



GCHP Medi-Cal Clinical Guidelines

Immunoglobulins

**(AlygloTM, AscenivTM, BivigamTM, CutaquigTM, CuvitruTM,
Flebogamma DIFTM, GamaSTANTM, GammagardTM,
Gammagard S/DTM, GammakedTM, GammaplexTM, Gamunex-
CTM, HizentraTM, HyqviaTM, OctagamTM, PanzygaTM,
PrivigenTM, XembifyTM)**

PA Criteria	Criteria Details
Covered Uses (FDA approved indication)	<p>GamastanTM</p> <ul style="list-style-type: none"> Anti-viral prophylaxis <p>Gammagard LiquidTM, GammakedTM, Gamunex-CTM, HizentraTM, HyQviaTM, PanzygaTM, PrivigenTM</p> <ul style="list-style-type: none"> Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) <p>OctagamTM 10%</p> <ul style="list-style-type: none"> Dermatomyositis/polymyositis, severe, life-threatening, or refractory <p>AlygloTM, AscenivTM, BivigamTM, CutaquigTM, CuvitruTM, Flebogamma DIFTM, GamaSTANTM, GammagardTM, Gammagard S/DTM, GammakedTM, GammaplexTM, Gamunex-CTM, HizentraTM, HyqviaTM, OctagamTM, PanzygaTM, PrivigenTM, XembifyTM</p> <ul style="list-style-type: none"> Hypogammaglobulinemia, prophylaxis against bacterial infection. <p>Carimune NFTM, Flebogamma 10% DIFTM, Gammagard S/DTM, GammakedTM, GammaplexTM, Gamunex-CTM, Octagam 10%TM, PanzygaTM, PrivigenTM</p> <ul style="list-style-type: none"> Chronic Immune thrombocytopenia <p>Gammagard S/DTM</p> <ul style="list-style-type: none"> Kawasaki syndrome <p>GammagardTM Liquid</p> <ul style="list-style-type: none"> Multifocal motor neuropathy <p>AlygloTM, AscenivTM, BivigamTM, CutaquigTM, CuvitruTM, Flebogamma DIFTM, GamaSTANTM, GammagardTM, Gammagard S/DTM, GammakedTM, GammaplexTM, Gamunex-CTM, HizentraTM, HyqviaTM, OctagamTM, PanzygaTM, PrivigenTM, XembifyTM</p>



	<ul style="list-style-type: none"> • Primary Humoral Immunodeficiency (PI) in patients 2 years of age and older. <p>Medically accepted off-label indications supported in compendia or treatment guidelines, such as but not limited to:</p> <ul style="list-style-type: none"> • Acquired red cell aplasia • Acute disseminated encephalomyelitis • Allogeneic hematopoietic stem cell transplant • Autoimmune bullous disease • Autoimmune encephalitis • Antibody-mediated rejection, treatment • Fetal-neonatal alloimmune thrombocytopenia • Hematologic malignancy • Hemolytic disease of newborn • Hemolytic transfusion reaction • Hemophagocytic syndrome • HIV-positive status • Guillain-Barré syndrome • Kidney transplant • Lambert-Eaton myasthenic syndrome • Multiple sclerosis • Multisystem inflammatory syndrome in child • Myasthenia gravis, acute exacerbation • Opsoclonus-myoclonus syndrome • Post-transfusion purpura • Pregnancy-associated idiopathic (immune) thrombocytopenic purpura • Rasmussen encephalitis • Stevens-Johnson syndrome • Stiff-person syndrome • Systemic lupus erythematosus
Exclusion Criteria	None.
Required Medical Information	<p>Antiviral Prophylaxis</p> <p>One of the following diagnoses</p> <ol style="list-style-type: none"> 1. Hep A infection prophylaxis <ol style="list-style-type: none"> a. Virus exposure within previous 14 days OR b. Traveler to area with high or intermediate hepatitis A endemicity AND one or more of the following <ol style="list-style-type: none"> i. Age younger than 12 months (six months for travelling prophylaxis) or 40 years or older ii. Chronic liver disease iii. Hepatitis A vaccine contraindicated iv. Immunocompromised patient 2. Measles infection prophylaxis <ol style="list-style-type: none"> a. Measles exposure within previous six days AND



