MEDICAL PRIOR AUTHORIZATION

Abraxane (paclitaxel, albumin-bound)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Metastatic Breast Cancer
      Abraxane is indicated for the treatment of metastatic breast cancer after failure of combination chemotherapy for metastatic disease or relapse within 6 months of adjuvant chemotherapy. Prior therapy should have included an anthracycline unless clinically contraindicated.

   2. Non-Small Cell Lung Cancer
      Abraxane is indicated for the first-line treatment of locally advanced or metastatic non-small cell lung cancer, in combination with carboplatin, in patients who are not candidates for curative surgery or radiation therapy.

   3. Adenocarcinoma of the Pancreas
      Abraxane is indicated for the first-line treatment of patients with metastatic adenocarcinoma of the pancreas, in combination with gemcitabine.

B. Compendial Uses
   1. Breast cancer
   2. NSCLC
   3. Pancreatic adenocarcinoma
   4. Melanoma
   5. Epithelial ovarian cancer/fallopian tube cancer/primary peritoneal cancer
   6. Bladder cancer
   7. AIDS-related Kaposi sarcoma
   8. Endometrial carcinoma

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Pancreatic adenocarcinoma
   Authorization of 12 months may be granted for treatment of pancreatic adenocarcinoma.

B. Breast cancer
   Authorization of 12 months may be granted for treatment of recurrent or metastatic breast cancer.
C. **Non-small cell lung cancer (NSCLC)**  
Authorization of 12 months may be granted for treatment of recurrent, locally advanced, or metastatic NSCLC.

D. **Melanoma**  
Authorization of 12 months may be granted for treatment of metastatic or unresectable melanoma.

E. **Epithelial ovarian cancer/fallopian tube cancer/primary peritoneal cancer**  
Authorization of 12 months may be granted for treatment of persistent or recurrent disease.

F. **Bladder cancer**  
Authorization of 12 months may be granted for treatment of bladder cancer.

G. **AIDS-related Kaposi sarcoma**  
Authorization of 12 months may be granted for treatment of relapsed or refractory advanced AIDS-related Kaposi sarcoma.

H. **Endometrial carcinoma**  
Authorization of 12 months may be granted for treatment of endometrial carcinoma.

### III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

### IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

ADCETRIS (brentuximab vedotin)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Classical Hodgkin Lymphoma (CHL)
      a. Treatment of classical HL after failure of autologous hematopoietic stem cell transplantation (auto-HSCT) or after failure of at least two prior multi-agent chemotherapy regimens in patients who are not auto-HSCT candidates
      b. Classical HL at high risk of relapse or progression as post-auto-HSCT consolidation
   2. Systemic anaplastic large cell lymphoma (sALCL)
      a. Treatment of sALCL after failure of at least one prior multi-agent chemotherapy regimen

B. Compendial Uses
   Non-Hodgkin’s Lymphoma (NHL)
   1. CD30+ adult T-cell leukemia/lymphoma
   2. Breast implant-associated anaplastic large cell lymphoma (ALCL)
   3. Mycosis Fungoides (MF)/Sezary Syndrome (SS)
   4. Systemic anaplastic large cell lymphoma, CD30+ peripheral T-cell lymphoma (PTCL), or CD30+ angioimmunoblastic T-cell lymphoma
   5. Primary cutaneous CD30+ T-cell lymphoproliferative disorders

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Classical Hodgkin lymphoma (CHL)
   Authorization of 12 months may be granted for treatment of CHL.

B. Non-Hodgkin’s lymphoma (NHL)
   Authorization of 12 months may be granted for treatment of NHL with any of the following subtypes:
   1. Anaplastic large cell lymphoma (ALCL)
   2. CD30+ adult T-cell leukemia/lymphoma
   3. Mycosis fungoides (MF)
   4. Sezary syndrome (SS)
   5. CD30+ peripheral T-cell lymphoma (PTCL)
   6. CD30+ angioimmunoblastic T-cell lymphoma
   7. Lymphomatoid papulosis (LyP)
III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES
MEDICAL PRIOR AUTHORIZATION

ALIMTA (pemetrexed)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Nonsquamous non-small cell lung cancer (NSCLC)
      a. Alimta is indicated in combination with cisplatin therapy for the initial treatment of patients with locally advanced or metastatic nonsquamous NSCLC.
      b. Alimta is indicated for the maintenance treatment of patients with locally advanced or metastatic nonsquamous NSCLC whose disease has not progressed after four cycles of platinum-based first-line chemotherapy.
      c. Alimta is indicated as a single agent for the treatment of patients with locally advanced or metastatic nonsquamous NSCLC after prior chemotherapy.

      Limitations of use: Alimta is not indicated for the treatment of patients with squamous cell NSCLC.

   2. Malignant pleural mesothelioma (MPM)
      Alimta in combination with cisplatin is indicated for the treatment of patients with MPM whose disease is unresectable or who are otherwise not candidates for curative surgery.

B. Compendial Uses
   1. Bladder cancer, primary carcinoma of the urethra, upper genitourinary (GU) tract tumors, and urothelial carcinoma of the prostate
   2. Malignant pleural mesothelioma
   3. Nonsquamous NSCLC
   4. Ovarian cancer (epithelial histology), fallopian tube cancer, and primary peritoneal cancer
   5. Primary central nervous system (CNS) lymphoma
   6. Thymoma and thymic carcinoma

All other indications are considered experimental/investigational and are not a covered benefit.

II. EXCLUSIONS

Coverage will not be provided for members with any of the following exclusions: Squamous cell NSCLC

III. CRITERIA FOR INITIAL APPROVAL

A. Bladder Cancer, Primary Carcinoma of the Urethra, Upper Genitourinary Tract Tumors, or Urothelial Carcinoma of the Prostate
   1. Bladder Cancer

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Authorization of 12 months may be granted for treatment of bladder cancer.

2. **Primary Carcinoma of the Urethra, Upper Genitourinary Tract Tumors, or Urothelial Carcinoma of the Prostate**
   Authorization of 12 months may be granted for treatment of recurrent or metastatic primary carcinoma of the urethra, upper genitourinary tract tumors, or urothelial carcinoma of the prostate.

B. **Malignant Pleural Mesothelioma (MPM)**
   Authorization of 12 months may be granted for treatment of MPM.

C. **Non-Small Cell Lung Cancer (Non-Squamous)**
   Authorization of 12 months may be granted for treatment of non-squamous non-small cell lung cancer.

D. **Ovarian Cancer (Epithelial)/Fallopian Tube Cancer/Primary Peritoneal Cancer**
   Authorization of 12 months may be granted for treatment of persistent or recurrent epithelial ovarian cancer, fallopian tube cancer, or primary peritoneal cancer.

E. **Primary CNS Lymphoma**
   Authorization of 12 months may be granted for treatment of progressive or recurrent primary CNS lymphoma.

F. **Thymoma and Thymic Carcinoma**
   Authorization of 12 months may be granted for treatment of thymoma or thymic carcinoma.

IV. **CONTINUATION OF THERAPY**

   All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

V. **DOSAGE AND ADMINISTRATION**

   Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

VI. REFERENCES


SPECIALTY GUIDELINE MANAGEMENT

ALIQOPA (copanlisib)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications

Aliqopa is indicated for the treatment of adult patients with relapsed follicular lymphoma (FL) who have received at least two prior systemic therapies.

Accelerated approval was granted for this indication based on overall response. Continued approval for this indication may be contingent upon verification and description of clinical benefit in a confirmatory trial.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Follicular lymphoma

Authorization of 12 months may be granted for treatment of relapsed follicular lymphoma (FL) when the member has received at least two prior systemic therapies.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

ARANESP (darbepoetin alfa)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications

1. Treatment of anemia due to chronic kidney disease (CKD), including patients on dialysis and patients not on dialysis.
2. Treatment of anemia in patients with non-myeloid malignancies where anemia is due to the effect of concomitant myelosuppressive chemotherapy, and upon initiation, there is a minimum of two additional months of planned chemotherapy.

Limitations of Use:

1. Aranesp has not been shown to improve quality of life, fatigue, or patient well-being.
2. Aranesp is not indicated for use:
   - In patients with cancer receiving hormonal agents, biologic products, or radiotherapy, unless also receiving concomitant myelosuppressive chemotherapy.
   - In patients with cancer receiving myelosuppressive chemotherapy when the anticipated outcome is cure.
   - As a substitute for RBC transfusions in patients who require immediate correction of anemia.

B. Compendial Uses

1. Symptomatic anemia in patients with myelodysplastic syndromes (MDS)
2. Anemia in patients whose religious beliefs forbid blood transfusions
3. Symptomatic anemia in patients with primary myelofibrosis, post-polycythemia vera myelofibrosis, and post-essential thrombocythemia myelofibrosis

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Note: Requirements regarding pretreatment hemoglobin level exclude values due to a recent transfusion.

A. Anemia Due to CKD

Authorization of 12 weeks may be granted for members with pretreatment hemoglobin < 10 g/dL.

B. Anemia Due to Myelosuppressive Chemotherapy

Authorization of 12 weeks may be granted for members with nonmyeloid malignancy who meet ALL of the following criteria:

1. The intent of chemotherapy is non-curative
2. Pretreatment hemoglobin < 10 g/dL
C. Anemia in MDS
Authorization of 12 weeks may be granted for members with pretreatment hemoglobin < 10 g/dL.

D. Anemia in Members Whose Religious Beliefs Forbid Blood Transfusions
Authorization of 12 weeks may be granted for members with pretreatment hemoglobin < 10 g/dL.

E. Anemia in Primary Myelofibrosis (MF), Post-polycythemia Vera MF, and Post-Essential Thrombocytthemia MF
Authorization of 12 weeks may be granted for members who meet ALL of the following criteria:
1. Member has symptomatic anemia
2. Pretreatment hemoglobin < 10 g/dL
3. Pretreatment serum erythropoietin level < 500 mU/mL

III. CONTINUATION OF THERAPY

Note: Requirements regarding current hemoglobin level exclude values due to a recent transfusion.

For all indications below: all members (including new members) requesting authorization for continuation of therapy after at least 12 weeks of ESA treatment must show a response with a rise in hemoglobin of ≥ 1 g/dL. Members who completed less than 12 weeks of ESA treatment and have not yet responded with a rise in hemoglobin of ≥ 1 g/dL may be granted authorization of up to 12 weeks to allow for sufficient time to demonstrate a response.

A. Anemia due to CKD
Authorization of 12 weeks may be granted for continuation of treatment when the current hemoglobin is ≤ 12 g/dL.

B. Anemia Due to Myelosuppressive Chemotherapy
Authorization of 12 weeks may be granted for continuation of treatment in members with nonmyeloid malignancy who meet BOTH of the following criteria:
1. The intent of chemotherapy is non-curative
2. Current hemoglobin is < 11 g/dL

C. Anemia in MDS
Authorization of 12 weeks may be granted for continuation of treatment when the current hemoglobin is ≤ 12 g/dL.

D. Anemia in members whose religious beliefs forbid blood transfusions
Authorization of 12 weeks may be granted for continuation of treatment when the current hemoglobin is ≤ 12 g/dL.

E. Anemia in Primary Myelofibrosis, Post-polycythemia Vera Myelofibrosis, and Post-Essential Thrombocytthemia Myelofibrosis
Authorization of 12 weeks may be granted for continuation of treatment when the current hemoglobin is ≤ 12 g/dL.

IV. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

BAVENCIO (avelumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
A. Treatment of adults and pediatric patients 12 years and older with metastatic Merkel cell carcinoma
B. Treatment of patients with locally advanced or metastatic urothelial carcinoma who have disease progression during or following platinum-containing chemotherapy or have disease progression within 12 months of neoadjuvant or adjuvant treatment with platinum-containing chemotherapy

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Merkel Cell Carcinoma
   Authorization of 12 months may be granted for the treatment of metastatic Merkel cell carcinoma.

B. Urothelial Carcinoma
   Authorization of 12 months may be granted for treatment of locally advanced or metastatic urothelial carcinoma when either of the following criteria are met:
   1. Member experienced disease progression during or following platinum-containing chemotherapy.
   2. Member experienced disease progression within 12 months of neoadjuvant or adjuvant treatment with platinum-containing chemotherapy.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

BELEODAQ (belinostat)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   Treatment of patients with relapsed or refractory peripheral T-cell lymphoma (PTCL)

B. Compendial Uses
   Non-Hodgkin’s Lymphoma (NHL)
   1. Adult T-cell leukemia/lymphoma (ATLL)
   2. Mycosis fungoides (MF)/Sezary syndrome (SS)
   3. Primary cutaneous CD30+ T-cell lymphoproliferative disorders

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Peripheral T-cell lymphoma (PTCL)
   Authorization of 12 months may be granted for treatment of PTCL.

B. Adult T-cell leukemia/lymphoma (ATLL)
   Authorization of 12 months may be granted for treatment of ATLL.

C. Mycosis fungoides (MF)/Sezary syndrome (SS)
   Authorization of 12 months may be granted for treatment of mycosis fungoides (MF)/Sezary syndrome (SS).

D. Primary cutaneous CD30+ T-cell lymphoproliferative disorders
   Authorization of 12 months may be granted for treatment of cutaneous anaplastic large cell lymphoma (ALCL).

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.
IV. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

BENLYSTA (belimumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication
Treatment of adult patients with active, autoantibody-positive, systemic lupus erythematosus (SLE) who are receiving standard therapy.

Limitations of Use
The efficacy of Benlysta has not been evaluated in patients with severe active lupus nephritis or severe active central nervous system lupus. Benlysta has not been studied in combination with other biologics or intravenous cyclophosphamide. Use of Benlysta is not recommended in these situations.

All other indications are considered experimental/investigational and are not a covered benefit.

II. EXCLUSIONS

Coverage will not be provided for members with any of the following exclusions:
A. Severe active lupus nephritis
B. Severe active central nervous system lupus

III. CRITERIA FOR INITIAL APPROVAL

Systemic Lupus Erythematosus (SLE)
Authorization of 12 months may be granted for treatment of active SLE when all of the following criteria are met:
1. Prior to initiating therapy, the member is autoantibody-positive.
2. The member is currently receiving standard therapy for SLE (see Appendix) or has tried and had an inadequate response or intolerance to standard therapy for SLE.

IV. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.
V. APPENDIX

Standard Therapy for SLE
- Antimalarials (e.g., hydroxychloroquine)
- Azathioprine
- Corticosteroids
- Leflunomide
- Methotrexate
- Mycophenolate mofetil
- Non-steroidal anti-inflammatory drugs

VI. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

BLINCYTO (blinatumomab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
Philadelphia chromosome-negative relapsed or refractory B-cell precursor acute lymphoblastic leukemia (ALL)

B. Compendial Uses
Relapsed/refractory Philadelphia chromosome-positive B-cell precursor ALL

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Authorization of 9 months may be granted for treatment of precursor B-cell acute lymphoblastic leukemia (ALL).

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

BOTOX (onabotulinumtoxinA)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications

1. Overactive bladder with symptoms of urge urinary incontinence, urgency, and frequency, in adults who have an inadequate response to or are intolerant of an anticholinergic medication
2. Urinary incontinence due to detrusor overactivity associated with a neurologic condition (e.g., spinal cord injury, multiple sclerosis) in adults who have an inadequate response to or are intolerant of an anticholinergic medication
3. Prophylaxis of headaches in adult patients with chronic migraine (≥15 days per month with headache lasting 4 hours a day or longer)
4. Treatment of upper limb spasticity in adult patients
5. Treatment of lower limb spasticity in adult patients
6. Cervical dystonia in adults, to reduce the severity of abnormal head position and neck pain
7. Severe primary axillary hyperhidrosis that is inadequately managed with topical agents in adult patients
8. Strabismus and blepharospasm associated with dystonia, including benign essential blepharospasm or VII nerve disorders in patients 12 years of age and above

B. Compendial Uses

1. Achalasia
2. Chronic anal fissures
3. Essential tremor
4. Excessive salivation secondary to advanced Parkinson’s disease
5. Hemifacial spasm
6. Spasmodic dysphonia (laryngeal dystonia)

All other indications are considered experimental/investigational and are not a covered benefit.

II. EXCLUSIONS

Coverage will not be provided for cosmetic use.

III. CRITERIA FOR INITIAL APPROVAL

A. Blepharospasm

Authorization of 24 months may be granted for treatment of blepharospasm.
B. Cervical dystonia
Authorization of 24 months may be granted for treatment of cervical dystonia (e.g., torticollis).

C. Chronic migraine prophylaxis
Authorization of 6 months (two injection cycles) may be granted for treatment of chronic migraine prophylaxis when all of the following criteria are met:
1. Member experiences headaches ≥ 15 days per month
2. Member completed adequate trial (≥ 8 weeks) of an oral migraine preventative therapy such as:
   a. Divalproex sodium (Depakote, Depakote ER)
   b. Topiramate (Topamax)
   c. Gabapentin (Neurontin)
   d. Amitriptyline (Elavil)
   e. Venlafaxine (Effexor)
   f. Atenolol/Metoprolol/Propranolol/Timolol/Nadolol
   g. Nimodipine/Verapamil
   h. Naproxen/other NSAID

D. Overactive bladder with urinary incontinence
Authorization of 12 months may be granted for treatment of overactive bladder with urinary incontinence when the member has had an inadequate response or experienced intolerance to an anticholinergic medication (e.g., Vesicare [solifenacin], Enablex [darifenacin], Toviaz [fesoterodine], Detrol/Detrol LA [tolterodine], Sanctura/Sanctura XR [trospium], Ditropan XL [oxybutynin]).

E. Primary axillary hyperhidrosis
Authorization of 12 months may be granted for treatment of primary axillary hyperhidrosis.

F. Strabismus
Authorization of 12 months may be granted for treatment of strabismus.

G. Upper limb spasticity
Authorization of 24 months may be granted for treatment of upper limb spasticity.

H. Lower limb spasticity
Authorization of 24 months may be granted for treatment of lower limb spasticity.

I. Urinary incontinence associated with a neurologic condition (e.g., spinal cord injury, multiple sclerosis)
Authorization of 24 months may be granted for treatment of urinary incontinence associated with a neurologic condition (e.g., spinal cord injury, multiple sclerosis) when the member has had an inadequate response or experienced intolerance to an anticholinergic medication (e.g., Vesicare [solifenacin], Enablex [darifenacin], Toviaz [fesoterodine], Detrol/Detrol LA [tolterodine], Sanctura/Sanctura XR [trospium], Ditropan XL [oxybutynin]).

J. Achalasia
Authorization of 24 months may be granted for treatment of achalasia.

K. Chronic anal fissures
Authorization of 12 months may be granted for treatment of chronic anal fissures.

L. Essential tremor
Authorization of 24 months may be granted for treatment of essential tremor.
M. Excessive salivation due to advanced Parkinson’s disease
   Authorization of 24 months may be granted for treatment of excessive salivation due to advanced Parkinson’s disease.

N. Hemifacial spasm
   Authorization of 24 months may be granted for treatment of hemifacial spasm.

O. Spasmodic dysphonia (laryngeal dystonia)
   Authorization of 24 months may be granted for treatment of spasmodic dysphonia (laryngeal dystonia).

IV. CONTINUATION OF THERAPY

A. All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria for all approvable conditions other than chronic migraine prophylaxis.

B. Authorization of 12 months may be granted for treatment of chronic migraine prophylaxis when the member has achieved or maintained a 50% reduction in monthly headache frequency since starting therapy with Botox.

V. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

CEREZYME (imiglucerase)

POLICY

I. INDICATIONS
The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
Cerezyme is indicated for long-term enzyme replacement therapy (ERT) for pediatric and adult patients with a confirmed diagnosis of type 1 Gaucher disease that results in one or more of the following conditions: anemia, thrombocytopenia, bone disease, hepatomegaly, or splenomegaly.

