Medica (formerly WellFirst Health)

Immune Globulins (immunoglobulin): Asceniv™; Alyglo™; Bivigam®; Flebogamma®; Gamunex-C®; Gammagard® Liquid; Gammagard® S/D; Gammaked™; Gammaplex®; Octagam®; Privigen®; Panzyga®; Yimmugo® (Intravenous)

Document Number: IC-0071

Last Review Date: 07/02/2024
Date of Origin: 07/20/2010
Dates Reviewed: 09/2010, 12/2010, 02/2011, 03/2011, 06/2011, 09/2011, 10/2011, 12/2011, 03/2012, 06/2012, 09/2012, 12/2012, 03/2013, 05/2013, 06/2013, 09/2013, 12/2013, 03/2014, 06/2014, 09/2014, 12/2014, 03/2015, 06/2015, 09/2015, 12/2015, 03/2016, 06/2016, 09/2016, 12/2016, 03/2017, 06/2017, 09/2017, 12/2017, 03/2018, 06/2018, 09/2018, 10/2018, 05/2019, 10/2019, 10/2020, 01/2021, 04/2021, 10/2021, 11/2022, 11/2023, 02/2024, 07/2024

I. Length of Authorization

- Initial and renewal authorization periods vary by specific covered indication.
- Unless otherwise specified, the initial authorization will be provided for 6 months and may be renewed annually.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

Drug	Vial size in IgG grams	# of vials	
		One time only	per 28 days
		LOAD	MAINTENANCE
Asceniv	5	18	18
Alyglo	5, 10, 20	1	1
Bivigam	5	1	1
	10	23	23
Flebogamma 10% DIF	5, 10, 20	1	1
	20	11	11
Flebogamma 5% DIF	0.5, 2.5, 5, 10	1	1
	20	11	11
Gamunex-C	1, 2.5, 5, 10, 20	1	1
	40	6	6
	1, 2.5, 5, 10, 20	1	1

Gammagard Liquid	30	8	8
Gammagard S/D	5	1	1
	10	23	23
Gammaked	1, 2.5, 5, 10	1	1
	20	11	11
Gammaplex (5% and 10%)	5, 10	1	1
	20	11	11
Octagam 10%	2, 5, 10, 20	1	1
	30	8	8
Octagam 5%	1, 2.5, 5, 10	1	1
	25	9	9
Privigen	5, 10, 20	1	1
	40	6	6
Panzyga	1, 2.5, 5, 10, 20	1	1
	30	8	8
Yimmugo	5, 10, 20	1	1

B. Max Units (per dose and over time) [HCPCS Unit]:

Indication	Billable Units	Per # days *(unless otherwise specified)*
PID and Supportive Care after Rethymic transplant	180	21
IgG Subclass Deficiency	90	14
CIDP	Load: 460	5
	Maintenance: 230	21
Immune thrombocytopenia/ITP	460	28
FAIT	230	7
Kawasaki's Disease	460	2 doses only
Multifocal Motor Neuropathy	460	28
CLL/MM	90	21
ALL	90	21
HIV (Pediatric Patients only)	46	14
Guillain-Barré	460	5 (*for two courses only)*
Myasthenia Gravis	460	28
Auto-immune blistering diseases	460	28
Allogeneic Bone Marrow or Stem Cell Transplant	Load: 120	7 (for 90 days)
	Maintenance: 120	21

Dermatomyositis/Polymyositis	460	28
Complications of transplanted solid organ or bone marrow transplant	460	28
Stiff Person Syndrome	460	28
Toxic Shock Syndrome	460	5 *(for one cycle only)*
NAIT	20	2 doses only
Management of Immune Checkpoint Inhibitor Related Toxicity	460	5 *(for one cycle only)*
Management of CAR T-Cell-Related Toxicity	120	28

III. Initial Approval Criteria [1-16,71]

Coverage is provided for the following conditions:

Patient is required to meet Site of Service specialty infusion program requirements (refer to the Medica (formerly Wellfirst Health Plan) Site of Service Policy

Primary Immunodeficiency (PID) † [1-16,38,54,56,57,70,103]

Such as: Wiskott-Aldrich syndrome, x-linked agammaglobulinemia, common variable immunodeficiency, transient hypogammaglobulinemia of infancy, antibody deficiency with near normal immunoglobulin levels, and combined deficiencies (severe combined immunodeficiencies, ataxia-telangiectasia, x-linked lymphoproliferative syndrome) ***[list not all inclusive]***

- Patient has an IgG level < 200 mg/dL; **OR**
- Patient meets both of the following:
  - Patient has a history of multiple hard to treat infections as indicated by at least one of the following:
    - Four or more ear infections within 1 year
    - Two or more serious sinus infections within 1 year
    - Two or more months of antibiotics with little effect
    - Two or more pneumonias within 1 year
    - Recurrent, deep skin or organ abscesses
    - Persistent thrush in the mouth or fungal infections on the skin
    - Need for intravenous antibiotics to clear infections
    - Two or more deep-seated infections including septicemia
    - Family history of PID; **AND**
  - Patient has a deficiency in producing antibodies in response to vaccination; **AND**
    - Titers were drawn before challenging with vaccination; **AND**
    - Titers were drawn between 4 and 8 weeks of vaccination

IgG Subclass Deficiency ‡ [57,70,98-100]

- Patient has an IgG level < 400 mg/dL; **AND**
- Patient has a history of recurrent infections; **AND**
- Patient is receiving prophylactic antibiotic therapy

Immune Thrombocytopenia/Idiopathic Thrombocytopenia Purpura (ITP) † (**Φ** for Gammaplex) [2,5-9,11-13,32,37,39,81]

For acute ITP:

- Used to manage acute bleeding due to severe thrombocytopenia (platelet count < 30 X 10^9/L); **OR**
- Used to increase platelet counts prior to invasive surgical procedures such as splenectomy (platelet count < 100 X 10^9/L); **OR**
- Patient has severe thrombocytopenia (platelet count < 20 X 10^9/L)

Note: Authorization is valid for 1 month only and cannot be renewed

For chronic ITP:

- Patient is at increased risk for bleeding as indicated by a platelet count < 30 X 10^9/L; **AND**
- Patient has a history of failure, contraindication, or intolerance to corticosteroids; **AND**
- Duration of illness > 6 months

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) † (**Φ** for Gamunex-C) [4,6,7,12,13,18-22,24-26,42,44,72,116]

- Patient's disease course is progressive or relapsing and remitting for >2 months; **AND**
- Patient has abnormal or absent deep tendon reflexes in upper or lower limbs; **AND**
- Electrodiagnostic testing indicating demyelination:
  - Partial motor conduction block in at least 2 motor nerves or in 1 nerve plus one other demyelination criterion listed here in at least 1 other nerve; **OR**
  - Distal CMAP duration increase in at least 1 nerve plus one other demyelination criterion listed here in at least 1 other nerve; **OR**
  - Abnormal temporal dispersion conduction must be present in at least 2 motor nerves; **OR**
  - Reduced motor conduction velocity in at least 2 motor nerves; **OR**
  - Prolonged distal motor latency in at least 2 motor nerves; **OR**
  - Absent F wave in at least 2 motor nerves plus one other demyelination criterion listed here in at least 1 other nerve; **OR**
  - Prolonged F wave latency in at least 2 motor nerves; **AND**
- Patient is refractory or intolerant to corticosteroids (e.g., prednisolone, prednisone, etc.) given in therapeutic doses over at least three months; **AND**

- Baseline in strength/weakness has been documented using an objective clinical measuring tool (e.g., INCAT, Medical Research Council (MRC) muscle strength, 6-MWT, Rankin, Modified Rankin, etc.)

Note: Initial authorization is valid for 3 months

Guillain-Barré Syndrome (Acute inflammatory polyneuropathy) ‡ [19,21,22,24,30,31,58,70,77,115]

- Patient has severe disease (i.e., patient requires assistance to ambulate); **AND**
- Onset of symptoms are recent (i.e., less than 1 month); **AND**
- Patient has abnormal or absent deep tendon reflexes in upper or lower limbs; **AND**
- Patient diagnosis is confirmed using a cerebrospinal fluid (CSF) analysis; **AND**
- Approval will be granted for a maximum of 2 courses of therapy within 6 weeks of onset

Note: Authorization is valid for 2 months only and cannot be renewed

Multifocal Motor Neuropathy † (**Φ** for Gammagard Liquid) [4,19,21,22,24,25]

- Patient has progressive, focal, asymmetric limb weakness (without sensory symptoms) for >1 month; **AND**
- Patient has complete or partial conduction block or abnormal temporal dispersion conduction in at least 2 motor nerves; **AND**
- Patient has normal sensory nerve conduction on all nerves tested; **AND**
- Baseline in strength/weakness has been documented using an objective clinical measuring tool (e.g., INCAT, Medical Research Council (MRC) muscle strength, 6-MWT, Rankin, Modified Rankin, etc.)

Note: Initial authorization is valid for 3 months

HIV Infected Children: Bacterial Control or Prevention ‡ [27,28,37,89]

- Patient ≤ 13 years of age; **AND**
- Patient has an IgG level < 400 mg/dL

Myasthenia Gravis ‡ [53,78,85]

- Patient has a positive serologic test for anti-acetylcholine receptor (AchR) antibodies; **AND**
- Patient has an acute exacerbation resulting in impending myasthenic crisis (i.e., respiratory compromise, acute respiratory failure, and/or bulbar compromise); **AND**
- Patient is failing on conventional immunosuppressant therapy alone (e.g., corticosteroids, azathioprine, cyclosporine, mycophenolate, methotrexate, tacrolimus, cyclophosphamide, etc.); **AND**
- Patient will be on combination therapy with corticosteroids or other immunosuppressant (e.g., azathioprine, mycophenolate, cyclosporine, methotrexate, tacrolimus, cyclophosphamide, etc.)