- b. Patient nonimmune to measles OR with severe immunocompromised
- 3. Rubella infection prophylaxis
 - a. Rubella exposure within previous three days AND pregnant patient
- 4. Varicella infection prophylaxis
 - a. High-risk patients (e.g., immunocompromised, perinatally infected neonates, preterm neonates exposed postnatally, pregnant patients) AND
 - b. Nonimmune to varicella
 - i. Exposure to varicella within the past four days

Chronic inflammatory demyelinating polyneuropathy (CIDP)

All of the following:

- 1. Clinic notes confirming patients' diagnosis AND
- 2. Documented failure of glucocorticoids or reasons for non – use

Dermatomyositis or polymyositis refractory to other immunosuppressive therapy or if immune globulin is being used as steroid-sparing option

- 1. Clinic notes confirming patients' diagnosis

Hypogammaglobulinemia, prophylaxis against bacterial infection

One or more of the following:

- 1. Age 18 years or younger AND serum IgG level less than 400mg/dL OR
- 2. Stem cell transplant AND serum IgG less than 400mg/dL AND severe infection

Chronic Immune Thrombocytopenia

- 1. Clinic notes confirming patients' diagnosis AND the need for rapid rise in platelet count to prevent or control bleeding

Kawasaki Disease

- 1. Clinic notes confirming patients' diagnosis

Multifocal motor neuropathy (MMN)

- 1. Clinic notes confirming patients' diagnosis

Primary Immunodeficiency (PI)

- 1. Clinic notes confirming diagnosis of PI required IgG replacement therapy due to hypogammaglobulinemia or agammaglobulinemia and diagnosis is defined by one of the following
 - Based on European Society for Immunodeficiencies (ESID) and Pan-American Group for Immunodeficiency OR

- Diagnosis is based on the following criteria and patient requires IgG therapy to treat the primary immunodeficiency diseases (PIDDs)

Primary Immunodeficiencies diseases (PIDDs)

A. Common variable immunodeficiency (CVID)

All the following:

- Recurrent bacterial infections of the ears, nasal sinuses, bronchi, or lungs.
- Pretreatment IgG level is less than 500 mg/dL or equal to or greater than two standard deviations (SD) below the mean for age.
- Low levels of IgA and/or IgM (more than two SD below mean for age).
- Lack of functional antibody response to vaccines
- Other causes of immune deficiency have been excluded

B. Chronic Granulomatous Disease (CGD):

1. Abnormal Nitroblue Tetrazolium (NBT) reduction test or respiratory burst in activated neutrophils (less than five percent of control) with **one** of the following:
 - a. Genetic testing showing mutation in gp91, p22, p47 or p67 phox.
 - b. Absent mRNA for one of the above genes by Northern blot analysis.
 - c. Maternal cousins, uncles, or nephews with an abnormal NBT or respiratory burst.
 - d. Recurrent bacterial or fungal infections of lung, skin, lymph nodes, and liver, etc. (CGD-type infections include Staphylococcus aureus, Burkholderia cepacia complex, Serratia marcescens, Nocardia and Aspergillus).
 - e. Formation of granulomata in tissues or organs.
 - f. Failure to thrive and hepatosplenomegaly or lymphadenopathy.

C. DiGeorge syndrome:

1. Patient has reduced numbers of CD3+ T cells (less than 500/mm³) and two out of three of a-c below or d alone or e alone:
 - a. Genetic testing showing deletion of chromosome 22q11.2.
 - b. Hypocalcemia of greater than three weeks' duration that requires therapy.
 - c. Conotruncal cardiac defect (truncus arteriosus, tetralogy of Fallot, interrupted aortic arch or aberrant right subclavian); or



- d. Patient has reduced numbers of CD3+ T cells (less than 1500/mm³) and a deletion of chromosome 22q11.2; or
- e. Patient has recurrent infections and classic features such as abnormal facial features, cardiac defect, hypoplastic thymus, hypocalcemia, and cleft palate.

