<table>
<thead>
<tr>
<th>Brand Name</th>
<th>Generic Name</th>
<th>Length of Authorization</th>
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<tbody>
<tr>
<td>Bivigam</td>
<td>IVIG</td>
<td>Per Medical Guidelines</td>
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<tr>
<td>Carimune</td>
<td>IVIG</td>
<td>Per Medical Guidelines</td>
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<tr>
<td>Flebogamma</td>
<td>IVIG</td>
<td>Per Medical Guidelines</td>
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<tr>
<td>Gammagard</td>
<td>IVIG/SCIG</td>
<td>Per Medical Guidelines</td>
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<td>Gammaked</td>
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<tr>
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<td>Hizentra</td>
<td>SCIG</td>
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<td>HyQvia</td>
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<td>Octagam</td>
<td>IVIG</td>
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<td>Privigen</td>
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<tr>
<td>Vivaglobin</td>
<td>SCIG</td>
<td>Per Medical Guidelines</td>
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</table>

**Approvable Criteria:**

1. **Immunodeficiency, Primary Humoral (Treatment).** Approve in patients who meet the following criteria (a and b):
   a. IVIG is prescribed by or in consultation with one of the following physician specialists: an allergist/immunologist, immunologist, otolaryngologist (ear nose and throat [ENT] physician), pulmonologist, or an infectious diseases physician who treats patients with primary immune deficiencies; **AND**
   b. The patient has one of the following primary humoral or combined immune deficiencies (i, ii, iii, iv, v, vi, or vii):
      i. **Common Variable Immunodeficiency (CVID) OR unspecified hypogammaglobulinemia** **AND** the patient meets the following criteria (1, 2, 3 and 4):
         i. Patient has a documented history of significant recurrent or persistent, severe bacterial infections (such as recurrent pneumonias, frequent episodes of bacterial infections such as sinusitis, otitis, bronchitis, skin structure infections, or infections of the gastrointestinal tract) according to the prescribing physician; **AND**
         ii. Infections are responding inadequately to treatment with antibiotics and/or appropriate prophylaxis with antibiotics OR the patient has multiple antibiotic hypersensitivities that interfere with treatment according to the prescribing physician; **AND**
         iii. Other disorders that may increase susceptibility to infection such as allergy or anatomic defects, have been sought out and treated aggressively if present according to the prescribing physician; **AND**
         iv. The patient has at least ONE of the following according to the prescribing physician:
            • Reduced total serum IgG level (age-adjusted and according to the normal reference range for the reporting laboratory); **OR**

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IMMUNE GLOBULIN (IVIG AND SCIG)

- Reduced IgG1 and IgG3 subclass levels or reduced IgG1 alone according to the normal reference range for the reporting laboratory (Patients with low levels of IgG1 and/or IgG2 will almost always have reduced levels of total IgG.); OR
- Markedly impaired antibody response to protein (e.g., tetanus, diphtheria) antigen OR antibody testing with a polysaccharide antigen (pneumococcus) can be used instead of a protein if the patient already has antibodies to tetanus and diphtheria.

OR

ii. X-linked agammaglobulinemia (Bruton’s agammaglobulinemia, congenital agammaglobulinemia); OR

iii. Severe Combined Immunodeficiency (SCID); OR

iv. Wiskott-Aldrich syndrome; OR

2. Idiopathic (Immune) Thrombocytopenic Purpura (ITP) or Immune Thrombocytopenia (IT), Acute and Chronic. Approve if the patient meets the following criteria (a, b, or c):
   a. Adults and adolescents (> 17 years of age) with ITP/IT. Approve for ONE of the following (i, ii, or iii):
      i. Acute bleeding in a patient who is newly diagnosed or requiring therapy for the first time OR in patients with persistent or chronic ITP. Approve IVIG for 1 month if the patient meets the following criteria (1, 2 and 3):
         i. IVIG is prescribed by or in consultation with a hematologist; AND
         ii. One of the following applies:
            • The patient has tried a systemic corticosteroid (e.g., prednisone) for ITP/IT; OR
            • There is an urgent need to increase the platelet count quickly AND IVIG will be started with a systemic corticosteroid; OR
            • A corticosteroid is contraindicated according to the prescribing physician; AND
         iii. The platelet count is < 30,000 per mm$^3$ (microliter).
      OR
      ii. To increase platelet counts before surgical procedures (e.g., splenectomy) or dental procedures, approve IVIG for 1 month if the patient meets the following criteria (1 and 2):
         i. IVIG is prescribed by or in consultation with a hematologist; AND
         ii. The platelet count is < 50,000 per mm$^3$ OR if the patient is undergoing major surgery (e.g., central nervous system or cardiac surgery) and the platelet count is < 75,000 per mm$^3$.
      OR
      iii. The patient has persistent (3 to 12 months duration) or chronic (≥ 12 months duration) ITP/IT. Approve IVIG if the patient meets the following criteria (1, 2 and 3):
         i. IVIG is prescribed by or in consultation with a hematologist; AND

