AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name:	Subcutaneous Immune Globulins	Page:	1 of 21
Effective Date:	3/30/2026	Last Review Date:	11/21/2025

Applies to:	☒Illinois	☒Florida Kids	☒New Jersey
	☒Pennsylvania Kids	☐Virginia	☒Kentucky PRMD

Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Subcutaneous Immune Globulins under the patient's prescription drug benefit.

Description:

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. Cutaquig (Immune Globulin Subcutaneous [Human] - hipp, 16.5% Solution)
      Cutaquig is indicated as replacement therapy for primary humoral immunodeficiency (PI) in adults and pediatric patients 2 years of age and older.
   2. Cuvitru (Immune Globulin Subcutaneous [Human], 20% Solution)
      Cuvitru is indicated as replacement therapy for primary humoral immunodeficiency in adult and pediatric patients two years of age and older.
   3. Hizentra (Immune Globulin Subcutaneous [Human], 20% Liquid)
      a. Hizentra is indicated as replacement therapy for primary humoral immunodeficiency in adults and pediatric patients 2 years of age and older.
      b. 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.
   4. 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.
   5. Xembify (Immune Globulin Subcutaneous [Human} – klhw, 20% Solution)
      Xembify is indicated for treatment of primary humoral immunodeficiency (PI) in patients 2 years of age and older.

B. Compendial Uses
   1. Idiopathic thrombocytopenic purpura (ITP)

2. Multifocal motor neuropathy
3. Kawasaki syndrome
4. B-cell chronic lymphocytic leukemia (CLL)
5. Prophylaxis of bacterial infections in pediatric human immunodeficiency virus (HIV) infection
6. Bone marrow transplant (BMT)/hematopoietic stem cell transplant (HSCT)
7. Dermatomyositis
8. Polymyositis
9. Myasthenia gravis
10. Guillain-Barré syndrome
11. Lambert-Eaton myasthenic syndrome
12. Fetal/neonatal alloimmune thrombocytopenia
13. Parvovirus B19-induced pure red cell aplasia
14. Stiff-person syndrome
15. Management of immune checkpoint inhibitor-related toxicities
16. Acquired red cell aplasia
17. Acute disseminated encephalomyelitis
18. Autoimmune mucocutaneous blistering diseases
19. Autoimmune hemolytic anemia
20. Autoimmune neutropenia
21. Birdshot retinochoroidopathy
22. BK virus associated nephropathy
23. Churg-Strauss Syndrome
24. Enteroviral meningoencephalitis
25. Hematophagocytic lymphohistiocytosis (HLH) or macrophage activation syndrome (MAS)
26. Hemolytic disease of newborn
27. HIV-associated thrombocytopenia
28. Hyperimmunoblobulinemia E Syndrome
29. Hypogammaglobulinemia from chimeric antigen receptor T (CAR-T) therapy
30. Multiple myeloma
31. Neonatal hemochromatosis, prophylaxis
32. Opsoclonus-myoclonus
33. Paraneoplastic opsonus-myoclonus ataxia associated with neuroblastoma
34. Pediatric Acute-onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS)
35. Post-transfusion purpura
36. Rasmussen encephalitis
37. Renal transplantation from a live donor with ABO incompatibility or positive cross match
38. Retinocochleocerebral vasculopathy, Central nervous system-predominant

39. Secondary immunosuppression associated with major surgery, hematological malignancy, major burns, and collagen-vascular diseases
40. Solid organ transplantation, for allosensitized members
41. Toxic epidermal necrolysis and Stevens-Johnson syndrome
42. Toxic shock syndrome
43. Systemic lupus erythematosus (SLE)
44. Toxic necrotizing fasciitis due to group A streptococcus
45. Measles (Rubeola) prophylaxis
46. Tetanus treatment and prophylaxis
47. Varicella prophylaxis

All other indications are considered experimental/investigational and not medically necessary.