D. IgA Deficiency:

- 1. Patient is over four years of age with **one** of the following:
 - a. Serum IgA of less than 7 mg/dl (0.07 g/L) but normal serum IgG and IgM and other causes of hypogammaglobulinemia have been excluded (Patient has a normal IgG antibody response to vaccination).
 - b. Serum IgA at least two SD below normal for age but normal serum IgG and IgM, and other causes of hypogammaglobulinemia have been excluded (Patient has a normal IgG antibody response to vaccination).
 - c. Frequent upper respiratory tract infections, persistent or recurrent infections, autoimmune disease, and allergies.

E. IgG subclass deficiency

- 1. Patient is seven years or older with **ALL** of the following:
 - a. Recurrent / severe ear and/or sinus infections.
 - b. Measurement of IgG subclass level showing deficiency (based on lab and age) or equal to or greater than two SD below the mean for age. Repeated at least once in separate sample. Normal levels of IgM and IgA.
 - c. Poor response to some vaccines (for example, Pneumovax).

F. Severe Combined Immunodeficiency (SCID):

- 1. Patient has at least one of the following:
 - a. Molecular or genetic confirmation of mutation in the cytokine common gamma chain (γ_c) or in one of these genes; JAK3, RAG1 or RAG2, IL-7R α .
 - b. ADA activity of less than 2% of control or mutations in both alleles of ADA.
 - c. Autologous CD3+ T cells less than 300 cells/microL in typical SCID and 300 to less than 1500 cells/microL in leaky SCID.
 - d. Detection of T-cells of maternal origin with normal lymphocyte count.
 - e. Serious or life-threatening infections, especially viral infections, which may result in pneumonia and chronic diarrhea, failure to thrive.
 - f. Absent or extremely low T cell mitogen response.
 - g. Very low levels of IgA and IgM; absent to elevated IgE.



- h. Positive family history of SCID or positive SCID newborn screening test.
- i. Pretreatment IgG level less than 200 mg/dL

G. Wiskott-Aldrich Syndrome (WAS):

1. Patient is male with congenital thrombocytopenia (less than 70,000 platelets/mm³), small platelets, and at least one of the following:
 - a. Genetic testing showing mutation of the WAS gene.
 - b. Absent WAS messenger RNA (mRNA) on Northern blot analysis of lymphocytes.
 - c. Absence of WAS protein (WASP) in lymphocytes.
 - d. Maternal male cousins, uncles, or nephews with small platelets and thrombocytopenia.
 - e. Eczema (localized or generalized).
 - f. Unusual bleeding and bruises, congenital or early onset thrombocytopenia, and small platelet size.
 - g. Defective antibody responses to some vaccine antigens (for example, Pneumovax).
 - h. Recurrent bacterial or viral infections.
 - i. Elevated IgA and IgE, low to normal IgG and IgM levels.
 - j. Autoimmune diseases, lymphoma, leukemia, or brain tumor.

H. X-linked agammaglobulinemia (XLA; Bruton's Agammaglobulinemia or Congenital Agammaglobulinemia):

1. Male patient with less than 2% CD19+ B cells and at least one of the following:
 - a. Genetic testing with mutation in Bruton's Tyrosine Kinase (BTK).
 - b. Absent BTK mRNA on Northern blot analysis of neutrophils or monocytes.
 - c. Absent BTK protein in monocytes or platelets.
 - d. Maternal cousins, uncles, or nephews with less than two percent CD19+ B cells.
 - e. Recurrent or severe bacterial infections, especially with small or absent tonsils and lymph nodes.
 - f. Onset of recurrent bacterial infections in the first five years of life, serum IgG, IgM, and IgA more than two SD below normal for age, absent isohemagglutinins and /or poor response to vaccines, and other causes of hypogammaglobulinemia have been excluded.

I. X-linked hyper IgM syndrome (XHIM):

1. Patient is male and has a serum IgG concentration at least two SD below normal for age and one of the following:
 - a. Genetic testing with a mutation in the CD40L gene.



