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|---|---|
| **Clinical Policy Title:** | immune Globulins |
| **Policy Number:** | RxA.143 |
| **Drug(s) Applied:** | Asceniv™, Bivigam™, Cutaquig®, Cuvitru™, Flebogamma® DIF, Gamastan® S/D, Gammagard® liquid, Gammagard® S/D, Gammaked™, Gammaplex®, Gamunex®-C, Hizentra®, Hyqvia®, Octagam®, Panzyga®, Privigen®, Xembify® |
| **Original Policy Date:** | 02/07/2020 |
| **Last Review Date:** | 10/19/2023 |
| **Line of Business Policy Applies to:** | All lines of business (except Medicare) |

## Criteria

**I. Initial Approval Criteria**

**A. B-Cell Chronic Lymphocytic Leukemia Infection Prophylaxis** (must meet all)**:**

1. Diagnosis of B-cell CLL;
2. Prescribed by or in consultation with a hematologist, oncologist, or immunologist;
3. Current (within the last 6 months) hypogammaglobulinemia as evidenced by two separate measurements of immunoglobulin G (IgG) level less than 500 mg/dL;
4. Member has had recurrent serious bacterial infections (e.g., requiring intravenous antibiotics, hospitalization, or consultation with an infectious disease specialist) within the past 12 months;
5. Dose does not exceed one of the following (a or b) :
   a. 400 mg per kg intravenously every 3 to 4 weeks;
   b. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

   *Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months
**Medicaid**: 6 months

**B. Dermatomyositis, Polymyositis (off-label)** (must meet all)**:**

1. Diagnosis of dermatomyositis (DM) or polymyositis (PM);
2. Request for Octagam 10%;
3. Prescribed by or in consultation with a dermatologist, rheumatologist, neurologist, or neuromuscular specialist;
4. Trial and failure of 4-months of a systemic corticosteroid (e.g., prednisone) in combination with one of the following immunosuppressive agents, both at up to maximally indicated doses unless contraindicated or clinically significant adverse effects are experienced: methotrexate, azathioprine, cyclophosphamide, mycophenolate mofetil, tacrolimus, cyclosporine ;
5. For dermatomyositis requests only: Trial and failure of rituximab, unless contraindicated, clinically significant adverse effects are experienced, or the member is diagnosed with juvenile dermatomyositis plus calcinosis;
6. Dose does not exceed one of the following (a or b) :

a. 2 gm per kg intravenously per month;
b. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

*Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months
**Medicaid**: 6 months

**C. Fetal/Neonatal Alloimmune Thrombocytopenia (off-label)** (must meet all)**:**
1. Diagnosis of fetal/neonatal alloimmune thrombocytopenia (FNAIT);
2. Prescribed by or in consultation with a hematologist, immunologist, perinatologist, or neonatologist;
3. Meets one of the following (a, b, c, or d):
   a. Previous pregnancy affected by FNAIT;
   b. Serological confirmation of FNAIT as evidenced by maternal-fetal HPA incompatibility;
   c. Nadir platelet count < 100 x $10^9$/L at birth or within 7 days after birth of the affected child;
   d. Fetal intracranial hemorrhage;
4. Dose does not exceed one of the following (a or b) ) :
   a. 2 gm per kg intravenously per week;
   b. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

   *Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial**: 6 months
**Medicaid:** 6 months

**D. Inflammatory Demyelinating Polyneuropathy (Acute/Guillain-Barre Syndrome or Chronic)** (must meet all):
1. Diagnosis of acute inflammatory demyelinating polyneuropathy (AIDP)/Guillain- Barre Syndrome (GBS) or CIDP;
2. Prescribed by or in consultation with a neurologist or neuromuscular specialist;
3. Member meets one of the following (a - b):
   a. Diagnosis is AIDP/GBS and member meets one of the following (i-vii):
      i. Inability to stand or walk at least 30 feet without assistance;
      ii. ICU admission required for aspiration or mechanical ventilation;
      iii. Miller-Fisher syndrome;
      iv. Inability to raise head against gravity;
      v. Severe bulbar palsy (e.g., impaired gag reflex, dysarthria and/or dysphagia);
      vi. Bilateral facial weakness;
      vii. Autonomic dysfunction (e.g., unexplained dysrhythmia, blood pressure fluctuations, significant bowel or bladder involvement);
   b. Diagnosis is CIDP and member meets all of the following (i-v):
      i. Disease is progressive or relapsing for more than 2 months;
      ii. Member has either of the following (a or b):
         a. Both of the following, characterizing typical CIDP (1 and 2):
            1. Chronically progressive, stepwise, or recurrent symmetric proximal and distal weakness and sensory dysfunction of all extremities;
            2. Absent or reduced tendon reflexes in all extremities;
         b. One of the following, characterizing atypical CIDP (1-3):
            1) Predominantly distal (distal acquired demyelinating symmetric, DADS) or asymmetric