B. Compendial Uses
Gaucher disease type 3

II. CRITERIA FOR INITIAL APPROVAL

A. Gaucher disease type 1
Indefinite authorization may be granted for treatment of Gaucher disease type 1 when the diagnosis of Gaucher disease was confirmed by enzyme assay demonstrating a deficiency of beta-glucocerebrosidase (glucosidase) enzyme activity or by genetic testing.

A. Gaucher disease type 3
Indefinite authorization may be granted for treatment of Gaucher disease type 3 when the diagnosis of Gaucher disease was confirmed by enzyme assay demonstrating a deficiency of beta-glucocerebrosidase (glucosidase) enzyme activity or by genetic testing.

III. CONTINUATION OF THERAPY
All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

CINQAIR (reslizumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication
Cinqaír is indicated for the add-on maintenance treatment of patients with severe asthma aged 18 years and older with an eosinophilic phenotype.

Limitations of Use:
- Not for treatment of other eosinophilic conditions.
- Not for the relief of acute bronchospasm or status asthmaticus.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Authorization of 12 months may be granted for treatment of eosinophilic asthma when ALL of the following criteria are met:
A. Member is 18 years of age or older
B. Member has baseline blood eosinophil count of at least 400 cells per microliter
C. Member has a history of severe asthma despite current treatment with both of the following medications at optimized doses:
   1. Inhaled corticosteroid
   2. Additional controller (long acting β2-agonist, leukotriene modifier, or sustained-release theophylline)

III. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for treatment of eosinophilic asthma when ALL of the following criteria are met:
A. Member is 18 years of age or older
B. Asthma control has improved on Cinqaír treatment, demonstrated by a reduction in the frequency and/or severity of symptoms and exacerbations

IV. REFERENCES


SPECIALTY GUIDELINE MANAGEMENT

CYRAMZA (ramucirumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Gastric Cancer: Cyramza as a single agent, or in combination with paclitaxel, is indicated for the treatment of patients with advanced or metastatic, gastric or gastro-esophageal junction (GEJ) adenocarcinoma with disease progression on or after prior fluoropyrimidine-or platinum-containing chemotherapy.
   2. Non-Small Cell Lung Cancer (NSCLC): Cyramza, in combination with docetaxel, is indicated for the treatment of patients with metastatic NSCLC with disease progression on or after platinum-based chemotherapy. Patients with EGFR or ALK genomic tumor aberrations should have disease progression on FDA-approved therapy for these aberrations prior to receiving Cyramza.
   3. Colorectal Cancer: Cyramza, in combination with FOLFIRI (irinotecan, folinic acid, and 5-fluorouracil), is indicated for the treatment of patients with metastatic colorectal cancer with disease progression on or after prior therapy with bevacizumab, oxaliplatin, and a fluoropyrimidine.

B. Compendial Uses
   1. Esophageal adenocarcinoma
   2. Colorectal cancer, advanced

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Gastric, Gastro-esophageal Junction (GEJ), and Esophageal Adenocarcinoma
   Authorization of 12 months may be granted for treatment of gastric, gastro-esophageal junction (GEJ), and esophageal adenocarcinoma.

B. Non-Small Cell Lung Cancer (NSCLC)
   Authorization of 12 months may be granted for treatment of metastatic NSCLC.

C. Colorectal Cancer
   Authorization of 12 months may be granted for treatment of advanced or metastatic colorectal cancer.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.
IV. REFERENCES

Specialty Guideline Management

DARZALEX (daratumumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered covered benefits provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
A. In combination with lenalidomide and dexamethasone, or bortezomib and dexamethasone, for the treatment of patients with multiple myeloma who have received at least one prior therapy
B. In combination with pomalidomide and dexamethasone for the treatment of patients with multiple myeloma who have received at least two prior therapies including lenalidomide and a proteasome inhibitor
C. As monotherapy, for the treatment of patients with multiple myeloma who have received at least three prior lines of therapy including a proteasome inhibitor and an immunomodulatory agent or who are double-refractory to a proteasome inhibitor and an immunomodulatory agent

All other indications are considered experimental/investigational and are not covered benefits.

II. CRITERIA FOR INITIAL APPROVAL

Authorization of 12 months may be granted for the treatment of multiple myeloma when the member has received at least one prior therapy.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT
DYSPORT (abobotulinumtoxinA)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Treatment of cervical dystonia in adults
   2. Treatment of spasticity (upper and/or lower limb) in adults
   3. Treatment of lower limb spasticity in pediatric patients 2 years of age and older

B. Compendial Uses
   1. Treatment of benign essential blepharospasm

All other indications are considered experimental/investigational and are not a covered benefit.

II. EXCLUSIONS

Coverage will not be provided for cosmetic use.

III. CRITERIA FOR INITIAL APPROVAL

A. Cervical dystonia
   Authorization of 24 months may be granted for treatment of cervical dystonia (e.g., torticollis).

B. Upper limb spasticity
   Authorization of 24 months may be granted for treatment of upper limb spasticity.

C. Lower limb spasticity
   Authorization of 24 months may be granted for treatment of lower limb spasticity (e.g., cerebral palsy, multiple sclerosis).

D. Blepharospasm
   Authorization of 24 months may be granted for treatment of benign essential blepharospasm.

IV. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.
V. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

EMPLICITI (elotuzumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications

Empliciti is indicated in combination with lenalidomide and dexamethasone for the treatment of patients with multiple myeloma who have received one to three prior therapies.

B. Compendial Uses

Therapy for patients who have received one to three prior therapies for relapse or for progressive or refractory multiple myeloma in combination with bortezomib and dexamethasone

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Authorization of 12 months may be granted for the treatment of multiple myeloma for members who have received at least one prior therapy.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

EPOGEN, PROCRIT (epoetin alfa)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications

1. Epoetin alfa is indicated for the treatment of anemia due to chronic kidney disease (CKD), including patients on dialysis and not on dialysis to decrease the need for red blood cell (RBC) transfusion.
2. Epoetin alfa is indicated for the treatment of anemia due to zidovudine administered at ≤ 4200 mg/week in HIV-infected patients with endogenous serum erythropoietin levels of ≤ 500 mUnits/mL.
3. Epoetin alfa is indicated for the treatment of anemia in patients with non-myeloid malignancies where anemia is due to the effect of concomitant myelosuppressive chemotherapy, and upon initiation, there is a minimum of two additional months of planned chemotherapy.
4. Epoetin alfa is indicated to reduce the need for allogeneic RBC transfusions among patients with perioperative hemoglobin > 10 to ≤ 13 g/dL who are at high risk for perioperative blood loss from elective, noncardiac, nonvascular surgery. Epoetin alfa is not indicated for patients who are willing to donate autologous blood preoperatively.

Limitations of Use:

1. Epoetin alfa has not been shown to improve quality of life, fatigue, or patient well-being.
2. Epoetin alfa is not indicated for use:
   - In patients with cancer receiving hormonal agents, biologic products, or radiotherapy, unless also receiving concomitant myelosuppressive chemotherapy.
   - In patients with cancer receiving myelosuppressive chemotherapy when the anticipated outcome is cure.
   - In patients scheduled for surgery who are willing to donate autologous blood.
   - In patients undergoing cardiac or vascular surgery.
   - As a substitute for RBC transfusions in patients who require immediate correction of anemia.

B. Compendial Uses

1. Symptomatic anemia in patients with myelodysplastic syndromes (MDS)
2. Anemia in congestive heart failure
3. Anemia in rheumatoid arthritis
4. Anemia due to hepatitis C treatment with ribavirin in combination with either interferon alfa or peginterferon alfa
5. Anemia in patients whose religious beliefs forbid blood transfusions
6. Symptomatic anemia in patients with primary myelofibrosis, post-polycythemia vera myelofibrosis, and post-essential thrombocythemia myelofibrosis

All other indications are considered experimental/investigational and are not a covered benefit.
II. CRITERIA FOR INITIAL APPROVAL

Note: Requirements regarding pretreatment hemoglobin level exclude values due to a recent transfusion.

A. Anemia Due to CKD
Authorization of 12 weeks may be granted for members with pretreatment hemoglobin < 10 g/dL.

B. Anemia Due to Myelosuppressive Chemotherapy
Authorization of 12 weeks may be granted for members with nonmyeloid malignancy who meet ALL of the following criteria:
1. The intent of chemotherapy is non-curative
2. Pretreatment hemoglobin < 10 g/dL

C. Anemia in MDS
Authorization of 12 weeks may be granted for members with pretreatment hemoglobin < 10 g/dL.

D. Reduction of Allogeneic Red Blood Cell Transfusion in Patients Undergoing Elective, Noncardiac, Nonvascular Surgery
Authorization of 12 weeks may be granted for members scheduled to have an elective, noncardiac, nonvascular surgery when the pretreatment hemoglobin is > 10 to ≤ 13 g/dL.

E. Anemia in Congestive Heart Failure (CHF)
Authorization of 12 weeks may be granted for members with pretreatment hemoglobin < 9 g/dL.

F. Anemia in Rheumatoid Arthritis (RA)
Authorization of 12 weeks may be granted for members with pretreatment hemoglobin < 10 g/dL.

G. Anemia Due to Hepatitis C Treatment
Authorization of 12 weeks may be granted for members with pretreatment hemoglobin < 10 g/dL who are receiving ribavirin in combination with either interferon alfa or peginterferon alfa.

H. Anemia Due to Zidovudine in HIV-infected Patients
Authorization of 12 weeks may be granted for members currently receiving zidovudine with pretreatment hemoglobin < 10 g/dL.

I. Anemia in Members Whose Religious Beliefs Forbid Blood Transfusions
Authorization of 12 weeks may be granted for members with pretreatment hemoglobin < 10 g/dL.

J. Anemia in Primary Myelofibrosis (MF), Post-polycythemia Vera MF, and Post-Essential Thrombocythemia MF
Authorization of 12 weeks may be granted for members who meet ALL of the following criteria:
1. Member has symptomatic anemia
2. Pretreatment hemoglobin < 10 g/dL
3. Pretreatment serum erythropoietin level < 500 mU/mL

III. CONTINUATION OF THERAPY

Note: Requirements regarding current hemoglobin level exclude values due to a recent transfusion.
For all indications below: all members (including new members) requesting authorization for continuation of therapy after at least 12 weeks of ESA treatment must show a response with a rise in hemoglobin of $\geq 1$ g/dL. Members who completed less than 12 weeks of ESA treatment and have not yet responded with a rise in hemoglobin of $\geq 1$ g/dL may be granted authorization of up to 12 weeks to allow for sufficient time to demonstrate a response.

A. Anemia Due to CKD
Authorization of 12 weeks may be granted for continuation of therapy when the current hemoglobin is $\leq 12$ g/dL.

B. Anemia Due to Myelosuppressive Chemotherapy
Authorization of 12 weeks may be granted for the continuation of therapy in members with nonmyeloid malignancy who meet BOTH of the following criteria:
1. The intent of chemotherapy is non-curative
2. Current hemoglobin is $< 11$ g/dL

C. Anemia in MDS
Authorization of 12 weeks may be granted for continuation of treatment when the current hemoglobin is $\leq 12$ g/dL.

D. Anemia in CHF, RA
Authorization of 12 weeks may be granted for continuation of treatment when the current hemoglobin is $\leq 12$ g/dL.

E. Anemia Due to Hepatitis C Treatment
Authorization of 12 weeks may be granted for continuation of treatment when the member meets ALL of the following criteria:
1. The member is receiving ribavirin in combination with either interferon alfa or peginterferon alfa
2. The current hemoglobin is $\leq 12$ g/dL.

F. Anemia Due to Zidovudine in HIV-infected Patients
Authorization of 12 weeks may be granted for continuation of therapy in members receiving zidovudine when the current hemoglobin is $\leq 12$ g/dL.

G. Anemia in Members Whose Religious Beliefs Forbid Blood Transfusions
Authorization of 12 weeks may be granted for continuation of treatment when the current hemoglobin is $\leq 12$ g/dL.

H. Anemia in Primary Myelofibrosis, Post-polycythemia Vera Myelofibrosis, and Post-Essential Thrombocythemia Myelofibrosis
Authorization of 12 weeks may be granted for continuation of treatment when the current hemoglobin is $\leq 12$ g/dL.

IV. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

epoprostenol for injection (generic)
Flolan (epoprostenol for injection)
Veletri (epoprostenol for injection)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication
Epoprostenol/Flolan/Veletri is indicated for the treatment of pulmonary arterial hypertension (WHO Group I) to improve exercise capacity.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Indefinite authorization may be granted for treatment of PAH when ALL of the following criteria are met:
A. Member has PAH defined as WHO Group 1 class of pulmonary hypertension (refer to Appendix).
B. PAH was confirmed by either criterion (1) or criterion (2) below:
   1. Pretreatment right heart catheterization with all of the following results:
      i. mPAP ≥ 25 mmHg
      ii. PCWP ≤ 15 mmHg
      iii. PVR > 3 Wood units
   2. For infants less than one year of age with any of the following conditions, PAH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed:
      i. Post cardiac surgery
      ii. Chronic heart disease
      iii. Chronic lung disease associated with prematurity
      iv. Congenital diaphragmatic hernia

III. CONTINUATION OF THERAPY

Indefinite authorization may be granted for members with PAH who are currently receiving epoprostenol/Flolan/Veletri therapy through a paid pharmacy or medical benefit.

IV. APPENDIX

WHO Classification of Pulmonary Hypertension
WHO Group 1. Pulmonary Arterial Hypertension (PAH)
1.1 Idiopathic (IPAH)
1.2 Heritable PAH
   1.2.1 Germline mutations in the bone morphogenetic protein receptor type 2 (BMPR2)
   1.2.2 Activin receptor-like kinase type 1 (ALK1), endoglin (with or without hereditary hemorrhagic telangiectasia), Smad 9, caveolin-1 (CAV1), potassium channel super family K member-3 (KCNK3)
   1.2.3 Unknown
1.3 Drug- and toxin-induced
1.4. Associated with:
   1.4.1 Connective tissue diseases
   1.4.2 HIV infection
   1.4.3 Portal hypertension
   1.4.4 Congenital heart diseases
   1.4.5 Schistosomiasis
1.‘ Pulmonary veno-occlusive disease (PVOD) and/or pulmonary capillary hemangiomatosis (PCH)
1". Persistent pulmonary hypertension of the newborn (PPHN)

WHO Group 2. Pulmonary Hypertension Owing to Left Heart Disease
2.1 Systolic dysfunction
2.2 Diastolic dysfunction
2.3 Valvular disease
2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies

WHO Group 3. Pulmonary Hypertension Owing to Lung Disease and/or Hypoxia
3.1 Chronic obstructive pulmonary disease
3.2 Interstitial lung disease
3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
3.4 Sleep-disordered breathing
3.5 Alveolar hypoventilation disorders
3.6 Chronic exposure to high altitude
3.7 Developmental abnormalities

WHO Group 4. Chronic Thromboembolic Pulmonary Hypertension (CTEPH)
WHO Group 5. Pulmonary Hypertension with Unclear Multifactorial Mechanisms
5.1 Hematologic disorders: Chronic hemolytic anemia, myeloproliferative disorders, splenectomy
5.2 Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis: lymphangioleiomyomatosis, neurofibromatosis, vasculitis
5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
5.4 Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure on dialysis, segmental PH

V. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

BEBULIN, PROFILNINE
(factor IX complex [human])

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication
   Hemophilia B

B. Compendial Uses
   1. Bleeding due to low levels of liver-dependent coagulation factors
   2. Factor X deficiency (Bebulin only)
   3. Factor II deficiency (Profilnine only)

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

1. Hemophilia B
   Indefinite authorization may be granted for treatment of hemophilia B.

2. Bleeding Due to Low Levels of Liver-dependent Coagulation Factors
   Indefinite authorization may be granted for treatment of bleeding due to low levels of liver-dependent coagulation factors.

3. Factor X Deficiency
   Indefinite authorization of Bebulin may be granted for treatment of factor X deficiency.

4. Factor II Deficiency
   Indefinite authorization of Profilnine may be granted for treatment of factor II deficiency.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

REBINYN (coagulation factor IX [recombinant], glycoPEGylated)

IDELVION (coagulation factor IX [recombinant], albumin fusion protein)

ALPROLIX (coagulation factor IX [recombinant], Fc fusion protein)

BENEFIX, IXINITY, RIXUBIS (coagulation factor IX [recombinant])

ALPHANINE SD, MONONINE (coagulation factor IX [human])

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication

Hemophilia B

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Hemophilia B

Indefinite authorization may be granted for treatment of hemophilia B.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

IV. REFERENCES


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SPECIALTY GUIDELINE MANAGEMENT

FACTOR VIII CONCENTRATES

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

Table: Factor VIII Concentrates and Covered Uses

<table>
<thead>
<tr>
<th>Brand</th>
<th>Generic</th>
<th>FDA-Approved Indication(s)</th>
<th>Compendial Indication(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Advate</td>
<td>antihemophilic factor [recombinant]</td>
<td>Hemophilia A</td>
<td>Acquired Hemophilia A</td>
</tr>
<tr>
<td>Afstyla</td>
<td>antihemophilic factor [recombinant], single chain</td>
<td>Hemophilia A</td>
<td></td>
</tr>
<tr>
<td>Helixate FS</td>
<td>antihemophilic factor [recombinant]</td>
<td>Hemophilia A</td>
<td>Acquired Hemophilia A</td>
</tr>
<tr>
<td>Kogenate FS</td>
<td>antihemophilic factor [recombinant]</td>
<td>Hemophilia A</td>
<td>Acquired Hemophilia A</td>
</tr>
<tr>
<td>Kovaltry</td>
<td>antihemophilic factor [recombinant]</td>
<td>Hemophilia A</td>
<td></td>
</tr>
<tr>
<td>Novoeight</td>
<td>antihemophilic factor [recombinant]</td>
<td>Hemophilia A</td>
<td></td>
</tr>
<tr>
<td>Nuwiq</td>
<td>antihemophilic factor [recombinant]</td>
<td>Hemophilia A</td>
<td></td>
</tr>
<tr>
<td>Recombinate</td>
<td>antihemophilic factor [recombinant]</td>
<td>Hemophilia A</td>
<td>Acquired Hemophilia A</td>
</tr>
<tr>
<td>Xyntha</td>
<td>antihemophilic factor [recombinant]</td>
<td>Hemophilia A</td>
<td>Acquired Hemophilia A</td>
</tr>
</tbody>
</table>

Prolonged Half-life Recombinant Factor VIII Concentrate

<table>
<thead>
<tr>
<th>Brand</th>
<th>Generic</th>
<th>FDA-Approved Indication(s)</th>
<th>Compendial Indication(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adynovate</td>
<td>antihemophilic factor [recombinant], PEGylated</td>
<td>Hemophilia A</td>
<td></td>
</tr>
<tr>
<td>Eloctate</td>
<td>antihemophilic factor [recombinant], Fc fusion protein</td>
<td>Hemophilia A</td>
<td></td>
</tr>
</tbody>
</table>

Human Plasma-Derived Factor VIII Concentrates

<table>
<thead>
<tr>
<th>Brand</th>
<th>Generic</th>
<th>FDA-Approved Indication(s)</th>
<th>Compendial Indication(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemofil M</td>
<td>antihemophilic factor [human] monoclonal antibody purified</td>
<td>Hemophilia A</td>
<td>Acquired Hemophilia A</td>
</tr>
<tr>
<td>Monoclate-P</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Human Plasma-Derived Factor VIII Concentrates That Contain Von Willebrand Factor

<table>
<thead>
<tr>
<th>Brand</th>
<th>Generic</th>
<th>FDA-Approved Indication(s)</th>
<th>Compendial Indication(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Humate-P</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Koate</td>
<td>antihemophilic factor [human]</td>
<td>Hemophilia A</td>
<td>Acquired Hemophilia A, von Willebrand Disease</td>
</tr>
</tbody>
</table>

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All other indications are considered experimental/investigational and are not a covered benefit.