Note: Authorization is valid for 1 course (1 month) only and cannot be renewed

Dermatomyositis † (**Φ** for Octagam 10%) **/ Polymyositis ‡** [11,19,21,22,24,65,66,70,82,87]

- Patient has severe active disease; **AND**
- Patient has proximal weakness in all upper and/or lower limbs; **AND**
- Diagnosis has been confirmed by muscle biopsy; **AND**
- Patient has failed a trial of corticosteroids (i.e., prednisone); **AND**
- Patient has failed a trial of an immunosuppressant (e.g., methotrexate, azathioprine, etc.); **AND**
- Patient will be on combination therapy with corticosteroids or other immunosuppressants; **AND**
- Patient has a documented baseline physical exam and muscular strength/function

Note: Initial authorization is valid for 3 months

Complications of Transplanted Solid Organ (kidney, liver, lung, heart, pancreas) and Bone Marrow Transplant ‡ [59-62,70,102]

Coverage is provided for one or more of the following (list not all-inclusive):

- Suppression of panel reactive anti-human leukocyte antigen (HLA) antibodies prior to transplantation
- Treatment of antibody-mediated rejection of solid organ transplantation
- Prevention or treatment of viral infections (e.g., cytomegalovirus, Parvo B-19 virus, Polyoma BK virus, etc.)

Stiff-Person Syndrome ‡ [21,24,64,114]

- Patient has anti-glutamic acid decarboxylase (GAD) antibodies; **AND**
- Patient has failed ≥ 2 of the following treatments: benzodiazepines (e.g., diazepam, clonazepam, alprazolam, lorazepam, oxazepam, temazepam, etc.), anti-spasticity agents (e.g., baclofen, tizanidine, etc.) or anti-epileptics (e.g., gabapentin, valproate, tiagabine, levetiracetam, etc.); **AND**
- Patient has a documented baseline on physical exam

Allogeneic Bone Marrow or Stem Cell Transplant ‡ [76,102,113]

- Used for prevention of acute Graft-Versus-Host-Disease (aGVHD) or infection; **AND**
- Patient's bone marrow (BMT) or hematopoietic stem cell (HSCT) transplant was allogeneic; **AND**
- Patient has an IgG level < 400 mg/dL

Note: Initial authorization is valid for 3 months

Kawasaki's Disease † [5,83]

Note: Authorization is valid for 1 course (1 month) only and cannot be renewed

Fetal Alloimmune Thrombocytopenia (FAIT) ‡ [32,37,47,84,90]

- Patient has a history of one or more of the following:
  - Previous FAIT pregnancy

- Family history of the disease
- Screening reveals platelet alloantibodies

Note: Authorization is valid through the delivery date only and cannot be renewed

Neonatal Alloimmune Thrombocytopenia (NAIT) ‡ [35-37,84]

Note: Authorization is valid for 1 course (1 month) only and cannot be renewed

Autoimmune Mucocutaneous Blistering Diseases ‡ [34,40,41,67-69,91,110-112]

- Patient has been diagnosed with one of the following:
  - Pemphigus vulgaris
  - Pemphigus foliaceus
  - Bullous Pemphigoid
  - Mucous Membrane Pemphigoid (a.k.a. Cicatricial Pemphigoid)
  - Epidermolysis bullosa aquisita
  - Pemphigus gestationis (Herpes gestationis)
  - Linear IgA dermatosis; **AND**
- Patient has severe disease that is extensive and debilitating; **AND**
- Diagnosis has been confirmed by biopsy; **AND**
- Patient has progressive disease; **AND**
- Disease is refractory to a trial of conventional therapy with corticosteroids and concurrent immunosuppressive treatment (e.g., azathioprine, cyclophosphamide, mycophenolate mofetil, etc.); **AND**
- Patient has a documented baseline on physical exam

Acquired Immune Deficiency Secondary to Acute Lymphoblastic Leukemia (ALL) ‡ or Multiple Myeloma ‡ [37,70,79,92,106]

- Used for prevention of infection; **AND**
- Patient has an IgG level < 400 mg/dL

Acquired Immune Deficiency Secondary to Chronic Lymphocytic Leukemia † ‡ or Small Lymphocytic Lymphoma ‡ [5,37,70,88,103,107]

- Patient has an IgG level < 200 mg/dL; **OR**
- Patient has an IgG level < 500 mg/dL**; AND**
  - Patient has recurrent sinopulmonary infections requiring IV antibiotics or hospitalization; **OR**
- Patient meets both of the following:
  - Patient has a history of multiple hard to treat infections as indicated by at least one of the following:
    - Four or more ear infections within 1 year

        - Two or more serious sinus infections within 1 year
        - Two or more months of antibiotics with little effect
        - Two or more pneumonias within 1 year
        - Recurrent, deep skin or organ abscesses
        - Persistent thrush in the mouth or fungal infections on the skin
        - Need for intravenous antibiotics to clear infections
        - Two or more deep-seated infections including septicemia; **AND**
    - The patient has a deficiency in producing antibodies in response to vaccination; **AND**
        - Titers were drawn before challenging with vaccination; **AND**
        - Titers were drawn between 4 and 8 weeks of vaccination

Note: Other secondary immunodeficiencies resulting in hypogammaglobulinemia and/or B-cell aplasia will be evaluated on a case-by-case basis

Toxic Shock Syndrome ‡ [46,93,94]

Note: Authorization is valid for 1 course (1 month) only and cannot be renewed

Management of Immune-Checkpoint-Inhibitor Related Toxicity ‡ [73,80]

- Patient has been receiving therapy with an immune checkpoint inhibitor (e.g., nivolumab, pembrolizumab, atezolizumab, avelumab, durvalumab, cemiplimab, ipilimumab, dostarlimab, tremelimumab, retifanlimab, etc.); **AND**
- Patient has one of the following toxicities related to their immunotherapy:
    - Severe (G3) or life-threatening (G4) bullous dermatitis as an adjunct to rituximab
    - Stevens-Johnson syndrome (SJS)
    - Toxic epidermal necrolysis (TEN)
    - Severe (G3-4) myasthenia gravis
    - Demyelinating disease (optic neuritis, transverse myelitis, acute demyelinating encephalomyelitis)
    - Myocarditis as further intervention if no improvement within 24-48 hours of starting high-dose methylprednisolone
    - Moderate (G2) or severe (G3-4) Guillain-Barré Syndrome or severe (G3-4) peripheral neuropathy used in combination with high-dose methylprednisolone
    - Moderate (G2) pneumonitis if no improvement after 48-72 hours of corticosteroids
    - Severe (G3-4) pneumonitis if no improvement after 48 hours of methylprednisolone
    - Encephalitis used in combination with high-dose methylprednisolone for severe or progressing symptoms
    - Moderate, severe, or life-threatening steroid-refractory myositis (proximal muscle weakness, neck flexor weakness, with or without myalgias) for significant dysphagia, life-threatening situations, or cases refractory to corticosteroids

Management of CAR T-Cell-Related Toxicity ‡ [73,80,86,95,96,104,105]

- Patient has received treatment with anti-CD19 CAR T-cell therapy (e.g., axicabtagene ciloleucel, brexucabtagene autoleucel, lisocabtagene maraleucel, tisagenlecleucel, etc.); **AND**
  - Used for the management of G4 cytokine release syndrome (CRS) that is refractory to high-dose corticosteroids and anti-IL-6 therapy (e.g., tocilizumab); **OR**
  - Patient has hypogammaglobulinemia as confirmed by serum IgG levels <600 mg/dL and serious or recurrent infections; **OR**
- Patient has received treatment with BCMA-targeted CAR T-cell therapy (e.g., idecabtagene vicleucel, ciltacabtagene autoleucel, etc.); **AND**
  - Used for the management of G4 cytokine release syndrome (CRS) that is refractory to high-dose corticosteroids and anti-IL-6 therapy (e.g., tocilizumab); **OR**
  - Patient has hypogammaglobulinemia as confirmed by serum IgG levels <400 mg/dL; **OR**
- Used as prophylactic therapy prior to receiving treatment with anti-CD19 or BCMA-targeted CAR T-cell therapy (e.g., axicabtagene ciloleucel, brexucabtagene autoleucel, idecabtagene vicleucel, lisocabtagene maraleucel, tisagenlecleucel, ciltacabtagene autoleucel, etc.); **AND**
  - Patient has hypogammaglobulinemia as confirmed by serum IgG levels ≤400 mg/dL and serious, persistent, or recurrent bacterial infections

Supportive Care after Rethymic transplant ‡ [97]

- Used as immunoglobulin replacement therapy in pediatric patients with congenital athymia after surgical implantation of Rethymic; **OR**
- Used as re-initiation of treatment 2 months after stopping immunoglobulin replacement therapy in pediatric patients who have an IgG trough level lower than normal range for age

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); Φ Orphan Drug

For Reference Use Only

Brand Name/ Formulation	FDA Indication	Contraindications	Product Specs	Comments
Asceniv 10%	PID (≥12yo)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	• **IgA**: ≤200 mcg/mL • **Osmolality**: 370 to 510 mOsm/kg • **Stabilizer**: Glycine	Other stabilizer used is Polysorbate 80
Alyglo 10%	PID (adults)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	• **IgA**: ≤100 mcg/mL • **Osmolality**: N/A • **Stabilizer**: Glycine	
Bivigam 10% (liquid)	PID (peds ≥2)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	• **IgA**: ≤200 mcg/mL • **Osmolality**: 370 to 510 mOsm/kg • **Stabilizer**: glycine	
Flebogamma 5% (liquid)	PID (peds ≥2)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	**IgA**: <50 mcg/mL **Osmolarity**: 240 to 370 mOsm/kg	

