

1. Patient's disease is stable, defined as unchanged degree of curvature for at least three months
2. Patient has a stable curvature of the penis that is between 30 and 90 degrees with a palpable plaque cord, or a cord that is documented through ultrasound
3. Patient has intact erectile function, with or without the use of medications
4. Documentation of a functional impairment that is expected to improve with treatment (e.g. inability to have intercourse despite intact erectile function, due to curvature)
5. Documentation showing the patient does not have any of the following:
 - a. Significant pain with palpation of the plaque
 - b. Lack of full erectile response to prostaglandin E1 during curvature measurement
 - c. Isolated hourglass deformity
 - d. Ventral curvature
 - e. Calcified plaque
 - f. Plaque located proximal to the base of the penis
6. Documentation that the patient has been counseled on expectations of treatment (e.g., expected average curvature reduction is 17 degrees without reduction in pain or erectile dysfunction, potential for adverse effects)

Reauthorization after the initial two (2) treatment cycles (four (4) injections) will require documentation that

the curvature of the penis remains greater than 15 degrees

AGE RESTRICTION

Approved for 18 years and older

PRESCRIBER RESTRICTION

N/A

COVERAGE DURATION

For Dupuytren's contracture: Authorization will be approved for three (3) months for a maximum of two (2) treatment courses.

For Peyronie's disease: Initial authorization will be approved for three (3) months, not to exceed four (4) injections. Reauthorization will be approved for six (6) months, not to exceed eight (8) injections per lifetime.

OTHER CRITERIA

N/A

XOLAIR - MEDICAL BENEFIT

MEDICATION(S)

XOLAIR

COVERED USES

N/A

EXCLUSION CRITERIA

Concurrent use with another therapeutic immunomodulator agent utilized for the same indication

REQUIRED MEDICAL INFORMATION

For asthma, must meet all of the following criteria:

1. Diagnosis of moderate to severe persistent allergic asthma
2. IgE baseline levels greater than 30 IU/ml
3. Positive skin test to a common perennial aeroallergens
4. Documentation that in the past three months patient is adherent to a combination of a medium/high-dose inhaled corticosteroids and a long-acting inhaled beta2-agonist. (This may be verified by pharmacy claims information)
5. Documentation of inadequate asthma control despite above therapy, defined as one of the following:
 - a. Asthma Control Test (ACT) score less than 20 or Asthma Control Questionnaire (ACQ) score greater than or equal to 1.5
 - b. At least two exacerbations requiring oral systemic corticosteroids in the last 12 months
 - c. At least one exacerbation requiring hospitalization

Reauthorization for asthma requires documentation of response to therapy, such as attainment and maintenance of remission or decrease in number of relapses

For chronic idiopathic urticaria, must meet all of the following criteria:

1. Documentation that the condition is idiopathic and that secondary causes of urticaria (e.g. offending allergens, physical contact, etc.) have been ruled out

AND

2. Trial and failure of a second-generation non-sedating H1 antihistamine (e.g., levocetirizine, loratadine, cetirizine, fexofenadine)

AND

3. Trial and failure of one additional medication from the following classes: leukotriene receptor antagonists (e.g., montelukast), first generation H1 antihistamine (e.g., diphenhydramine), or histamine H2-receptor antagonist (e.g., famotidine, ranitidine)

Reauthorization for chronic idiopathic urticaria will require documentation of response to therapy (e.g. reduction in flares or oral steroid dose).

For nasal polyps, must meet all the following criteria:

1. Evidence of bilateral nasal polyposis by direct examination, endoscopy or sinus CT scan
2. Documentation of one of the following:
 - a. Patient had an inadequate response to sinonasal surgery or is not a candidate for sinonasal surgery
 - b. Patient has tried and had an inadequate response to, or has an intolerance or contraindication to, oral systemic corticosteroids
3. Patient has tried and had an inadequate response to a three month trial of intranasal corticosteroids (e.g., fluticasone) or has a documented intolerance or contraindication to ALL intranasal corticosteroids
4. Documentation that patient will continue standard maintenance therapy (e.g., intranasal corticosteroids, nasal saline irrigation) in combination with omalizumab

Reauthorization for nasal polyps requires documentation of positive clinical response to therapy such as symptom improvement

AGE RESTRICTION

Treatment of asthma: Approved for six years of age or older.