[multifocal acquired demyelinating sensory and motor neuropathy (MADSAM), Lewis-Sumner syndrome] or focal (e.g., involvement of the brachial or lumbosacral plexus or of one or more peripheral nerves in one upper or lower limb) disease;
2) Pure motor symptoms;
3) Pure sensory symptoms (including chronic immune sensory polyradiculopathy affecting the central process of the primary sensory neuron);

iii. Diagnosis has been confirmed via electrodiagnostic testing;
iv. Member does not have any of the following (1-6):
1) Borrelia burgdorferi infection (Lyme disease), diphtheria, drug or toxin exposure probably to have caused the neuropathy;
2) Hereditary demyelinating neuropathy;
3) Prominent sphincter disturbance;
4) Diagnosis of multifocal motor neuropathy;
5) IgM monoclonal gammopathy with high titre antibodies to myelin-associated glycoprotein;
6) Other causes for a demyelinating neuropathy including POEMS syndrome, osteosclerotic myeloma, diabetic and nondiabetic lumbosacral radiculoplexus neuropathy;
v. For members who do not have pure motor symptoms, failure of at least one corticosteroid (e.g., prednisone) at up to maximally indicated doses unless contraindicated or clinically significant adverse effects are experienced;

4. Dose does not exceed one of the following (a, b, c, or d):
a. For AIDP/GB: 0.4 g per kg per day intravenously for 5 days;
b. For CIDP: Gammaked™, Gamunex®-C; Panzyga®, Privigen: Loading dose 2 gm per kg Intravenously given in divided doses over two to four consecutive days, following by maintenance dose of 1 gm per kg intravenous every 3 weeks;
c. For CIDP: Hizentra 0.2 gm per kg body weight subcutaneously per week, starting 1 week after last IVIG infusion or 0.4 gm per kg body weight subcutaneously per week if evidence is submitted demonstrating worsening symptoms;
d. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).
*Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months
**Medicaid:** 6 months

**E. Idiopathic Thrombocytopenic Purpura (Acute or Chronic)** (must meet all)**:**
1. Diagnosis of ITP;
2. Member meets one of the following (a or b):
a. Chronic ITP: If request is for Flebogamma® 10%, Gammagard S/D®, Gammaked™, Gammaplex®, Gamunex C®, Octagam 10%, Panzyga®, Privigen®
b. Acute ITP: Gammaked™, Gamunex C®
3. Member meets one of the following (a or b or c):
a. Octagam 10% , Gammaked®, Gammaplex®, Gammagard S/D®, Gamunex C®, Panzyga®: Age ≥ 18 years;
b. Privigen®: Age at least 15 years or older;
c. Flebogamma®: 10%: ≥ 2 years of age;
4. Prescribed by or in consultation with a hematologist;
5. Member meets one of the following (a or b):

a. Trial and failure of one of the following at up to maximally indicated doses unless contraindicated or clinically significant adverse effects are experienced (i or ii):
   i. Systemic corticosteroids (e.g., prednisone);
   ii. Rho(D) immune globulin (RhIG);
   *Prior authorization is required for RhIG
b. Pregnant;

6. Member meets one of the following (a – e):
   a. Acute bleeding due to severe thrombocytopenia (platelet count less than 30,000/µL);
   b. In patients with severe thrombocytopenia (platelet counts less than 20,000/µL) considered to be at risk for intracerebral hemorrhage;
   c. Platelet counts persistently at or below 20,000/µl (For CITP);
   d. Splenectomy is scheduled;
   e. Pregnant;
7. Dose does not exceed one of the following (a or b):
   a. Dose does not exceed FDA recommended maximum dose and frequency (refer to dosing information section for product specific dosing and frequency or manufacturer's prescribing information.
   b. Dose is supported by practice guidelines or peer-reviewed literatures for the relevant off-label use (prescriber must submit supporting evidence).