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IMMUNE GLOBULIN (IVIG AND SCIG)

ii. One of the following applies:
   • The patient has tried a systemic corticosteroid (e.g., prednisone) for ITP/IT; OR
   • There is an urgent need to increase the platelet count quickly AND IVIG will be started with a systemic corticosteroid; OR
   • A corticosteroid is contraindicated according to the prescribing physician; AND

   iii. IVIG is required to prevent bleeding.

b. Children and adolescents (≤ 17 years of age) with ITP/IT. Approve for one of the following (i, ii, or iii):
   i. Acute bleeding in a patient who is newly diagnosed or requiring therapy for the first time OR in patients with persistent or chronic ITP. Approve for one month if the patient meets the following criteria (1 and 2):
      1. IVIG is prescribed by or in consultation with a hematologist; AND
      2. There is significant acute mucous membrane bleeding or other noncutaneous bleeding.
   OR
   ii. The patient has **persistent** (3 to 12 months) or **chronic** (≥ 12 months) ITP/IT. Approve if the patient meets the following criteria (1 and 2):
      i. IVIG is prescribed by or in consultation with a hematologist; AND
      ii. IVIG is required to prevent bleeding.
   OR
   iii. To increase the platelet count before major surgery such as splenectomy, or before other surgery, dental extraction(s), or other procedures likely to cause blood loss. Approve for 1 month if IVIG is prescribed by or in consultation with a hematologist.

3. B-Cell Chronic Lymphocytic Leukemia (CLL) for Prevention of Bacterial Infections. Approve in patients who meet the following criteria (a, b, and c):
   a. The patient has an IgG level < 500 mg/dL (5.0 g/L); **AND**
   b. The patient has a history of a serious bacterial infection that required IV antibiotic therapy or hospitalization; **AND**
   c. IVIG is prescribed by or in consultation with an oncologist, hematologist, or infectious diseases physician.

4. Hematopoietic Cell Transplantation (HCT) to Prevent Bacterial Infection. Approve IVIG for 6 months in patients who meet the following criteria (a, b, c, and d):
   a. IVIG is prescribed by or in consultation with a hematologist, oncologist or infectious diseases physician; **AND**
   b. The patient has had a HCT within the previous year; **AND**

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c. The patient has an IgG level < 500 mg/dL OR if the patient has multiple myeloma or malignant macroglobulinemia they are not required to have IgG level < 500 mg/dL (In the professional opinion of a specialist physician, we have adopted this criterion for IgG level < 500 mg/dL); **AND**
d. According to the prescribing physician the patient has a significant risk of having frequent and/or severe bacterial infections despite antibiotic therapy.

5. **Kawasaki Disease.** Approve IVIG for 1 day in patients who meet the following criteria (a and b):
   a. IVIG is prescribed by or in consultation with a pediatric cardiologist or a pediatric infectious diseases physician; **AND**
   b. The patient has persistent or recrudescent (recurring) fever or signs of inflammation 24 to 48 hours after completing the initial IVIG infusion(s).
      *These criteria assume that the first dose was given in the hospital. Patients should receive a single dose of IVIG together with aspirin within the first 10 days of illness, and if possible, within 7 days of illness.*

6. **Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) or Polyradiculoneuropathy.** Approve if IVIG is prescribed by or in consultation with a neurologist.

7. **Multifocal Motor Neuropathy (MMN) (Treatment).** Approve if IVIG is prescribed by or in consultation with a neurologist.