Treatment of urticaria: Approved for 12 years of age or older.

Treatment of nasal polyps: Approved for 18 years of age or older.

PRESCRIBER RESTRICTION

Urticaria: Must be prescribed by, or in consultation with, a dermatologist, allergist or immunologist

Asthma: Must be prescribed by, or in consultation with an asthma specialist (such as a pulmonologist, immunologist, or allergist)

Nasal polyps: Must be prescribed by, or in consultation with, an otolaryngologist, allergist, pulmonologist or immunologist

COVERAGE DURATION

Urticaria and nasal polyps: Initial authorization will be for one year and reauthorization will be approved until no longer eligible with the plan, subject to formulary or benefit changes

Asthma: Authorization will be approved until no longer eligible with the plan, subject to formulary or benefit changes

OTHER CRITERIA

N/A

ZINPLAVA - MEDICAL BENEFIT

MEDICATION(S)

ZINPLAVA

COVERED USES

N/A

EXCLUSION CRITERIA

Patients with existing heart failure

REQUIRED MEDICAL INFORMATION

All of the following criteria must be met for Clostridioides difficile infection (CDI):

1. Previous trial of standard-of-care antibiotic regimen for recurrent CDI (e.g., oral vancomycin, fidaxomicin)

AND

2. Patient has at least one risk factor for higher likelihood of recurrent CDI (e.g. an age of 65 years or older, a history of C. difficile infection in the previous six months, compromised immunity, clinically severe C. difficile infection (defined as a Zar score greater than or equal to 2, scores range from 1 to 8, with higher scores indicating more severe infection))

AND

3. Bezlotoxumab (Zinplava®) must be used in combination with standard-of-care antibiotics for treatment (e.g., oral vancomycin, fidaxomicin)

Reauthorization requires:

1. Previous dose was at least 12 months prior

AND

2. Patient must have had documented benefit from previous infusion, defined as reduction in frequency of recurrences of CDI from baseline

AND

3. Bezlotoxumab (Zinplava®) is used in combination with standard-of-care antibiotics for treatment (e.g., oral vancomycin, fidaxomicin)

AGE RESTRICTION

Approved for 18 years of age and older

PRESCRIBER RESTRICTION

Must be prescribed by or in consultation with an infectious disease specialist or gastroenterology specialist

COVERAGE DURATION

Initial authorization and reauthorization will be approved for a one-time intravenous dose at 10 mg/kg

(subject to audit).

OTHER CRITERIA

N/A

ZOLGENSMA

MEDICATION(S)

ZOLGENSMA

COVERED USES

N/A

EXCLUSION CRITERIA

- Use in combination with nusinersen (Spinraza®) or risdiplam (Evrysdi®) therapy
- Repeat infusion of onasemnogene abeparvovec
- Advanced symptoms of SMA (e.g., complete paralysis of limbs, tracheostomy or ongoing invasive ventilator support in the absence of an acute reversible illness)

REQUIRED MEDICAL INFORMATION

1. Confirmed genetic diagnosis of Spinal Muscular Atrophy (SMA) with documentation of bi-allelic mutations in the survival motor neuron 1 (SMN1) gene and less than or equal to three copies of SMN2
2. Documentation that premedication with prednisolone 1 mg/kg/day (or equivalent) will be started 24 hours prior to infusion and continue for at least 30 days
3. Documentation of baseline anti-AAV9 antibody titers of less than or equal to 1:50
4. Documentation of baseline tests for liver function, platelet count, and troponin-I

AGE RESTRICTION

May be covered for patients two years of age and under

PRESCRIBER RESTRICTION

Must be prescribed by, or in consultation with, a neurologist

COVERAGE DURATION

Authorization will be approved for a one-time infusion

OTHER CRITERIA

N/A