   *Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months
**Medicaid:** 6 months

**F. Kawasaki Syndrome Aneurysm Prevention** (must meet all)**:**
1. Diagnosis of Kawasaki Syndrome or Incomplete (Atypical) Kawasaki Disease;
2. Prescribed by or in consultation with a cardiologist, allergist, immunologist, infectious disease specialist, or rheumatologist;
3. Prescribed concurrently with aspirin therapy, unless contraindicated or clinically significant adverse effects are experienced;
4. Dose does not exceed one of the following (a or b):
   a. Dose does not exceed FDA recommended maximum dose and frequency (refer to dosing information section for product specific dosing and frequency or manufacturer's prescribing information);
   b. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

   *Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 1 month (one time approval)
**Medicaid:** 1 month (one time approval)

**G. Kidney Transplant (off-label)** (must meet all)**:**
1. Member meets one of the following (a or b):
   a) If prescribed prior to kidney transplant, member has high levels of "anti-donor" antibodies (i.e., member is highly sensitized to the tissue of the majority of living or cadaveric donors because of "non-self" human leukocyte antigen (HLA) or ABO incompatibility);
   b) If prescribed following kidney transplant, used for the treatment of antibody- mediated rejection;

2. Prescribed by or in consultation with a nephrologist, transplant specialist, or hematologist;
3. Dose does not exceed one of the following (a or b):
   a. Dose does not exceed FDA recommended maximum dose and frequency (refer to dosing information section for product specific dosing and frequency or manufacturer's prescribing information);
   b. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

*Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months
**Medicaid:** 6 months

**H. Multifocal Motor Neuropathy** (must meet all)**:**
1. Diagnosis of MMN;
2. Prescribed by or in consultation with a neurologist or neuromuscular specialist;
3. Dose does not exceed one of the following (a or b):
   a. 2.4 gm per kg intravenously per month;
   b. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

*Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months
**Medicaid:** 6 months

**I. Multiple Myeloma Infection Prophylaxis (off-label)** (must meet all)**:**
1. Diagnosis of multiple myeloma (MM) with stable plateau phase disease;
2. Prescribed by or in consultation with a hematologist, oncologist, or immunologist;
3. Current (within the last 6 months) hypogammaglobulinemia as evidenced by two separate measurements of immunoglobulin G (IgG) level less than 400 mg/dL;
4. Member has had recurrent serious bacterial infections (e.g., requiring intravenous antibiotics, hospitalization, or consultation with an infectious disease specialist) within the past 12 months;
5. Documented failure or inability to tolerate chemotherapy or radiation therapy;
6. Dose does not exceed one of the following (a or b):
   a. 400 mg per kg intravenously every 3 weeks;
   b. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

*Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months
**Medicaid:** 6 months

**J. Multiple Sclerosis (off-label)** (must meet all)**:**
1. Diagnosis of relapsing-remitting multiple sclerosis (MS);
2. Prescribed by or in consultation with a neurologist;
3. Trial and failure of three FDA-approved disease-modifying MS therapies (e.g., Avonex, Betaseron, Copaxone, Vumerity, Kesimpta) at up to maximally indicated doses unless contraindicated or clinically significant side effects are experienced;
   *Prior authorization is required for MS therapies

4. Dose does not exceed one of the following (a or b):
   a. Initial loading dose of 400 mg per kg intravenously for 5 days, followed by maintenance dose of 1 g per kg intravenously per month;
   b. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

   *Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months
**Medicaid:** 6 months

**K. Myasthenia Gravis (MG)/Lambert Eaton Myasthenic Syndrome (LEMS) (off-label)** (must meet all)**:**
1. Diagnosis of myasthenia gravis (MG) or Lambert Eaton myasthenic syndrome (LEMS);
2. Prescribed by or in consultation with a neurologist or neuromuscular specialist;
3. Member meets one of the following (a, b, or c):
   a) Acute crisis (e.g., vital capacity less than 1 L/min, inability to walk 100 ft without assistance, intubation, dysphagia with aspiration, mechanical ventilation);
   b) Thymectomy surgery is scheduled;
   c) Failure of both of the following at up to maximally indicated doses, unless contraindicated or clinically significant adverse effects are experienced (i and ii):
      i. Amifampridine or a cholinesterase inhibitor (e.g., pyridostigmine); for LEMS;
      ii. Systemic corticosteroid (e.g., prednisone) or immunosuppressant (e.g., azathioprine) for MG;

