Pre-PA Allowance
None

Prior-Approval Requirements

Diagnoses

Patient must have ONE of the following documented indications:

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 < 500 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) Selective IgM deficiency: IgM level < 30 mg/dL with normal IgG and IgA levels
         4) 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
         5) 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 < 200 mg/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
IVIG (intravenous immunoglobulin)
Asceniv, Bivigam, Flebogamma, Gammagard, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Panzyga, Privigen

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

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

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

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

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
IVIG (intravenous immunoglobulin)
Ascending, Bivigam, Flebogamma, Gammagard, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Panzyga, Privigen

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:
      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 **ONE** 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
   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. Electrodagnostic 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. Electrodagnostic studies are consistent with multifocal demyelinating abnormalities

18. Autoimmune encephalitis
   a. Confirmation of diagnosis with **TWO** of the following tests:
      i. Neuroimaging
      ii. Electroencephalography (EEG)
      iii. Lumbar puncture
      iv. Serologic testing

19. Lambert-Eaton Myasthenic syndrome (LEMS)
IVIG (intravenous immunoglobulin)
Asceniv, Bivigam, Flebogamma, Gammagard, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Panzyga, Privigen

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 for **ALL** indications:
   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

   **AND** the following for **ALL** indications:
   a. **NO** concurrent therapy with another IVIG / SCIG product

**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
IVIG (intravenous immunoglobulin)
Asceniv, Bivigam, Flebogamma, Gammagard, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Panzyga, Privigen

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 IVIG 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

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
   a. Improvement in disability and maintenance of improvement since initiation confirmed by neurological exam

19. Lambert-Eaton Myasthenic syndrome (LEMS)

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

21. Stiff-person Syndrome

AND ONE of the following for ALL indications:
   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

AND the following for ALL indications:
   a. NO concurrent therapy with another IVIG / SCIG product

Prior – Approval Renewal Limits
Same as above