Applicable Drug List:

PREFERRED AGENT:
Hizentra

NON-PREFERRED AGENTS:
HyQvia
Cutaquig
Cuvitru
Xembify

Policy/Guideline:

Documentation for all indications:

Requests for non-preferred subcutaneous immune globulin (SCIG) products required that the patient is unable to take ONE preferred subcutaneous immune globulin (SCIG) product or ONE intravenous immune globulin (IVIG) for the given diagnosis due to a trial and inadequate treatment response or intolerance, or a contraindication. Documentation is required for approval.

Documentation:

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

A. Primary immunodeficiency
   1. Diagnostic test results
      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 IG therapy

B. Myasthenia gravis
1. Clinical records describing standard treatments tried and failed

C. Secondary hypogammaglobulinemia (CLL, HIV, BMT/HSCT recipient)
1. Copy of laboratory report with pre-treatment serum IgG level

D. Chronic inflammatory demyelinating polyneuropathy (CIDP) and multifocal motor neuropathy (MMN)
1. Pre-treatment electrodiagnostic studies (electromyography [EMG] or nerve conduction studies [NCS])

E. Dermatomyositis and polymyositis
1. Clinical records describing standard treatments tried and failed

F. Lambert-Eaton Myasthenic Syndrome (LEMS)
1. Neurophysiology studies (e.g., electromyography)
2. A positive anti- P/Q type voltage-gated calcium channel antibody test

G. Idiopathic thrombocytopenic purpura
1. Laboratory report with pre-treatment/current platelet count
2. Chronic/persistent ITP: copy of medical records supporting trial and failure with corticosteroid or anti-D therapy (unless contraindicated)

H. Parvovirus B19-indicated Pure Red Cell Aplasia (PRCA)
1. Copy of test result confirming presence of parvovirus B19

I. Pediatric Acute-onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS)
1. Medical records confirming the diagnosis and previous treatment with systemic corticosteroids for those initiating the IG therapy
2. Medical records documenting a clinical response for those continuing with IG therapy

J. Stiff-person syndrome
1. Anti-glutamic acid decarboxylase (GAD) antibody testing results
2. Clinical records describing standard treatments tried and failed

K. Toxic shock syndrome or toxic necrotizing fasciitis due to group A streptococcus
1. Documented presence of fasciitis (toxic necrotizing fasciitis due to group A streptococcus only)
2. Microbiological data (culture or Gram stain)

Criteria for Initial Approval:

A. Primary Immunodeficiency

Initial authorization of 6 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 2 years or older, and
   b. Other causes of immune deficiency have been excluded (eg, drug induced, genetic disorders, infectious diseases such as HIV, malignancy), and
   c. Pretreatment IgG level < 500 mg/dL or ≥ 2 SD below the mean for age, and
   d. History of recurrent bacterial infections, and
   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, and
   b. Impaired antibody response to pneumococcal polysaccharide vaccine (see Appendix A), and
   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 12 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 IG 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 IG and consider a dose adjustment (when appropriate).

B. Myasthenia Gravis

1. Authorization of 1 month may be granted to members who are prescribed IG 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 6 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. Disease course is progressive or relapsing/remitting for 2 months or longer
   b. Moderate to severe functional disability
   c. The diagnosis was confirmed by electrodiagnostic studies
2. Re-authorization of 6 months may be granted when the following criteria are met:
   a. Significant improvement in disability and maintenance of improvement since initiation of IG therapy
   b. IG 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. Member has at least 4 of the following:
      i. Proximal muscle weakness (upper or lower extremity and trunk)

ii. Elevated serum creatine kinase (CK) or aldolase level
iii. Muscle pain on grasping or spontaneous pain
iv. Myogenic changes on EMG (short-duration, polyphasic motor unit potentials with spontaneous fibrillation potentials)
v. Positive for anti-synthetase antibodies (e.g., anti-Jo-1, also called histadyl tRNA synthetase)
vi. Non-destructive arthritis or arthralgias
vii. Systemic inflammatory signs (fever: more than 37°C at axilla, elevated serum CRP level or accelerated ESR of more than 20 mm/h by the Westergren method)
viii. Pathological findings compatible with inflammatory myositis (inflammatory infiltration of skeletal evidence of active regeneration may be seen), and

b. Standard first-line treatments (corticosteroids) and second-line treatments (immunosuppressants) have been tried but were unsuccessful or not tolerated, or
c. Member is unable to receive standard first-line and second-line therapy because of a contraindication or other clinical reason.