- b. Patient's maternal cousins, uncles, or nephews have confirmed diagnosis of XHIM.
- c. One or more of the following infections or complications:
 - a. Recurrent bacterial infections in the first five years of life
 - b. Pneumocystis carinii infection in the first year of life
 - c. Neutropenia
 - d. Cryptosporidium-related diarrhea
 - e. Sclerosing cholangitis
 - f. Parvovirus-induced aplastic anemia
- d. Absent CD40 ligand cell surface staining on activated CD41 T cells as assessed by binding to soluble CD40 or by binding of monoclonal antibody to CD40 ligand.
- e. Serum concentration of IgG is less than 200 mg/dL; IgM may be low, normal, or elevated.

Medically accepted off-label indications

Acquired red cell aplasia

One or more of the following:

1. Immunocompromised patient with aplasia associated with parvovirus B19 infection
2. Failure of or inability to tolerate first-line therapy (e.g., prednisone, cyclophosphamide, cyclosporine)

Acute disseminated encephalomyelitis

1. Failure of treatment with corticosteroids or corticosteroids contraindicated

Allogeneic hematopoietic stem cell transplant

1. Serum IgG less than 400 mg/dL (4 g/L) AND Severe infection (sinopulmonary infection requiring hospitalization)

Autoimmune bullous diseases

All of the following:

1. Confirmed diagnosis of ONE of the following dermatologic conditions
 - a. Bullous pemphigoid
 - b. Epidermolysis bullosa acquisita
 - c. Linear IgA bullous dermatosis Mucous membrane (cicatricial) pemphigoid
 - d. Pemphigoid gestationis
 - e. Pemphigus foliaceus
 - f. Pemphigus vulgaris
2. Disease severity documented as extensive and debilitating
3. Documentation of Contraindications to, failure of, or significant side effects from systemic corticosteroids or immunosuppressive treatment AND



Autoimmune encephalitis

1. Clinic notes confirming patients' diagnosis

Autoimmune hemolytic anemia

1. Clinic notes confirming patients' diagnosis

Encephalomyelitis, acute disseminated

1. Clinic notes confirming patients' diagnosis AND failure of treatment with corticosteroids, or corticosteroids contraindicated

Fetal-neonatal alloimmune thrombocytopenia

One or more of the following:

1. Thrombocytopenia persisting in Newborn after transfusion of antigen-negative compatible platelets
2. Pregnant female with one or more: family history of disease, previous affected pregnancy, or platelet alloantibodies

Guillain-Barré syndrome

All of the following:

1. Four weeks or less since symptom onset
2. Progressive symptoms OR patient only able to walk with assistance OR Worse symptom severity
3. Hypogammaglobulinemia, prophylaxis against bacterial infection, hematopoietic cell transplantation

Hematologic malignancy

One or more of the following:

1. Chronic lymphocytic leukemia and one or more of the following
 - a. Corticosteroid-refractory autoimmune hemolytic anemia
 - b. History of recurrent or severe infection (e.g., sinopulmonary infection requiring hospitalization), and serum IgG less than 500 mg/dL (5 g/L)
2. Multiple myeloma and recurrent life-threatening infection

Hemolytic disease of newborn

One or more of the following:

1. Total serum bilirubin level within 2 mg/dL (34 micromoles/L) of age-adjusted and gestation-adjusted threshold for initiation of exchange transfusion OR
2. Total serum bilirubin still rising despite intensive phototherapy

Hemolytic transfusion reaction

One or more of the following:

1. After incompatible blood transfusion for severe life-threatening disease unresponsive to other therapies
2. Sickle cell disease and life-threatening post-transfusion hemolysis



Hemophagocytic syndrome

1. Clinic notes confirming patients' diagnosis AND immune globulins use for severe life-threatening disease unresponsive to other therapies

HIV-positive status

One or more of the following:

1. Active bleeding and platelet count less than 10,000/mm³ (10 x10⁹/L)
2. Hypogammaglobulinemia and ALL of the following:
 - a. Age 18 years or younger
 - b. Serum IgG level less than 400 mg/dL (4 g/L)