   *Prior authorization may be required for amifampridine
4. Dose does not exceed one of the following (a or b):
   a) 2 gm per kg intravenously for 2 to 5 days per treatment course;
   b) Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

   *Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months
**Medicaid:** 6 months

**L. Paraneoplastic Neurological Syndrome (off-label)** (must meet all)**:**
1. Diagnosis of one of the following subtypes of paraneoplastic neurological syndrome (a or b):
   a. Opsoclonus-myoclonus syndrome;
   b. Anti-NMDA encephalitis;
2. Prescribed by or in consultation with a neurologist, neuromuscular specialist, or oncologist;
3. For opsoclonus-myoclonus syndrome: Trial and failure of at least one systemic corticosteroid (e.g., prednisone) at up to maximally indicated doses, unless contraindicated or clinically significant adverse effects are experienced;
4. Dose does not exceed one of the following (a or b) :
   a. Dose does not exceed FDA recommended maximum dose and frequency (refer to dosing information section for product specific dosing and frequency or manufacturer's prescribing information);
   b. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

   *Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months

**Medicaid:** 6 months

**M. Parvovirus B19 Infection and Anemia (off-label)** (must meet all)**:**

1. Diagnosis of anemia secondary to chronic parvovirus B19 infection;
2. Prescribed by or in consultation with a hematologist, infectious disease specialist, or immunologist;
3. Current (within the last 30 days) severe anemia (i.e., Hgb <10 or Hct < 30) due to bone marrow suppression;
4. Dose does not exceed one of the following (a or b):
   a. Initial dose of 2 gm per kg per day for up to 5 days, followed by maintenance dose of 400 mg per kg intravenously every 4 weeks;
   b. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

*Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months
**Medicaid:** 6 months

**N. Pediatric Human Immunodeficiency Virus (HIV) Infection Prophylaxis (off-label)** (must meet all)**:**

1. Prescribed for prophylaxis of serious bacterial infection in a child who has human immunodeficiency virus (HIV);
2. Prescribed by or in consultation with an HIV or infectious disease specialist;
3. Current (within the last 6 months) hypogammaglobulinemia as evidenced by two separate measurements of serum IgG concentration less than 250 mg/dL;
4. Member meets one of the following (a - e):
   a) Recurrent serious bacterial infections (defined as two or more infections such as bacteremia, meningitis, or pneumonia in a 12-month period);
   b) Inadequate antibody response to protein/polysaccharide antigens (e.g., measles, pneumococcal, and/or Haemophilus influenzae type b);
   c) Lives in an area where measles is highly prevalent and has not developed an antibody response after two doses of measles, mumps, and rubella virus live vaccine;
   d) Exposure to measles (requires a single dose);
   e) Chronic bronchiectasis that is suboptimally responsive to antimicrobial and pulmonary therapy;
5. Dose does not exceed one of the following (a or b):
   a) 400 mg per kg intravenously every 2 to 4 weeks;
   b) Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

*Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months
**Medicaid:** 6 months

**O. Pemphigus Vulgaris, Pemphigus Foliaceus, Bullous Pemphigoid, Mucous Membrane Pemphigoid (a.k.a. Cicatricial Pemphigoid), Epidermolysis Bullosa Acquisita** (off-label) (must meet all):

1. Diagnosis of one of the following (a, b, c, d, or e):
   a) Pemphigus vulgaris;
   b) Pemphigus foliaceus;
   c) Bullous pemphigoid;

d) Mucous membrane pemphigoid (a.k.a. cicatricial pemphigoid);
e) Epidermolysis bullosa acquisita;
2. Prescribed by or in consultation with a dermatologist;
3. Trial and failure of at least one corticosteroid (e.g., prednisone) at up to maximally indicated doses unless contraindicated or clinically significant adverse effects are experienced;
4. Trial and failure of at least one immunosuppressive agent (e.g., cyclophosphamide, azathioprine, mycophenolate mofetil) at up to maximally indicated doses unless contraindicated or clinically significant adverse effects are experienced;
5. Trial and failure of Rituxan® unless contraindicated or clinically significant adverse effects are experienced;
*Prior authorization is required for Rituxan
6. Dose does not exceed one of the following (a, b, c, or d):
a) 2 gm per kg intravenously every 4 weeks;
b) 400 mg per kg per day intravenously for 5 days (1 cycle only; may repeat up to three times in a 6-month period);
c) 300 mg per kg per day intravenous for 5 days at monthly intervals (for up to 3 cycles);
d) Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).
*Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial**: 6 months
**Medicaid:** 6 months

