**Pre - PA Allowance**

None

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**Prior-Approval Requirements**

**Diagnoses**

Patient must have **ONE** of the following:

1. Primary Immunodeficiency Disease (PID) with **ONE** of the following:
   a. Hypogammaglobulinemia, IgG subclass deficiency, selective IgA deficiency, selective IgM deficiency, or specific antibody deficiency with **ALL** of the following:
      i. Documented history of recurrent bacterial and viral infections
      ii. Impaired antibody response to pneumococcal vaccine
      iii. **ONE** of the following pre-treatment laboratory findings:
         1) Hypogammaglobulinemia: IgG < mg/dL or ≥ 2 SD below the mean age
         2) Selective IgA deficiency: IgA level < 7 mg/dL with normal IgG and IgM levels
         3) IgG subclass deficiency: IgG1, IgG2, or IgG3 ≥ 2 SD below the mean age assessed on at least 2 occasions; normal IgG (total) and IgM levels, normal/ low IgA levels
         4) Specific antibody deficiency: normal IgG, IgA and IgM levels
   b. SCID (severe combined immunodeficiency disease) or Agammaglobulinemia with **ONE** of the following
      i. Confirmed diagnosis by genetic or molecular testing
      ii. Pretreatment IgG level < 200mg/dL
      iii. Absence or very low number of T cells (CD3 T cells < 300/microliter) or presence of maternal T cells in the circulation (SCID only)
   c. Wiskott-Aldrich syndrome, DiGeorge syndrome, or ataxia-telangiectasia (or other non SCID combined immunodeficiency) with **ALL** of the following:
      i. Confirmed diagnosis by genetic or molecular testing
      ii. Documented history of recurrent bacterial and viral infections
      iii. Impaired antibody response to pneumococcal vaccine
IVIG (intravenous immunoglobulin)
Bivigam, Carimune NF, Flebogamma, Gammagard, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Privigen

d. CVID (common variable Immunodeficiency disease) with ALL of the following:
   i. Age 4 years and older
   ii. Documented history of recurrent bacterial and viral infections
   iii. Impaired antibody response to pneumococcal vaccine
   iv. Other causes of immune deficiency have been excluded (eg, drug induced, genetic disorders, infectious diseases such as HIV, malignancy)
   v. Pretreatment IgG level < 500mg/dL or ≥ 2 SD below the mean for the age

2. Idiopathic thrombocytopenic purpura (ITP)
   a. Newly diagnosed ITP (diagnosed with in the past 3 months) must have ONE of the following:
      i. Children (<18 years of age) with ONE of the following:
         1) Significant bleeding symptoms (mucosal bleeding or moderate/severe bleeding)
         2) High risk for bleeding
         3) Rapid increase in platelets is required (eg. Surgery or procedure)

      ii. Adults (≥ 18 years of age) with ONE of the following:
         1) Platelet count < 30,000/mcL
         2) Platelet count < 50,000/mcL and significant bleeding symptoms, high risk for bleeding or rapid increase in platelets is required

      AND the following:
         1) Corticosteroid therapy is contraindicated and IVIG will be used alone or IVIG will be used in combination with corticosteroid therapy

   b. Chronic/persistent ITP (> 3 months from diagnosis)

      AND ONE of the following:
      i. Platelet count < 30,000/mcL
      ii. Platelet count < 50,000/mcL and significant bleeding symptoms, high risk for bleeding or rapid increase in platelets is required
IVIG (intravenous immunoglobulin)
Bivigam, Carimune NF, Flebogamma, Gammagard, Gammagard S/D, Gammaked, 
Gammaplex, Gamunex-C, Octagam, Privigen

**AND** the following:
  
i. Relapse after previous response to IVIG or inadequate response, intolerance or contraindication to corticosteroid therapy

**c.** ITP unresponsive to first-line therapy

**AND ONE** of the following:
  
i. Platelet count < 30,000/mcL

  ii. Platelet count < 50,000/mcL and significant bleeding symptoms, high risk for bleeding or rapid increase in platelets is required

**AND** the following:
  
i. Relapse after previous response to IVIG or inadequate response, intolerance or contraindication to corticosteroid therapy

**d.** Adults with refractory ITP after splenectomy must have ONE of the following:
  
i. Platelet count < 30,000/mcL

  ii. Significant bleeding symptoms

**e.** ITP in pregnant women

3. B-cell chronic lymphocytic leukemia with ALL of the following:
   
a. IVIG is prescribed for prophylaxis of bacterial and viral infections

   b. Documented history of recurrent sinopulmonary infections requiring intravenous antibiotics or hospitalization

   c. Pretreatment serum IgG level < 500 mg/dL

4. Kawasaki syndrome

5. Prophylaxis of bacterial and viral infections in Bone marrow transplantation (BMT) / Hematopoietic stem cell transplantation (HSCT) Recipients with ALL of the following:
   
a. IVIG is prescribed for prophylaxis of bacterial and viral infections

   b. **ONE** of the following:
IVIG (intravenous immunoglobulin)
Bivigam, Carimune NF, Flebogamma, Gammagard, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Privigen

i. IVIG is requested within the first 100 days post-transplant
ii. Pretreatment serum IgG level < 400 mg/dL

6. Peripheral blood progenitor cell (PBPC) collection

7. Umbilical Cord Stem Cell Transplantation

8. Prophylaxis of bacterial and viral infections in HIV-Infected Pediatric patients with ALL of the following:
   a. Member is < 12 years of age
   b. Primary prophylaxis:
      i. Pretreatment serum IgG level < 400 mg/dL
   c. Secondary prophylaxis:
      i. Documented recurrent bacterial and viral infections (> 2 serious infections in a year)
      ii. NOT able to take combination antiretroviral therapy
      iii. Antibiotic prophylaxis NOT effective