CRITERIA FOR INITIAL APPROVAL

A. Hemophilia A
   Indefinite authorization of Advate, Adynovate, Afstyla, Alphanate, Eloctate, Helixate FS, Hemofil M, Humate-P, Koate, Kogenate FS, Kovaltry, Monoclate-P, Novoeight, Nuwiq, Recombinate or Xyntha may be granted for treatment of hemophilia A when either of the following criteria is met:
   1. Member has mild disease (see Appendix A) and has had an insufficient response to desmopressin or a documented clinical reason for not using desmopressin (see Appendix B).
   2. Member has moderate to severe disease (see Appendix A).

B. Von Willebrand Disease
   Indefinite authorization of Alphanate, Humate-P or Koate may be granted for treatment of vWD when any of the following criteria is met:
   1. Member has type 1, 2A, 2M, or 2N vWD and has had an insufficient response to desmopressin or a documented clinical reason for not using desmopressin (see Appendix B).
   2. Member has type 2B or type 3 vWD.

C. Acquired Hemophilia A
   Indefinite authorization of Advate, Alphanate, Helixate FS, Hemofil M, Humate-P, Koate, Kogenate FS, Monoclate-P, Recombinate or Xyntha may be granted for treatment of acquired hemophilia A.

D. Acquired von Willebrand Syndrome
   Indefinite authorization of Alphanate or Humate-P may be granted for treatment of acquired von Willebrand syndrome.

II. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

III. APPENDICES

Appendix A: Classification of Hemophilia by Clotting Factor Level (% Activity) and Bleeding Episodes

<table>
<thead>
<tr>
<th>Severity</th>
<th>Clotting Factor Level % activity*</th>
<th>Bleeding Episodes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Severe</td>
<td>&lt;1%</td>
<td>Spontaneous bleeding episodes, predominantly into joints and muscles</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Severe bleeding with trauma, injury or surgery</td>
</tr>
<tr>
<td>Moderate</td>
<td>1% to 5%</td>
<td>Occasional spontaneous bleeding episodes</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Severe bleeding with trauma, injury or surgery</td>
</tr>
</tbody>
</table>
Appendix B: Clinical Reasons For Not Utilizing Desmopressin in Patients with Hemophilia A and Type 1, 2A, 2N and 2M vWD

A. Age < 2 years
B. Pregnancy
C. Fluid/electrolyte imbalance
D. High risk for cardiovascular or cerebrovascular disease (especially the elderly)
E. Predisposition to thrombus formation
F. Trauma requiring surgery
G. Life-threatening bleed
H. Contraindication or intolerance to desmopressin
I. Severe type 1 von Willebrand disease

IV. REFERENCES


SPECIALTY GUIDELINE MANAGEMENT

FEIBA (anti-inhibitor coagulant complex [human])

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication
   Hemophilia A and hemophilia B with inhibitors

B. Compendial Use
   Acquired hemophilia A

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Hemophilia A with Inhibitors
   Indefinite authorization may be granted for treatment of hemophilia A with inhibitors (see Appendix) when the inhibitor titer is ≥ 5 Bethesda units per milliliter (BU/mL).

B. Hemophilia B with Inhibitors
   Indefinite authorization may be granted for treatment of hemophilia B with inhibitors (see Appendix) when the inhibitor titer is ≥ 5 Bethesda units per milliliter (BU/mL).

C. Acquired Hemophilia A
   Indefinite authorization may be granted for treatment of acquired hemophilia A.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

IV. APPENDIX

Appendix: Inhibitors - Bethesda Units (BU)
The presence of inhibitors is confirmed by a specific blood test called the Bethesda inhibitor assay.

- High-titer inhibitors:
  - ≥ 5 BU/mL
  - Inhibitors act strongly and quickly neutralize factor
- Low-titer inhibitors:
  - < 5 BU/mL
  - Inhibitors act weakly and slowly neutralize factor

V. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

GAZYVA (obinutuzumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Chronic Lymphocytic Leukemia (CLL)
      Gazyva, in combination with chlorambucil, is indicated for the treatment of patients with previously untreated CLL.
   2. Follicular Lymphoma
      a. Gazyva, in combination with bendamustine followed by Gazyva monotherapy, is indicated for the treatment of patients with follicular lymphoma who relapsed after, or are refractory to, a rituximab-containing regimen.
      b. Gazyva, in combination with chemotherapy followed by Gazyva monotherapy in patients achieving at least a partial remission, is indicated for the treatment of adult patients with previously untreated stage II bulky, III or IV follicular lymphoma.

B. Compendial Uses
   1. Chronic lymphocytic leukemia, relapsed or refractory disease
   2. Small lymphocytic lymphoma (SLL) (managed in the same manner as CLL)
   3. Gastric MALT lymphoma, recurrent or progressive disease
   4. Non-gastric MALT lymphoma, refractory or progressive disease
   5. Nodal and splenic marginal zone lymphoma, refractory or progressive disease
   6. Primary cutaneous B-cell lymphomas: primary cutaneous marginal zone or follicle center lymphoma

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Chronic Lymphocytic Leukemia (CLL) and Small Lymphocytic Lymphoma (SLL)
   Authorization of 12 months may be granted for the treatment of CD20-positive CLL/SLL.

B. Follicular Lymphoma
   Authorization of 30 months total may be granted for the treatment of CD20-positive follicular lymphoma.

C. Gastric MALT Lymphoma, Non-gastric MALT Lymphoma, Nodal and Splenic Marginal Zone Lymphoma
   Authorization of 30 months total may be granted for the treatment of recurrent, refractory, or progressive CD20-positive gastric MALT lymphoma, non-gastric MALT lymphoma, nodal marginal zone lymphoma, or splenic marginal zone lymphoma.
D. Primary Cutaneous Marginal Zone or Follicle Center Lymphoma

Authorization of 30 months total may be granted for the treatment of CD20-positive primary cutaneous marginal zone or follicle center lymphoma.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

HERCEPTIN (trastuzumab)
OGIVRI (trastuzumab-dkst)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications

1. Adjuvant breast cancer
   Treatment of human epidermal growth factor receptor 2 (HER2)-overexpressing node positive or node negative (estrogen receptor (ER)/progesterone receptor (PR) negative or with one high risk feature) breast cancer:
   a. As part of a treatment regimen consisting of doxorubicin, cyclophosphamide, and either paclitaxel or docetaxel
   b. As part of a treatment regimen with docetaxel and carboplatin
   c. As a single agent following multi-modality anthracycline based therapy

2. Metastatic breast cancer
   a. In combination with paclitaxel for first-line treatment of HER2-overexpressing metastatic breast cancer
   b. As a single agent for treatment of HER2-overexpressing breast cancer in patients who have received one or more chemotherapy regimens for metastatic disease

3. Metastatic gastric or gastroesophageal junction cancer
   In combination with cisplatin and capecitabine or 5-fluorouracil, for the treatment of patients with HER2-overexpressing metastatic gastric or gastroesophageal junction adenocarcinoma, who have not received prior treatment for metastatic disease.

B. Compendial Uses

1. HER2-positive breast cancer
   a. Neoadjuvant therapy
   b. Treatment of recurrent disease

2. Leptomeningeal metastases from HER2-positive breast cancer

3. HER2-positive esophageal and esophagogastric cancer

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Breast Cancer

1. Authorization of 6 months may be granted for neoadjuvant treatment of HER2-positive breast cancer.

2. Authorization of up to 12 months total may be granted for adjuvant treatment of HER2-positive breast cancer.

3. Authorization of 12 months may be granted for treatment of HER2-positive metastatic or recurrent breast cancer.

4. Authorization of 12 months may be granted for treatment of leptomeningeal metastases from HER2-positive breast cancer.
B. **Esophageal, Gastric, or Gastroesophageal Junction Cancer**\(^1-3,5\)

Authorization of 12 months may be granted for treatment of HER2-positive esophageal, gastric, or gastroesophageal junction cancer.

III. **CONTINUATION OF THERAPY**

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. **REFERENCES**

SPECIALTY GUIDELINE MANAGEMENT

Subcutaneous Immune Globulin (SCIG):
Hizentra®, HyQvia® and Cuvitru™

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications

A. Cuvitru (Immune Globulin Subcutaneous [Human], 20% Solution)
   Cuvitru is indicated as replacement therapy for primary humoral immunodeficiency (PI) in adult and pediatric patients two years of age and older.

B. Hizentra (Immune Globulin Subcutaneous [Human], 20% Liquid)
   1. Hizentra is indicated for the treatment of primary immunodeficiency in adults and pediatric patients 2 years of age and older.
   2. Hizentra is indicated for the treatment of adult patients with chronic inflammatory demyelinating polyneuropathy (CIDP) as maintenance therapy to prevent relapse of neuromuscular disability and impairment.

   Limitations of Use:
   Hizentra maintenance therapy in CIDP has been systematically studied for 6 months and for a further 12 months in a follow-up study. Maintenance therapy beyond these periods should be individualized based upon the patient’s response and need for continued therapy.

C. HyQvia (Immune Globulin Infusion 10% [Human] with Recombinant Human Hyaluronidase)
   HyQvia is indicated for the treatment of primary immunodeficiency in adults.

   Limitation of use: Safety and efficacy of chronic use of recombinant human hyaluronidase in HyQvia have not been established in conditions other than primary immunodeficiency.

All other indications are considered experimental/investigational and are not a covered benefit.

II. REQUIRED DOCUMENTATION

The following information is necessary to initiate the prior authorization review (for primary immunodeficiency only):

A. Diagnostic test results (when applicable)
   1. Copy of laboratory report with serum immunoglobulin levels: IgG, IgA, IgM, and IgG subclasses
   2. Vaccine response to pneumococcal polysaccharide vaccine (post- vaccination Streptococcus pneumoniae antibody titers)
III. CRITERIA FOR INITIAL APPROVAL

A. Primary Immunodeficiency

Initial authorization of 12 months may be granted for members with any of the following diagnoses:

1. Severe combined immunodeficiency (SCID) or congenital agammaglobulinemia (eg, X-linked or autosomal recessive agammaglobulinemia):
   a. Diagnosis confirmed by genetic or molecular testing, or
   b. Pretreatment IgG level < 200 mg/dL, or
   c. Absence or very low number of T cells (CD3 T cells < 300/microliter) or the presence of maternal T cells in the circulation (SCID only)

2. Wiskott-Aldrich syndrome, DiGeorge syndrome, or ataxia-telangiectasia (or other non-SCID combined immunodeficiency):
   a. Diagnosis confirmed by genetic or molecular testing (if applicable), and
   b. History of recurrent bacterial infections (eg, pneumonia, otitis media, sinusitis, sepsis, gastrointestinal), and
   c. Impaired antibody response to pneumococcal polysaccharide vaccine (see Appendix)

3. Common variable immunodeficiency (CVID):
   a. Age 4 years or older
   b. Other causes of immune deficiency have been excluded (eg, drug induced, genetic disorders, infectious diseases such as HIV, malignancy)
   c. Pretreatment IgG level < 500 mg/dL or ≥ 2 SD below the mean for age
   d. History of recurrent bacterial infections
   e. Impaired antibody response to pneumococcal polysaccharide vaccine (see Appendix)

4. Hypogammaglobulinemia (unspecified), IgG subclass deficiency, selective IgA deficiency, selective IgM deficiency, or specific antibody deficiency:
   a. History of recurrent bacterial infections
   b. Impaired antibody response to pneumococcal polysaccharide vaccine (see Appendix)
   c. Any of the following pre-treatment laboratory findings:
      i. Hypogammaglobulinemia: IgG < 500 mg/dL or ≥ 2 SD below the mean for age
      ii. Selective IgA deficiency: IgA level < 7 mg/dL with normal IgG and IgM levels
      iii. Selective IgM deficiency: IgM level < 30 mg/dL with normal IgG and IgA levels
      iv. IgG subclass deficiency: IgG1, IgG2, or IgG3 ≥ 2 SD below mean for age assessed on at least 2 occasions; normal IgG (total) and IgM levels, normal/low IgA levels
      v. Specific antibody deficiency: normal IgG, IgA and IgM levels

5. Other predominant antibody deficiency disorders must meet a., b., and c.i. in section 4. above.

6. Other combined immunodeficiency must meet criteria in section 2. above.

B. Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) (Hizentra only)

Initial authorization of 3 months may be granted for the maintenance treatment of CIDP in members currently receiving intravenous immune globulin (IVIG) therapy.
IV. CONTINUATION OF THERAPY

The following criteria apply to members who are currently receiving SCIG therapy through a paid pharmacy or medical benefit. All other members (including new members) must meet initial authorization criteria.

A. Primary Immunodeficiency
Authorization of 24 months may be granted when the following criteria are met:
1. A reduction in the frequency of bacterial infections has been demonstrated since initiation of SCIG therapy, AND
2. IgG trough levels are monitored at least yearly and maintained at or above the lower range of normal for age (when applicable for indication), OR
3. The prescriber will re-evaluate the dose of SCIG and consider a dose adjustment (when appropriate).

B. Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) (Hizentra only)
Authorization of 24 months may be granted when the following criteria are met:
1. Maintenance of response from previous IVIG therapy
2. SCIG is being used at the lowest effective dose

V. APPENDIX

Impaired Antibody Response to Pneumococcal Polysaccharide Vaccine:
- Age 2 years and older: impaired antibody response demonstrated to vaccination with a pneumococcal polysaccharide vaccine
- Not established for children less than 2 years of age
- Excludes the therapy initiated in the hospital setting

VI. REFERENCES

Medical Department Procedure Manual

Section: Chapter 7A Prescription Medication Prior Authorization

Title: corticotropin (H.P. Acthar® Gel)

Purpose:

To provide guidelines and criteria for the review and decision determination of requests for medications that requires prior authorization.

Implementation Information:

1.0 Under the supervision of the Clinical Pharmacy Management (CPM) Director, the CPM staff is responsible for the development of guidelines and criteria for use by the Medical Department:

   1.1 Medical Department staff has access to the Medical Department Procedure Manual and receives notice from management when procedures are developed, updated and/or revised, or archived.

2.0 Staff utilizing this procedure is monitored, as indicated, via individual departmental audit process(es).

3.0 Medical Department staff has access to the Medical Department Procedure Manual and receives notice from management when procedures are developed, revised, or archived:

Background Information:

Reference Statement

- Guidelines are compiled from available US Food and Drug Administration (FDA) approved indications, general practice guidelines, and/or evidence-based uses established through phase III clinical studies without published conflicting data. Only clinical studies published in their entirety in reputable peer-reviewed journals will be evaluated.
Medical Department Procedure Manual

Section: Chapter 7A Prescription Medication Prior Authorization            Number: 07.161

Title: corticotropin (H.P. Acthar® Gel)                              Page 2 of 5

Background Information, continued:

Medication Summary

- Corticotropin is a parental preparation of adrenocorticotropic hormone (ACTH).
- Corticotropin is a hormone naturally secreted by cells in the anterior lobe of the pituitary gland.
- Corticotropin is available as H.P Acthar® Gel 80units/ml Repository Injection. Corticotropin repository gel is contraindicated for intravenous administration.
- BSA is calculated using the following formula: \( \sqrt{\frac{\text{weight (kg)} \times \text{height (cm)}}{3600}} \).
- AvMed considers repository corticotropin (H.P. Acthar® Gel) medically necessary for West syndrome (infantile spasms).

Additional Information

- AvMed’s Clinical Pharmacists are licensed by the State of Florida.
- AvMed’s Medical Directors are Board Certified physicians licensed by the State of Florida.

Coverage Guidelines

- Member must be eligible and have applicable benefit coverage.
- Prior authorization requests that do not meet clinical criteria in this Procedure will be forwarded to a Clinical Pharmacist for review.
Background Information, continued:

Exclusion Criteria

- Members with primary adrenal insufficiency or hypercortisolism.
- Members with pre-existing osteoporosis.
- Members with peptic ulcer disease.
- Members with scleroderma.
- Members who have had recent surgery.
- Members with a history of hypersensitivity to corticotropin therapy.
- Members with systemic fungal infection or ocular herpes infection.
- Very young pediatric Members with a suspected congenital infection.
- Members with porcine protein hypersensitivity.
- AvMed considers repository corticotropin not medically necessary for diagnostic testing of adrenocortical function because it has not been shown to be superior to cosyntropin for this purpose.
- AvMed considers repository corticotropin not medically necessary for corticosteroid-responsive conditions because it has not been proven to be more effective than corticosteroids for these indications.
- AvMed considers repository corticotropin experimental and investigational for all other indications because its effectiveness for these indications has not been established.
Medical Department Procedure Manual

Section: Chapter 7A Prescription Medication Prior Authorization Number: 07.161
Title: corticotropin (H.P. Acthar® Gel) Page 4 of 5

Procedure:

1.0 Request for initial therapy with corticotropin (H.P. Acthar®) for infantile spasms (West Syndrome) requires documentation from the Member’s medical records maintained by the requesting independent practitioner verifying the following:

1.1 Prescriber must be board certified neurologist or epileptologist; AND

1.2 Member must be less than two (2) years of age; AND

1.3 Definitive diagnosis as evidenced by hypsarrhythmia made by electroencephalogram (EEG);

1.4 If criteria are met, H.P. Acthar® Gel may be approved for four (4) weeks treatment total at the recommended dose of 150 units/m² daily intramuscularly in two (2) evenly divided doses for two (2) weeks and then gradually tapered and discontinued over a two (2) week period (suggested tapering schedule 30U/m² in the morning for 3 days; 15U/m² in the morning for 3 days; 10U/m² in the morning for 3 days; and 10U/m² every other morning for 6 days):

1.4.1 H.P. Acthar® Gel is typically dosed based on body surface area (BSA) for infantile spasms. To calculate body surface area (BSA), see http://www-users.med.cornell.edu/~spon/picu/calc/bsacalc.htm.

References:


Title: corticotropin (H.P. Acthar® Gel)

References, continued:


SPECIALTY GUIDELINE MANAGEMENT

IMFINZI (durvalumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication
A. Locally advanced or metastatic urothelial carcinoma in patients with disease progression during or following platinum-containing chemotherapy or with disease progression within 12 months of neoadjuvant or adjuvant treatment with platinum-containing chemotherapy.
B. Unresectable, stage III non-small cell lung cancer (NSCLC) whose disease has not progressed following concurrent platinum-based chemotherapy and radiation therapy

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Urothelial carcinoma
Authorization of 12 months may be granted for treatment of locally advanced or metastatic urothelial carcinoma when any of the following criteria is met:
1. Member experienced disease progression during or following platinum-containing chemotherapy.
2. Member experienced disease progression within 12 months of neoadjuvant or adjuvant treatment with platinum-containing chemotherapy.

B. Non-small cell lung cancer
Authorization of up to 12 months may be granted for treatment of unresectable, stage III NSCLC following concurrent platinum-based chemotherapy and radiation therapy.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

PRIOR AUTHORIZATION CRITERIA

BRAND NAME
IMLYGIC
(Generic) (talimogene laherparepvec)

Status: CVS Caremark Criteria
Type: Initial Prior Authorization

FDA-APPROVED INDICATION
Imlygic is indicated for the local treatment of unresectable cutaneous, subcutaneous, and nodal lesions in patients with melanoma recurrent after initial surgery.