2. Re-authorization of 6 months may be granted when the following criterion is met:
a. Significant improvement in disability and maintenance of improvement since initiation of IG therapy

E. Idiopathic Thrombocytopenic Purpura ITP/(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
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 IG will be used alone or IG 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 IG 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 all of the following criteria are met:
   a. IG 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 a reduction in the frequency of bacterial infections has been demonstrated since initiation of IG 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 any of the following criteria are met:
   a. IG is prescribed for primary prophylaxis of bacterial infections and pretreatment serum IgG < 400 mg/dL, or
   b. IG 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), or
   c. Member has failed to form antibodies to common antigens, such as measles, pneumococcal, and/or Haemophilus influenzae type b vaccine, or
   d. Member lives in an area where measles is highly prevalent and who have not developed an antibody response after two doses of measles, mumps, and rubella virus vaccine live, or
   e. Member has been exposed to measles and request is for a single dose, or
   f. Member has chronic bronchiectasis that is suboptimally responsive to antimicrobial and pulmonary therapy
2. Re-authorization of 6 months may be granted when a reduction in the frequency of bacterial infections has been demonstrated since initiation of IG therapy.

H. Bone marrow transplant/hemopoietic stem cell transplant (BMT/HSCT)

1. Initial authorization of 6 months may be granted to members who are BMT/HSCT recipients when the following criteria are met:
   a. Therapy will be used to prevent the risk of acute graft-versus-host disease, associated interstitial pneumonia (infectious or idiopathic), septicemia, and other infections (e.g., cytomegalovirus infections [CMV], recurrent bacterial infection)
   b. Either of the following:
      i. IG 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 a reduction in the frequency of bacterial infections has been demonstrated since initiation of IG therapy.

I. Multifocal Motor Neuropathy (MMN)

1. Initial authorization of 3 months may be granted when the following criteria are met:
   a. Member experienced progressive, multifocal, asymmetrical weakness without objective sensory loss in 2 or more nerves for at least 1 month
   b. The diagnosis was confirmed by electrodiagnostic studies
2. Re-authorization of 6 months may be granted when significant improvement in disability and maintenance of improvement have occurred since initiation of IG therapy

J. Guillain-Barre Syndrome (GBS)

Authorization of 1 month total may be granted for GBS when the following criteria are met:

1. Member has severe disease with significant weakness (e.g., inability to stand or walk without aid, respiratory weakness)
2. Onset of neurologic symptoms occurred less than 4 weeks from the anticipated start of therapy

K. Lambert-Eaton Myasthenic Syndrome (LEMS)

1. Initial authorization of 6 months may be granted for LEMS when the following criteria are met:
   a. Diagnosis has been confirmed by either of the following:
      i. Neurophysiology studies (e.g., electromyography)
      ii. A positive anti- P/Q type voltage-gated calcium channel antibody test
   b. Anticholinesterases (eg pyridostigmine) and amifampridine (e.g., 3,4-diaminopyridine phosphate, Firdapse) have been tried but were unsuccessful or not tolerated
   c. Weakness is severe or there is difficulty with venous access for plasmapheresis

2. Re-authorization of 6 months may be granted when member is responding to therapy (i.e., there is stability or improvement in symptoms relative to the natural course of 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 severe, refractory anemia associated with bone marrow suppression, with parvovirus B19 viremia.