Antibody-mediated rejection, treatment, kidney transplant

One of the following

1. Postoperative immune globulin needed for Planned plasmapheresis OR
2. Preoperative and perioperative immune globulin needed for kidney transplant recipient anti-HLA antibody titer less than 1:16 of donor kidney AND Living donor transplant

Lambert-Eaton myasthenic syndrome

1. Failure of steroids and other immunosuppressive treatments in controlling symptoms

Multiple_sclerosis

ALL of the following

1. Clinical findings of multiple sclerosis, as indicated by 1 or more of the following:
 - a. Relapsing-remitting multiple sclerosis
 - b. Severe refractory optic neuritis and 1 or more of the following:
 - i. Corticosteroid therapy contraindicated
 - ii. No vision recovery after 3 months of corticosteroid therapy
2. Treatment with standard immunomodulatory drugs (i.e., interferon beta or glatiramer acetate) is not indicated due to 1 or more of the following:
 - a. Failure of previous treatment
 - b. Intolerance to attempted previous treatment
 - c. Patient pregnant, post-partum, or breast-feeding

Myasthenia gravis, acute exacerbation

All of the following

1. Immune globulin not to be used for chronic maintenance therapy AND



2. Treatment of myasthenia gravis needed for neonatal, juvenile, or adult myasthenia gravis

Multisystem inflammatory syndrome in child in setting of coronavirus disease 2019 (COVID-19) infection

1. Clinic notes confirming patients' diagnosis

Opsoclonus-mycoclonus syndrome

1. Clinic notes confirming patients' diagnosis

Post-transfusion purpura

1. Clinic notes confirming patients' diagnosis

Pregnancy-associated idiopathic (immune) thrombocytopenic purpura

One or more of the following:

1. Any bleeding during pregnancy
2. Platelet count less than 30,000/mm³ (30 x10⁹/L) at any time during pregnancy Platelet count less than level considered safe for invasive procedures (e.g., cesarean birth, neuraxial anesthesia)

Rasmussen encephalitis

1. Clinic notes confirming patients' diagnosis AND use of immunoglobulin for short-term amelioration of symptoms

Stevens-Johnson syndrome or toxic epidermal necrolysis, for life-threatening disease

1. Clinic notes confirming patients' diagnosis

Stiff-person syndrome

1. Clinic notes confirming patients' diagnosis AND failure of, or inability to receive or tolerate, GABA agonists

Systemic_lupus_erythematosus

1. Clinic notes confirming patients' diagnosis AND purpose of immunoglobulin is for disease unresponsive to other immunosuppressive therapies

Continued therapy:

1. Patient continues to meet initial coverage criteria.
2. Patient has experienced positive clinical response as evidenced by at least one of the following:
 - a. Patient has a decrease in the frequency of infections
 - b. Patient has a decrease in the severity of infections
 - c. Patient previously received intravenous immune globulin or is continuing therapy with subcutaneous immune globulin