CRITERIA FOR APPROVAL

<table>
<thead>
<tr>
<th>1</th>
<th>Does the patient have a diagnosis of melanoma?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
<td>No</td>
</tr>
</tbody>
</table>

Guidelines for Approval

Duration of Approval: 12 months

Set 1: Melanoma
Yes to question(s): No to question(s)
1: None

Mapping Instructions

<table>
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<tr>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Approve, 12 months</td>
<td>Deny</td>
</tr>
</tbody>
</table>

RATIONALE
The intent of the criteria is to ensure that patients follow selection elements noted in labeling and/or practice guidelines in order to decrease the potential for inappropriate utilization.

REFERENCES

DOCUMENT HISTORY
Written: Specialty Clinical Development (HY) 09/2016 (converted SGM criteria)
Revised: IP 03/2017, JL 12/2017
Reviewed: CDPR/ ME 05/2016; SAD 03/2017
External Review: 06/2016, 05/2017
SPECIALTY GUIDELINE MANAGEMENT

INTRON A (interferon alfa-2b)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Malignant melanoma
   2. Condylomata acuminata
   3. Hairy cell leukemia
   4. AIDS-related Kaposi's sarcoma
   5. Chronic hepatitis B virus infection
   6. Chronic hepatitis C virus infection
   7. Follicular non-Hodgkin's lymphoma

B. Compendial Uses
   1. Non-Hodgkin's lymphoma
      a. Adult T-cell leukemia/lymphoma (ATLL)
      b. Mycosis fungoides (MF)/Sezary syndrome (SS)
   2. Polycythemia vera
   3. Renal cell carcinoma
   4. Chronic myelogenous leukemia (CML)
   5. Giant cell tumor of the bone
   6. Acute hepatitis C virus infection
   7. Desmoid tumors (soft tissue sarcoma)
   8. Myeloproliferative neoplasms

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Malignant melanoma
   Authorization of 12 months may be granted for treatment of malignant melanoma.

B. Non-Hodgkin's lymphoma
   Authorization of 12 months may be granted for treatment of NHL with any of the following subtypes:
   1. Adult T-cell leukemia/lymphoma (ATLL)
   2. Mycosis fungoides (MF)/Sezary syndrome (SS)
   3. Hairy cell leukemia
   4. Follicular lymphoma (clinically aggressive)
C. **Polycythemia vera**
   Authorization of 12 months may be granted for treatment of polycythemia vera.

D. **Renal cell carcinoma**
   Authorization of 12 months may be granted for treatment of renal cell carcinoma.

E. **Condylomata acuminata**
   Authorization of 12 months may be granted for treatment of condylomata acuminata.

F. **AIDS-related Kaposi’s sarcoma**
   Authorization of 12 months may be granted for treatment of AIDS-related Kaposi’s sarcoma.

G. **Chronic myelogenous leukemia (CML)**
   Authorization of 12 months may be granted for treatment of CML.

H. **Giant cell tumor of the bone**
   Authorization of 12 months may be granted for treatment of giant cell tumor of the bone.

I. **Desmoid tumors (soft tissue sarcoma)**
   Authorization of 12 months may be granted for treatment of desmoid tumors.

J. **Acute and chronic hepatitis C virus infection**
   Authorization of up to 48 weeks may be granted for treatment of acute and chronic hepatitis C virus infection.

K. **Chronic hepatitis B (including hepatitis D virus co-infection) virus infection**
   Authorization of 48 weeks may be granted for treatment of chronic hepatitis B (including hepatitis D virus co-infection) virus infection.

L. **Myeloproliferative neoplasms**
   Authorization of 12 months may be granted for treatment of symptomatic low-risk myelofibrosis.

III. **CONTINUATION OF THERAPY**

   All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. **REFERENCES**

SPECIALTY GUIDELINE MANAGEMENT

Intravenous Immune Globulin (IVIG):
Bivigam®, Carimune® NF, Flebogamma® DIF, Gammagard® Liquid, Gammagard® S/D, Gammaked™, Gammaplex®, Gamunex®-C, Octagam®, and Privigen®

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Primary immunodeficiency
   2. Idiopathic thrombocytopenic purpura (ITP)
   3. Chronic inflammatory demyelinating polyneuropathy
   4. Multifocal motor neuropathy
   5. Kawasaki syndrome
   6. B-cell chronic lymphocytic leukemia (CLL)

B. Compendial Uses
   1. Prophylaxis of bacterial infections in pediatric human immunodeficiency virus (HIV) infection
   2. Prophylaxis of bacterial infections in bone marrow transplant (BMT)/hematopoietic stem cell transplant (HSCT) recipients
   3. Dermatomyositis
   4. Polymyositis
   5. Myasthenia gravis
   6. Guillain-Barre syndrome
   7. Lambert-Eaton myasthenic syndrome
   8. Fetal/neonatal alloimmune thrombocytopenia
   9. Parvovirus B19-induced pure red cell aplasia
   10. Stiff-person syndrome

   All other indications are considered experimental/investigational and are not a covered benefit.

II. REQUIRED DOCUMENTATION

The following information is necessary to initiate the prior authorization review:

A. Primary immunodeficiency
   1. Diagnostic test results (when applicable)
      a. Copy of laboratory report with serum immunoglobulin levels: IgG, IgA, IgM, and IgG subclasses
      b. Vaccine response to pneumococcal polysaccharide vaccine (post-vaccination Streptococcus pneumoniae antibody titers)
      c. Pertinent genetic or molecular testing in members with a known genetic disorder
      d. Copy of laboratory report with lymphocyte subset enumeration by flow cytometry
   2. IgG trough level for those continuing with IVIG therapy
B. Secondary hypogammaglobulinemia (CLL, HIV, BMT/HSCT recipients)
   1. Copy of laboratory report with pre-treatment serum IgG level (when applicable)
C. Chronic inflammatory demyelinating polyneuropathy (CIDP) and multifocal motor neuropathy (MMN)
   1. Pre-treatment electrodiagnostic studies (electromyography [EMG] or nerve conduction studies [NCS])
   2. For CIDP, pre-treatment cerebrospinal fluid (CSF) analysis (when available)
D. Dermatomyositis and polymyositis
   1. Pre-treatment electrodiagnostic studies (EMG/NCS)
   2. Pre-treatment muscle biopsy report (when available)

III. CRITERIA FOR INITIAL APPROVAL

A. Primary Immunodeficiency
   Initial authorization of 12 months may be granted for members with any of the following diagnoses:
   1. Severe combined immunodeficiency (SCID) or congenital agammaglobulinemia (eg, X-linked or autosomal recessive agammaglobulinemia):
      a. Diagnosis confirmed by genetic or molecular testing, or
      b. Pretreatment IgG level < 200 mg/dL, or
      c. Absence or very low number of T cells (CD3 T cells < 300/microliter) or the presence of maternal T cells in the circulation (SCID only)
   2. Wiskott-Aldrich syndrome, DiGeorge syndrome, or ataxia-telangiectasia (or other non-SCID combined immunodeficiency):
      a. Diagnosis confirmed by genetic or molecular testing (if applicable), and
      b. History of recurrent bacterial infections (eg, pneumonia, otitis media, sinusitis, sepsis, gastrointestinal), and
      c. Impaired antibody response to pneumococcal polysaccharide vaccine (see Appendix A)
   3. Common variable immunodeficiency (CVID):
      a. Age 4 years or older
      b. Other causes of immune deficiency have been excluded (eg, drug induced, genetic disorders, infectious diseases such as HIV, malignancy)
      c. Pretreatment IgG level < 500 mg/dL or ≥ 2 SD below the mean for age
      d. History of recurrent bacterial infections
      e. Impaired antibody response to pneumococcal polysaccharide vaccine (see Appendix A)
   4. Hypogammaglobulinemia (unspecified), IgG subclass deficiency, selective IgA deficiency, selective IgM deficiency, or specific antibody deficiency:
      a. History of recurrent bacterial infections
      b. Impaired antibody response to pneumococcal polysaccharide vaccine (see Appendix A)
      c. Any of the following pre-treatment laboratory findings:
         i. Hypogammaglobulinemia: IgG < 500 mg/dL or ≥ 2 SD below the mean for age
         ii. Selective IgA deficiency: IgA level < 7 mg/dL with normal IgG and IgM levels
         iii. Selective IgM deficiency: IgM level < 30 mg/dL with normal IgG and IgA levels
         iv. IgG subclass deficiency: IgG1, IgG2, or IgG3 ≥ 2 SD below mean for age assessed on at least 2 occasions; normal IgG (total) and IgM levels, normal/low IgA levels
         v. Specific antibody deficiency: normal IgG, IgA and IgM levels
   5. Other predominant antibody deficiency disorders must meet a., b., and c.i. in section 4. above.
   6. Other combined immunodeficiency must meet criteria in section 2. above.

Re-authorization of 24 months may be granted when the following criteria are met:
   1. A reduction in the frequency of bacterial infections has been demonstrated since initiation of IVIG therapy, AND
2. IgG trough levels are monitored at least yearly and maintained at or above the lower range of normal for age (when applicable for indication), OR
3. The prescriber will re-evaluate the dose of IVIG and consider a dose adjustment (when appropriate).

Gammagard Liquid, Gamunex-C, and Gammaked may be administered intravenously or subcutaneously for primary immunodeficiency.

B. Myasthenia Gravis
1. Authorization of 1 month may be granted to members who are prescribed IVIG for worsening weakness, acute exacerbation, or in preparation for surgery.
   a. Worsening weakness includes an increase in any of the following symptoms: diplopia, ptosis, blurred vision, difficulty speaking (dysarthria), difficulty swallowing (dysphagia), difficulty chewing, impaired respiratory status, fatigue, and limb weakness. Acute exacerbations include more severe swallowing difficulties and/or respiratory failure
   b. Pre-operative management (eg, prior to thymectomy)
2. Authorization of 3 months may be granted to members with refractory myasthenia gravis who have tried and failed 2 or more of standard therapies (eg, corticosteroids, azathioprine, cyclosporine, mycophenolate mofetil, rituximab).

C. Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)
1. Initial authorization of 3 months may be granted when the following criteria are met:
   a. Moderate to severe functional disability
   b. The diagnosis was confirmed by electrodiagnostic studies and the evaluation of cerebrospinal fluid (CSF)
2. Re-authorization of 24 months may be granted when the following criteria are met:
   a. Significant improvement in disability and maintenance of improvement since initiation of IVIG therapy
   b. IVIG is being used at the lowest effective dose and frequency

D. Dermatomyositis or Polymyositis
1. Initial authorization of 3 months may be granted when the following criteria are met:
   a. Diagnosis established by clinical features (eg, proximal weakness, rash), elevated muscle enzyme levels, electrodiagnostic studies, and muscle biopsy (when available); supportive diagnostic tests include autoantibody testing and muscle imaging (eg, MRI), and
   b. Standard first-line treatments (corticosteroids or immunosuppressants) have been tried but were unsuccessful or not tolerated, or
   c. Member is unable to receive standard first-line therapy because of a contraindication or other clinical reason.
2. Re-authorization of 12 months may be granted when the following criterion is met:
   a. Significant improvement in disability and maintenance of improvement since initiation of IVIG therapy

E. Idiopathic Thrombocytopenic Purpura (Immune Thrombocytopenia)
1. Newly diagnosed ITP (diagnosed within the past 3 months) or initial therapy: authorization of 1 month may be granted when the following criteria are met:
   a. Children (< 18 years of age)
      i. Significant bleeding symptoms (mucosal bleeding or other moderate/severe bleeding) or
      ii. High risk for bleeding* (see Appendix B), or
      iii. Rapid increase in platelets is required* (eg, surgery or procedure)
   b. Adults (≥ 18 years of age)
      i. Platelet count < 30,000/mcL, or

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*Indicates conditions requiring urgent medical attention.
ii. Platelet count < 50,000/mcL and significant bleeding symptoms, high risk for bleeding or rapid increase in platelets is required*, and
iii. Corticosteroid therapy is contraindicated and IVIG will be used alone or IVIG will be used in combination with corticosteroid therapy

2. Chronic/persistent ITP (≥ 3 months from diagnosis) or ITP unresponsive to first-line therapy: authorization of 6 months may be granted when the following criteria are met:
   a. Platelet count < 30,000/mcL, or
   b. Platelet count < 50,000/mcL and significant bleeding symptoms, high risk for bleeding or rapid increase in platelets is required*, and
   c. Relapse after previous response to IVIG or inadequate response/intolerance/contraindication to corticosteroid or anti-D therapy

3. Adults with refractory ITP after splenectomy: authorization of 6 months may be granted when either of the following criteria is met:
   a. Platelet count < 30,000/mcL, or
   b. Significant bleeding symptoms

4. ITP in pregnant women: authorization through delivery may be granted to pregnant women with ITP.

   * The member’s risk factor(s) for bleeding (see Appendix B) or reason requiring a rapid increase in platelets must be provided.

F. B-cell Chronic Lymphocytic Leukemia (CLL)

1. Initial authorization of 6 months may be granted when the following criteria are met:
   a. IVIG is prescribed for prophylaxis of bacterial infections.
   b. Member has a history of recurrent sinopulmonary infections requiring intravenous antibiotics or hospitalization.
   c. Member has a pretreatment serum IgG level <500 mg/dL.

2. Re-authorization of 6 months may be granted when the following criterion is met:
   a. A reduction in the frequency of bacterial infections has been demonstrated since initiation of IVIG therapy.

G. Prophylaxis of Bacterial Infections in HIV-Infected Pediatric Patients

1. Initial authorization of 6 months may be granted to pediatric members with HIV infection when the following criteria are met:
   a. Member is ≤ 12 years of age.
   b. IVIG is prescribed for primary prophylaxis of bacterial infections and pretreatment serum IgG < 400 mg/dL, or
   c. IVIG is prescribed for secondary prophylaxis of bacterial infections for members with a history of recurrent bacterial infections (> 2 serious bacterial infections in a 1-year period)

2. Re-authorization of 6 months may be granted when the following criterion is met:
   a. A reduction in the frequency of bacterial infections has been demonstrated since initiation of IVIG therapy.

H. Prophylaxis of Bacterial Infections in BMT/HSCT Recipients

1. Initial authorization of 6 months may be granted to members who are BMT/HSCT recipients when the following criteria are met:
   a. IVIG is prescribed for prophylaxis of bacterial infections.
   b. Either of the following:
      i. IVIG is requested within the first 100 days post-transplant.
      ii. Member has a pretreatment serum IgG < 400 mg/dL.

2. Re-authorization of 6 months may be granted when the following criterion is met:
   a. A reduction in the frequency of bacterial infections has been demonstrated since initiation of IVIG therapy.
I. Multifocal Motor Neuropathy (MMN)
   1. Initial authorization of 3 months may be granted when the following criteria are met:
      a. Weakness without objective sensory loss in 2 or more nerves
      b. The diagnosis was confirmed by electrodiagnostic studies
   2. Re-authorization of 24 months may be granted when the following criterion is met:
      a. Significant improvement in disability and maintenance of improvement since initiation of IVIG therapy

J. Guillain-Barre Syndrome (GBS)
   Authorization of 2 months total may be granted for the treatment of GBS.

K. Lambert-Eaton Myasthenic Syndrome (LEMS)
   Authorization of 24 months may be granted for LEMS.

L. Kawasaki Syndrome
   Authorization of 1 month may be granted for pediatric members with Kawasaki syndrome.

M. Fetal/Neonatal Alloimmune Thrombocytopenia (F/NAIT)
   Authorization of 6 months may be granted for treatment of F/NAIT.

N. Parvovirus B19-induced Pure Red Cell Aplasia (PRCA)
   Authorization of 6 months may be granted for parvovirus B19-induced PRCA.

O. Stiff-person Syndrome
   Authorization of 6 months may be granted for treatment of stiff-person syndrome.

IV. CONTINUATION OF THERAPY

Reauthorization criteria apply to members who are currently receiving IVIG therapy through a paid pharmacy or medical benefit. All other members (including new members) must meet initial authorization criteria.

V. OTHER

When Gammagard Liquid, Gamunex-C and Gammaked will be administered subcutaneously, they may be approved for primary immunodeficiency only.

VI. APPENDICES

Appendix A: Impaired Antibody Response to Pneumococcal Polysaccharide Vaccine
- Age 2 years and older: impaired antibody response demonstrated to vaccination with a pneumococcal polysaccharide vaccine
- Not established for children less than 2 years of age
- Excludes the therapy initiated in the hospital setting

Appendix B: Examples of Risk Factors for Bleeding (not all inclusive)
- Undergoing a medical or dental procedure where blood loss is anticipated
- Comorbidity (e.g., peptic ulcer disease, hypertension)
- Mandated anticoagulation therapy
• Profession or lifestyle predisposes patient to trauma (eg, construction worker, fireman, professional athlete)

VII. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

JEVTANA (cabazitaxel)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication

Jevtana is indicated in combination with prednisone for the treatment of patients with hormone-refractory metastatic prostate cancer previously treated with a docetaxel-containing treatment regimen.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Authorization of 24 months may be granted for the treatment of metastatic prostate cancer.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

Kadcyla (ado-trastuzumab emtansine)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications

Kadcyla, as a single agent, is indicated for the treatment of patients with human epidermal growth factor receptor 2 (HER2)-positive, metastatic breast cancer who previously received trastuzumab and a taxane, separately or in combination. Patients should have either received prior therapy for metastatic disease, or developed disease recurrence during or within six months of completing adjuvant therapy.

B. Compendial Use

1. Recurrent HER2-positive breast cancer
2. Non-small cell lung cancer with HER2 mutations

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Breast cancer

Authorization of 12 months may be granted for treatment of HER2-positive breast cancer.

B. Non-small cell lung cancer

Authorization of 12 months may be granted for treatment of lung cancer with HER2 mutations.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

KANUMA (sebelipase alfa)

POLICY

I. INDICATIONS
The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
Kanuma is indicated for the treatment of patients with a diagnosis of Lysosomal Acid Lipase (LAL) deficiency.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL
Lysosomal acid lipase (LAL) deficiency
Indefinite authorization may be granted for treatment of LAL deficiency when the diagnosis of LAL deficiency was confirmed by enzyme assay demonstrating a deficiency of lysosomal acid lipase enzyme activity or by genetic testing.

III. CONTINUATION OF THERAPY
All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

KEYTRUDA (pembrolizumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications

1. Melanoma
   Keytruda is indicated for the treatment of patients with unresectable or metastatic melanoma.

2. Non-Small Cell Lung Cancer
   - Keytruda, as a single agent, is indicated for the first-line treatment of patients with metastatic non-small cell lung cancer (NSCLC) whose tumors have high PD-L1 expression [Tumor Proportion Score (TPS) ≥50%] as determined by an FDA-approved test, with no EGFR or ALK genomic tumor aberrations.
   - Keytruda, as a single agent, is indicated for the treatment of patients with metastatic NSCLC whose tumors express PD-L1 (TPS ≥1%) as determined by an FDA approved test, with disease progression on or after platinum-containing chemotherapy. Patients with EGFR or ALK genomic tumor aberrations should have disease progression on FDA-approved therapy for these aberrations prior to receiving Keytruda.
   - Keytruda, in combination with pemetrexed and carboplatin, is indicated for the first-line treatment of patients with metastatic nonsquamous NSCLC.

3. Head and Neck Cancer
   Keytruda is indicated for the treatment of patients with recurrent or metastatic head and neck squamous cell carcinoma (HNSCC) with disease progression on or after platinum-containing chemotherapy.

4. Classical Hodgkin Lymphoma
   Keytruda is indicated for the treatment of adult and pediatric patients with refractory classical Hodgkin lymphoma (cHL), or who have relapsed after three or more prior lines of therapy.