9. Polymyositis or Dermatomyositis with ALL of the following:
   a. Documented clinical features of diagnosis (eg. elevated muscle enzymes, muscle biopsy, supportive diagnostic tests)
   b. Inadequate response, intolerance or contraindication to first–line treatments (corticosteroids or immunosuppressants)

10. Inclusion-body myositis

11. Guillain-Barre Syndrome (GBS) with ALL of the following:
    a. Physical mobility is severely affected such that patient requires an aid to walk
    b. IVIG therapy will be initiated within 2 weeks of symptom onset

12. Fetal alloimmune thrombocytopenia (F/NAIT)

13. Myasthenia gravis with ALL of the following:
    a. Worsening weakness includes an increase in any of the following symptoms:
       i. Diplopia
       ii. Ptosis
       iii. Blurred vision
       iv. Dysarthria
v. Dysphagia
vi. Difficulty chewing
vii. Impaired respiratory status
viii. Fatigue
ix. Limb weakness
x.

b. Pre-operative management

14. Multiple sclerosis

15. Multifocal motor neuropathy (MMN) with ALL of the following:
   a. Weakness without objective sensory loss in 2 or more nerves
   b. Electrodiagnostic studies are consistent with motor conduction block
   c. Normal sensory nerve conduction studies

16. Neoplastic disease

17. Chronic inflammatory demyelinating polyneuropathy (CIDP) with ALL of the following:
   a. Moderate to severe functional disability
   b. Electrodiagnostic studies are consistent with multifocal demyelinating abnormalities

18. Autoimmune encephalitis

19. Lambert-Eaton Myasthenic syndrome (LEMS)

20. Parvovirus B 19-induced pure red cell aplasia (PRCA)

21. Stiff-person Syndrome with ALL of the following:
   a. Inadequate response, intolerance or contraindication to first–line treatments (benzodiazepine or baclofen)

AND ONE of the following:
   a. Monitor patients carefully for signs and symptoms of thrombosis both at the time of infusion and after infusion
   b. Patients or caregivers have been instructed on how to monitor for signs and symptoms of thrombosis when self-administering the medication
IVIG (intravenous immunoglobulin)  
Bivigam, Carimune NF, Flebogamma, Gammagard, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Privigen

Prior - Approval Limits  
Duration 12 months

Prior – Approval Renewal Requirements

Diagnoses

Patient must have **ONE** of the following:

1. Primary Immunodeficiency Disease (PID) with **ONE** of the following:  
   a. Hypogammaglobulinemia, IgG subclass deficiency, selective IgA deficiency, selective IgM deficiency, or specific antibody deficiency  
   b. SCID (severe combined immunodeficiency disease) or Agammaglobulinemia  
   c. Wiskott-Aldrich syndrome, DiGeorge syndrome, or ataxia-telangiectasia (or other non SCID combined immunodeficiency)  
   d. CVID (common variable Immunodeficiency disease)  
      i. Age 4 years and older

   **AND ALL** of the following:  
   a. Reduction in frequency of bacterial and viral infections has been documented since initiation  
   b. IgG trough levels are monitored at least yearly and maintained at or above the lower range of normal for age (when applicable for indication)  
   c. The prescriber will re-evaluate the dose of the SCIG and reconsider a dose adjustment

2. Idiopathic thrombocytopenic purpura (ITP)

3. B-cell chronic lymphocytic leukemia  
   a. Reduction in frequency of bacterial and viral infections has been documented since initiation

4. Kawasaki syndrome

5. Prophylaxis of bacterial and viral infections in Bone marrow transplantation (BMT) / Hematopoietic stem cell transplantation (HSCT) Recipients  
   a. Reduction in frequency of bacterial and viral infections has been documented since initiation

IVIG FEP Clinical Criteria
IVIG (intravenous immunoglobulin)
Bivigam, Carimune NF, Flebogamma, Gammagard, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Privigen

6. Peripheral blood progenitor cell (PBPC) collection

7. Umbilical Cord Stem Cell Transplantation

8. Prophylaxis of bacterial and viral infections in HIV-Infected Pediatric
   a. Reduction in frequency of bacterial and viral infections has been documented since initiation

9. Polymyositis or Dermatomyositis
   a. Significant improvement in disability and maintenance of improvement since initiation

10. Inclusion-body myositis

11. Guillain-Barre Syndrome (GBS)

12. Fetal alloimmune thrombocytopenia (F/NAIT)

13. Myasthenia gravis

14. Multiple sclerosis

15. Multifocal motor neuropathy (MMN) with ALL of the following:
   a. Significant improvement in disability and maintenance of improvement since initiation

16. Neoplastic disease

17. Chronic inflammatory demyelinating polyneuropathy (CIDP) with ALL of the following:
   a. Significant improvement in disability and maintenance of improvement since initiation
   b. IVIG is being used at the lowest effective dose and frequency
   c. Chronic stable patients have been tapered and/or treatment withdrawn to determine whether continued treatment is necessary

18. Autoimmune encephalitis

19. Lambert-Eaton Myasthenic syndrome (LEMS)
20. Parvovirus B 19-induced pure red cell aplasia (PRCA)

21. Stiff-person Syndrome

Prior – Approval *Renewal Limits*

**Duration**  12 months