8. **Human Immunodeficiency Virus (HIV)-Infected Infants and Children to Prevent Recurrent Bacterial Infections.** Approve in patients who meet the following criteria (a, b, c, and d):
   a. IVIG is prescribed by or in consultation with an infectious diseases specialist or an immunologist; **AND**
   b. The patient is < 13 years of age; **AND**
   c. The patient is receiving cART (Note: cART is a combination of usually three or more antiretroviral drugs as part of an HIV treatment regimen that is designed to achieve virologic suppression.); **AND**
   d. The patient has ONE of the following (i, ii or iii):
      i. Hypogammaglobulinemia (IgG < 400 mg/dL); **OR**
      ii. Functional antibody deficiency is demonstrated by poor specific antibody titers (that is, the patient does not develop specific antibody responses against protein and polysaccharide antigens); **OR**
      iii. Functional antibody deficiency is demonstrated by the patient having recurrent (two or more per year), serious bacterial infections (e.g., bacteremia, meningitis, pneumonia) despite administration of cART and appropriate antimicrobial prophylaxis.

9. **Autoimmune Mucocutaneous Blistering Diseases (Pemphigus Vulgaris, Pemphigus Foliaceus, Bullous Pemphigoid, Mucous Membrane Pemphigoid [Cicatricial Pemphigoid], and Epidermolysis Bullosa Acquisita).** Approve in children or adults who meet the following criteria (a and b):
   a. IVIG is prescribed by or in consultation with a dermatologist; **AND**

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b. The patient meets ONE of the following criteria (i, ii, or iii):
   i. The patient has tried a systemic corticosteroid OR has a contraindication to treatment with a corticosteroid according to the prescribing physician AND has tried an immunosuppressive agent (e.g., azathioprine, cyclophosphamide, dapsone, methotrexate [MTX], cyclosporine, mycophenolate mofetil, tacrolimus) OR has a contraindication to trying an immunosuppressive agent according to the prescribing physician; OR
   ii. The patient has rapid, debilitating, progressive disease, that cannot be controlled with a systemic corticosteroid and an immunosuppressive agent; OR
   iii. The disease is so serious that there is inadequate time for therapy with a systemic corticosteroid and an immunosuppressive agent to have a rapid enough effect.

10. Dermatomyositis or Polymyositis. Approve in patients who meet the following criteria (a, b and c):
   a. IVIG is prescribed by or in consultation with a neurologist or a rheumatologist; AND
   b. The patient has tried a systemic corticosteroid OR a corticosteroid is contraindicated according to the prescribing physician; AND
   c. The patient has tried an immunosuppressant agent (e.g., azathioprine, MTX, cyclosporine, cyclophosphamide, mycophenolate mofetil) OR an immunosuppressant agent is contraindicated according to the prescribing physician.

11. Desensitization Therapy Prior to and Immediately after Solid Organ (Kidney, Heart, Lung, Liver, Intestinal) Transplantation. Approve IVIG if prescribed by or in consultation with a physician affiliated with a transplant center.

12. Guillain-Barré Syndrome (GBS). Approve for 1 month in patients who meet the following criteria (a and b):
   a. IVIG is prescribed by or in consultation with a neurologist or a specialist with experience in diagnosing and treating patients with GBS; AND
   b. The patient meets one of the following criteria (i or ii):
      i. IVIG is initiated within 2 weeks and no longer than 4 weeks of onset of neuropathic symptoms (weakness, inability to stand or walk without assistance, respiratory or bulbar weakness; patients are hospitalized); OR
      ii. The patient has had a relapse, but had an initial response to IVIG.

13. Multiple Myeloma. Approve in patients who meet the following criteria (a, b and c):
   a. The patient has stable (plateau phase) disease (> 3 months from diagnosis); AND
   b. The patient has severe recurrent bacterial infections according to the prescribing physician; AND
   c. IVIG is prescribed by or in consultation with a hematologist, oncologist, or infectious diseases specialist.

14. Multiple Sclerosis (MS), Acute Severe Exacerbation. Approve for 5 days in patients who meet the following criteria (a, b, and c):

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a. IVIG is prescribed by or in consultation with, a neurologist or a physician who specializes in the treatment of MS; AND

b. The patient has a documented inadequate response to an appropriate trial with an immunomodulatory drug (Avonex, Betaseron/Extavia, Rebif, or Copaxone) [Compliance must be verified]; AND

c. The patient has either not responded to or has had a significant adverse reaction AND is continuing to deteriorate despite therapy with ONE of the following (1, 2 or 3):
   - Oral or IV corticosteroids (e.g., methylprednisolone sodium succinate injection); OR
   - Plasma exchange; OR
   - Acthar® H.P. gel (Acthar) [repository corticotropin injection; adrenocorticotropic hormone {ACTH}];