5. Urothelial Carcinoma
   Keytruda is indicated for the treatment of patients with locally advanced or metastatic urothelial carcinoma who:
   - Are not eligible for cisplatin-containing chemotherapy, or
   - Have disease progression during or following platinum-containing chemotherapy or within 12 months of neoadjuvant or adjuvant treatment with platinum-containing chemotherapy.

6. Microsatellite Instability-High Cancer
   Keytruda is indicated for the treatment of adult and pediatric patients with unresectable or metastatic, microsatellite instability-high (MSI-H) or mismatch repair deficient solid tumors that have progressed following prior treatment and who have no satisfactory alternative treatment options, or
ii. Colorectal cancer that has progressed following treatment with a fluoropyrimidine, oxaliplatin, and irinotecan.

Limitation of Use: The safety and effectiveness of Keytruda in pediatric patients with MSI-H central nervous system cancers have not been established.

7. Gastric Carcinoma

Keytruda is indicated for the treatment of patients with recurrent, locally advanced, metastatic gastric or gastroesophageal junction adenocarcinoma whose tumors express PD-L1 [Combined Positive Score (CPS) ≥ 1] as determined by an FDA approved test, with disease progression on or after two or more prior lines of therapy including fluoropyrimidine and platinum containing chemotherapy and if appropriate, HER2/neu targeted therapy.

B. Compendial Uses

1. Non-small cell lung cancer
2. Unresectable advanced or metastatic microsatellite instability-high colorectal cancer
3. Malignant pleural mesothelioma
4. Merkel cell carcinoma

All other indications are considered experimental/investigational and are not a covered benefit.

II. EXCLUSIONS

Coverage will not be provided for pediatric patients with microsatellite instability-high (MSI-H) central nervous system cancers.

III. CRITERIA FOR INITIAL APPROVAL

A. Melanoma

Authorization of 12 months may be granted for treatment of unresectable or metastatic melanoma.

B. Non-small cell lung cancer (NSCLC)

Authorization of 12 months may be granted for treatment of metastatic NSCLC in either of the following settings:

1. First-line treatment
   i. The tumor has high PD-L1 expression [Tumor Proportion Score (TPS) ≥50%] and EGFR, ALK, or ROS1 genomic tumor markers are negative or unknown, OR
   ii. The patient has nonsquamous NSCLC and Keytruda will be used in combination with pemetrexed and carboplatin.

2. Subsequent therapy
   i. The patient’s tumor is positive for the PD-L1 protein, AND
   ii. Keytruda is requested for disease progression on a first-line cytotoxic regimen or for further progression on other systemic therapy.

C. Head and Neck Cancer

Authorization of 12 months may be granted for the treatment of patients with recurrent or metastatic head and neck squamous cell carcinoma (HNSCC) with disease progression on or after platinum-containing chemotherapy.
D. Classical Hodgkin Lymphoma
Authorization of 12 months may be granted for treatment of refractory or relapsed classical Hodgkin lymphoma.

E. Urothelial carcinoma
Authorization of 12 months may be granted for treatment of locally advanced or metastatic urothelial carcinoma when any of the following criteria is met:
1. Patient is not eligible for cisplatin-containing chemotherapy.
2. Patient experienced disease progression during or following platinum-containing chemotherapy.
3. Patient experienced disease progression within 12 months of neoadjuvant or adjuvant treatment with platinum-containing chemotherapy.

F. Microsatellite Instability-High Cancer
Authorization of 12 months may be granted for treatment of unresectable or metastatic microsatellite instability-high (MSI-H) or mismatch repair deficient solid tumors when either of the following criteria are met:
1. The patient has colorectal cancer
2. For other solid tumors: Member experienced disease progression following prior treatment and has no satisfactory alternative treatment options.

G. Malignant Pleural Mesothelioma
Authorization 12 months may be granted for treatment of malignant pleural mesothelioma.

H. Merkel Cell Carcinoma
Authorization of 12 months may be granted for treatment of Merkel cell carcinoma.

I. Gastric Carcinoma
Authorization of 12 months may be granted for treatment of recurrent locally advanced, metastatic gastric or gastroesophageal junction adenocarcinoma when all of the following criteria are met:
1. Tumor expresses PD-L1 [Combined Positive Score (CPS) greater than equal to 1].
2. Patient experienced disease progression on or after two or more prior lines of therapy including fluoropyrimidine- and platinum-containing chemotherapy.
3. If HER2 positive, patient received HER2/neu-targeted therapy.

IV. CONTINUATION OF THERAPY
All patients (including new patients) requesting authorization for continuation of therapy must meet all initial authorization criteria.

V. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

KRISTEXXA (pegloticase)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication
   Krystexxa is a PEGylated uric acid specific enzyme indicated for the treatment of chronic gout in adult patients refractory to conventional therapy.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Chronic gout
Authorization of 12 months may be granted for members with a diagnosis of chronic gout when ALL of the following criteria are met:
A. Krystexxa will NOT be used concomitantly with oral urate-lowering therapies
B. Member has had an inadequate response to or a clinical reason for not completing at least a three-month trial (see Appendix) with ALL of the following medications at the medically appropriate maximum doses:
   1. Allopurinol
   2. Febuxostat
   3. Probenecid (alone or in combination with allopurinol or febuxostat)

III. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for all members (including new members) with a diagnosis of chronic gout that meet ALL initial authorization criteria and have NOT had two consecutive uric acid levels above 6 mg/dL since starting treatment with Krystexxa.

IV. APPENDIX: Clinical reasons for not completing a three-month trial with allopurinol, febuxostat, and probenecid (examples):
A. Member experienced a severe allergic reaction to the medication
B. Member experienced toxicity with the medication
C. Member could not tolerate the medication
D. Member’s current medication regimen has a significant drug interaction
E. Member has severe renal dysfunction (allopurinol)
F. Member has known blood dyscrasias or uric acid kidney stones (probenecid)
G. Member has renal insufficiency (i.e., glomerular filtration rate 30 mL/minute or less) (probenecid)
V. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

KYPROLIS (carfilzomib)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications

1. In combination with dexamethasone or with lenalidomide plus dexamethasone for the treatment of patients with relapsed multiple myeloma who have received one to three prior lines of therapy.
2. As a single agent for the treatment of patients with relapsed or refractory multiple myeloma who have received one or more prior lines of therapy.

B. Compendial Uses

Waldenström’s macroglobulinemia/lymphoplasmacytic lymphoma

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Multiple myeloma

Authorization of 12 months may be granted for treatment of multiple myeloma.

B. Waldenström’s macroglobulinemia/lymphoplasmacytic lymphoma

Authorization of 12 months may be granted for treatment of Waldenström’s macroglobulinemia/lymphoplasmacytic lymphoma.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

LARTRUVO (olaratumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication
   Lartruvo is indicated, in combination with doxorubicin, for the treatment of adult patients with soft tissue sarcoma (STS) with a histologic subtype for which an anthracycline-containing regimen is appropriate and which is not amenable to curative treatment with radiotherapy or surgery.

   This indication is approved under accelerated approval. Continued approval for this indication may be contingent upon verification and description of clinical benefit in the confirmatory trial.

B. Compendial Use
   Lartruvo is indicated, in combination with doxorubicin, for the treatment of adult patients with uterine sarcoma.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Soft Tissue Sarcoma
   Authorization of 12 months may be granted for treatment of soft tissue sarcoma (STS) when Lartruvo is used in combination with doxorubicin.

B. Uterine Sarcoma
   Authorization of 12 months may be granted for the treatment of uterine sarcoma when Lartruvo is used in combination with doxorubicin.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

LEMTRADA (alemtuzumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered covered benefits provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

**FDA-Approved Indication:** Lemtrada is indicated for the treatment of patients with relapsing forms of multiple sclerosis (MS). Because of its safety profile, the use of Lemtrada should generally be reserved for patients who have had an inadequate response to two or more drugs indicated for the treatment of MS.

All other indications are considered experimental/investigational and are not covered benefits.

II. CRITERIA FOR APPROVAL

A. **First Course – Relapsing forms of multiple sclerosis**
   Authorization of 30 days (5 doses) may be granted to members with a diagnosis of a relapsing form of multiple sclerosis who have had an inadequate response to two or more drugs indicated for multiple sclerosis.

B. **Second Course – Relapsing forms of multiple sclerosis**
   Authorization of 30 days (3 doses) may be granted to members with a diagnosis of a relapsing form of multiple sclerosis who have completed one previous course of therapy.

III. REFERENCE

SPECIALTY GUIDELINE MANAGEMENT

LEUKINE (sargramostim)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Use Following Induction Chemotherapy in Acute Myelogenous Leukemia
      a. Leukine is indicated for use following induction chemotherapy in older adult patients with acute myelogenous leukemia to shorten time to neutrophil recovery and to reduce the incidence of severe and life-threatening infections and infections resulting in death.
   2. Use in Mobilization and Following Transplantation of Autologous Peripheral Blood Progenitor Cells
      a. Leukine is indicated for the mobilization of hematopoietic progenitor cells into peripheral blood for collection by leukapheresis. Mobilization allows for the collection of increased numbers of progenitor cells capable of engraftment as compared with collection without mobilization. After myeloablative chemotherapy, the transplantation of an increased number of progenitor cells can lead to more rapid engraftment, which may result in a decreased need for supportive care. Myeloid reconstitution is further accelerated by administration of Leukine following peripheral blood progenitor cell transplantation.
   3. Use in Myeloid Reconstitution After Autologous Bone Marrow Transplantation
      a. Leukine is indicated for acceleration of myeloid recovery in patients with non-Hodgkin's lymphoma (NHL), acute lymphoblastic leukemia (ALL) and Hodgkin's disease undergoing autologous bone marrow transplantation (BMT).
   4. Use in Myeloid Reconstitution After Allogeneic Bone Marrow Transplantation
      a. Leukine is indicated for acceleration of myeloid recovery in patients undergoing allogeneic BMT from HLA-matched related donors.
   5. Use in Bone Marrow Transplantation Failure or Engraftment Delay
      a. Leukine is indicated in patients who have undergone allogeneic or autologous BMT in whom engraftment is delayed or has failed.

B. Compendial Uses
   1. Prophylaxis and treatment of chemotherapy-induced febrile neutropenia in non-myeloid malignancies
   2. Treatment of neutropenia in patients with myelodysplastic syndromes (MDS)
   3. AML following consolidation chemotherapy
   4. ALL following induction or consolidation chemotherapy
   5. Agranulocytosis
   6. Aplastic anemia
   7. Neutropenia related to HIV/AIDS
   8. Stem cell transplantation-related indications

All other indications are considered experimental/investigational and are not a covered benefit.
II. CRITERIA FOR INITIAL APPROVAL

A. Neutropenia in cancer patients receiving myelosuppressive chemotherapy
   Authorization of 6 months may be granted for prevention or treatment of febrile neutropenia when both of the following criteria are met:
   a. Member has a non-myeloid malignancy and has received, is currently receiving, or will be receiving myelosuppressive anti-cancer therapy
   b. Leukine will not be administered less than 24 hours before or after chemotherapy or radiotherapy

B. Other indications
   Authorization of 6 months may be granted for members with any of the following indications:
   1. Agranulocytosis
   2. Aplastic anemia
   3. Neutropenia related to HIV/AIDS
   4. Acute myeloid leukemia
   5. Myelodysplastic syndrome
   6. Stem cell transplantation-related indications

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

MYOBLOC (rimabotulinumtoxinB)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication

Cervical dystonia in adults to reduce the severity of abnormal head position and neck pain associated with cervical dystonia

All other indications are considered experimental/investigational and are not a covered benefit.

II. EXCLUSIONS

Coverage will not be provided for cosmetic use.

III. CRITERIA FOR INITIAL APPROVAL

Authorization of 24 months may be granted for treatment of cervical dystonia (e.g., torticollis).

IV. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

V. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

NEULASTA (pegfilgrastim)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication

Neulasta is indicated to decrease the incidence of infection, as manifested by febrile neutropenia, in patients with non-myeloid malignancies receiving myelosuppressive anti-cancer drugs associated with a clinically significant incidence of febrile neutropenia.

B. Compendial Use

Stem cell transplantation-related indications

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Prevention of neutropenia in cancer patients receiving myelosuppressive chemotherapy

Authorization of 6 months may be granted for prevention of febrile neutropenia when both of the following criteria are met:

1. Member has a non-myeloid malignancy and is currently receiving, or will be receiving myelosuppressive anti-cancer therapy
2. Neulasta will not be administered less than 24 hours before or after chemotherapy or radiotherapy

B. Stem cell transplantation-related indications

Authorization of 6 months may be granted for stem cell transplantation-related indications.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES


SPECIALTY GUIDELINE MANAGEMENT

NEUPOGEN (filgrastim)
GRANIX (tbo-filgrastim)
ZARXIO (filgrastim-sndz)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications

**Neupogen**

1. Patients with Cancer Receiving Myelosuppressive Chemotherapy
   Neupogen is indicated to decrease the incidence of infection, as manifested by febrile neutropenia, in patients with non-myeloid malignancies receiving myelosuppressive anti-cancer drugs associated with a significant incidence of severe neutropenia with fever.

2. Patients With Acute Myeloid Leukemia Receiving Induction or Consolidation Chemotherapy
   Neupogen is indicated for reducing the time to neutrophil recovery and the duration of fever, following induction or consolidation chemotherapy treatment of adults with acute myeloid leukemia.

3. Patients with Cancer Receiving Bone Marrow Transplant
   Neupogen is indicated to reduce the duration of neutropenia and neutropenia-related clinical sequelae, (e.g., febrile neutropenia) in patients with non-myeloid malignancies undergoing myeloablative chemotherapy followed by marrow transplantation.

4. Patients Undergoing Autologous Peripheral Blood Progenitor Cell Collection and Therapy
   Neupogen is indicated for the mobilization of autologous hematopoietic progenitor cells into the peripheral blood for collection by leukapheresis.

5. Patients With Severe Chronic Neutropenia
   Neupogen is indicated for chronic administration to reduce the incidence and duration of sequelae of neutropenia (e.g., fever, infections, oropharyngeal ulcers) in symptomatic patients with congenital neutropenia, cyclic neutropenia, or idiopathic neutropenia.

**Granix**

Granix is indicated to reduce the duration of severe neutropenia in patients with non-myeloid malignancies receiving myelosuppressive anti-cancer drugs associated with a clinically significant incidence of febrile neutropenia.

**Zarxio**

1. Patients with Cancer Receiving Myelosuppressive Chemotherapy
a. Zarxio is indicated to decrease the incidence of infection, as manifested by febrile neutropenia, in patients with non-myeloid malignancies receiving myelosuppressive anti-cancer drugs associated with a significant incidence of severe neutropenia with fever.

2. Patients With Acute Myeloid Leukemia Receiving Induction or Consolidation Chemotherapy
   a. Zarxio is indicated for reducing the time to neutrophil recovery and the duration of fever, following induction or consolidation chemotherapy treatment of adults with acute myeloid leukemia.

3. Patients with Cancer Undergoing Bone Marrow Transplant
   a. Zarxio is indicated to reduce the duration of neutropenia and neutropenia-related clinical sequelae, (e.g., febrile neutropenia) in patients with non-myeloid malignancies undergoing myeloablative chemotherapy followed by marrow transplantation.

4. Patients Undergoing Autologous Peripheral Blood Progenitor Cell Collection and Therapy
   a. Zarxio is indicated for the mobilization of autologous hematopoietic progenitor cells into the peripheral blood for collection by leukapheresis.

5. Patients With Severe Chronic Neutropenia
   a. Zarxio is indicated for chronic administration to reduce the incidence and duration of sequelae of neutropenia (e.g., fever, infections, oropharyngeal ulcers) in symptomatic patients with congenital neutropenia, cyclic neutropenia, or idiopathic neutropenia.

B. Compendial Uses (Neupogen/Granix/Zarxio)
   1. Treatment of chemotherapy-induced febrile neutropenia in patients with non-myeloid malignancies
   2. Treatment of anemia in patients with myelodysplastic syndromes (MDS)
   3. Treatment of neutropenia in patients with MDS
   4. Following chemotherapy for acute lymphocytic leukemia (ALL)
   5. Stem cell transplantation-related indications
   6. Agranulocytosis
   7. Aplastic anemia
   8. Neutropenia related to HIV/AIDS
   9. Neutropenia related to renal transplantation

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Neutropenia in cancer patients receiving myelosuppressive chemotherapy
   Authorization of 6 months may be granted for prevention or treatment of febrile neutropenia when both of the following criteria are met:
   1. Member has a non-myeloid malignancy and has received, is currently receiving, or will be receiving myelosuppressive anti-cancer therapy
   2. Neupogen/Granix/Zarxio will not be administered less than 24 hours before or after chemotherapy or radiotherapy

B. Other indications
   Authorization of 6 months may be granted for members with any of the following indications:
   1. Agranulocytosis
   2. Aplastic anemia
   3. Neutropenia related to HIV/AIDS
   4. Neutropenia related to renal transplantation
   5. Acute myeloid leukemia
   6. Stem cell transplantation-related indications
   7. Severe chronic neutropenia (congenital, cyclic, or idiopathic)
8. Myelodysplastic syndrome (anemia or neutropenia)

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

NPLATE (romiplostim)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication
   Treatment of thrombocytopenia in patients with chronic immune thrombocytopenia (ITP) who have had an insufficient response to corticosteroids, immunoglobulins, or splenectomy

B. Compendial Use
   Cyclic thrombocytopenia

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Chronic or persistent primary immune thrombocytopenia (ITP)
   Authorization of 6 months may be granted to members with chronic or persistent ITP who meet all of the following criteria:
   1. Inadequate response or intolerance to prior therapy with corticosteroids, immunoglobulins, or splenectomy
   2. Untransfused platelet count at time of diagnosis is less than $30 \times 10^9/L$ OR $30 \times 10^9/L$ to $50 \times 10^9/L$ with symptomatic bleeding (e.g., significant mucous membrane bleeding, gastrointestinal bleeding or trauma) or risk factors for bleeding (see Section IV).

B. Cyclic thrombocytopenia
   Authorization of 12 months may be granted to members who are prescribed Nplate for the treatment of cyclic thrombocytopenia.

III. CONTINUATION OF THERAPY

Chronic or persistent ITP
A. Authorization of 12 months may be granted to members with current platelet count less than or equal to $200 \times 10^9/L$.
B. Authorization of 12 months may be granted to members with current platelet count greater than $200 \times 10^9/L$ for whom Nplate dosing will be adjusted to achieve a platelet count sufficient to avoid clinically important bleeding.

IV. APPENDIX
Examples of risk factors for bleeding (not all inclusive)
- Undergoing a medical or dental procedure where blood loss is anticipated
- Comorbidity (e.g., peptic ulcer disease, hypertension)
- Mandated anticoagulation therapy
- Profession (e.g., construction worker) or lifestyle (e.g., plays contact sports) that predisposes patient to trauma

V. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

NUCALA (mepolizumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications

1. Maintenance Treatment of Severe Asthma
   Nucala is an interleukin-5 antagonist monoclonal antibody (IgG1 kappa) indicated for add-on maintenance treatment of patients with severe asthma aged 12 years and older, and with an eosinophilic phenotype.

   Limitations of Use: Not for relief of acute bronchospasm or status asthmaticus

2. Eosinophilic Granulomatosis with Polyangiitis
   Nucala is indicated for the treatment of adult patients with eosinophilic granulomatosis with polyangiitis (EGPA).