	Other Off-label indications: 1) The requested unlabeled use must represent reasonable and current prescribing practices based on current medical literature, provider organizations, or academic & professional specialists. 2) In addition, one of the following is required: a. Documentation of trial & failure (or contraindication) to on-label treatments, or b. There are no FDA-approved drug treatments for the diagnosis.																			
Age Restriction	2 years of age and older (For ages 2 – 21, check for CCS eligibility)																			
Prescriber Restrictions	<ul style="list-style-type: none"> • PIDDs: Immunologists, Allergists • CIDP: Neurologists • GBS: Neurologists • Kawasaki disease: Cardiologist, Rheumatologist, Infectious Disease specialist • ITP: Hematologists • Hypogammaglobulinemia: Hematologists, Oncologist • Autoimmune hemolytic anemia: Hematologists • LEMS: Neurologists • Dermatomyositis: Rheumatologists, Dermatologist, Neurologists • Post-Transplant Immunodeficiency: Hematologists, Infection Disease specialists, Transplant specialists, • Pemphigus Vulgaris and Other Autoimmune Blistering Diseases: Dermatologists • Other Approved and Off-Label Indications: The diagnosis and treatment must be within the scope of the treating physician's board-certified specialty. 																			
Coverage Duration	Initial authorization is for three months. Reauthorization is for six months																			
Other Criteria / Information	Criteria adapted from DHCS OCT 2024 & MCG A) Accepted HCPCS codes (with an approved TAR): <table border="1"> <thead> <tr> <th>Product</th><th>HCPCS</th><th>HCPCS Description</th></tr> </thead> <tbody> <tr> <td colspan="3"><i>Intravenous Infusion</i></td></tr> <tr> <td>Asceniv</td><td>J1554</td><td>Injection, immune globulin (asceniv), 500 mg</td></tr> <tr> <td>Bivigam</td><td>J1556</td><td>Injection, immune globulin (bivigam), 500 mg</td></tr> <tr> <td>Flebogamma; Flebogamma DIF</td><td>J1572</td><td>Injection, immune globulin, (flebogamma/flebogamma dif), intravenous, non-lyophilized (e.g., liquid), 500 mg</td></tr> <tr> <td>Gammagard S/D; Carimune NF</td><td>J1566</td><td>Injection, immune globulin, intravenous, lyophilized (e.g., powder), not otherwise specified, 500 mg</td></tr> </tbody> </table>		Product	HCPCS	HCPCS Description	<i>Intravenous Infusion</i>			Asceniv	J1554	Injection, immune globulin (asceniv), 500 mg	Bivigam	J1556	Injection, immune globulin (bivigam), 500 mg	Flebogamma; Flebogamma DIF	J1572	Injection, immune globulin, (flebogamma/flebogamma dif), intravenous, non-lyophilized (e.g., liquid), 500 mg	Gammagard S/D; Carimune NF	J1566	Injection, immune globulin, intravenous, lyophilized (e.g., powder), not otherwise specified, 500 mg
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<i>Intravenous Infusion</i>																				
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Bivigam	J1556	Injection, immune globulin (bivigam), 500 mg																		
Flebogamma; Flebogamma DIF	J1572	Injection, immune globulin, (flebogamma/flebogamma dif), intravenous, non-lyophilized (e.g., liquid), 500 mg																		
Gammagard S/D; Carimune NF	J1566	Injection, immune globulin, intravenous, lyophilized (e.g., powder), not otherwise specified, 500 mg																		



	Gammaflex	J1557	Injection, immune globulin, (gammaflex), intravenous, nonlyophilized (e.g., liquid), 500 mg
	Octagam	J1568	Injection, immune globulin, (octagam), intravenous, nonlyophilized (e.g., liquid), 500 mg
	Panzyga	J1576	Injection, immune globulin (panzyga), intravenous, nonlyophilized (e.g., liquid), 500 mg
	Privigen	J1459	Injection, immune globulin (privigen), intravenous, nonlyophilized (e.g., liquid), 500 mg
	Intramuscular Injection		
	Gammagard	J1569	Injection, immune globulin, (gammagard liquid), nonlyophilized, (e.g., liquid), 500 mg • IV or SC: Primary Immunodeficiency • IV only: All other indications
	Gammaked; Gamunex-C	J1561	Injection, immune globulin, (gamunex-c/gammaked), nonlyophilized (e.g., liquid), 500 mg • IV or SC: Primary Immunodeficiency • IV only: All other indications
	Subcutaneous Infusion		
	GamaSTAN S/D	J1460	Injection, gamma globulin, intramuscular, per 1 cc
		J1560	Injection, gamma globulin, intramuscular, per 10 cc
	Cutaquig	J1551	Injection, immune globulin (cutaquig), 100 mg
	Cuvitru	J1555	Injection, immune globulin (cuvitru), 100 mg
	Hizentra	J1559	Injection, immune globulin (hizentra), 100 mg
	Hyqvia	J1575	Injection, immune globulin/hyaluronidase, (hyqvia), 100 mg immune globulin
	Xembify	J1558	Injection, immune globulin (xembify), 100 mg
	Indication		Dosing
	Antiviral prophylaxis		Dosing and frequency determined by wt. (kg), type (Hepatitis A, measles, varicella) and time of potential exposure, current IVIG products used to treat the patient.