15. Myasthenia Gravis. Approve IVIG for 5 days in patients who meet the following criteria (a and b).

a. IVIG is prescribed by or in consultation with a neurologist; AND

b. The patient meets ONE of the following criteria (i, ii, or iii):
   - The patient has an exacerbation of myasthenia gravis; OR
   - The patient requires stabilization of myasthenia gravis before surgery; OR
   - The patient has been started on an immunosuppressive drug (e.g., azathioprine, cyclosporine, cyclophosphamide, mycophenolate mofetil) and is waiting for full effect.

SPECIALTY PHARMACY PRODUCT

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# IMMUNE GLOBULIN (IVIG AND SCIG)

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<tr>
<th>Indication</th>
<th>Recommended Dosing Regimen (based on Ideal Body Weight)</th>
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</thead>
<tbody>
<tr>
<td><strong>Primary immunodeficiency</strong></td>
<td><em>Dose recommendations may vary by manufacturer.</em></td>
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<tr>
<td>- Congenital agammaglobulinemia</td>
<td>200-400 mg/kg monthly (adjust for trough IgG of 400-500 mg/dL, and correlate with patient response)</td>
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<tr>
<td>- Common variable immunodeficiency</td>
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<tr>
<td>- Wiskott-Aldrich Syndrome</td>
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<td>- X-linked agammaglobulinemia</td>
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<td>- Severe combined immunodeficiency</td>
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<tr>
<td><strong>Idiopathic thrombocytopenia purpura (ITP)</strong></td>
<td>Initial: 400mg/kg/day for 5 days or 1000mg/kg/day for 1-2 days. Maintenance: 400-1000mg/kg intermittently to maintain platelet count &gt;20,000/mm³ (may consider Rho-gam if Rh(+) 50 – 75 mcg/kg as an alternative)</td>
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<tr>
<td>- platelets less than 20,000 or actively bleeding</td>
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<tr>
<td><strong>B-cell chronic lymphocytic leukemia (CLL)</strong></td>
<td>400 mg/kg/dose every 3-4 weeks</td>
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<tr>
<td><strong>Bone marrow transplantation</strong></td>
<td>500 mg/kg 7 and 2 days before transplant, then 500 mg/kg weekly until 90 days after transplant (Investigators have confirmed that the half-life of IgG is significantly reduced (30 hours to 10 days) in bone marrow transplant recipients compared to normal individuals (Rand et al, 1989; Hagenbeek et al, 1987).)</td>
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<tr>
<td><strong>HIV-infected children</strong></td>
<td>400 mg/kg every 28 days</td>
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<tr>
<td><strong>Kawasaki syndrome</strong></td>
<td>400 mg/kg for 4 consecutive days, or single dose of 2,000 mg/kg; oral aspirin 80-100 mg/kg daily (in 4 divided doses) orally until the 14th day of illness, then 3-5 mg/kg for a period of 5 weeks</td>
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<tr>
<td><strong>Guillain-Barre syndrome</strong></td>
<td>IVIG is recommended as an equivalent alternative to plasma exchange in children and adults. 400mg/kg/day for 5 days or 1 gm/kg/day x 2 days</td>
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<tr>
<td><strong>Chronic inflammatory demyelinating polyneuropathy (CIDP)</strong></td>
<td>IVIG is recommended as an equivalent alternative to plasma exchange in children and adults. Loading dose: 2000mg/kg given in divided doses over 2-4 days Maintenance: 1000mg/kg over 1 day every 3 weeks</td>
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<td><strong>Dermatomyositis</strong></td>
<td>2 g/kg IV in two separate doses once per month for 3 months.</td>
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<tr>
<td><strong>Multiple Sclerosis, acute exacerbation</strong></td>
<td>Either a single 1 g/kg dose or 0.4 g/kg daily for 5 days.</td>
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<tr>
<td><strong>Multifocal Motor Neuropathy (MMN)</strong></td>
<td>0.5 – 2.4 g/kg/month IV.</td>
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<tr>
<td><strong>Myasthenia Gravis</strong></td>
<td>400 mg/kg IV once daily for 5 days.</td>
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</table>

*References:
- Adapted from Express Scripts Prior Authorization Policy. Last updated 02/04/2015.*