			Stabilizer: sorbitol	
Flebogamma 10% (liquid)	PID (peds ≥2) cITP (peds ≥2)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	**IgA**: <32 mcg/mL **Osmolarity**: 240 to 370 mOsm/L **Stabilizer**: sorbitol	
Gammagard 10% (liquid)	PID (peds ≥2) MMN (adults) CIDP (adults)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	**IgA**: 37 mcg/mL **Osmolality**: 240 to 300 mOsm/kg **Stabilizer**: glycine	May be used SC (see SCIG policy for criteria)
Gammagard S/D 5%(lyophilized)	PID (peds ≥2) cITP (adult) CLL Kawasaki (peds)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	**IgA**: ≤2.2 mcg/mL **Osmolality**: 636 mOsm/L **Stabilizer**: glycine	Contains some sugar (20mg/mL when prepared)
Gammaked 10% (liquid)	PID (peds ≥2) aITP or cITP (peds/adults) CIDP (adults)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	**IgA**: 46 mcg/mL **Osmolality**: 258 mOsm/kg **Stabilizer**: glycine	May be used SC (see SCIG policy for criteria)
Gammaplex 5% (liquid)	PID (peds ≥2) cITP (peds/adults)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies Fructose intolerance	**IgA**: <10 mcg/mL **Osmolality**: 460 to 500 mOsm/kg **Stabilizer**: glycine	Other stabilizer used is Polysorbate 80
Gammaplex 10% (liquid)	PID (peds ≥2) cITP (adults)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	**IgA**: <20 mcg/mL **Osmolality**: 280 mOsm/kg **Stabilizer**: glycine	Other stabilizer used is Polysorbate 80
Gamunex-C 10% (liquid)	PID (peds ≥2) aITP or cITP (peds/adults) CIDP (adults)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	**IgA**: 46 mcg/mL **Osmolality**: 258 mOsm/kg **Stabilizer**: glycine	May be used SC (see SCIG policy for criteria)
Octagam 5% (liquid)	PID (peds ≥6)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies Corn allergy	**IgA**: ≤100 mcg/mL **Osmolality**: 310 to 380 mOsm/kg **Stabilizer**: maltose	
Octagam 10% (liquid)	cITP (adults) Dermatomyositis (adult)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	**IgA**: 106 mcg/mL **Osmolality**: 310 to 380 mOsm/kg **Stabilizer**: maltose	
Panzyga 10% (liquid)	PID (peds ≥2) cITP (adults) CIDP (adults)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	**IgA**: ≤100 mcg/mL **Osmolality**: 240 to 310 mOsm/kg **Stabilizer**: glycine	
Privigen 10% (liquid)	PID (peds ≥3) cITP (ped ≥15) CIDP (adults)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies Hyperprolinemia	**IgA**: ≤25 mcg/mL **Osmolality**: 320 mOsm/kg **Stabilizer**: L-proline	
Yimmugo 10% (liquid)	PID (peds ≥2)	History of anaphylaxis to IgG IgA-deficient with IgA antibodies	**IgA**: ≤300 mcg/mL **Osmolality**: 280 to 380 mOsm/kg **Stabilizer**: N/A	Does not contain carbohydrate stabilizers (e.g., sucrose, maltose) or preservatives

- All intravenous immunoglobulins are derived from human plasma.
- Products with higher IgA content pose a greater risk for anaphylactic reactions, especially in patients with IgA deficiencies.
- All products may predispose patients to nephrotoxicity especially those with sugar-based or proline-based stabilizers. To lower risks, lower concentration products and infusions rates should be used as well as using products with osmolality/osmolarity that is near physiologic range (around 300 mOsm/kg or mOsm/L).
- Premedications (e.g., acetaminophen, antihistamine, etc.) are recommended to reduce the risk of infusion related reactions.

Adapted from:
- Professional Resource, Comparison of IVIG Products. Pharmacist's Letter/Prescriber's Letter. December 2016.
- Product package inserts
- Characteristics of Immunoglobulin Products Used to Treat Primary Immunodeficiencies (PI). Immune Deficiency Foundation. April 2020

IV. Renewal Criteria [1-16,57,71]

Coverage can be renewed based upon the following criteria:

Note: unless otherwise specified, renewal authorizations are provided for 1 year

- Patient continues to meet indication-specific relevant criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: renal dysfunction and acute renal failure, thrombosis, hemolysis, severe hypersensitivity reactions, pulmonary adverse reactions/transfusion-related acute lung injury (TRALI), hyperproteinemia, increased serum viscosity, hyponatremia, aseptic meningitis syndrome, hypertension, volume overload, etc.; **AND**

Primary Immunodeficiency (PID) [1-16,38,54,56,57,70]

- Disease response as evidenced by one or more of the following:
  - Decrease in the frequency of infection
  - Decrease in the severity of infection

IgG Subclass Deficiency [70,98,100]

- Disease response as evidenced by one or more of the following:
  - Decrease in the frequency of infection
  - Decrease in the severity of infection; **AND**
- Continued treatment is necessary to decrease the risk of infection

Immune Thrombocytopenia/Idiopathic Thrombocytopenia Purpura (ITP) [2,5-9,11-13,32,37,39,81]

- Acute ITP:
  - May not be renewed.
- Chronic ITP:
  - Disease response as indicated by the achievement and maintenance of a platelet count of ≥ 30 X 109/L and at least doubling the baseline platelet count

Chronic Inflammatory Demyelinating Polyneuropathy [4,6,7,12,13,18-22,24-26,42,44,72,116]

- Renewals will be authorized for patients that have demonstrated a clinical response to therapy based on an objective clinical measuring tool (e.g., INCAT, Medical Research Council (MRC) muscle strength, 6-MWT, Rankin, Modified Rankin, etc.)

Guillain-Barre Syndrome (Acute inflammatory polyneuropathy) [58]

- May not be renewed.

Multifocal Motor Neuropathy [1-14,19,21,22,24,25]

- Renewals will be authorized for patients that have demonstrated a clinical response to therapy based on an objective clinical measuring tool (e.g., INCAT, Medical Research Council (MRC) muscle strength, 6-MWT, Rankin, Modified Rankin, etc.)

HIV Infected Children: Bacterial Control or Prevention [27,28,37,89]

- Disease response as evidenced by one or more of the following:
  - Decrease in the frequency of infection
  - Decrease in the severity of infection; **AND**
- Patient continues to be at an increased risk of infection necessitating continued therapy as evidenced by an IgG level < 400 mg/dL

Myasthenia Gravis [53,78,85]

- May not be renewed.

Dermatomyositis/Polymyositis [19,21,22,24,65,66,70,82]

- Patient had an improvement from baseline on physical exam and/or muscular strength and function

Note: Renewal authorizations are provided for 6 months

Complications of Transplanted Solid Organ (kidney, liver, lung, heart, pancreas) and Bone Marrow Transplant [59-62,70,102]

- Disease response as evidenced by one or more of the following:
  - Decrease in the frequency of infection
  - Decrease in the severity of infection; **AND**
- Continued treatment is necessary to decrease the risk of infection

Stiff Person Syndrome [21,24,64]

- Documented improvement from baseline on physical exam

Allogeneic Bone Marrow or Stem Cell Transplant [76,102]

- Patient continues to be at an increased risk of infection necessitating continued therapy as evidenced by an IgG level < 400 mg/dL

Note: Renewal authorizations are provided for 3 months

Kawasaki's Disease [5,83]

- May not be renewed.

Fetal Alloimmune Thrombocytopenia (FAIT) [33,38,48,85,90]

- Authorization is valid through the delivery date only and cannot be renewed

Neonatal Alloimmune Thrombocytopenia [35-37,84]

- May not be renewed.

Autoimmune Mucocutaneous Blistering Diseases [34,40,41,67-69,91,110-112]

- Documented improvement from baseline on physical exam

Note: Renewal authorizations are provided for 6 months

Acquired Immune Deficiency Secondary to Acute Lymphoblastic Leukemia (ALL), Chronic Lymphocytic Leukemia (CLL), Small Lymphocytic Lymphoma (SLL), or Multiple Myeloma (MM) [37,70,79,92]

- Disease response as evidenced by one or more of the following:
  - Decrease in the frequency of infection
  - Decrease in the severity of infection; **AND**
- Continued treatment is necessary to decrease the risk of infection

Toxic Shock Syndrome [46,93,94]

- May not be renewed.

Management of Immune Checkpoint Inhibitor Related Toxicity [73,80]

- May not be renewed.

Management of CAR T-Cell-Related Toxicity [73,80,86,104,105]

- Patient has received treatment with anti-CD19 CAR T-cell therapy (e.g., axicabtagene ciloleucel, brexucabtagene autoleucel, lisocabtagene maraleucel, tisagenlecleucel, etc.); **AND**
  - Patient has serum IgG levels <600 mg/dl; **OR**
- Patient is has received treatment with BCMA-targeted CAR T-cell therapy (e.g., idecabtagene vicleucel, ciltacabtagene autoleucel, etc.); **AND**
  - Patient has serum IgG levels <400 mg/dL

Supportive Care after Rethymic transplant ‡ [97]

- Renewals for use as initial immunoglobulin replacement therapy will be authorized until all of the following criteria are met:
  - Patient is no longer on immunosuppression (at least 10% of CD3+ T cells are naïve in phenotype); **AND**
  - Patient is at least 9 months post-treatment; **AND**
  - Patient's phytohemagglutinin (PHA) response within normal limits; **OR**
- Renewals for use as re-initiation of treatment after stopping immunoglobulin replacement therapy for patients with an IgG trough level lower than normal range will be continued for 1 year before being retested using the above guidelines.