O. Stiff-person Syndrome
Authorization of 6 months may be granted for stiff-person syndrome when the following criteria are met:
1. Diagnosis has been confirmed by anti-glutamic acid decarboxylase (GAD) antibody testing
2. Member had an inadequate response to first-line treatment (benzodiazepines and/or baclofen)

P. Management of immune checkpoint inhibitor-related toxicities
Authorization of 1 month may be granted for management of immune checkpoint-inhibitor toxicities when all of the following criteria are met:
1. Member has experienced a moderate or severe adverse event to a PD-1 or PD-L1 inhibitor (eg, pembrolizumab, nivolumab, atezolizumab, avelumab, durvalumab)
2. The offending medication has been held or discontinued
3. Member experienced one or more of the following nervous system adverse events: myocarditis, bullous dermatitis, Stevens-Johnson syndrome, toxic epidermal necrolysis, pneumonitis, myasthenia gravis, peripheral neuropathy, encephalitis, transverse myelitis, severe inflammatory arthritis, Guillain-Barre syndrome, or steroid-refractory myalgias or myositis

Q. Acquired Red Cell Aplasia
Authorization of 6 months may be granted for acquired red cell aplasia.

R. Acute Disseminated Encephalomyelitis
Authorization of 1 month may be granted for acute disseminated encephalomyelitis in members who have had an insufficient response or a contraindication to intravenous corticosteroid treatment.

S. Autoimmune Mucocutaneous Blistering Disease

Authorization of 6 months may be granted for autoimmune mucocutaneous blistering disease (includes pemphigus vulgaris, pemphigus foliaceus, bullous pemphigoid, mucous membrane pemphigoid, and epidermolysis bullosa aquisita) when the following criteria are met:

1. Diagnosis has been proven by biopsy and confirmed by pathology report, and
2. Condition is rapidly progressing, extensive or debilitating, and
3. Member has failed or experienced significant complications (eg diabetes, steroid-induced osteoporosis) from standard treatment (corticosteroids, immunosuppressive agents).

T. Autoimmune Hemolytic Anemia

Authorization of 6 months may be granted for warm-type autoimmune hemolytic anemia in members who do not respond or have a contraindication to corticosteroids or splenectomy.

U. Autoimmune Neutropenia

Authorization of 6 months may be granted for autoimmune neutropenia where treatment with G-CSF (granulocyte colony stimulating factor) is not appropriate.

V. Birdshot Retinochoroidopathy

Authorization of 6 months may be granted for birdshot (vitiliginous) retinochoroidopathy that is not responsive to immunosuppressives (eg corticosteroids, cyclosporine).

W. BK Virus Associated Nephropathy

Authorization of 6 months may be granted for BK virus associated nephropathy.

X. Churg-Strauss Syndrome

Authorization of 6 months may be granted for severe, active Churg-Strauss syndrome as adjunctive therapy for members who have experienced failure, intolerance, or are contraindicated to other interventions.

Y. Enteroviral Meningoencephalitis

Authorization of 6 months may be granted for severe cases of enteroviral meningoencephalitis.

Z. Hematophagocytic Lymphohistiocytosis (HLH) or Macrophage Activation Syndrome (MAS)

Authorization of 6 months may be granted for treatment of hypogammaglobulinemia in HLH or MAS when total IgG is less than 400 mg/dL or two standard deviations below the mean for age.

AA. Hemolytic Disease of Newborn
Authorization of 6 months may be granted for isoimmune hemolytic disease in neonates.

BB. HIV-associated Thrombocytopenia
Authorization of 6 months may be granted for HIV-associated thrombocytopenia when the following criteria are met:
1. Pediatric members with IgG < 400 mg/dL and has one of the following:
   a. 2 or more bacterial infections in a 1-year period despite antibiotic chemoprophylaxis with TMP-SMZ or another active agent, or
   b. Received 2 doses or measles vaccine and lives in a region with a high prevalence or measles, or
   c. HIV-associated thrombocytopenia despite anti-retroviral therapy, or
   d. Chronic bronchiectasis that is suboptimally responsive to antimicrobial and pulmonary therapy, or
   e. T4 cell count ≥ 200/mm^3
2. Adult members with significant bleeding, platelet count < 20,000/mcL, and failure of RhIG in Rh-positive patients

CC. Hyperimmunoglobulinemia E Syndrome
Authorization of 6 months may be granted to treat severe eczema in hyperimmunoglobulinemia E syndrome.