	Chronic inflammatory demyelinating polyneuropathy	Initial: 2 g/kg IV divided in doses over two to five days or 400 mg/kg IV once a day for five days (max daily dose of 1 g/kg). Maintenance: 1 g/kg IV divided over one to two days every three weeks. Transitioning to SC: Start one week after last IVIG infusion, at 200 mg/kg – 400 mg/kg per week, over one to two sessions over one to two days.
	Dermatomyositis/polymyositis, severe, life threatening or refractory	1 g/kg per day IV x two days every four weeks or 1 g/kg per day once every two weeks
	Hypogammaglobulinemia prophylaxis against bacterial infection	Acquired secondary to malignancy: 200 mg/kg – 400 mg/kg IV once every three to four weeks Primary humoral immunodeficiency disorder: 200 mg/kg – 800 mg/kg IV once every three to four weeks
	Immune thrombocytopenia	≥ 18 yrs: 1 g/kg IV once a day for one to two days, may hold second dose with adequate platelet response (e.g., plt > 50,000 mm ³) after 24 hrs or 400 mg/kg IV daily x five days 2-17 years: Dose is dependent on product used for treatment, age, wt (kg) and dosing frequency chosen for acute or chronic treatment.
	Kawasaki Syndrome	Infants and children (specific age range in not referenced): 2000 mg/kg IV over 8-12 hr, given within 10 days of disease onset. If signs and symptoms persist ≥ 36 hrs, 1000 mg/kg – 2000 mg/kg may be considered.
	Multifocal motor neuropathy	Initial dosing: 2 g/kg IV divided over two to five consecutive days or 400 mg/kg IV once a day x five days (max daily dose: 1 g/kg) and maintenance dose of 1 g/kg – 2 g/kg every two to six weeks or if high dose was tolerated dosing 1 g/kg IV once daily x two days can be considered.
	Primary humoral immunodeficiency disorders (PI)	400 to 600 mg/kg IV as a single dose once every three to four weeks.
Off-Label Indications		



	Encephalitis, immune checkpoint inhibitor-induced, severe, or progressive	400 mg/kg IV once daily for five days (in combination with IV methylprednisolone)
	Encephalomyelitis, acute disseminated	400 mg/kg IV once daily for five days.
	Fetal and neonatal alloimmune thrombocytopenia (maternal administration)	1 to 2 g/kg IV per week, with or without glucocorticoids (doses >1 g/kg are typically divided into two doses and given over two days)
	Pemphigus foliaceus and vulgaris, refractory	2 g/kg IV given in divided doses over 2-5 days or 400 mg/kg IV once a day x five days. May repeat every four to six weeks based on clinical response.
	Guillain-Barré syndrome	Start treatment within four weeks of symptoms. 400 mg/kg IV x five days only. Retreatment is not recommended.
	Lambert-Eaton myasthenic syndrome	2 g/kg IV administered in divided doses over 2 to 5 consecutive days. If responding, repeat every four to 12 weeks
	Myasthenia gravis, acute exacerbation	2 g/kg IV administered in divided doses given over two to five consecutive days or 400 mg/kg IV once a day x five days or 1 g/kg IV once a day for two days.
	Multiple sclerosis	Children and adolescents: 1,000 mg/kg/dose IV once monthly, with or without an induction of 400 mg/kg/day for five days
	Toxic shock syndrome, streptococcal (adjunctive agent)	1 g/kg IV on day one, followed by 500 mg/kg IV once daily on days two and three
	Transplant	Kidney transplantation: >one year after transplant: 200 mg/kg IV every two weeks for three doses Kidney transplantation: ≤ one year after transplant: 200 mg/kg IV every two weeks for three doses



STATUS	DATE REVISED	REVIEW DATE	APPROVED / REVIEWED BY	EFFECTIVE DATE
Approved	N/A	11/14/2024	Pharmacy & Therapeutics (P&T) Committee	5/1/2025