Dosing Recommendations:

- Patient's dose should be reduced to the lowest necessary to maintain benefit for their condition. Patients who are stable, or who have reached the maximum therapeutic response, should have a trial of dose reduction (e.g., 25-50% reduction in dose every 3 months).
- Patients who have tolerated dose reduction and continue to show sustained improvement (i.e., remission) should have a trial of treatment discontinuation; with the following exceptions:
    - PID would be excluded from a trial of discontinuation
    - HIV-infected children should show satisfactory control of the underlying disease [e.g., undetectable viral load, CD4 counts elevated above 200 or >15% (ages 9 months – 5 years) on antiretroviral therapy, etc.]
    - Solid organ transplant, CLL, SLL, ALL, and MM patients should not be at an increased risk of infection

V. Dosage/Administration [1-16,24,25,32,41,53,58,63,64,76,78-80,83,84,89-94,99,101,102,106,110,111,116]

Dosing should be calculated using adjusted body weight if one or more of the following criteria are met:

- Patient's body mass index (BMI) is 30 kg/m^2 or more; **OR**
- Patient's actual body weight is 20% higher than his or her ideal body weight (IBW)

Use the following dosing formulas to calculate the adjusted body weight (round dose to nearest 5 gram increment in adult patients):

Dosing formulas
BMI = 703 x (weight in pounds/height in inches^2)
IBW (kg) for males = 50 + [2.3 (height in inches – 60)]
IBW (kg) for females = 45.5 + [2.3 x (height in inches – 60)]
Adjusted body weight = IBW + 0.4 (actual body weight – IBW)

This information is not meant to replace clinical decision making when initiating or modifying medication therapy and should only be used as a guide. Patient-specific variables should be taken into account.

Indication	Dose ¤
PID and Supportive Care after Rethymic transplant	200 to 800 mg/kg every 21 to 28 days
IgG Subclass Deficiency	300 to 400 mg/kg every 14 days

Indication	Dose ¤
CIDP	2 g/kg divided over 2-5 days initially, then 1 g/kg administered in 1-2 infusions every 21 days
ITP	2 g/kg divided over 5 days or 1 g/kg once daily for 2 consecutive days in a 28-day cycle
Fetal Alloimmune thrombocytopenia (FAIT)	1 g/kg/week until delivery
Kawasaki's Disease	1 g/kg to 2 g/kg x 1 dose, may be repeated once if needed
Multifocal Motor Neuropathy	Up to 2 g/kg divided over 5 days in a 28-day cycle
Acquired immune deficiency: CLL, SLL, MM, and ALL	400 mg/kg every 3 to 4 weeks
HIV Infected Children	400 mg/kg every 2 to 4 weeks
Guillain-Barré	2 g/kg divided over 5 days x 1 course. May be repeated once within 6 weeks of onset if needed
Myasthenia Gravis	1-2 g/kg divided as either 0.5 g/kg daily x 2 days or 0.4 g/kg daily x 5 days x 1 course
Auto-immune blistering diseases	Up to 2 g/kg divided over 5 days in a 28-day cycle
Dermatomyositis/Polymyositis	2 g/kg divided over 2 to 5 days in a 28-day cycle
Allogeneic Bone Marrow or Stem Cell Transplant	500 mg/kg once weekly x 90 days, then 500 mg/kg every 3 to 4 weeks
Complications of transplanted solid organ (kidney, liver, lung, heart, pancreas) and bone marrow transplant	2 g/kg divided over 5 days in a 28-day cycle
Stiff Person Syndrome	2 g/kg divided over 5 days in a 28-day cycle
Toxic Shock Syndrome	2 g/kg divided over 5 days x 1 course
Neonatal Alloimmune Thrombocytopenia (NAIT)	1 g/kg x 1 dose, may be repeated once if needed
Management of Immune Checkpoint Inhibitor Related Toxicity	2 g/kg divided over 5 days x 1 course
Management of CAR T-Cell-Related Toxicity	400-500 mg/kg every 28 days
¤ *Dosing for IVIG is highly variable depending on numerous patient specific factors, indication(s), and the specific product selected. For specific dosing regimens refer to current prescribing literature.*	

VI. Billing Code/Availability Information

HCPCS Code & NDC:

Drug	Manufacturer	HCPCS Code	1 Billable Unit Equivalent	IgG (grams) per SDV	NDC
Asceniv*	ADMA Biologics	J1554	500 mg	5	69800-0250-XX
Alyglo	GC Biopharma	J1599	N/A	5, 10, 20	61476-0104-XX
Bivigam*	ADMA Biologics	J1556	500 mg	5	69800-6502-XX
				10	69800-6503-XX
Flebogamma 10% DIF*	Instituto Grifols, S.A.	J1572	500 mg	5, 10, 20	61953-0005-XX
Flebogamma 5% DIF*				0.5, 2.5, 5, 10, 20	61953-0004-XX
Gamunex-C	Grifols Therapeutics	J1561	500 mg	1, 2.5, 5, 10, 20, 40	13533-0800-XX
Gammagard Liquid*	Baxalta	J1569	500 mg	1, 2.5, 5, 10, 20, 30	00944-2700-XX
Gammagard S/D*	Baxalta	J1566	500 mg	5	00944-2656-XX
				10	00944-2658-XX
Gammaked*	Grifols Therapeutics	J1561	500 mg	1, 2.5, 5, 10, 20	76125-0900-XX
Gammaplex 5%*	Bio Products Laboratory	J1557	500 mg	5, 10, 20	64208-8234-XX
Gammaplex 10%*				5, 10, 20	64208-8235-XX
Octagam 10%*	Octapharma USA Inc	J1568	500 mg	2, 5, 10, 20, 30	68982-0850-XX
Octagam 5%*				1, 2.5, 5, 10, 25	68982-0840-XX
Privigen*	CSL Behring AG	J1459	500 mg	5	44206-0436-XX
				10	44206-0437-XX
				20	44206-0438-XX
				40	44206-0439-XX

Panzyga*	Octapharma USA Inc	J1576	500mg	1, 2.5, 5, 10, 20, 30	68982-0820-XX
Yimmugo	Biotest AG	J1599	N/A	5, 10, 20	83372-0605-XX
Injection, immune globulin, intravenous, non-lyophilized (e.g., liquid), not otherwise specified	N/A	J1599	500 mg	N/A	N/A
**90283 – immune globulin (IgIV), human, for intravenous use*					

VII. References

1. Bivigam® [package insert]. Boca Raton, FL; ADMA Biologics, Inc.; December 2023. Accessed June 2024.
2. Flebogamma® 10% DIF [package insert]. Barcelona, Spain; Instituto Grifols, S.A.; September 2019. Accessed October 2023.
3. Flebogamma® 5% DIF [package insert]. Barcelona, Spain; Instituto Grifols, S.A.; September 2019. Accessed October 2023.
4. Gammagard® Liquid [package insert]. Lexington, MA; Baxalta US Inc.; January 2024. Accessed June 2024.
5. Gammagard® S/D Less IgA [package insert]. Lexington, MA; Baxalta US Inc.; March 2023. Accessed October 2023.
6. Gamunex®-C [package insert]. Research Triangle, NC; Grifols Therapeutics, Inc.; January 2020. Accessed October 2023.
7. Gammaked™ [package insert]. Research Triangle, NC; Grifols Therapeutics, Inc; January 2020. Accessed October 2023.
8. Gammaplex® 5% [package insert]. Durham, NC; Bio Products Laboratory Ltd.; September 2019. Accessed October 2023.
9. Gammaplex® 10% [package insert]. Durham, NC; Bio Products Laboratory Ltd.; November 2021. Accessed October 2023.
10. Octagam® 5% [package insert]. Paramus, NJ; Octapharma USA Inc; April 2022. Accessed October 2023.
11. Octagam® 10% [package insert]. Paramus, NJ; Octapharma USA Inc; April 2022. Accessed October 2023.
12. Privigen® [package insert]. Berne, Switzerland; CSL Behring AG March 2022. Accessed October 2023.
13. Panzyga® [package insert]. Paramus, NJ; Octapharma USA Inc; February 2021. Accessed October 2023.
14. Asceniv™ [package insert]. Boca Raton, FL; ADMA Biologics; April 2019. Accessed October 2023.

15. Alyglo™ [package insert]. Republic of Korea; GC Biopharma Corp; December 2023. Accessed December 2023.
16. Yimmugo® [package insert]. Dreieich, Germany; Biotest AG; June 2024. Accessed June 2024.
17. Skeie GO, Apostolski S, Evoli A, et al. Guidelines for the treatment of autoimmune neuromuscular transmission disorders. Eur J Neurol. 2010;17(7):893-902.
18. Van den Bergh PY, Hadden RD, Bouche P, et al. European Federation of Neurological Societies/Peripheral Nerve Society guideline on management of chronic inflammatory demyelinating polyradiculoneuropathy: report of a joint task force of the European Federation of Neurological Societies [trunc]. Eur J Neurol 2010 Mar;17(3):356-63.
19. Patwa HS, Chaudhry V, Katzberg H, et al. Evidence-based guideline: intravenous immunoglobulin in the treatment of neuromuscular disorders: report of the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology. Neurology. 2012 Mar 27;78(13):1009-15.
20. French CIDP Study Group. Recommendations on diagnostic strategies for chronic inflammatory demyelinating polyradiculoneuropathy. J Neurol Neurosurg Psychiatry 2008; 79: 115–118.
21. Donofrio PD, Berger A, Brannagan TH, et al. Consensus statement: The use of intravenous immunoglobulin in the treatment of neuromuscular conditions report of the AANEM ad hoc committee. Muscle Nerve. 2009;40:890-900.
22. Feasby T, Banwell B, Benstead T, et al. Guidelines on the use of intravenous immune globulin for neurologic conditions. Transfus Med Rev. 2007;21(2 suppl 1):S57-107.
23. Gajdos P, Tranchant C, Clair B, et al; Myasthenia Gravis Clinical Study Group. Treatment of myasthenia gravis exacerbation with intravenous immunoglobulin: a randomized double-blind clinical trial. Arch Neurol. 2005;62(11):1689-1693.
24. Elovaara I, et al. EFNS guidelines for the use of intravenous immunoglobulin in treatment of neurological diseases: EFNS task force on the use of intravenous immunoglobulin in treatment of neurological diseases. European Journal of Neurology 2008;15(9):893-908.
25. Joint Task Force of the EFNS and the PNS. European Federation of Neurological Societies/Peripheral Nerve Society guideline on management of multifocal motor neuropathy. Report of a joint task force of the European Federation of Neurological Societies and the Peripheral Nerve Society--first revision. J Peripher Nerv Syst. 2010 Dec;15(4):295-301. doi: 10.1111/j.1529-8027.2010.00290.x.
26. Hahn AF, Bolton CF, Pillay N, et al. Plasma exchange therapy in chronic inflammatory demyelinating polyneuropathy. A double-blind, sham controlled, cross-over study. Brain 1996;119:1055–66.
27. The National Institute of Child Health and Human Developments Intravenous Immunoglobulin Study Group. Intravenous immune globulin for the prevention of bacterial infections in children with symptomatic human immunodeficiency virus infection. N Engl J Med. 1991 Jul 11;325(2):73-80.
28. Silberry GK, Abzug MJ, Nachman, S, et al. Guidelines for the Prevention and Treatment of Opportunistic Infections in HIV-Exposed and HIV-Infected Children: Recommendations from the National Institutes of Health, Centers for Disease Control and Prevention, the HIV Medicine

Association of the Infectious Diseases Society of America, the Pediatric Infectious Diseases Society, and the American Academy of Pediatrics. J Pediatric Infect Dis Soc. 2013 Nov; 32 Suppl 2: i-KK4.