**P. Primary Immunodeficiencies** (must meet all)**:**
1. Diagnosis of primary immunodeficiencies (PI), including any of the following (a - h):
a) Agammaglobulinemia (e.g., X-linked, congenital);
b) Common variable immunodeficiency (CVID);
c) Congenital hypogammaglobulinemia;
d) Immunodeficiency with near/normal IgM (absent IgG, IgA) (also known as Hyper IgM syndrome);
e) Selective immunodeficiency (e.g., selective IgA, IgM, or IgG subclass);
f) Severe combined immunodeficiency disorders (SCID) (e.g., X-SCID, jak3, ZAP70, adenosine deaminase (ADA) deficiency, PNP, RAG defects, Ataxia Telangiectasia, Wiskott-Aldrich syndrome, DiGeorge syndrome);
g) Subclass deficiency;
h) Functional/specific antibody deficiency;
2. Prescribed by or in consultation with an immunologist or hematologist;
3. Meet one of the following (a, b, c or d):
a) Asceniv™: Age ≥ 12 years;
b) Octagam®, Bivigam™: Age ≥ 6 years ;
c) Xembify®, Panzyga®, Hizentra®, Gamunex®-C, Gammaplex®, Gammaked™, Gammagard® S/D, Gammagard® liquid, Flebogamma® DIF, Cuvitru™ Cutaquig®: Age ≥ 2 years;
d) Privigen: Age ≥ 3 years;
4. Member meets one of the following (a or b):
a) For functional/specific antibody deficiency, meets all of the following (i, ii, and iii):
i. Normal immune globulin levels;
ii. Inadequate antibody response to polysaccharide antigens (e.g., pneumococcal);
iii. Recurrent sinopulmomary infections within the past 12 months;

b) Current (within the last 6 months) total or subclass immune globulin deficiency (below normal for age) as evidenced by two separate measurements of immunoglobulin level and one of the following (i, ii, iii, or iv):
   i. For ADA-SCID: failure (defined as experiencing continued recurrent serious bacterial infections) of Revcovi®, or hematopoietic stem cell transplant, unless contraindicated or clinically significant adverse effects are experienced;
      *Prior authorization is required for Adagen® and Revcovi®
   ii. SCID (not including ADA-SCID);
   iii. Recurrent serious bacterial infections (e.g., requiring intravenous antibiotics, hospitalization, or consultation with an infectious disease specialist) within the past 12 months;
   iv. Inadequate antibody response to protein/polysaccharide antigens (e.g., tetanus, diphtheria, pneumococcal);

5. Dose does not exceed one of the following (a or b) :
   a. Dose does not exceed FDA recommended maximum dose and frequency (refer to dosing information section for product specific dosing and frequency or manufacturer's prescribing information);
   b. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

   *Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial:** 6 months
**Medicaid:** 6 months

**Q. Stiff Person Syndrome (off-label)** (must meet all)**:**
1. Diagnosis of stiff person syndrome (also known as Moersch-Woltmann syndrome);
2. Prescribed by or in consultation with a neurologist or neuromuscular specialist;
3. Trial and failure of a benzodiazepine (e.g., diazepam) or baclofen at up to maximally indicated doses, unless contraindicated or clinically significant adverse effects are experienced;
4. Dose does not exceed one of the following (a or b):
   a. 2 gm per kg given over two to three intravenously infusions, each separated by three to five days per treatment course;
   b. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

   *Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**
**Commercial**: 6 months
**Medicaid:** 6 months