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Severe asthma with an eosinophilic phenotype
   Authorization of 12 months may be granted for treatment of severe asthma with an eosinophilic phenotype when all of the following criteria are met:
   1. Member is 12 years of age or older.
   2. Member has a baseline eosinophil count of at least 150 cells per microliter.
   3. Member has a history of severe asthma despite current treatment with both of the following medications at optimized doses:
      1. Inhaled corticosteroid
      2. Additional controller (long acting beta2-agonist, leukotriene modifier, or sustained-release theophylline)

B. Eosinophilic Granulomatosis with Polyangiitis
   Authorization of 12 months may be granted for treatment of eosinophilic granulomatosis with polyangiitis when all of the following criteria are met:
   1. Member is 18 years of age or older.
   2. Member has a history or the presence of an eosinophil count of more than 1000 cells per microliter or a blood eosinophil level of greater than 10%.
III. CONTINUATION OF THERAPY

A. Severe asthma with an eosinophilic phenotype
   Authorization of 12 months may be granted for continuation of treatment of severe asthma with an eosinophilic phenotype when all of the following criteria are met:
   1. Member is 12 years of age or older.
   2. Asthma control has improved on Nucala treatment as demonstrated by either:
      a. A reduction in the frequency or severity of symptoms and exacerbations, or
      b. A reduction in the daily maintenance oral corticosteroid dose

B. Eosinophilic Granulomatosis with Polyangiitis
   Authorization of 12 months may be granted for continuation of treatment of eosinophilic granulomatosis with polyangiitis when all of the following criteria are met:
   1. Member is 18 years of age or older.
   2. Member has beneficial response to treatment with Nucala as demonstrated by any of the following:
      a. A reduction in the frequency of relapses, or
      b. A reduction in the daily oral corticosteroid dose, or
      c. No active vasculitis

IV. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

OBIZUR (antihemophilic factor [recombinant], porcine sequence)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication
Obizur is indicated for the treatment of bleeding episodes in adults with acquired hemophilia A.

Limitations of Use:
A. Safety and efficacy of Obizur has not been established in patients with a baseline anti-porcine factor VIII inhibitor titer of greater than 20 BU.
B. Obizur is not indicated for the treatment of congenital hemophilia A or von Willebrand disease

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Acquired hemophilia A
Authorization of 1 month may be granted for treatment of acquired hemophilia A.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

IV. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

OCREVUS (ocrelizumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered covered benefits provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications:

Ocrevus is indicated for the treatment of adult patients with relapsing or primary progressive forms of multiple sclerosis (MS).

All other indications are considered experimental/investigational and are not covered benefits.

II. PRESCRIBER SPECIALTIES

This medication must be prescribed by or in consultation with a neurologist.

III. CRITERIA FOR INITIAL APPROVAL

A. Relapsing Forms of Multiple Sclerosis

Authorization of 24 months may be granted to members who are 18 years of age or older for the treatment of relapsing forms of MS when at least ONE of the following criteria is met:

1. The member is newly diagnosed with MS.
2. The member is new to treatment with disease modifying therapy.
3. For members who have previously received or are currently receiving disease modifying therapy: The member’s disease is not currently stabilized on existing disease modifying therapy as evidenced by disease worsening or occurrence of an intolerable adverse event.

B. Primary Progressive Multiple Sclerosis

Authorization of 24 months may be granted to members who are 18 years of age or older for the treatment of primary progressive MS.

IV. CONTINUATION OF THERAPY

A. Relapsing Forms of Multiple Sclerosis

Authorization of 24 months may be granted to members requesting continuation of therapy for the treatment of relapsing forms of MS when the member has experienced disease improvement or slowing of
disease worsening (eg, decrease in the number of relapses, improvement or no decline in Kurtzke Expanded Disability Status Scale [EDSS] or in MRI findings) since initiating Ocrevus therapy.

B. Primary Progressive Multiple Sclerosis

Authorization of 24 months may be granted to members requesting continuation of therapy for the treatment of primary progressive MS when the member has experienced slowing of disease worsening (eg, no decline in EDSS or MRI findings) since initiating Ocrevus therapy.

V. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

SANDOSTATIN (octreotide acetate injection)
SANDOSTATIN LAR DEPOT (octreotide acetate for injectable suspension)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. octreotide acetate/Sandostatin:
      a. Indicated to reduce blood levels of growth hormone and IGF-1 (somatomedin C) in acromegaly patients who have had inadequate response to or cannot be treated with surgical resection, pituitary irradiation, and bromocriptine mesylate at maximally tolerated doses.
      b. Indicated for the symptomatic treatment of patients with metastatic carcinoid tumors where it suppresses or inhibits the severe diarrhea and flushing episodes associated with the disease.
      c. Indicated for the treatment of the profuse watery diarrhea associated with vasoactive intestinal peptide (VIP)-secreting tumors.

2. Sandostatin LAR: Sandostatin LAR Depot is indicated in patients in whom initial treatment with Sandostatin injection has been shown to be effective and tolerated.
   a. Indicated for long-term maintenance therapy in acromegalic patients who have had an inadequate response to surgery and/or radiotherapy, or for whom surgery and/or radiotherapy is not an option.
   b. Indicated for long-term treatment of the severe diarrhea and flushing episodes associated with metastatic carcinoid tumors.
   c. Indicated for long-term treatment of the profuse watery diarrhea associated with vasoactive intestinal peptide (VIP)-secreting tumors.

B. Compendial Uses
   1. Neuroendocrine tumors (NETs):
      a. Adrenal gland tumors
      b. Tumors of the gastrointestinal (GI) tract, lung, and thymus (carcinoid tumors)
      c. Tumors of the pancreas
   2. Meningiomas
   3. Thymomas and thymic carcinomas
   4. Congenital hyperinsulinism (CHI)/persistent hyperinsulinemic hypoglycemia of infancy (PHHI) (octreotide and Sandostatin only)

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Acromegaly
   Authorization of 24 months may be granted for the treatment of acromegaly when all of the following criteria are met:
1. Member has a high pretreatment insulin-like growth factor-1 (IGF-1) level for age and/or gender based on the laboratory reference range.
2. Member had an inadequate or partial response to surgery or radiotherapy OR there is a clinical reason why the member has not had surgery or radiotherapy

B. Neuroendocrine tumors (NETs)/carcinoid syndrome
1. Tumors of the gastrointestinal (GI) tract (carcinoid tumor)
   Authorization of 24 months may be granted for treatment of metastatic or unresectable NETs of the GI tract.
2. Tumors of the thymus (carcinoid tumor)
   Authorization of 24 months may be granted for treatment of metastatic or unresectable NETs of the thymus.
3. Tumors of the lung (carcinoid tumor)
   Authorization of 24 months may be granted for treatment of metastatic or unresectable NETs of the lung.
4. Tumors of the pancreas
   Authorization of 24 months may be granted for treatment of NETs of the pancreas.
5. Tumors of the adrenal gland
   Authorization of 24 months may be granted for treatment of NETs of the adrenal gland.

C. Meningiomas
   Authorization of 24 months may be granted to members for treatment of unresectable meningioma.

D. Thymomas and thymic carcinomas
   Authorization of 24 months may be granted for treatment of thymomas and thymic carcinomas.

E. Congenital hyperinsulinism (CHI)/persistent hyperinsulinemic hypoglycemia of infancy (octreotide and Sandostatin only)
   Authorization of 6 months may be granted for treatment of CHI and persistent hyperinsulinemic hypoglycemia in an infant.

III. CONTINUATION OF THERAPY

A. Acromegaly
   Authorization of 24 months may be granted for continuation of therapy for acromegaly when the member's IGF-1 level has decreased or normalized since initiation of therapy.

B. All other indications
   Members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES


SPECIALTY GUIDELINE MANAGEMENT

OPDIVO (nivolumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications

1. Unresectable or metastatic melanoma
   i. As a single agent for the treatment of patients with BRAF V600 wild-type unresectable or metastatic melanoma.
   ii. As a single agent for the treatment of patients with BRAF V600 mutation-positive unresectable or metastatic melanoma.
   iii. In combination with ipilimumab for the treatment of patients with unresectable or metastatic melanoma.

2. Adjuvant treatment of melanoma
   Opdivo is indicated for the adjuvant treatment of melanoma with lymph node involvement or metastatic disease who have undergone complete resection

3. Metastatic non-small cell lung cancer (NSCLC)
   Opdivo is indicated for the treatment of patients with metastatic NSCLC with progression on or after platinum-based chemotherapy. Patients with EGFR or ALK genomic tumor aberrations should have disease progression on FDA-approved therapy for these aberrations prior to receiving Opdivo.

4. Renal cell carcinoma (RCC)
   Opdivo is indicated for the treatment of patients with advanced RCC who have received prior anti-angiogenic therapy.

5. Classical Hodgkin lymphoma (cHL)
   Opdivo is indicated for the treatment of patients with cHL that has relapsed or progressed after:
   i. Autologous hematopoietic stem cell transplantation (HSCT) and brentuximab vedotin
   ii. 3 or more lines of therapy that includes autologous HSCT

6. Squamous Cell Carcinoma of the Head and Neck
   Opdivo is indicated for the treatment of patients with recurrent or metastatic squamous cell carcinoma of the head and neck (SCCHN) with disease progression on or after platinum-based therapy.

7. Urothelial Carcinoma
   Opdivo is indicated for the treatment of patients with locally advanced or metastatic urothelial carcinoma who:
   i. Have disease progression during or following platinum-containing chemotherapy
   ii. Have disease progression within 12 months of neoadjuvant or adjuvant treatment with platinum-containing chemotherapy.
8. Colorectal Cancer
Opdivo is indicated for adult and pediatric (12 years and older) patients with microsatellite instability-high or mismatch repair deficient metastatic colorectal cancer that has progressed following treatment with a fluoropyrimidine, oxaliplatin, and irinotecan.

9. Hepatocellular Carcinoma
Opdivo is indicated for the treatment of hepatocellular carcinoma who have been previously treated with sorafenib.

B. Compendial Uses
1. Classical Hodgkin lymphoma
2. Renal cell carcinoma
3. Malignant pleural mesothelioma
4. NSCLC
5. Small cell lung cancer

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Unresectable or metastatic melanoma
Authorization of 12 months may be granted for the treatment of unresectable or metastatic melanoma.

B. Adjuvant treatment of melanoma
Authorization of 12 months may be granted for the adjuvant treatment of melanoma with lymph node involvement or metastatic disease who have undergone complete resection

C. Non-small cell lung cancer (NSCLC)
Authorization of 12 months may be granted for treatment of metastatic NSCLC when Opdivo is requested for disease progression on or after a first-line cytotoxic regimen or for further progression on other systemic therapy.

D. Renal cell carcinoma
Authorization of 12 months may be granted for treatment of advanced, relapsed or unresectable renal cell carcinoma.

E. Classical Hodgkin lymphoma (cHL)
Authorization of 12 months may be granted for treatment of cHL.

F. Squamous cell carcinoma of the head and neck (SCCHN)
Authorization of 12 months may be granted for treatment of recurrent or metastatic SCCHN in members with disease progression on or after platinum-based therapy.

G. Urothelial carcinoma
Authorization of 12 months may be granted for treatment of locally advanced or metastatic urothelial carcinoma when any of the following criteria is met:
1. Member experienced disease progression during or following platinum-containing chemotherapy.
2. Member experienced disease progression within 12 months of neoadjuvant or adjuvant treatment with platinum-containing chemotherapy.

H. Colorectal cancer
Authorization of 12 months may be granted for treatment of unresectable locally advanced or metastatic colorectal cancer with defective mismatch repair or high microsatellite instability.

I. Malignant pleural mesothelioma
   Authorization of 12 months may be granted for treatment of malignant pleural mesothelioma

J. Small cell lung cancer
   Authorization of 12 months may be granted for treatment of small cell lung cancer.

K. Hepatocellular carcinoma
   Authorization of 12 months may be granted for treatment of hepatocellular carcinoma for members who have been previously treated with sorafenib.

III. CONTINUATION OF THERAPY

   All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

RADICAVA (edaravone)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
Radicava is indicated for the treatment of amyotrophic lateral sclerosis (ALS).

All other indications are considered experimental/investigational and are not a covered benefit.

II. PRESCRIBER SPECIALTIES

This medication must be prescribed by or in consultation with a neuromuscular specialist.

III. CRITERIA FOR INITIAL APPROVAL

Authorization of 6 months may be granted for treatment of ALS when all of the following criteria are met:
A. Diagnosis of definite or probable ALS
B. Duration of ALS is 2 years or less
C. Functional ability is retained for most activities of daily living (ADLs)
D. Ventilatory support, noninvasive or invasive, is not required

IV. CONTINUATION OF THERAPY

Authorization of 6 months may be granted for members continuing with Radicava therapy when the following criteria are met:
A. Diagnosis of definite or probable ALS
B. There is a clinical benefit from Radicava therapy such as stabilization of functional ability and maintenance of ADLs
C. Invasive ventilation is not required

V. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

Remodulin (treprostinil injection)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
1. **Pulmonary Arterial Hypertension**
   Remodulin is indicated for the treatment of pulmonary arterial hypertension (PAH) (WHO Group 1) to diminish symptoms associated with exercise.

2. **Pulmonary Arterial Hypertension in Patients Requiring Transition from Flolan**
   In patients with PAH requiring transition from Flolan (epoprostenol sodium), Remodulin is indicated to diminish the rate of clinical deterioration. The risks and benefits of each drug should be carefully considered prior to transition.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Indefinite authorization may be granted for treatment of PAH when ALL of the following criteria are met:

A. Member has PAH defined as WHO Group 1 class of pulmonary hypertension (refer to Appendix).

B. PAH was confirmed by either criterion (1) or criterion (2) below:
   1. Pretreatment right heart catheterization with all of the following results:
      - mPAP ≥ 25 mmHg
      - PCWP ≤ 15 mmHg
      - PVR > 3 Wood units
   2. For infants less than one year of age with any of the following conditions, PAH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed:
      - Post cardiac surgery
      - Chronic heart disease
      - Chronic lung disease associated with prematurity
      - Congenital diaphragmatic hernia

III. CONTINUATION OF THERAPY

Indefinite authorization may be granted for members with PAH who are currently receiving Remodulin therapy through a paid pharmacy or medical benefit.

IV. APPENDIX

**WHO Classification of Pulmonary Hypertension**
WHO Group 1. Pulmonary Arterial Hypertension (PAH)
1.1 Idiopathic (IPAH)
1.2 Heritable PAH
   1.2.1 Germline mutations in the bone morphogenetic protein receptor type 2 (BMPR2)
   1.2.2 Activin receptor-like kinase type 1 (ALK1), endoglin (with or without hereditary hemorrhagic telangiectasia), Smad 9, caveolin-1 (CAV1), potassium channel super family K member-3 (KCNK3)
   1.2.3 Unknown

1.3 Drug- and toxin-induced

1.4. Associated with:
   1.4.1 Connective tissue diseases
   1.4.2 HIV infection
   1.4.3 Portal hypertension
   1.4.4 Congenital heart diseases
   1.4.5 Schistosomiasis

1'. Pulmonary veno-occlusive disease (PVOD) and/or pulmonary capillary hemangiomatosis (PCH)

1". Persistent pulmonary hypertension of the newborn (PPHN)

WHO Group 2. Pulmonary Hypertension Owing to Left Heart Disease

2.1 Systolic dysfunction
2.2 Diastolic dysfunction
2.3 Valvular disease
2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies

WHO Group 3. Pulmonary Hypertension Owing to Lung Disease and/or Hypoxia

3.1 Chronic obstructive pulmonary disease
3.2 Interstitial lung disease
3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
3.4 Sleep-disordered breathing
3.5 Alveolar hypoventilation disorders
3.6 Chronic exposure to high altitude
3.7 Developmental abnormalities

WHO Group 4. Chronic Thromboembolic Pulmonary Hypertension (CTEPH)

WHO Group 5. Pulmonary Hypertension with Unclear Multifactorial Mechanisms

5.1 Hematologic disorders: Chronic hemolytic anemia, myeloproliferative disorders, splenectomy
5.2 Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis: lymphangioleiomyomatosis, neurofibromatosis, vasculitis
5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
5.4 Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure on dialysis, segmental PH

V. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

RITUXAN (rituximab)
Treatment of Hematologic and Oncologic Conditions

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications

1. Non-Hodgkin’s Lymphoma (NHL) in patients with:
   a. Relapsed or refractory, low-grade or follicular, CD20-positive, B-cell NHL as a single agent
   b. Previously untreated follicular, CD20-positive, B-cell NHL in combination with first line chemotherapy and, in patients achieving a complete or partial response to Rituxan in combination with chemotherapy, as single-agent maintenance therapy
   c. Non-progressing (including stable disease), low-grade, CD20-positive, B-cell NHL, as a single agent after first-line CVP (cyclophosphamide, vincristine, and prednisone) chemotherapy
   d. Previously untreated diffuse large B-cell, CD20-positive NHL in combination with cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) or other anthracycline-based chemotherapy regimens

2. Chronic Lymphocytic Leukemia (CLL), in combination with fludarabine and cyclophosphamide (FC), for the treatment of patients with previously untreated and previously treated CD20-positive CLL.

3. Granulomatosis with polyangiitis (Wegener’s Granulomatosis) and Microscopic Polyangiitis (MPA) (Not addressed in this policy – Refer to Rituxan-RA SGM)

4. Rheumatoid Arthritis (Not addressed in this policy – Refer to Rituxan-RA SGM)

B. Compendial Uses

1. Sjögren’s syndrome (Not addressed in this policy – Refer to Rituxan-RA SGM)

2. Multiple sclerosis (Not addressed in this policy – Refer to Rituxan-RA SGM)

3. Non-Hodgkin’s lymphoma
   a. Small lymphocytic lymphoma (SLL)
   b. Mantle cell lymphoma
   c. Marginal zone lymphomas (nodal, splenic, MALT)
   d. Burkitt lymphoma
   e. Primary cutaneous B-cell lymphoma
   f. Castleman’s disease
   g. Acquired immunodeficiency syndrome (AIDS)-related B-cell lymphoma
   h. Hairy cell leukemia
   i. Post-transplant lymphoproliferative disorder (PTLD)
   j. Lymphoblastic lymphoma

4. Relapsed/refractory immune or idiopathic thrombocytopenic purpura (ITP)

5. Autoimmune hemolytic anemia

6. Waldenström’s macroglobulinemia/lymphoplasmacytic lymphoma (LPL)

7. Thrombotic thrombocytopenic purpura

8. Myasthenia gravis, refractory

9. Hodgkin’s lymphoma, nodular lymphocyte-predominant
10. Chronic graft-versus-host disease (GVHD)
11. Central nervous system (CNS) cancers
   a. Leptomeningeal metastases from lymphomas
   b. Primary CNS lymphoma
12. Acute lymphoblastic leukemia (ALL)
13. Prevention of Epstein-Barr virus (EBV)-related PTLD in high risk patients

All other indications are considered experimental/investigational and are not a covered benefit.