29. Wolfe GI, Barohn RJ, Foster BM, et al; Myasthenia Gravis-IVIG Study Group. Randomized, controlled trial of intravenous immunoglobulin in myasthenia gravis. Muscle Nerve. 2002;26(4):549-552.
30. Hughes RA, Wijdicks EF, Barohn R, et al. Quality Standards Subcommittee of the American Academy of Neurology. Practice parameter: immunotherapy for Guillain-Barré syndrome: report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology. 2003 (reaffirmed in 2016);61(6):736-740.
31. Hughes RA, Swan AV, Raphael JC, et al. Immunotherapy for Guillain-Barré syndrome: a systematic review. Brain. 2007;130(pt 9):2245-2257.
32. Bussel, JB et al. Antenatal management of alloimmune thrombocytopenia with Intravenous Immunoglobulin: A randomized trial of low dose steroid to intravenous immunoglobulin. Am J Obstet Gynecol 1996; 174 1414-23.
33. Ratko TA, Burnett DA, The Univ Hospital Consortium Expert Panel for the Off-label Use of Polyvalent Intravenously Administered Immunoglobulin Preparations, et al. Recommendations for the off-label use of intravenously administered immunoglobulin preparations. JAMA 1995; 273:1865-70.
34. Ahmed AR, Spigelman Z, Cavacine LA et al. Treatment of pemphigus vulgaris with rituximab and intravenous immune globulin. N Eng J Med 2006; 1772-9.
35. American Academy of Pediatrics Subcommittee on Hyperbilirubinemia. Management of hyperbilirubinemia in the newborn infant 35 or more weeks of gestation. Pediatrics 2004; 114:297-316.
36. Gottstein R, Cooke R. Systematic Review of intravenous immunoglobulin in haemolytic disease of the newborn. Arch Dis Child Fetal Neonatal Ed 2003; 88:F6-10
37. Anderson D, Ali K, Blanchette V, et al. Guidelines on the use of intravenous immune globulin for hematologic conditions. Transfus Med Rev. 2007;21(2 Suppl 1):S9-56.
38. Orange J, Hossny E, Weiler C, et al. Use of intravenous immunoglobulin in human disease: A review of evidence by members of the Primary Immunodeficiency Committee of the American Academy of Allergy, Asthma and Immunology. J Allergy Clin Immunol 2006;117(4 Suppl): S525-53.
39. Stasi R, Evangelista ML, Stipa E, et al. Idiopathic thrombocytopenic purpura: current concepts in pathophysiology and management. Thrombosis and Haemostasis 2008;99(1):4-13.
40. Amagai M, Ikeda S, Shimizu H, et al. A randomized, double-blind trial of intravenous immunoglobulin for pemphigus. J Am Acad Dermatol 2009; 60:595-602.
41. Ahmed AR. Intravenous immunoglobulin therapy in the treatment of patients with pemphigus vulgaris unresponsive to conventional immunosuppressive treatment. J Am Acad Dermatol 2001; 45:679-90.

42. Hughes R, Bensa S, Willison H, Vet al. Inflammatory Neuropathy Cause and Treatment (INCAT) Group. Randomized controlled trial of intravenous immunoglobulin versus oral prednisolone in chronic inflammatory demyelinating polyradiculoneuropathy. Ann Neurol. 2001 Aug;50(2):195-201.
43. Zinman L, Ng E, Bril V. IV immunoglobulin in patients with myasthenia gravis: a randomized controlled trial. Neurology. 2007 Mar 13;68(11):837-41.
44. Koski CL, Baumgarten M, Magder LS, et al. Derivation and validation of diagnostic criteria for chronic inflammatory demyelinating polyneuropathy. Journal of the Neurological Sciences 2009; 277:1-8.
45. Sullivan KM, Storek J, Kopecky KJ, et al. A controlled trial of long-term administration of intravenous immunoglobulin to prevent late infection and chronic graft-vs.-host disease after marrow transplantation: clinical outcome and effect on subsequent immune recovery. Biol Blood Marrow Transplant 1996;2:44-53.
46. Alejandria MM, Lansang MA, Dans LF, Mantaring JB. Intravenous immunoglobulin for treating sepsis and septic shock. Cochrane Database Syst Rev 2002;CD001090.
47. American College of Obstetricians and Gynecologists (ACOG), Committee on Practice Bulletins -- Obstetrics. Thrombocytopenia in pregnancy. ACOG Practice Pattern No. 6. Washington, DC: ACOG; September 1999.
48. Centers for Disease Control and Prevention. Guidelines for preventing opportunistic infections among hematopoietic stem cell transplant recipients: recommendations of CDC, the Infectious Disease Society of America, and the American Society of Blood and Marrow Transplantation. MMWR 2000;49(No. RR-10):1-128.
49. Emerson GG, Herndon CN, Sreih AG. Thrombotic complications after intravenous immunoglobulin therapy in two patients. Pharmacotherapy. 2002;22:1638-1641.
50. Department of Health (London). Clinical Guidelines for Immunoglobulin Use: Update to Second Edition. August, 2011.
51. Provan, Drew, et al. "Clinical guidelines for immunoglobulin use." Department of Health Publication, London (2008).
52. Sussman J, Farrugia ME, Maddison P, et al. Myasthenia gravis: Association of British Neurologists' management guidelines. Pract Neurol 2015; 15: 199-206.
53. Sanders DB, Wolfe GI, Benatar M, et al. International consensus guidance for management of myasthenia gravis-Executive Summary. Neurology. 2016 Jul 26; 87(4): 419-25.
54. Orange JS, Ballow M, Stiehm, et al. Use and interpretation of diagnostic vaccination in primary immunodeficiency: A working group report of the Basic and Clinical Immunology Interest Section of the American Academy of Allergy, Asthma & Immunology. J Allergy Clin Immunol Vol 130 (3).
55. Neunert C, Lim W, Crowther M, et al. The American Society of Hematology 2011 Evidence-based practice guidelines for immune thrombocytopenia. Blood April 2011; Vol 117 (16).
56. Jeffrey Modell Foundation Medical Advisory Board, 2013. 10 Warning Signs of Primary Immunodeficiency. Jeffrey Modell Foundation, New York, NY.
57. Bonilla FA, Khan DA, Ballas ZK, et al. Practice Parameter for the diagnosis and management of primary immunodeficiency. J Allergy Clin Immunol 2015 Nov;136(5):1186-205.e1-78.

58. Kuitwaard K, de Gelder J, Tio-Gillen AP, et al. Pharmacokinetics of intravenous immunoglobulin and outcome in Guillain-Barré syndrome. Ann Neurol. 2009;66(5):597.
59. Shehata N, Palda VA, Meyer RM, et al. The use of immunoglobulin therapy for patients undergoing solid organ transplantation: an evidence-based practice guideline. Transfus Med Rev 2010; 24 Suppl 1:S7-S27.
60. Jordan SC, Tyan D, Stablein D, et al. Evaluation of intravenous immunoglobulin as an agent to lower allosensitization and improve transplantation in highly sensitized adult patients with end-stage renal disease: report of the NIH IG02 trial. J Am Soc Nephrol 2004; 15(12):3256-3262.
61. Yuan XP, Wang CX, Gao W, et al. Kidney transplant in highly sensitized patients after desensitization with plasmapheresis and low-dose intravenous immunoglobulin. Exp Clin Transplant 2010; 8(2):130-135.
62. Jordan SC, Quartel AW, Czer LSC, et al. Posttransplant therapy using high-dose human immunoglobulin (intravenous gamma globulin) to control acute humoral rejection in renal and cardiac allograft recipients and potential mechanism of action. Transplantation 1998; 66(6):800-805.
63. Sullivan KM, Kopecky KJ, Jocom J, et al. Immunomodulatory and antimicrobial efficacy of intravenous immunoglobulin in bone marrow transplantation. N Engl J Med 1990; 323:705-712.
64. Bhatti AB, Gazali ZA. Recent Advances and Review on Treatment of Stiff Person Syndrome in Adults and Pediatric Patients. Cureus. 2015 Dec 22;7(12):e427
65. Tanimoto K, Nakano K, Kano S, et al. Classification criteria for polymyositis and dermatomyositis. J Rheumatol. 1995 Apr;22(4):668-74.
66. Kyriakides T, Angelini C, Schaefer J, et al. EFNS guidelines on the diagnostic approach to pauci- or asymptomatic hyperCKemia. Eur J Neurol. 2010 Jun 1;17(6):767-73.
67. Feliciani C, Joly P, Jonkman MF, et al. Management of bullous pemphigoid: the European Dermatology Forum consensus in collaboration with the European Academy of Dermatology and Venereology. Br J Dermatol. 2015 Apr;172(4):867-77.
68. Hertl M, Jedlickova H, Karpati S, et al. Pemphigus. S2 Guideline for diagnosis and treatment--guided by the European Dermatology Forum (EDF) in cooperation with the European Academy of Dermatology and Venereology (EADV). J Eur Acad Dermatol Venereol. 2015 Mar;29(3):405-14.
69. Harman KE, Albert S, Black MM; British Association of Dermatologists. Guidelines for the management of pemphigus vulgaris. Br J Dermatol. 2003 Nov;149(5):926-37.
70. Perez EE, Orange JS, Bonilla F, et al. Update on the use of immunoglobulin in human disease: A review of evidence. J Allergy Clin Immunol. 2017 Mar;139(3S):S1-S46.
71. Dantal J. Intravenous Immunoglobulins: In-Depth Review of Excipients and Acute Kidney Injury Risk. Am J Nephrol 2013;38:275-284.
72. Rajabally YA et al. Validity of diagnostic criteria for chronic inflammatory demyelinating polyneuropathy: A multicentre European study. J Neurol Neurosurg Psychiatry 2009 Dec; 80:1364.
73. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) Management of Immunotherapy-Related Toxicities, Version 2.2023. National Comprehensive Cancer Network, 2023. The NCCN Compendium® is a derivative work of the

NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc." To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed October 2023.