DD. Hypogammaglobulinemia from CAR-T therapy
Authorization of 6 months may be granted for members with IgG < 400 mg/dL receiving treatment with CAR-T therapy (including but not limited to idecabtagene vicleucel [Abecma], tisagenlecleucel [Kymriah], or axicabtagene ciloleucel [Yescarta]).

EE. Multiple Myeloma
Authorization of 6 months may be granted for multiple myeloma in members who have recurrent, serious infections despite the use of prophylactic antibiotics.

FF. Neonatal Hemochromatosis
Authorization of 6 months may be granted for prophylaxis in members who are pregnant with a history of pregnancy ending in documented neonatal hemochromatosis.

GG. Opsoclonus-myoclonus
Authorization of 6 months may be granted for treatment of either of the following:
1. Paraneoplastic opsoclonus-myoclonus-ataxia associated with neuroblastoma
2. Refractory opsoclonus-myoclonus, as last-resort treatment

HH. Pediatric Acute-onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS) (See Appendix C)

Initial Authorization of 6 months may be granted when the following criteria are met:

1. Child meets PANS Research Consortium Diagnostic Criteria for PANS/PANDAS:
    - Documentation of abrupt, dramatic onset (within less than one month) of obsessive-compulsive disorder or severely restricted food intake; and
    - Documentation of concurrent presence of additional neuropsychiatric symptoms, with similarly severe and acute onset, from at least two of the following seven categories:
        - Anxiety
        - Emotional lability and/or depression
        - Irritability, aggression, and/or severely oppositional behaviors
        - Behavioral (developmental) regression
        - Deterioration in school performance (related to attention deficit/hyperactivity disorder (ADHD)-like symptoms, memory deficits, cognitive changes)
        - Sensory or motor abnormalities
        - Somatic signs and symptoms, including sleep disturbances, enuresis, or urinary frequency; and
            - Onset of symptoms occurs between 3 years of age and puberty; and
            - Documentation that other causes of symptoms have been ruled out.
2. Child has tried and failed treatment with systemic corticosteroids.
3. Documented objective assessment of baseline symptoms has been submitted (e.g., Children's Yale-Brown Obsessive-Compulsive Scale [CY-BOCS], Clinical Global Impression of Severity [CGIS], Parent-Rated Pediatric Acute Neuropsychiatric Symptom Scale [PANS Scale]).

Re-authorization of 6 months maybe granted when documentation of objective clinical response to therapy has been submitted (e.g., Children's Yale-Brown Obsessive-Compulsive Scale [CY-BOCS], Clinical Global Impression of Severity [CGIS], Parent-Rated Pediatric Acute Neuropsychiatric Symptom Scale [PANS Scale]).

II. Post-transfusion Purpura

Authorization of 1 month may be granted for post-transfusion purpura.

JJ. Rasmussen Encephalitis

Authorization of 6 months may be granted for Rasmussen encephalitis in members whose symptoms do not improve with anti-epileptic drugs and corticosteroids.

KK. Renal Transplantation

Authorization of 6 months may be granted for a member undergoing renal transplantation from a live donor with ABO incompatibility or positive cross match.

LL. Retinocochleocerebral vasculopathy, Central nervous system-predominant
Authorization of 6 months may be granted for treatment of central nervous system (CNS)-predominant Retinocochleocerebral vasculopathy (Susac syndrome (SuS)) when used as an adjunctive therapy to corticosteroid and other treatment options

MM. Secondary Immunosuppression Associated with Major Surgery, Hematological Malignancy, Major Burns, and Collagen-Vascular Diseases
Authorization of 6 months may be granted to prevent or modify recurrent bacterial or viral infections in members with secondary immunosuppression (IgG < 400 mg/dL) associated with major surgery, hematological malignancy, extensive burns, or collagen-vascular disease.

NN. Solid Organ Transplantation
Authorization of 6 months may be granted for solid organ transplantation for allosensitized members.