**R. Viral Prophylaxis for Hepatitis A, Measles, Varicella, Rubella Viruses** (must meet all):
1. Request is for intramuscular formulation;
2. Request is for one of the following indications (a, b, c, or d):
   a) Hepatitis A post-exposure/high-risk prophylaxis and meets both of the following (i and ii):
      i. Hepatitis A exposure or at high risk for exposure as evidenced by (a or b):
         a. Exposure to hepatitis A in the past 2 weeks (e.g., household contact, sexual contact, sharing illicit drugs with someone positive for hepatitis A, regular babysitters/caretakers, food handlers at the same establishment as one who is positive for hepatitis A) and does not have clinical manifestations of hepatitis A;

b. Traveling to or working in an area endemic for hepatitis A;

ii. Meets at least one of the following (a, b, or c):
   a. Hepatitis A vaccine is locally unavailable;
   b. History of severe allergic reaction (anaphylaxis) to the hepatitis A vaccine;
   c. If either exposed to the virus or traveling in ≤ 2 weeks to an area endemic for hepatitis A, then (1, 2, or 3):
      1. Age < 1 year or > 40 years;
      2. Chronic liver disease or other chronic medical condition;
      3. Immunocompromised;

b) Measles (rubella) post-exposure prophylaxis and meets all of the following (i, ii, iii, and iv):
   i. Exposure to measles within the past 6 days;
   ii. Member has not previously received a measles vaccine;
   iii. Member has not previously had measles;
   iv. Meets at least one of the following (a - f):
      a. Measles vaccine is locally unavailable;
      b. History of severe allergic reaction (anaphylaxis) to the measles vaccine;
      c. Pregnancy;
      d. Immunocompromised;
      e. Has been > 3 days since exposure;
      f. Age < 12 months;

c) Chickenpox (varicella) post-exposure prophylaxis and meets all of the following (i, ii, iii, and iv):
   i. Exposure to varicella within the past 10 days;
   ii. Member lacks immunity to varicella;
   iii. Varicella zoster immune globulin (VZIG) is currently unavailable;
   iv. Meets any of the following (a - e):
      a. Varicella vaccine is locally unavailable;
      b. History of a severe allergic reaction (anaphylaxis) to the varicella vaccine;
      c. Pregnancy;
      d. Immunocompromised;
      e. Newborn of mother who had varicella from 5 days before to 2 days after delivery;

d) Rubella post-exposure prophylaxis (i and ii):
   i. Recent exposure to rubella;
   ii. Member is pregnant;

2. Dose does not exceed one of the following (a – e):
   a. Hepatitis A (i, ii, or iii):
      i. 0.1 mL/kg intramuscularly once;
      ii. For anticipated exposure up to 2 months: 0.2 mL/kg intramuscularly once;
      iii. For anticipated exposure 2 months or longer: 0.2 mL/kg intramuscularly every 2 months;
   b. Measles: 15 mL intramuscularly once;
   c. Varicella: 1.2 mL/kg intramuscularly once;
   d. Rubella: 0.55 mL/kg intramuscularly once;
   e. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

   *Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval duration**

**Commercial:** Hepatitis A: Up to 6 months

**Medicaid:** Hepatitis A: Up to 6 months

All other indications: One-time approval (1 month)

**S. Management of Immunotherapy-Related Toxicities (CAR T-Cell-Related Toxicities)** (off-label) (must meet all):

1. Diagnosis of Immunotherapy-Related Toxicities (CAR T-Cell-Related Toxicities);
2. Prescribed by immunologist;
3. Member has serum IgG levels <400-600 mg/dL;
4. Serious or recurrent infections [particularly bacterial];
5. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

*Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval Duration**
**Commercial:** Not applicable
**Medicaid:** Not applicable

**T. Management of Immunotherapy-Related Toxicities (Immune Checkpoint Inhibitor-Related Toxicities**) (off-label) (must meet all):

1. Diagnosis of Immunotherapy-Related Toxicities (Immune Checkpoint Inhibitor-Related Toxicities);
2. Prescribed by immunologist;
3. Meet any one of the following:
   a. Additional therapy for suspected myocarditis if no improvement within 24 hours of starting pulse-dose methylprednisolone;
   b. Severe (G3) or life-threatening (G4) bullous dermatitis;
   c. Stevens-Johnson syndrome, or toxic epidermal necrolysis;
   d. Moderate, severe, or life-threatening steroid-refractory myalgias or myositis;
   e. Severe (G3-4) myasthenia gravis;
   f. Moderate (G2) or severe (G3-4) Guillain-Barré Syndrome or severe (G3-4) peripheral neuropathy in combination with pulse-dose methylprednisolone;
   g. Encephalitis in combination with pulse-dose methylprednisolone if severe or progressing symptoms, or if oligoclonal bands present;
   h. Transverse myelitis;
   i. severe (G3-4) pneumonitis if no improvement after 48 hours of methylprednisolone;
4. Dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

*Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval Duration**
**Commercial:** Not applicable
**Medicaid:** Not applicable

**II. Continued Therapy Approval**

**A. Kawasaki Syndrome/Incomplete (Atypical) Kawasaki Disease, Viral Prophylaxis (Hep A, Measles, Varicella, Rubella), Management of Immunotherapy-Related Toxicities (Immune Checkpoint Inhibitor-Related Toxicities**), **Management of Immunotherapy-Related Toxicities (CAR T-Cell-Related Toxicities)**

1. Re-authorization is not permitted. Members must meet the initial approval criteria.

**Approval Duration**
**Commercial:** Not applicable
**Medicaid:** Not applicable

**B. All Other Indications in Section I (must meet all):**

1. Member is currently receiving medication that has been authorized by RxAdvance or the member has

met initial approval criteria listed in this policy;
2. Member is responding positively to therapy ;
3. If request is for a dose increase, request meets one of the following (a or b):
   a. Dose titration or conversion is appropriate per package insert labeling;
   b. New dose is supported by practice guidelines or peer-reviewed literature for the relevant off-label use (prescriber must submit supporting evidence).

*Prescribed regimen must be FDA-approved or recommended by NCCN

**Approval Duration**
**Commercial:** 6 months
**Medicaid:** 6 months

**References**


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**Inflammatory Demyelinating Polyneuropathy (Chronic)**

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| Review/Revision History | Review/Revision Date | P&T Approval Date |
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| Policy established. | 01/2020 | 02/07/2020 |
| Reviewed criteria and updated dosing information | 04/2020 | |
| Policy was reviewed: | 02/10/2021 | 03/09/2021 |

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| 1) Initial Approval Criteria-Approval duration updated for commercial<br>2) Continuation therapy criteria II.A.1. rephrased to "Member is currently receiving medication that has been authorized by RxAdvance or the member has met initial approval criteria listed in this policy<br>3) References were updated. | | |
| Policy was reviewed:<br>1. Policy updated to remove drug Carimune NF as it is no longer available.<br>2. Initial Approval Criteria,<br>a. I.E.1: Updated indication from Diagnosis of acute or chronic ITP to Diagnosis of ITP.<br>b. I.A.E.2: Updated to include new indication criteria Member meets one of the following (a-b):<br>a. Chronic ITP: If request is for Flebogamma® 10%, Gammagard S/D®, Gammaked™, Gammaplex®, Gamunex C®, Octagam 10%, Panzyga®, Privigen<br>b. Acute ITP: Gammaked™, Gamunex C®<br>c. I.D.5.b: Updated Dose criteria from For CIDP: Loading dose 2 g per kg Intravenous given in divided doses over two to four consecutive days, following by maintenance dose of 1 g per kg IV every 3 weeks;to For CIDP: Gammaked™, Gamunex®-C; Loading dose 2 g per kg Intravenous given in divided doses over two to four consecutive days, following by maintenance dose of 1 g per kg IV every 3 weeks.<br>d. I.B.2: Updated to include Request for Octagam 10%;<br>e. Initial Approval Criteria, I.E.3: Updated to include new age criteria Member meets one of the following (a or b or c):<br>a) Octagam 10% , Gammaked®, Gammagard S/D®, Gamunex C®: Age at least 18 years or older<br>b) Privigen®: Age at least 15 years or older<br>c) Flebogamma®, Gammaplex®: 10%: 2 years of age and older<br>f. I.P.3: Updated to include new age criteria Meet one of the following (a, b or c)<br>a. Asceniv™: Age 12 to 17 years<br>b. Octagam®, Bivigam™: Age at least 6 years or older<br>c. Xembify®, Panzyga®, Hizentra®, Gamunex®-C, Gammaplex®, Gammaked™, Gammagard® S/D, Gammagard® liquid, Flebogamma® DIF, Cuvitru™ Cutaquig®: Age at least 2 years or older | 12/15/2021 | 01/17/2022 |