74. Postow, MA. Managing Immune Checkpoint-Blocking Antibody Side Effects. American Society of Clinical Oncology Education Book. 2015; 76-83.
75. Williams TJ, Benavides DR, Patrice KA. Association of Autoimmune Encephalitis with combined immune checkpoint inhibitor treatment for metastatic cancer. JAMA Neurol .933-928:(8)73;2016 doi:10.1001/jamaneurol.2016.1399
76. Tomblyn M, Chiller T, Einsele H, et al. Guidelines for preventing infectious complications among hematopoietic cell transplantation recipients: a global perspective. Biol Blood Marrow Transplant. 2009;15(10):1143-1238. doi: 10.1016/j.bbmt.2009.06.019. [PubMed 19747629]
77. Willison HJ, Jacobs BS, van Doom PA. Guillain-Barré Syndrome. Lancet. 2016 Aug;388(10045):717-27. Epub 2016 Mar 2
78. Sanders DB, Wolfe GI, Benetar M, et al. International consensus guidance for management of myasthenia gravis. Neurology 2016;87:1–7
79. Van Winkle P, Burchette R, Kim R, et al. Prevalence and Safety of Intravenous Immunoglobulin Administration During Maintenance Chemotherapy in Children with Acute Lymphoblastic Leukemia in First Complete Remission: A Health Maintenance Organization Perspective. Perm J. 2018; 22: 17-141.
80. Referenced with permission from the NCCN Drugs and Biologics Compendium (NCCN Compendium®) Immune globulin. National Comprehensive Cancer Network, 2023. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed September 2023.
81. Neunert C, Terrell DR, Arnold DM, et al. American Society of Hematology 2019 guidelines for immune thrombocytopenia. Blood Adv. 2019; 3(23): 3829-3866.
82. Lundberg IE, Tjärnlund A, Bottai M, et al. 2017 European League Against Rheumatism/American College of Rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major. Ann Rheum Dis. 2017;76(12):1955-1964.
83. McCrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, treatment, and long-term management of Kawasaki disease: A scientific statement for health professionals from the American Heart Association. Circulation 2017;135:e927-e999.
84. Winkelhorst D, Murphy MF, Greinacher A, et al. Antenatal management in fetal and neonatal alloimmune thrombocytopenia: a systematic review. Blood. 2017;129(11):1538-1547.
85. Narayanaswami P, Sanders D, Wolfe G, Benatar M, et al. International consensus guidance for management of myasthenia gravis, 2020 update. Neurology® 2021;96:114-122. doi:10.1212/WNL.0000000000011124

86. Hill J, Giralt S, Torgerson T, et al. CAR-T– and a side order of IgG, to go? – Immunoglobulin Replacement in Patients Receiving CAR-T Cell Therapy. Blood Rev. 2019 Nov; 38: 100596. E-pub doi: 10.1016/j.blre.2019.100596
87. Aggarwal R, Schoeman C, Schessl J, et al. Prospective, double-blind, randomized, placebo-controlled phase III study evaluating efficacy and safety of octagam 10% in patients with dermatomyositis ("ProDERM Study").Clinical Trial Medicine (Baltimore) 2021 Jan 8;100(1):e23677. doi: 10.1097/MD.0000000000023677.
88. Chapel H, Dicato M, Gamm H, et al. Immunoglobulin replacement in patients with chronic lymphocytic leukaemia: a comparison of two dose regimes. Br J Haematol 1994 Sep;88(1):209-12. doi: 10.1111/j.1365-2141.1994.tb05002.x.
89. Panel on Opportunistic Infections in HIV-Exposed and HIV-Infected Children. Updates to Guidelines for the Prevention and Treatment of Opportunistic Infections in HIV-Exposed and HIV-Infected Children. Department of Health and Human Services. Available at https://clinicalinfo.hiv.gov/en/guidelines/pediatric-opportunistic-infection. Updated September 2023.
90. ACOG Practice Bulletin No. 207: Thrombocytopenia in Pregnancy. Obstet Gynecol. 2019 Mar;133(3):e181-e193. doi: 10.1097/AOG.0000000000003100.
91. Harman KE, Brown D, Exton LS, et al. British Association of Dermatologists' guidelines for the management of pemphigus vulgaris 2017. Br J Dermatol. 2017 Nov;177(5):1170-1201. doi: 10.1111/bjd.15930.
92. Ueda M, Berger M, Gale RP, Lazarus HM. Immunoglobulin therapy in hematologic neoplasms and after hematopoietic cell transplantation. Blood Rev. 2018 Mar;32(2):106-115. doi: 10.1016/j.blre.2017.09.003.
93. Alejandria MM, Lansang MA, Dans LF, Mantaring JB 3rd. Intravenous immunoglobulin for treating sepsis, severe sepsis and septic shock. Cochrane Database Syst Rev. 2013 Sep 16;2013(9):CD001090. Doi: 10.1002/14651858.CD001090.pub2.
94. Cawley MJ, Briggs M, Haith LR, et al: Intravenous immunoglobulin as adjunctive treatment for streptococcal toxic shock syndrome associated with necrotizing fasciitis: case report and review. Pharmacotherapy 1999; 19(9):1094-1098.
95. Hill JA, Seo SK. How I prevent infections in patients receiving CD19-targeted chimeric antigen receptor T cells for B-cell malignancies. Blood. 2020 Aug 20;136(8):925-935. Doi: 10.1182/blood.2019004000.
96. Derman BA, Schlei Z, Parsad S, et al. Changes in Intravenous Immunoglobulin Usage for Hypogammaglobulinemia After Implementation of a Stewardship Program. JCO Oncol Pract. 2021 Mar;17(3):e445-e453. doi: 10.1200/OP.20.00312.
97. Rethymic [package insert]. Cambridge, MA; Enzyvant Therapeutics, Inc.; February 2023. Accessed October 2023.
98. Abdou NI, Greenwell CA, Mehta R, et al. Efficacy of intravenous gammaglobulin for immunoglobulin G subclass and/or antibody deficiency in adults. Int Arch Allergy Immunol. 2009;149(3):267-74. doi: 10.1159/000199723.

99. Abrahamian F, Agrawal S, Gupta S. Immunological and clinical profile of adult patients with selective immunoglobulin subclass deficiency: response to intravenous immunoglobulin therapy. Clin Exp Immunol. 2010 Mar;159(3):344-50. doi: 10.1111/j.1365-2249.2009.04062.x.
100. Olinder-Nielsen AM, Granert C, Forsberg P, et al. Immunoglobulin prophylaxis in 350 adults with IgG subclass deficiency and recurrent respiratory tract infections: a long-term follow-up. Scand J Infect Dis. 2007;39(1):44-50. doi: 10.1080/00365540600951192.
101. Grindeland JW, Grindeland CJ, Moen C, Leedahl ND, Leedahl DD. Outcomes Associated With Standardized Ideal Body Weight Dosing of Intravenous Immune Globulin in Hospitalized Patients: A Multicenter Study. Ann Pharmacother. 2020 Mar;54(3):205-212. doi: 10.1177/1060028019880300. Epub 2019 Oct 3.
102. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) Prevention and Treatment of Cancer-Related Infections, Version 1.2023. National Comprehensive Cancer Network, 2023. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed September 2023.
103. Jeffrey Modell Foundation Medical Advisory Board, 2021. 10 Warning Signs of Primary Immunodeficiency. Jeffrey Modell Foundation, New York, NY. https://res.cloudinary.com/info4pi/image/upload/v1662306262/JMF_10_Signs_Generic_082421_v2_dcadf429cc.pdf?updated_at=2022-09-04T15:44:23.120Z. Accessed October 2023.
104. Abecma [package insert]. Summit, NJ; Celgene., Inc., March 2021. Accessed October 2023.
105. Carvykti [package insert]. Horsham, PA; Janssen Biotech, Inc., February 2023. Accessed October 2023.
106. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) Multiple Myeloma, Version 1.2024. National Comprehensive Cancer Network, 2023. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed October 2023.
107. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) Chronic Lymphocytic Leukemia/Small Lymphocytic Lymphoma, Version 3.2023. National Comprehensive Cancer Network, 2023. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed October 2023.
108. de Graeff N, Groot N, Ozen S, et al. European consensus-based recommendations for the diagnosis and treatment of Kawasaki disease - the SHARE initiative. Rheumatology (Oxford). 2019 Apr 1;58(4):672-682. doi: 10.1093/rheumatology/key344.