CRITERIA FOR INITIAL APPROVAL

A. Oncologic indications
   Authorization of 12 months may be granted for treatment of any of the following oncologic disorders that are CD20-positive as confirmed by testing or analysis:
   1. Non-Hodgkin’s lymphoma (NHL) with any of the following subtypes:
      a. Diffuse large B-cell lymphoma
      b. Chronic lymphocytic leukemia (CLL)/small lymphocytic lymphoma (SLL)
      c. Follicular lymphoma
      d. Mantle cell lymphoma
      e. Marginal zone lymphomas (nodal, splenic, MALT)
      f. Burkitt lymphoma
      g. Primary cutaneous B-cell lymphoma
      h. Castleman’s disease
      i. AIDS-related B-cell lymphoma
      j. Hairy cell leukemia
      k. Post-transplant lymphoproliferative disorder (PTLD)
      l. Lymphoblastic lymphoma
   2. Waldenström’s macroglobulinemia/lymphoplasmacytic lymphoma (LPL)
   3. Hodgkin’s lymphoma, nodular lymphocyte-predominant
   4. Central nervous system (CNS) cancers with either of the following:
      a. Leptomeningeal metastases from lymphomas
      b. Primary CNS lymphoma
   5. Acute lymphoblastic leukemia (ALL)

B. Hematologic indications
   Authorization of 12 months may be granted for treatment of any of the following indications:
   1. Refractory immune or idiopathic thrombocytopenic purpura (ITP)
   2. Autoimmune hemolytic anemia
   3. Thrombotic thrombocytopenic purpura
   4. Chronic graft-versus-host disease (GVHD)
   5. Prevention of Epstein-Barr virus (EBV)-related PTLD

C. Myasthenia gravis
   Authorization of 12 months may be granted for treatment of refractory myasthenia gravis.

II. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.
III. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

RITUXAN (rituximab)
Treatment of Rheumatoid Arthritis

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Moderately to severely active rheumatoid arthritis (RA)
      a. In combination with methotrexate in patients who have inadequate response to one or more TNF antagonist therapies
   2. Granulomatosis with Polyangiitis (GPA) (Wegener’s Granulomatosis) and Microscopic Polyangiitis (MPA)
      a. In combination with glucocorticoids
   3. Other FDA-approved indications (not addressed in this policy – Refer to Rituxan–Oncology SGM)
      a. Non-Hodgkin’s lymphoma (NHL)
      b. Chronic lymphocytic leukemia (CLL)

B. Compendial Uses
   1. Sjögren’s syndrome
   2. Multiple sclerosis, relapsing remitting
   3. For other compendial uses, refer to Rituxan–Oncology SGM

All other indications are considered experimental/investigational and are not a covered benefit.

II. EXCLUSIONS

Coverage will not be provided for requests for the treatment of rheumatoid arthritis when planned date of administration is less than 16 weeks since date of last dose received.

III. CRITERIA FOR INITIAL APPROVAL

Prior to initiating therapy, all members must be screened for hepatitis B virus infection with serologic assays, in addition to meeting the indication-specific criteria listed below.

A. Moderately to severely active rheumatoid arthritis (RA)
   1. Authorization of 24 months may be granted to members who have previously received any biologic DMARD or targeted synthetic DMARD (e.g., Xeljanz) indicated for the treatment of moderately to severely active rheumatoid arthritis OR have received at least two full doses of Rituxan for the treatment of RA, where the most recent dose was given within 6 months of the request. Rituxan must be prescribed in combination with methotrexate (MTX) unless the member has a contraindication or intolerance to MTX (see Appendix A).
2. Authorization of 24 months may be granted for treatment of moderately to severely active RA when all of the following criteria are met:
   a. Member is prescribed Rituxan in combination with MTX or has a contraindication or intolerance to MTX.
   b. Member meets any of the following criteria:
      i. Member has experienced an inadequate response to at least a 3-month trial of MTX despite adequate dosing (i.e., titrated to 20 mg/week)
      ii. Member has an intolerance or contraindication to MTX (see Appendix A)

B. Granulomatosis with Polyangiitis (GPA) (Wegener’s Granulomatosis) and Microscopic Polyangiitis (MPA)\(^1\)
   Authorization of 12 months may be granted for treatment of GPA or MPA.

C. Sjögren’s syndrome
   Authorization of 12 months may be granted for treatment of Sjögren’s syndrome.

D. Multiple sclerosis
   Authorization of 12 months may be granted for treatment of multiple sclerosis (MS) when both of the following criteria are met:
   1. Member has a diagnosis of relapsing remitting MS
   2. Member has had an inadequate response to two or more disease-modifying drugs indicated for MS despite adequate duration of treatment (see Appendix B)

IV. CONTINUATION OF THERAPY

A. Rheumatoid Arthritis
   Authorization of 24 months may be granted for all members (including new members) who meet all initial authorization criteria and achieve or maintain positive clinical response after at least two doses of therapy with Rituximab as evidenced by low disease activity or improvement in signs and symptoms of the condition.

B. Other indications
   Authorization of 12 months may be granted for all members (including new members) who meet all initial authorization criteria.

V. APPENDICES

Appendix A: Examples of contraindications to methotrexate
1. Alcoholism, alcoholic liver disease or other chronic liver disease
2. Breastfeeding
3. Blood dyscrasias (e.g., thrombocytopenia, leukopenia, significant anemia)
4. Elevated liver transaminases
5. History of intolerance or adverse event
6. Hypersensitivity
7. Interstitial pneumonitis or clinically significant pulmonary fibrosis
8. Myelodysplasia
9. Pregnancy or planning pregnancy (male or female)
10. Renal impairment
11. Significant drug interaction
Appendix B: Disease-modifying drugs indicated for multiple sclerosis
1. Aubagio (teriflunomide)
2. Avonex (interferon beta-1a)
3. Betaseron (interferon beta-1a)
4. Copaxone/Glatopa (glatiramer acetate)
5. Extavia (interferon beta-1a)
6. Gilenya (fingolimod)
7. Tecfidera (dimethyl fumarate)
8. Plegridy (peginterferon beta-1a)
9. Rebif (interferon beta-1a)
10. Tysabri (natalizumab)

VI. REFERENCES
2. DRUGDEX® System [Internet database]. Ann Arbor, MI: Truven Health Analytics. Updated periodically.
SPECIALTY GUIDELINE MANAGEMENT

sildenafil tablets (generic)
Revatio (sildenafil tablets and oral suspension)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication
Sildenafil/Revatio is indicated for the treatment of pulmonary arterial hypertension (WHO Group I) in adults to improve exercise ability and delay clinical worsening.

B. Compendial Use
Raynaud’s phenomenon secondary to systemic sclerosis (Tablets only)

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Pulmonary Arterial Hypertension
Authorization of 24 months may be granted for treatment of PAH when ALL of the following criteria are met:
1. Member has PAH defined as WHO Group 1 class of pulmonary hypertension (refer to Appendix).
2. PAH was confirmed by either criterion (1) or criterion (2) below:
   i. Pretreatment right heart catheterization with all of the following results:
      • mPAP ≥ 25 mmHg
      • PCWP ≤ 15 mmHg
      • PVR > 3 Wood units
   ii. For infants less than one year of age with any of the following conditions, PAH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed:
      • Post cardiac surgery
      • Chronic heart disease
      • Chronic lung disease associated with prematurity
      • Congenital diaphragmatic hernia

B. Secondary Raynaud’s Phenomenon
Authorization of 24 months may be granted for treatment Raynaud’s phenomenon secondary to systemic sclerosis when the patient has had an inadequate response to one of the following medications:
• Calcium channel blockers
• Angiotensin receptor blockers
• Selective serotonin reuptake inhibitors
• Alpha blockers
• Angiotensin converting enzyme inhibitors
• Topical nitrates

III. CONTINUATION OF THERAPY

Authorization of 24 months may be granted for members with PAH or secondary Raynaud’s phenomenon who are currently receiving sildenafil/Revatio therapy through a paid pharmacy or medical benefit.

IV. APPENDIX

WHO Classification of Pulmonary Hypertension

WHO Group 1. Pulmonary Arterial Hypertension (PAH)
1.1 Idiopathic (IPAH)
1.2 Heritable PAH
   1.2.1 Germline mutations in the bone morphogenetic protein receptor type 2 (BMPR2)
   1.2.2 Activin receptor-like kinase type 1 (ALK1), endoglin (with or without hereditary hemorrhagic telangiectasia), Smad 9, caveolin-1 (CAV1), potassium channel super family K member-3 (KCNK3)
   1.2.3 Unknown
1.3 Drug- and toxin-induced
1.4. Associated with:
   1.4.1 Connective tissue diseases
   1.4.2 HIV infection
   1.4.3 Portal hypertension
   1.4.4 Congenital heart diseases
   1.4.5 Schistosomiasis
1'. Pulmonary veno-occlusive disease (PVOD) and/or pulmonary capillary hemangiomatosis (PCH)
1". Persistent pulmonary hypertension of the newborn (PPHN)

WHO Group 2. Pulmonary Hypertension Owing to Left Heart Disease
2.1 Systolic dysfunction
2.2 Diastolic dysfunction
2.3 Valvular disease
2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies

WHO Group 3. Pulmonary Hypertension Owing to Lung Disease and/or Hypoxia
3.1 Chronic obstructive pulmonary disease
3.2 Interstitial lung disease
3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
3.4 Sleep-disordered breathing
3.5 Alveolar hypoventilation disorders
3.6 Chronic exposure to high altitude
3.7 Developmental abnormalities

WHO Group 4. Chronic Thromboembolic Pulmonary Hypertension (CTEPH)

WHO Group 5. Pulmonary Hypertension with Unclear Multifactorial Mechanisms
5.1 Hematologic disorders: Chronic hemolytic anemia, myeloproliferative disorders, splenectomy
5.2 Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis: lymphangioleiomyomatosis, neurofibromatosis, vasculitis
5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
5.4 Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure on dialysis, segmental PH
V. REFERENCES

Specialty Guideline Management

SOLIRIS (eculizumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered covered benefits provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications

1. Paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis
2. Atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy
3. Generalized myasthenia gravis (gMG) patients who are anti-acetylcholine receptor (AchR) antibody positive

Limitations of Use: Soliris is not indicated for the treatment of patients with Shiga toxin E. Coli related hemolytic uremic syndrome (STEC-HUS).

All other indications are considered experimental/investigational and are not covered benefits.

II. CRITERIA FOR INITIAL APPROVAL

A. Atypical hemolytic uremic syndrome
   Authorization of 24 months may be granted to members prescribed Soliris for the treatment of atypical hemolytic uremic syndrome which is not caused by Shiga toxin.

B. Paroxysmal nocturnal hemoglobinuria
   Authorization of 24 months may be granted to members prescribed Soliris for the treatment of paroxysmal nocturnal hemoglobinuria.

C. Generalized myasthenia gravis (gMG)
   Authorization of 24 months may be granted to members for treatment of generalized myasthenia gravis (gMG) who are anti-acetylcholine receptor (AchR) antibody positive.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

SPINRAZA (nusinersen)

POLICY

I. INDICATIONS
The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
Spinraza is indicated for the treatment of spinal muscular atrophy (SMA) in pediatric and adult patients.

All other indications are considered experimental/investigational and are not a covered benefit.

II. REQUIRED DOCUMENTATION
The following information is necessary to initiate the prior authorization review: Deletion or mutation at the SMN1 allele confirmed by genetic testing.

III. PRESCRIBER SPECIALTIES
This medication must be prescribed by or in consultation with a neurologist or neuromuscular specialist.

IV. CRITERIA FOR INITIAL APPROVAL
Authorization of 4 months may be granted for treatment of SMA when all of the following criteria are met:
A. Member has a diagnosis of SMA confirmed by genetic testing showing deletion or mutation at the SMN1 allele.
B. Member has Type 1, Type 2 or Type 3 SMA.
C. The diagnosis was made at or before 18 years of age.
D. Member is not on invasive or noninvasive ventilation support for more than 6 hours a day.

V. CONTINUATION OF THERAPY
Authorization of 12 months may be granted for members (including new members) when all of the following criteria are met:
A. Member meets initial authorization criteria
B. Member is receiving a clinical benefit from Spinraza therapy, as demonstrated by improvement or maintenance of motor skills or ability to sit, crawl, stand or walk, or new motor milestones.

VI. REFERENCES


SPECIALTY GUIDELINE MANAGEMENT

Supprelin LA (histrelin acetate)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication

Supprelin LA is indicated for the treatment of children with central precocious puberty.

B. Compendial Use

Gender Dysphoria (also known as gender non-conforming or transgender persons)

NOTE: Some plans may opt-out of coverage for gender dysphoria.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Central precocious puberty (CPP)

1. Authorization up to age 12 may be granted for the treatment of CPP in a female member when ALL of the following criteria are met:
   a. The diagnosis of CPP has been confirmed by a pubertal response to a gonadotropin releasing hormone (GnRH) agonist test or a pubertal level of a third generation luteinizing hormone (LH) assay
   b. The diagnosis of CPP has been confirmed by assessment of bone age versus chronological age
   c. The member was less than 8 years of age at the onset of secondary sexual characteristics

2. Authorization up to age 13 may be granted for the treatment of CPP in a male member when ALL of the following criteria are met:
   a. The diagnosis of CPP has been confirmed by a pubertal response to a GnRH agonist test or a pubertal level of a third generation LH assay
   b. The diagnosis of CPP has been confirmed by assessment of bone age versus chronological age
   c. The member was less than 9 years of age at the onset of secondary sexual characteristics

B. Gender dysphoria

1. Authorization of 12 months may be granted for pubertal suppression in preparation for gender reassignment in an adolescent member when ALL of the following criteria are met:
   a. The member has a diagnosis of gender dysphoria
   b. The member has reached Tanner stage 2 of puberty

2. Authorization of 12 months may be granted for gender reassignment in an adult member when ALL of the following criteria are met:
   a. The member has a diagnosis of gender dysphoria
   b. The member will receive Supprelin LA concomitantly with cross sex hormones
III. CONTINUATION OF THERAPY

A. CPP
1. Authorization up to age 12 may be granted for continuation of therapy for CPP in a female member if the member is currently less than 12 years of age.
2. Authorization up to age 13 may be granted for continuation of therapy for CPP in a male member if the member is currently less than 13 years of age.

B. Gender Dysphoria
All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

IV. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

SYLVANT (siltuximab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication
   Sylvant is indicated for the treatment of patients with multicentric Castleman’s disease who are human immunodeficiency virus (HIV) negative and human herpesvirus-8 (HHV-8) negative.

B. Compendial Use
   Relapsed/refractory unicentric Castleman’s disease

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Multicentric Castleman’s disease or relapsed/refractory unicentric Castleman’s disease.
Authorization of 12 months may be granted for the treatment of multicentric Castleman’s disease or relapsed/refractory unicentric Castleman’s disease.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

SYNAGIS (palivizumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication

Synagis is indicated for the prevention of serious lower respiratory tract disease caused by respiratory syncytial virus (RSV) in children at high risk* of RSV disease.

* In the absence of a specific definition of “high risk” in the Synagis package labeling, the American Academy of Pediatrics has endeavored to provide guidance for the use of Synagis. Refer to Appendix A for summary of recommendations.

The following points should be considered when prescribing Synagis:

- Safety and efficacy were established in children with bronchopulmonary dysplasia (BPD), infants with a history of premature birth (less than or equal to 35 weeks gestational age), and children with hemodynamically significant congenital heart disease (CHD).
- The safety and efficacy of Synagis have not been established for treatment of RSV disease.

B. Compendial Uses

RSV prophylaxis in infants with congenital abnormalities of the airway or neuromuscular disease that compromise handling of respiratory secretions

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Authorization of up to 5 doses may be granted for the prevention of serious lower respiratory tract disease caused by RSV when a member has any of the following diagnoses and meets the criteria pertaining to the diagnosis:

1. Prematurity
2. CHD (See Appendix B)
3. Chronic Lung Disease (CLD) of Prematurity
4. Congenital Airway Abnormality
5. Neuromuscular Condition

A. Prematurity

All of the following criteria are met:

1. Member’s gestational age is < 28 weeks, 6 days.
2. Member’s chronological age at the start of RSV season is <12 months.

B. CHD

All of the following criteria are met:

1. CHD is hemodynamically significant.
2. Member meets either of the following criteria:
   a. Member’s chronological age at the start of RSV season is < 12 months.
b. Member’s chronological age at the start of RSV season is between 12 to 24 months and the member will be undergoing cardiac transplantation during the RSV season.

C. CLD of prematurity
ALL of the following criteria must be met:
1. Member’s gestational age is < 31 weeks, 6 days.
2. Member requires > 21% oxygen for at least the first 28 days after birth.
3. Member meets either of the following criteria:
   a. Member’s chronological age at the start of their first RSV season is < 12 months.
   b. Member’s chronological age at the start of the subsequent RSV season is < 24 months and the member continues to require medical support (e.g., chronic corticosteroids, diuretic therapy, supplemental oxygen) during the 6-month period prior to the start of the RSV season.

D. Congenital airway abnormality
ALL of the following criteria must be met:
1. The condition compromises handling of respiratory secretions.
2. Member’s chronological age at the start of RSV season is < 12 months.

E. Neuromuscular condition
ALL of the following criteria must be met:
1. The condition compromises handling of respiratory secretions.
2. Member’s chronological age at the start of RSV season is < 12 months.

III. DOSAGE AND ADMINISTRATION

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines. The following limits apply:
A. RSV season requests: maximum of 5 doses per RSV season
B. Off-season requests: 1 dose per request up to a maximum of 5 doses per RSV season

CVS Caremark PBM Synagis Season for 2016-2017 will be November 1, 2016 to March 31, 2017. Other health plans may differ.

IV. OTHER

For all off-season Synagis requests, the RSV activity for the requested region must be ≥ 10% within 2 weeks of the intended dose according to the CDC National Respiratory and Enteric Virus Surveillance System (NREVSS). The local health department or the CDC NREVSS will be consulted to assess the RSV activity for that region (http://www.cdc.gov/surveillance/nrevss/rsv/index.html). Other Specialty Guideline Management criteria apply.

V. APPENDIX (if necessary)

Appendix A: Recommended Use of Synagis for Prevention of RSV Infection
Recommendations from the American Academy of Pediatrics for the prevention of RSV infection with Synagis are summarized in Table below. Synagis should be administered intramuscularly at a dose of 15 mg/kg once per month beginning prior to the onset of the RSV season, which typically occurs in November. Because 5 monthly doses of Synagis will provide more than 6 months of serum Synagis concentrations above the desired serum concentration for most infants, administration of more than 5 monthly doses is not recommended within the continental United States.
### Table. Recommended Use of Synagis for Prevention of RSV Infection

<table>
<thead>
<tr>
<th>Category</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prematurity</td>
<td>Preterm infants born at 28 weeks, 6 days of gestation or earlier who are younger than 12 months at the start of the RSV season</td>
</tr>
<tr>
<td>Congenital Heart Disease</td>
<td>Infants and children &lt; 12 months of age with hemodynamically significant CHD</td>
</tr>
<tr>
<td></td>
<td>Those most likely to benefit from prophylaxis include:</td>
</tr>
<tr>
<td></td>
<td>o Infants with acyanotic heart disease who are receiving medication to control congestive heart failure and will require cardiac surgical procedures</td>
</tr>
<tr>
<td></td>
<td>o Infants with moderate to severe pulmonary hypertension</td>
</tr>
<tr>
<td></td>
<td>Infants and children &lt; 24 months of age who undergo cardiac transplantation during the RSV season</td>
</tr>
<tr>
<td>Chronic Lung Disease of Prematurity</td>
<td>For the first RSV season during the first year of life:</td>
</tr>
<tr>
<td></td>
<td>Preterm infants who develop CLD of prematurity defined as:</td>
</tr>
<tr>
<td></td>
<td>o Gestational age ≤ 31 weeks, 6 days AND</td>
</tr>
<tr>
<td></td>
<td>o Requirement for &gt; 21% oxygen for at least the first 28 days after birth</td>
</tr>
<tr>
<td></td>
<td>For the second RSV season during the second year of life:</td>
</tr>
<tr>
<td></td>
<td>Preterm infants who:</td>
</tr>
<tr>
<td></td>
<td>o Satisfy the above definition of CLD of prematurity AND</td>
</tr>
<tr>
<td></td>
<td>o Continue to require medical support* for CLD during the 6-month period prior to the start of the second RSV season</td>
</tr>
<tr>
<td>Congenital Abnormality of the Airway/Neuromuscular Condition</td>
<td>Infants who have either a significant congenital abnormality of the airway or a neuromuscular condition that compromises handling of respiratory secretions for the first year of life</td>
</tr>
</tbody>
</table>

Abbreviations: CHD = congenital heart disease; CLD = chronic lung disease (formerly bronchopulmonary dysplasia); RSV = respiratory syncytial virus.