OO. Toxic Epidermal Necrolysis and Stevens-Johnson Syndrome
Authorization of 1 month may be granted for severe cases of toxic epidermal necrolysis or Stevens-Johnson syndrome.

PP. Toxic Shock Syndrome
Authorization of 1 month may be granted for staphylococcal or streptococcal toxic shock syndrome when the infection is refractory to several hours of aggressive therapy, an undrainable focus is present, or the member has persistent oliguria with pulmonary edema.

QQ. Systemic Lupus Erythematosus
Authorization of 6 months may be granted for severe, active SLE in members who have experienced inadequate response, intolerance or have a contraindication to first and second line therapies.

RR. Measles (Rubeola) prophylaxis
Authorization of 1 month may be granted for postexposure prophylaxis to prevent or modify symptoms of measles (rubeola) in susceptible members exposed to the disease less than 6 days previously.

SS. Tetanus treatment and prophylaxis
Authorization of 1 month may be granted for treatment or postexposure prophylaxis of tetanus as an alternative when tetanus immune globulin (TIG) is unavailable.

TT. Varicella prophylaxis
Authorization of 1 month may be granted for postexposure prophylaxis of varicella in susceptible individuals when varicella-zoster immune globulin (VZIG) is unavailable.

UU. Toxic Necrotizing Fasciitis Due To Group A Streptococcus
Authorization of 1 month may be granted for members with fasciitis due to invasive streptococcal infection.

Continuation of Therapy:

Authorization may be granted for continuation of therapy when either the following criteria is met:

A. For conditions with reauthorization criteria listed under section III: Members who are currently receiving IG therapy must meet the applicable reauthorization criteria for the member's condition.
B. For all other conditions, all members (including new members) must meet initial authorization criteria.

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 (eg, peptic ulcer disease, hypertension)
- Mandated anticoagulation therapy
- Profession or lifestyle predisposes patient to trauma (eg, construction worker, fireman, professional athlete)

Appendix C

- Members suspected of having PANS/PANDAS should be evaluated with the following tests, as indicated:
  - Complete blood cell counts with manual differential
  - Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP)
    - If there are elevated inflammatory markers, fatigue, rashes, or joint pain, antinuclear antibody (ANA) or fluorescent antinuclear antibody (FANA) should be obtained (if ANA is elevated, the member should be evaluated for lupus)
  - Comprehensive metabolic panel
    - If liver function tests are abnormal or Kayser–Fleisher rings are present, the member should be evaluated for Wilson's disease with ceruloplasmin and 24 urine copper tests.

  - Throat culture, anti-streptolysin O (ASO) and anti-DNAse B
  - Urinalysis (to assess hydration) and to rule out inflammation for children with urinary complaints
    - If urinalysis reveals pyuria, clean-catch urine culture
  - Antiphospholipid antibody work up if the member has chorea, petechiae, migraines, stroke, thrombosis, thrombocytopenia, or levido rash. Workup should include:
    - anticardiolipin antibody,
    - dilute Russell's viper venom time (dRVVT),
    - b 2-glycoprotein I antibodies.
- Members should be evaluated for other causes of symptoms, including:
  - Autism/Autistic Spectrum Disorder
  - Developmental Delay/Mental Retardation
  - Cerebral palsy
  - Genetic conditions and syndromes (such as Down Syndrome, Fragile X, Rett Syndrome, etc.)
  - Cerebral malformation syndromes
  - Inborn errors of metabolism
  - CNS infections
  - Toxic cerebral insults (such as kernicterus, chemo-radiation, etc.)
  - Obsessive compulsive disorder
  - Anorexia nervosa
  - Avoidant/restrictive food intake disorder (ARFID)
  - Tourette syndrome
  - Transient tic disorder
  - Bipolar disorder
  - Sydenham chorea
  - Autoimmune encephalitis
  - Systemic autoimmune disease
  - Wilson's disease

Approval Duration and Quantity Restrictions:

Initial Approval:
1 month:

- Myasthenia Gravis with worsening weakness, acute exacerbation, or in preparation for surgery
- New diagnosed or initial therapy for Idiopathic Thrombocytopenic Purpura ITP/(Immune Thrombocytopenia)
- Guillain-Barre Syndrome (GBS)
- Kawasaki syndrome
- Management of immune checkpoint inhibitor-related toxicities

- Acute disseminated encephalomyelitis
- Post-transfusion Purpura
- Toxic Epidermal Necrolysis and Stevens-Johnson Syndrome
- Toxic Shock Syndrome
- Measles (Rubeola) prophylaxis
- Tetanus treatment and prophylaxis
- Varicella prophylaxis
- Toxic Necrotizing Fasciitis Due To Group A Streptococcus

3 months:

- Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)
- Dermatomyositis or Polymyositis
- Multifocal Motor Neuropathy (MMN)

6 months:

- Severe combined immunodeficiency (SCID) or congenital agammaglobulinemia
- Wiskott-Aldrich syndrome, DiGeorge syndrome, or ataxia-telangiectasia
- Common variable immunodeficiency (CVID)
- Hypogammaglobulinemia (unspecified), IgG subclass deficiency, selective IgA deficiency, selective IgM deficiency, or specific antibody deficiency
- Myasthenia Gravis with refractory myasthenia gravis who have tried and failed 2 or more standard therapies
- Chronic/persistent ITP
- Adults with refractory ITP after splenectomy
- B-cell Chronic Lymphocytic Leukemia (CLL)
- Prophylaxis of Bacterial Infections in HIV-Infected Pediatric Patients
- Bone marrow transplant/hemopoietic stem cell transplant (BMT/HSCT)
- Lambert-Eaton Myasthenic Syndrome (LEMS)
- Pediatric Acute-onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS)
- Fetal/Neonatal Alloimmune Thrombocytopenia (F/NAIT)
- Parvovirus B19-induced Pure Red Cell Aplasia (PRCA)
- Stiff-person syndrome
- Acquired red cell aplasia
- Autoimmune mucocutaneous blistering disease
- Autoimmune Hemolytic Anemia
- Autoimmune neutropenia
- Birdshot Retinochoroidopathy
- BK virus associated nephropathy
- Churg-Strauss syndrome
- Enteroviral meningoencephalitis

- Hematophagocytic Lymphohistiocytosis (HLH) or Macrophage Activation Syndrome (MAS)
- Hemolytic Disease of Newborn
- HIV-associated Thrombocytopenia
- Hyperimmunoglobulinemia E Syndrome
- Hypogammaglobulinemia from CAR-T therapy
- Multiple Myeloma
- Neonatal Hemochromatosis
- Opsoclonus-myoclonus
- Rasmussen encephalitis
- Renal transplantation
- Retinocochleocerebral vasculopathy, Central nervous system-predominant
- Secondary Immunosuppression Associated with Major Surgery, Hematological Malignancy, Major Burns, and Collagen-Vascular Diseases
- Solid organ transplantation
- Systemic Lupus Erythematosus

Authorization through delivery:

- Idiopathic Thrombocytopenic Purpura in pregnant women

Renewal Approval:
6 months:

- Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)
- Dermatomyositis or Polymyositis
- B-cell Chronic Lymphocytic Leukemia (CLL)
- Prophylaxis of Bacterial Infections in HIV-Infected Pediatric Patients
- Bone marrow transplant/hemopoietic stem cell transplant (BMT/HSCT)
- Multifocal Motor Neuropathy (MMN)
- Lambert-Eaton Myasthenic Syndrome (LEMS)
- Pediatric Acute-onset Neuropsychiatric Syndrome (PANS)/Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infections (PANDAS)

12 months:

- Severe combined immunodeficiency (SCID) or congenital agammaglobulinemia
- Wiskott-Aldrich syndrome, DiGeorge syndrome, or ataxia-telangiectasia
- Common variable immunodeficiency (CVID)
- Hypogammaglobulinemia (unspecified), IgG subclass deficiency, selective IgA deficiency, selective IgM deficiency, or specific antibody deficiency

Quantity Level Limit: Reference Formulary for drug specific quantity level limits

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