109. Gorelik M, Chung SA, Ardalan K, et al. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Kawasaki Disease. Arthritis Care Res (Hoboken). 2022 Apr;74(4):538-548. doi: 10.1002/acr.24838.
110. Schmidt E, Rashid H, Marzano A, et al. European Guidelines (s3) on diagnosis and management of mucous membrane pemphigoid, initiated by the european academy of dermatology and venereology - part ii. J Eur Acad Dermatol Venereology (2021) 35:1926–48. doi: 10.1111/jdv.17395
111. Joly P, Horvath B, Patsatsi A, et al. Updated s2k guidelines on the management of pemphigus vulgaris and foliaceus initiated by the european academy of dermatology and venereology (eadv). J Eur Acad Dermatol Venereology (2020) 34:1900–13. doi: 10.1111/jdv.16752
112. Borradori L, Beek NV, Feliciani C, et al. Updated s2 k guidelines for the management of bullous pemphigoid initiated by the european academy of dermatology and venereology (eadv). J Eur Acad Dermatol Venereology (2022) 36:1689–704. doi: 10.1111/jdv.18220
113. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) Hematopoietic Cell Transplantation, Version 3.2023. National Comprehensive Cancer Network, 2023. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed October 2023.
114. Dalakas, M.C. Stiff-person Syndrome and GAD Antibody-spectrum Disorders: GABAergic Neuronal Excitability, Immunopathogenesis and Update on Antibody Therapies. Neurotherapeutics 19, 832–847 (2022).
115. Hughes, RAC, Wijdicks EFM, Barohn R, et al. Practice parameter: Immunotherapy for Guillain–Barré syndrome Report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology Sep 2003, 61 (6) 736-740; DOI: 10.1212/WNL.61.6.736
116. Van den Bergh PYK, van Doorn PA, Hadden RDM, et al. European Academy of Neurology/Peripheral Nerve Society guideline on diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy: Report of a joint Task Force-Second revision. J Peripher Nerv Syst. 2021 Sep;26(3):242-268. doi: 10.1111/jns.12455. Erratum in: J Peripher Nerv Syst. 2022 Mar;27(1):94. Erratum in: Eur J Neurol. 2022 Apr;29(4):1288.
117. National Coverage Determination (NCD) for Intravenous Immune Globulin for the Treatment of Autoimmune Mucocutaneous Blistering Diseases (250.3). Centers for Medicare and Medicaid Services, Inc. Updated on 10/01/2002 with effective date 10/01/2002. Accessed January 2024.
118. National Government Services, Inc. Local Coverage Article: Billing and Coding: Off-Label Use of Intravenous Immune Globulin (IVIG) (A59105). Centers for Medicare & Medicaid Services, Inc. Updated on 06/21/2023 with effective date 07/01/2023. Accessed January 2024.
119. Noridian Healthcare Solutions, LLC. Local Coverage Article: Billing and Coding: Intravenous Immune Globulin (IVIg)-NCD 250.3 (A54641, A54643). Centers for Medicare & Medicaid Services, Inc. Updated on 11/17/2023 with effective date 11/07/2015. Accessed January 2024.
120. Noridian Healthcare Solutions, LLC. Local Coverage Article: Billing and Coding: Coverage of Intravenous Immune Globulin for Treatment of Primary Immune Deficiency Diseases in the Home

– Medicare Benefit Policy Manual, Chapter 15, 50.6 (A54660, A54662). Centers for Medicare & Medicaid Services, Inc. Updated 11/17/2023 with effective date 08/13/2019. Accessed January 2024.

121. Noridian Healthcare Solutions, LLC. Local Coverage Article: Billing and Coding: Immune Globulin Intravenous (IVIg) (A57187). Centers for Medicare & Medicaid Services, Inc. Updated 11/16/2023 with effective date 07/01/2023. Accessed January 2024.
122. Noridian Healthcare Solutions, LLC. Local Coverage Article: Billing and Coding: Immune Globulin Intravenous (IVIg) (A57194). Centers for Medicare & Medicaid Services, Inc. Updated 11/16/2023 with effective date 07/01/2023. Accessed January 2024.
123. Wisconsin Physicians Service Insurance Corporation. Local Coverage Article: Billing and Coding: Immune Globulins (A57554). Centers for Medicare & Medicaid Services, Inc. Updated on 11/22/2022 with effective date 12/01/2022. Accessed January 2024.
124. CGS, Administrators, LLC. Local Coverage Article: Billing and Coding: Immune Thrombocytopenia (ITP) Therapy (A57160). Centers for Medicare & Medicaid Services, Inc. Updated on 11/07/2023 with effective date 11/16/2023. Accessed January 2024.
125. First Coast Service Options, Inc. Local Coverage Article: Billing and Coding: Immune Globulin (A57778). Centers for Medicare & Medicaid Services, Inc. Updated on 07/14/2023 with effective date 07/01/2023. Accessed January 2024.
126. Novitas Solutions, Inc. Local Coverage Article: Billing and Coding: Immune Globulin (A56786). Centers for Medicare & Medicaid Services, Inc. Updated on 07/14/2023 with effective date 07/01/2023. Accessed January 2024.
127. Palmetto GBA, LLC. Local Coverage Article: Intravenous Immunoglobulin (IVIG) (A56718). Centers for Medicare & Medicaid Services, Inc. Updated on 10/26/2023 with effective date 10/01/2023. Accessed January 2024.
128. CGS, Administrators, LLC. Local Coverage Article: Billing and Coding: Intravenous Immune Globulin (A56779). Centers for Medicare & Medicaid Services, Inc. Updated on 11/07/2023 with effective date 11/16/2023. Accessed January 2024.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
A48.3	Toxic shock syndrome
B20	Human immunodeficiency virus (HIV) disease
B25.0	Cytomegaloviral pneumonitis
B25.1	Cytomegaloviral hepatitis
B25.2	Cytomegaloviral pancreatitis
B25.8	Other cytomegaloviral diseases
B25.9	Cytomegaloviral disease, unspecified
C83.00	Small cell B-cell lymphoma, unspecified site

ICD-10	ICD-10 Description
C83.01	Small cell B-cell lymphoma, lymph nodes of head, face, and neck
C83.02	Small cell B-cell lymphoma, intrathoracic lymph nodes
C83.03	Small cell B-cell lymphoma, intra-abdominal lymph nodes
C83.04	Small cell B-cell lymphoma, lymph nodes of axilla and upper limb
C83.05	Small cell B-cell lymphoma, lymph nodes of inguinal region and lower limb
C83.06	Small cell B-cell lymphoma, intrapelvic lymph nodes
C83.07	Small cell B-cell lymphoma, spleen
C83.08	Small cell B-cell lymphoma, lymph nodes of multiple sites
C83.09	Small cell B-cell lymphoma, extranodal and solid organ sites
C91.10	Chronic lymphocytic leukemia of B-cell type not having achieved remission
C91.11	Chronic lymphocytic leukemia of B-cell type in remission
C91.12	Chronic lymphocytic leukemia of B-cell type in relapse
C90.00	Multiple Myeloma not having achieved remission
C90.01	Multiple Myeloma in remission
C90.02	Multiple Myeloma in relapse
C90.10	Plasma cell leukemia not having achieved remission
C90.11	Plasma cell leukemia in remission
C90.12	Plasma cell leukemia in relapse
C90.00	Acute lymphoblastic leukemia not having achieved remission
C90.01	Acute lymphoblastic leukemia, in remission
C90.02	Acute lymphoblastic leukemia, in relapse
D69.3	Immune thrombocytopenic purpura
D69.41	Evans syndrome
D69.42	Congenital and hereditary thrombocytopenic purpura
D69.49	Other primary thrombocytopenia
D69.59	Other secondary thrombocytopenia
D80.0	Hereditary hypogammaglobulinemia
D80.1	Nonfamilial hypogammaglobulinemia
D80.3	Selective deficiency of immunoglobulin G [IgG] subclasses
D80.5	Immunodeficiency with increased immunoglobulin M [IgM]
D80.7	Transient hypogammaglobulinemia of infancy
D81.0	Severe combined immunodeficiency [SCID] with reticular dysgenesis
D81.1	Severe combined immunodeficiency [SCID] with low T- and B-cell numbers
D81.2	Severe combined immunodeficiency [SCID] with low or normal B-cell numbers

ICD-10	ICD-10 Description
D81.6	Major histocompatibility complex class I deficiency
D81.7	Major histocompatibility complex class II deficiency
D81.89	Other combined immunodeficiencies
D81.9	Combined immunodeficiency, unspecified
D82.0	Wiskott-Aldrich syndrome
D82.1	DiGeorge's syndrome
D82.8	Immunodeficiency associated with other specified major defects
D83.0	Common variable immunodeficiency with predominant abnormalities of B-cell numbers and function
D83.2	Common variable immunodeficiency with autoantibodies to B- or T-cells
D83.8	Other common variable immunodeficiencies
D83.9	Common variable immunodeficiency, unspecified
D89.810	Acute graft-versus-host disease
D89.812	Acute on chronic graft-versus-host disease
D89.834	Cytokine release syndrome, grade 4
D89.839	Cytokine release syndrome, grade unspecified
G03.8	Meningitis due to other specified causes
G03.9	Meningitis, unspecified
G04.81	Other encephalitis and encephalomyelitis
G04.89	Other myelitis
G04.90	Encephalitis and encephalomyelitis, unspecified
G04.91	Myelitis, unspecified
G25.82	Stiff-man syndrome
G56.80	Other specified mononeuropathies of unspecified upper limb
G56.81	Other specified mononeuropathies of right upper limb
G56.82	Other specified mononeuropathies of left upper limb
G56.83	Other specified mononeuropathies of bilateral upper limbs
G56.90	Unspecified mononeuropathy of unspecified upper limb
G56.91	Unspecified mononeuropathy of right upper limb
G56.92	Unspecified mononeuropathy of left upper limb
G56.93	Unspecified mononeuropathy of bilateral upper limbs
G57.80	Other specified mononeuropathies of unspecified lower limb
G57.81	Other specified mononeuropathies of right lower limb
G57.82	Other specified mononeuropathies of left lower limb
G57.83	Other specified mononeuropathies of bilateral lower limbs