* Medical support includes supplemental oxygen, diuretic therapy, or chronic corticosteroid therapy.

**Appendix B: Examples of Congenital Heart Anomalies**

- Atrial or ventricular septal defect
- Coarctation of aorta
- Tetralogy of Fallot
- Pulmonary or aortic valve stenosis
- Tricuspid atresia
- Ebstein’s anomaly
- Pulmonary atresia
- Transposition of great arteries
- Truncus arteriosus
- Hypoplastic left/right ventricle
- Single ventricle
- Double-outlet right ventricle
  Total anomalous pulmonary venous return

*Must be hemodynamically significant. See Table above for examples of infants and children who are most likely to benefit from Synagis.*
VI. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

SYNRIBO (omacetaxine mepesuccinate)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication: Treatment of adult patients with chronic or accelerated phase chronic myeloid leukemia (CML) with resistance and/or intolerance to two or more tyrosine kinase inhibitors (TKIs)

B. Compendial Use: Treatment option for posttransplant relapse CML in patients with disease progression due to resistance and/or intolerance to two or more TKIs

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Chronic myeloid leukemia (CML)

Authorization of 12 months may be granted for members prescribed Synribo for the treatment of CML who have experienced resistance, toxicity, or intolerance to prior therapy with two or more TKIs (e.g., imatinib, dasatinib, nilotinib, bosutinib, ponatinib)

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

TECENTRIQ (atezolizumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Locally advanced or metastatic urothelial carcinoma
      Tecentriq is indicated for the treatment of patients with locally advanced or metastatic urothelial carcinoma who:
      a. Are not eligible for cisplatin-containing chemotherapy, or
      b. Have disease progression during or following any platinum-containing chemotherapy, or within 12 months of neoadjuvant or adjuvant chemotherapy
   2. Metastatic non-small cell lung cancer (NSCLC)
      Tecentriq is indicated for the treatment of patients with metastatic NSCLC who have disease progression during or following platinum-containing chemotherapy. Patients with EGFR or ALK genomic tumor aberrations should have disease progression on FDA-approved therapy for these aberrations prior to receiving Tecentriq

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Urothelial carcinoma
   Authorization of 12 months may be granted for treatment of locally advanced or metastatic urothelial carcinoma when either of the following criteria is met:
   1. Member is not eligible for cisplatin-containing chemotherapy
   2. The disease has progressed during or following platinum-containing chemotherapy

B. Non-small cell lung cancer (NSCLC)
   Authorization of 12 months may be granted for treatment of metastatic NSCLC when both of the following criteria are met:
   1. The disease has progressed during or following platinum-containing chemotherapy
   2. Members with positive epidermal growth factor receptor (EGFR) mutation or positive anaplastic lymphoma kinase (ALK) gene arrangement have had disease progression on FDA-approved therapy for these aberrations (e.g., erlotinib, afatinib, gefitinib, crizotinib, ceritinib) for these mutations prior to receiving Tecentriq

III. CONTINUATION OF THERAPY
All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCE
SPECIALTY GUIDELINE MANAGEMENT

TREANDA (bendamustine)
BENDEKA (bendamustine)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Chronic lymphocytic leukemia (CLL)
   2. Indolent B-cell non-Hodgkin lymphoma (NHL) that has progressed during or within six months of treatment with rituximab or a rituximab-containing regimen

B. Compendial Uses
   1. Classical Hodgkin lymphoma (CHL)
   2. Multiple myeloma (MM)
   3. Non-Hodgkin lymphoma (NHL)
      a. Adult T-cell leukemia/lymphoma (ATLL)
      b. Acquired immune deficiency syndrome (AIDS)-related B-cell lymphoma
      c. CLL/small lymphocytic lymphoma (SLL)
      d. Diffuse large B-cell lymphoma (DLBCL)
      e. Follicular lymphoma
      f. Marginal zone lymphoma
         i. Nodal marginal zone lymphoma
         ii. Gastric mucosa associated lymphoid tissue (MALT) lymphoma
         iii. Nongastric MALT lymphoma
         iv. Splenic marginal zone lymphoma
      g. Mantle cell lymphoma (MCL)
      h. Mycosis fungoides (MF)/Sezary syndrome (SS)
      i. Peripheral T-cell lymphoma (PTCL)
      j. Primary cutaneous B-cell lymphoma
      k. Primary cutaneous CD30+ T-cell lymphoproliferative disorder
   4. Waldenström’s macroglobulinemia/lymphoplasmacytic lymphoma

All other indications are considered experimental/investigational and are not a covered benefit.
II. CRITERIA FOR INITIAL APPROVAL

A. Non-Hodgkin lymphoma (NHL)
   Authorization of 12 months may be granted for treatment of NHL with any of the following subtypes:
   1. Follicular lymphoma
   2. Chronic lymphocytic leukemia/small lymphocytic leukemia (CLL/SLL) without chromosome 17p deletion
   3. Diffuse large B-cell lymphoma (DLBCL)
   4. Adult T-cell leukemia/lymphoma (ATLL)
   5. AIDS-related B-cell lymphoma
   6. Marginal zone lymphoma
      a. Nodal marginal zone lymphoma
      b. Gastric MALT lymphoma
      c. Nongastric MALT lymphoma
      d. Splenic marginal zone lymphoma
   7. Mantle cell lymphoma (MCL)
   8. Mycosis fungoides (MF)/Sezary syndrome (SS)
   9. Peripheral T-cell lymphoma (PTCL)
   10. Primary cutaneous B-cell lymphoma
   11. Cutaneous anaplastic large cell lymphoma (ALCL)

B. Waldenström’s macroglobulinemia/lymphoplasmacytic lymphoma
   Authorization of 12 months may be granted for treatment of Waldenström’s macroglobulinemia/lymphoplasmacytic lymphoma.

C. Multiple myeloma (MM)
   Authorization of 12 months may be granted for treatment of MM.

D. Classical Hodgkin lymphoma (CHL)
   Authorization of 12 months may be granted for treatment of CHL.

III. CONTINUATION OF THERAPY

   All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

TYSABRI (natalizumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
A. Moderately to severely active Crohn’s disease (CD)
B. Relapsing forms of multiple sclerosis (MS)

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Moderately to severely active Crohn’s disease (CD)
   1. Authorization of 24 months may be granted to members who have received Tysabri or any other biologic indicated for the treatment of Crohn’s disease.
   2. Authorization of 24 months may be granted for members who have an inadequate response, intolerance or contraindication to BOTH of the following:
      a. At least ONE conventional therapy option (See Appendix)
      b. At least ONE TNF-alpha inhibitor indicated for CD:
         i. Humira (adalimumab)
         ii. Remicade (infliximab)
         iii. Cimzia (certolizumab)

B. Relapsing forms of multiple sclerosis (MS)
   Authorization of 24 months may be granted to members who have been diagnosed with a relapsing form of multiple sclerosis.

III. CONTINUATION OF THERAPY

A. Crohn’s disease
   Authorization of 24 months may be granted for all members (including new members) who meet ALL initial authorization criteria and achieve or maintain positive clinical response after at least 3 months of therapy with Tysabri as evidenced by low disease activity or improvement in signs and symptoms of the condition.

B. Multiple sclerosis (MS)
   Authorization of 24 months may be granted for all members (including new members) who meet all initial authorization criteria.
IV. APPENDIX

Examples of Conventional Therapy Options for CD

1. Mild to moderate disease – induction of remission:
   a. Oral budesonide, oral mesalamine
   b. Alternatives: metronidazole, ciprofloxacin, rifaximin

2. Mild to moderate disease – maintenance of remission:
   a. Azathioprine, mercaptopurine
   b. Alternatives: oral budesonide, methotrexate intramuscularly (IM)

3. Moderate to severe disease – induction of remission:
   a. Prednisone, methylprednisolone intravenously (IV)
   b. Alternatives: methotrexate IM

4. Moderate to severe disease – maintenance of remission:
   a. Azathioprine, mercaptopurine
   b. Alternative: methotrexate IM

5. Perianal and fistulizing disease – induction of remission
   a. Metronidazole ± ciprofloxacin

6. Perianal and fistulizing disease – maintenance of remission
   a. Azathioprine, mercaptopurine
   b. Alternative: methotrexate IM

V. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

VECTIBIX (panitumumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications

Vectibix is indicated for the treatment of patients with wild-type RAS (defined as wild-type in both KRAS and NRAS as determined by an FDA-approved test for this use) metastatic colorectal cancer (mCRC):

1. As first-line therapy in combination with FOLFOX (fluorouracil, leucovorin, and oxaliplatin)
2. As monotherapy following disease progression after prior treatment with fluoropyrimidine-, oxaliplatin-, and irinotecan-containing chemotherapy

Limitation of Use: Vectibix is not indicated for the treatment of patients with RAS-mutant mCRC or for whom RAS mutation status is unknown.

B. Compendial Use

Colorectal cancer

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Colorectal Cancer (CRC)

Authorization of 12 months may be granted for the treatment of colorectal cancer when all of the following criteria are met:

A. The RAS (KRAS and NRAS) mutation status is negative (wild-type).
B. Member has not previously experienced clinical failure on cetuximab.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

VIMIZIM (elosulfase alfa)

POLICY

I. INDICATIONS
   The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

   FDA-Approved Indications
   Vimizim is indicated for patients with Mucopolysaccharidosis IVA (MPS IVA, Morquio A syndrome).

   All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

   Mucopolysaccharidosis IVA (MPS IVA)
   Indefinite authorization may be granted for treatment of MPS IVA when the diagnosis of MPS IVA was confirmed by enzyme assay demonstrating a deficiency of N-acetylgalactosamine 6-sulfatase enzyme activity or by genetic testing.

III. CONTINUATION OF THERAPY

   All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT
VIVITROL (naltrexone for extended-release injectable suspension)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
A. Vivitrol is indicated for the treatment of alcohol dependence in patients who are able to abstain from alcohol in an outpatient setting prior to initiation of treatment with Vivitrol. Patients should not be actively drinking at the time of initial Vivitrol administration.
B. Vivitrol is indicated for the prevention of relapse to opioid dependence, following opioid detoxification.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR APPROVAL

A. Alcohol Dependence
   Authorization of 24 months may be granted to members who are prescribed Vivitrol for the treatment of alcohol dependence.

B. Opioid Dependence
   Authorization of 24 months may be granted to members who are prescribed Vivitrol for the prevention of relapse to opioid dependence.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

IV. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

VONVENDI [von Willebrand factor (recombinant)]

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication
On-demand treatment and control of bleeding episodes in adults with von Willebrand disease (vWD)

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

Von Willebrand Disease
Indefinite authorization may be granted for treatment of vWD when any of the following criteria is met:
A. Member has type 1, 2A, 2M, or 2N vWD and has had an insufficient response to desmopressin or a documented clinical reason for not using desmopressin (see Appendix).
B. Member has type 2B or type 3 vWD.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

IV. APPENDIX

Clinical Reasons For Not Utilizing Desmopressin in Patients with Type 1, 2A, 2N and 2M vWD
A. Age < 2 years
B. Pregnancy
C. Fluid/electrolyte imbalance
D. High risk for cardiovascular or cerebrovascular disease (especially the elderly)
E. Predisposition to thrombus formation
F. Trauma requiring surgery
G. Life-threatening bleed
H. Contraindication or intolerance to desmopressin
I. Severe type 1 von Willebrand disease

V. REFERENCES


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SPECIALTY GUIDELINE MANAGEMENT

VPRIV (velaglucerase alfa)

POLICY

I. INDICATIONS
The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
VPRIV is indicated for long-term enzyme replacement therapy (ERT) for patients with type 1 Gaucher disease.

II. CRITERIA FOR INITIAL APPROVAL
Gaucher disease type 1
Indefinite authorization may be granted for treatment of Gaucher disease type 1 when the diagnosis of Gaucher disease was confirmed by enzyme assay demonstrating a deficiency of beta-glucocerebrosidase (glucosidase) enzyme activity or by genetic testing.

III. CONTINUATION OF THERAPY
All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

WILATE (von Willebrand factor/coagulation factor VIII complex [human])

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication
   Wilate is indicated in children and adults with von Willebrand Disease (vWD) for:
   1. On-demand treatment and control of bleeding episodes
   2. Perioperative management of bleeding

B. Compendial Use
   Acquired von Willebrand Syndrome

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Von Willebrand Disease
   Indefinite authorization may be granted for treatment of vWD when either of the following criteria is met:
   1. Member has type 1, 2A, 2M, or 2N vWD and has had an insufficient response to desmopressin or a documented clinical reason for not using desmopressin (see Appendix).
   2. Member has type 2B or type 3 vWD.

B. Acquired von Willebrand Syndrome
   Indefinite authorization may be granted for treatment of acquired von Willebrand syndrome.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

IV. APPENDIX

Clinical Reasons For Not Utilizing Desmopressin in Patients with Type 1, 2A, 2N and 2M vWD²-⁶-⁸

A. Age < 2 years
B. Pregnancy
C. Fluid/electrolyte imbalance
D. High risk for cardiovascular or cerebrovascular disease (especially the elderly)
E. Predisposition to thrombus formation
F. Trauma requiring surgery
G. Life-threatening bleed
H. Contraindication or intolerance to desmopressin
I. Severe type 1 von Willebrand disease

V. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT

XEOMIN (incobotulinumtoxinA)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
A. Cervical dystonia in adults in both botulinum toxin-naïve and previously treated patients
B. Blepharospasm in adults who were previously treated with onabotulinumtoxinA (Botox)
C. Upper limb spasticity in adults

All other indications are considered experimental/investigational and are not a covered benefit.

II. EXCLUSIONS

Coverage will not be provided for cosmetic use.

III. CRITERIA FOR INITIAL APPROVAL

A. Cervical dystonia
Authorization of 24 months may be granted for treatment of cervical dystonia (e.g., torticollis).

B. Blepharospasm
Authorization of 24 months may be granted for treatment of blepharospasm when the member has received a prior treatment with onabotulinumtoxinA.

C. Upper limb spasticity
Authorization of 24 months may be granted for treatment of upper limb spasticity.

IV. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

V. REFERENCES

SPECIALTY GUIDELINE MANAGEMENT

XOLAIR (omalizumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications
A. Allergic Asthma:
   1. Xolair is indicated for patients 6 years of age and older with moderate to severe persistent asthma who have a positive skin test or in vitro reactivity to a perennial aeroallergen and whose symptoms are inadequately controlled with inhaled corticosteroids.
   2. Limitations of use: Xolair is not indicated for the relief of acute bronchospasm or status asthmaticus, or for treatment of other allergic conditions.

B. Chronic Idiopathic Urticaria:
   1. Xolair is indicated for the treatment of adults and adolescents 12 years of age and older with chronic idiopathic urticaria (CIU) who remain symptomatic despite H1 antihistamine treatment.
   2. Limitations of use: Xolair is not indicated for treatment of other forms of urticaria.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Allergic Asthma
   Authorization of 12 months may be granted for treatment of allergic asthma when ALL of the following criteria are met:
   1. Member is 6 years of age or older
   2. Member has a positive skin or in vitro reactivity to at least one perennial aeroallergen
   3. Member has a pre-treatment IgE level greater than or equal to 30 IU/mL
   4. Member has inadequate asthma control despite current treatment with both of the following medications at optimized doses:
      a. Inhaled corticosteroid
      b. Additional controller (long acting beta-agonist, leukotriene modifier, or sustained-release theophylline)

B. Chronic Idiopathic Urticaria
   Authorization of 6 months may be granted for treatment of CIU when ALL of the following criteria are met:
   1. Member is 12 years of age or older
   2. Member has been evaluated for other causes of urticaria, including bradykinin-related angioedema and interleukin-1-associated urticarial syndromes (auto-inflammatory disorders, urticarial vasculitis)
   3. Member has experienced a spontaneous onset of wheals, angioedema, or both, for at least 6 weeks

III. CONTINUATION OF THERAPY
A. Allergic Asthma
Authorization of 12 months may be granted for treatment of allergic asthma when ALL of the following criteria are met:
1. Member is 6 years of age or older
2. Asthma control has improved on Xolair treatment as demonstrated by at least ONE of the following:
   a. A reduction in the frequency or severity of symptoms and exacerbations, OR
   b. An improvement in FEV₁ since initiation of therapy, OR
   c. A reduction in the daily maintenance oral corticosteroid dose¹⁰

B. CIU
Authorization of 12 months may be granted for continuation of treatment of CIU when ALL of the following criteria are met:
1. Member is 12 years of age or older
2. Member has experienced a response (e.g., improved symptoms) since initiation of therapy

IV. REFERENCES
SPECIALTY GUIDELINE MANAGEMENT
YERVOY (ipilimumab)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
   1. Yervoy is indicated for the treatment of unresectable or metastatic melanoma in adults and pediatric patients (12 years and older)
   2. Yervoy is indicated for the adjuvant treatment of patients with cutaneous melanoma with pathologic involvement of regional lymph nodes of more than 1 mm who have undergone complete resection, including total lymphadenectomy
   3. Yervoy is indicated for the treatment of patients with intermediate or poor risk, previously untreated advanced renal cell carcinoma, in combination with nivolumab.

B. Compendial Use
   1. Retreatment of melanoma in patients who experience disease control but who relapse or progress greater than 3 months after treatment discontinuation
   2. Central nervous system (CNS) metastases if active against primary tumor (melanoma) as a single agent or in combination with nivolumab
   3. Small cell lung cancer in combination with nivolumab
   4. Malignant pleural mesothelioma subsequent systemic therapy in combination with nivolumab

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Melanoma
   1. Authorization of 12 months may be granted for the treatment of unresectable or metastatic melanoma
   2. Authorization of 12 months may be granted for the adjuvant treatment of melanoma

B. CNS Metastases
   Authorization of 12 months may be granted for the treatment of CNS metastases in members with a diagnosis of melanoma when Yervoy was active against the primary melanoma tumor

C. Small Cell Lung Cancer
   Authorization of 12 months may be granted for the treatment of small cell lung cancer

D. Malignant pleural mesothelioma
   Authorization of 12 months may be granted for the treatment of malignant pleural mesothelioma

E. Renal Cell Carcinoma
Authorization of 12 months may be granted for the treatment of renal cell carcinoma in combination with nivolumab.

III. CONTINUATION OF THERAPY

A. Melanoma
   1. Authorization of 12 months may be granted for the treatment of unresectable or metastatic melanoma if the member had disease progression or relapse after stable disease of at least three months duration after their first course of Yervoy.
   2. Authorization of 12 months may be granted for the adjuvant treatment of melanoma when the member meets ALL initial authorization criteria.

B. CNS Metastases
   Authorization of 12 months may be granted for the treatment of CNS metastases when the member meets all initial authorization criteria.

C. Small Cell Lung Cancer
   Authorization of 12 months may be granted for the treatment of small cell lung cancer when the member meets all initial authorization criteria.

D. Malignant pleural mesothelioma
   Authorization of 12 months may be granted for the treatment of malignant pleural mesothelioma when the member meets all initial authorization criteria.

E. Renal Cell Carcinoma
   Authorization of 12 months may be granted for the treatment of renal cell carcinoma when the member meets all initial authorization criteria.

IV. REFERENCES