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| g. I.S: Updated to include approval criteria for indication, Management of Immunotherapy-Related Toxicities (CAR T-Cell-Related Toxicities)<br>h. I.T: Updated to include approval criteria for indication, Management of Immunotherapy-Related Toxicities (Immune Checkpoint Inhibitor-Related Toxicities)<br>i. E.6: Updated to add a.Acute bleeding due to severe thrombocytopenia (platelet count less than 30,000/ µL), b. In patients with severe thrombocytopenia( platelet counts less than 20,000/ µL) considered to be at risk for intracerebral hemorrhage; c. Platelets counts persistently at or below 20,000/ µL(for CITP);<br>j. I.5:Updated to add ,"documented failure or inability to tolerate chemotherapy or radiation therapy".<br>k. K.3.C.i and K.3.C.ii updated to add for LEMS and for MG respectively.<br>l. N.3 updated serum IgG concentration to less than 250 mg/dl from 400 mg/dl.<br>3. All Initial Approval Criteria were updated to remove requirement: Member meets one of the following (a or b): a.Request is for Gammagard unless there is a specific health plan-preferred* immune globulin product; b. Failure of Gammagard (or health plan-preferred* immune globulin product) unless contraindicated or clinically significant adverse effects are experienced; *Immune globulin products are generally interchangeable, and it is at the health plan's discretion to prefer a clinically appropriate alternative product based on the time of request.<br>4. Continued Therapy Approval Crtieria, IIA updated from Kawasaki Syndrome/Incomplete (Atypical) Kawasaki Disease, Viral Prophylaxis (Hep A, Measles, Varicella, Rubella) to Kawasaki Syndrome/Incomplete (Atypical) Kawasaki Disease, Viral Prophylaxis (Hep A, Measles, Varicella, Rubella), Management of Immunotherapy-Related Toxicities (Immune Checkpoint Inhibitor-Related Toxicities), Management of Immunotherapy-Related Toxicities (CAR T-Cell-Related Toxicities).<br>5. References were reviewed and updated. | | |
| Policy was reviewed:<br>1. Initial Approval Criteria, I.D.3: Updated to include new diagnostic criteria "Member meets one of the following (a - b):<br>a. Diagnosis is AIDP/GBS and member meets one of the | 11/03/2022 | 01/17/2023 |

following (i-vii):
i. Inability to stand or walk at least 30 feet without assistance;
ii. ICU admission required for aspiration or mechanical ventilation;
iii. Miller-Fisher syndrome;
iv. Inability to raise head against gravity;
v. Severe bulbar palsy (e.g., impaired gag reflex, dysarthria and/or dysphagia);
vi. Bilateral facial weakness;
vii. Autonomic dysfunction (e.g., unexplained dysrhythmia, blood pressure fluctuations, significant bowel or bladder involvement);

b. Diagnosis is CIDP and member meets all of the following (i-v):
i. Disease is progressive or relapsing for more than 2 months;
ii. Member has either of the following (a or b):
a. Both of the following, characterizing typical CIDP (1 and 2):
1. Chronically progressive, stepwise, or recurrent symmetric proximal and distal weakness and sensory dysfunction of all extremities;
2. Absent or reduced tendon reflexes in all extremities;
b. One of the following, characterizing atypical CIDP (1-3):
1. Predominantly distal (distal acquired demyelinating symmetric, DADS) or asymmetric [multifocal acquired demyelinating sensory and motor neuropathy (MADSAM), Lewis-Sumner syndrome] or focal (e.g., involvement of the brachial or lumbosacral plexus or of one or more peripheral nerves in one upper or lower limb) disease;
2. Pure motor symptoms;
3. Pure sensory symptoms (including chronic immune sensory polyradiculopathy affecting the central process of the primary sensory neuron);
iii. Diagnosis has been confirmed via electrodiagnostic testing.
iv. Member does not have any of the following (1-6):
1. Borrelia burgdorferi infection (Lyme disease), diphtheria, drug or toxin exposure probably to have caused the neuropathy;
2. Hereditary demyelinating neuropathy;
3. Prominent sphincter disturbance;
4. Diagnosis of multifocal motor neuropathy;
5. IgM monoclonal gammopathy with high titre

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| antibodies to myelin-associated glycoprotein;<br>6. Other causes for a demyelinating neuropathy including POEMS syndrome, osteosclerotic myeloma, diabetic and nondiabetic lumbosacral radiculoplexus neuropathy;<br>v. For members who do not have pure motor symptoms, failure of at least one corticosteroid (e.g., prednisone) at up to maximally indicated doses unless contraindicated or clinically significant adverse effects are experienced;<br>2. References were reviewed and updated. | | |
| Policy was reviewed. | 10/19/2023 | 10/19/2023 |