ICD-10	ICD-10 Description
G57.90	Unspecified mononeuropathy of unspecified lower limb
G57.91	Unspecified mononeuropathy of right lower limb
G57.92	Unspecified mononeuropathy of left lower limb
G57.93	Unspecified mononeuropathy of bilateral lower limbs
G61.0	Guillain-Barre syndrome
G61.1	Serum neuropathy
G61.81*	Chronic inflammatory demyelinating polyneuritis
G61.82	Multifocal motor neuropathy
G61.89	Other inflammatory polyneuropathies
G61.9	Inflammatory polyneuropathy, unspecified
G62.0	Drug-induced polyneuropathy
G62.89	Other specified polyneuropathies
G70.00	Myasthenia gravis without (acute) exacerbation
G70.01	Myasthenia gravis with (acute) exacerbation
H46.9	Unspecified optic neuritis
I30.8	Other forms of acute pericarditis
I30.9	Acute pericarditis, unspecified
I40.8	Other acute myocarditis
I40.9	Acute myocarditis, unspecified
J70.2	Acute drug-induced interstitial lung disorders
J70.4	Drug-induced interstitial lung disorders, unspecified
L10.0	Pemphigus vulgaris
L10.2	Pemphigus foliaceous
L12.0	Bullous pemphigoid
L12.1	Cicatricial pemphigoid
L12.30	Acquired epidermolysis bullosa, unspecified
L12.31	Epidermolysis bullosa due to drug
L12.35	Other acquired epidermolysis bullosa
L12.5	Other acquired epidermolysis bullosa
L13.8	Other specified bullous disorders
L13.9	Bullous disorder, unspecified
L51.1	Stevens-Johnson syndrome
L51.2	Toxic epidermal necrolysis [Lyell]
M30.3	Mucocutaneous lymph node syndrome [Kawasaki]

ICD-10	ICD-10 Description
M33.00	Juvenile dermatomyositis, organ involvement unspecified
M33.01	Juvenile dermatomyositis with respiratory involvement
M33.02	Juvenile dermatomyositis with myopathy
M33.03	Juvenile dermatomyositis without myopathy
M33.09	Juvenile dermatomyositis with other organ involvement
M33.10	Other dermatomyositis, organ involvement unspecified
M33.11	Other dermatomyositis with respiratory involvement
M33.12	Other dermatomyositis with myopathy
M33.13	Other dermatomyositis without myopathy
M33.19	Other dermatomyositis with other organ involvement
M33.20	Polymyositis, organ involvement unspecified
M33.21	Polymyositis with respiratory involvement
M33.22	Polymyositis with myopathy
M33.29	Polymyositis with other organ involvement
M33.90	Dermatopolymyositis, unspecified, organ involvement unspecified
M33.91	Dermatopolymyositis, unspecified with respiratory involvement
M33.92	Dermatopolymyositis, unspecified with myopathy
M33.93	Dermatopolymyositis, unspecified without myopathy
M33.99	Dermatopolymyositis, unspecified with other organ involvement
M36.0	Dermato(poly)myositis in neoplastic disease
M60.80	Other myositis, unspecified site
M60.811	Other myositis, right shoulder
M60.812	Other myositis, left shoulder
M60.819	Other myositis, unspecified shoulder
M60.821	Other myositis, right upper arm
M60.822	Other myositis, left upper arm
M60.829	Other myositis, unspecified upper arm
M60.831	Other myositis, right forearm
M60.832	Other myositis, left forearm
M60.839	Other myositis, unspecified forearm
M60.841	Other myositis, right hand
M60.842	Other myositis, left hand
M60.849	Other myositis, unspecified hand
M60.851	Other myositis, right thigh

ICD-10	ICD-10 Description
M60.852	Other myositis, left thigh
M60.859	Other myositis, unspecified thigh
M60.861	Other myositis, right lower leg
M60.862	Other myositis, left lower leg
M60.869	Other myositis, unspecified lower leg
M60.871	Other myositis, right ankle and foot
M60.872	Other myositis, left ankle and foot
M60.879	Other myositis, unspecified ankle and foot
M60.88	Other myositis, other site
M60.89	Other myositis, multiple sites
M60.9	Myositis, unspecified
M79.10	Myalgia, unspecified site
M79.11	Myalgia of mastication muscle
M79.12	Myalgia of auxiliary muscles, head and neck
M79.18	Myalgia, other site
O26.40	Herpes gestationis, unspecified trimester
O26.41	Herpes gestationis, first trimester
O26.42	Herpes gestationis, second trimester
O26.43	Herpes gestationis, third trimester
O36.8210	Fetal anemia and thrombocytopenia, first trimester, not applicable or unspecified
O36.8211	Fetal anemia and thrombocytopenia, first trimester, fetus 1
O36.8212	Fetal anemia and thrombocytopenia, first trimester, fetus 2
O36.8213	Fetal anemia and thrombocytopenia, first trimester, fetus 3
O36.8214	Fetal anemia and thrombocytopenia, first trimester, fetus 4
O36.8215	Fetal anemia and thrombocytopenia, first trimester, fetus 5
O36.8219	Fetal anemia and thrombocytopenia, first trimester, other fetus
O36.8220	Fetal anemia and thrombocytopenia, second trimester, not applicable or unspecified
O36.8221	Fetal anemia and thrombocytopenia, second trimester, fetus 1
O36.8222	Fetal anemia and thrombocytopenia, second trimester, fetus 2
O36.8223	Fetal anemia and thrombocytopenia, second trimester, fetus 3
O36.8224	Fetal anemia and thrombocytopenia, second trimester, fetus 4
O36.8225	Fetal anemia and thrombocytopenia, second trimester, fetus 5
O36.8229	Fetal anemia and thrombocytopenia, second trimester, other fetus
O36.8230	Fetal anemia and thrombocytopenia, third trimester, not applicable or unspecified

ICD-10	ICD-10 Description
O36.8231	Fetal anemia and thrombocytopenia, third trimester, fetus 1
O36.8232	Fetal anemia and thrombocytopenia, third trimester, fetus 2
O36.8233	Fetal anemia and thrombocytopenia, third trimester, fetus 3
O36.8234	Fetal anemia and thrombocytopenia, third trimester, fetus 4
O36.8235	Fetal anemia and thrombocytopenia, third trimester, fetus 5
O36.8239	Fetal anemia and thrombocytopenia, third trimester, other fetus
O36.8290	Fetal anemia and thrombocytopenia, unspecified trimester, not applicable or unspecified
O36.8291	Fetal anemia and thrombocytopenia, unspecified trimester, fetus 1
O36.8292	Fetal anemia and thrombocytopenia, unspecified trimester, fetus 2
O36.8293	Fetal anemia and thrombocytopenia, unspecified trimester, fetus 3
O36.8294	Fetal anemia and thrombocytopenia, unspecified trimester, fetus 4
O36.8295	Fetal anemia and thrombocytopenia, unspecified trimester, fetus 5
O36.8299	Fetal anemia and thrombocytopenia, unspecified trimester, other fetus
P61.0	Transient neonatal thrombocytopenia
T80.82XA	Complication of immune effector cellular therapy, initial encounter
T80.82XS	Complication of immune effector cellular therapy, sequela
T80.89XA	Other complications following infusion, transfusion and therapeutic injection, initial encounter
T80.89XS	Other complications following infusion, transfusion and therapeutic injection, sequela
T86.00	Unspecified complication of bone marrow transplant
T86.01	Bone marrow transplant rejection
T86.02	Bone marrow transplant failure
T86.03	Bone marrow transplant infection
T86.09	Other complications of bone marrow transplant
T86.10	Unspecified complication of kidney transplant
T86.11	Kidney transplant rejection
T86.12	Kidney transplant failure
T86.13	Kidney transplant infection
T86.19	Other complication of kidney transplant
T86.20	Unspecified complication of heart transplant
T86.21	Heart transplant rejection
T86.22	Heart transplant failure
T86.23	Heart transplant infection
T86.290	Cardiac allograft vasculopathy
T86.298	Other complications of heart transplant

ICD-10	ICD-10 Description
T86.30	Unspecified complication of heart-lung transplant
T86.31	Heart-lung transplant rejection
T86.32	Heart-lung transplant failure
T86.33	Heart-lung transplant infection
T86.39	Other complications of heart-lung transplant
T86.40	Unspecified complication of liver transplant
T86.41	Liver transplant rejection
T86.42	Liver transplant failure
T86.43	Liver transplant infection
T86.49	Other complications of liver transplant
T86.810	Lung transplant rejection
T86.811	Lung transplant failure
T86.812	Lung transplant infection
T86.818	Other complications of lung transplant
T86.819	Unspecified complication of lung transplant
T86.890	Other transplanted tissue rejection
T86.891	Other transplanted tissue failure
T86.892	Other transplanted tissue infection
T86.898	Other complications of other transplanted tissue
T86.899	Unspecified complication of other transplanted tissue
Z48.21	Encounter for aftercare following heart transplant
Z48.22	Encounter for aftercare following kidney transplant
Z48.23	Encounter for aftercare following liver transplant
Z48.24	Encounter for aftercare following lung transplant
Z48.280	Encounter for aftercare following heart-lung transplant
Z48.290	Encounter for aftercare following bone marrow transplant
Z94.0	Kidney transplant status
Z94.1	Heart transplant status
Z94.2	Lung transplant status
Z94.3	Heart and lungs transplant status
Z94.4	Liver transplant status
Z94.81	Bone marrow transplant status
Z94.83	Pancreas transplant status
Z94.84	Stem cells transplant status

**G61.81 is not payable when associated with diabetes mellitus, dysproteinemias, renal failure, or malnutrition*

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

The preceding information is intended for non-Medicare coverage determinations. Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications, including any preceding information, may be applied at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes		
Jurisdiction	**NCD/LCA/LCD Document (s)**	**Contractor**
E	A57187, A54660, A54641	Noridian Healthcare Solutions, LLC
F	A54643, A57194, A54662	Noridian Healthcare Solutions, LLC
H, L	A56786	Novitas Solutions, Inc.
J, M	A56718	Palmetto GBA
N	A57778	First Coast Service Options, Inc.
5, 8	A57554	Wisconsin Physicians Service Insurance Corporation
6, K	A59105	National Government Services, Inc. (NGS)
15	A56779, A57160	CGS Administrators, LLC
ALL	250.3	ALL

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corporation (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corporation (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC