

Medical Policy Bulletin

Title:

Immune Globulin Intravenous (IVIG), Subcutaneous (SCIG)

Policy #:

MA08.009z

The Company makes decisions on coverage based on the Centers for Medicare and Medicaid Services (CMS) regulations and guidance, benefit plan documents and contracts, and the member's medical history and condition. If CMS does not have a position addressing a service, the Company makes decisions based on Company Policy Bulletins. Benefits may vary based on contract, and individual member benefits must be verified. The Company determines medical necessity only if the benefit exists and no contract exclusions are applicable. Although the Medicare Advantage Policy Bulletin is consistent with Medicare's regulations and guidance, the Company's payment methodology may differ from Medicare.

When services can be administered in various settings, the Company reserves the right to reimburse only those services that are furnished in the most appropriate and cost-effective setting that is appropriate to the member's medical needs and condition. This decision is based on the member's current medical condition and any required monitoring or additional services that may coincide with the delivery of this service.

This Policy Bulletin document describes the status of CMS coverage, medical terminology, and/or benefit plan documents and contracts at the time the document was developed. This Policy Bulletin will be reviewed regularly and be updated as Medicare changes their regulations and guidance, scientific and medical literature becomes available, and/or the benefit plan documents and/or contracts are changed.

Policy

Coverage is subject to the terms, conditions, and limitations of the member's Evidence of Coverage.

The Company reserves the right to reimburse only those services that are furnished in the most appropriate and cost-effective setting that is appropriate to the member's medical needs and condition.

INDEX OF MEDICALLY NECESSARY INDICATIONS

This policy addresses numerous medically necessary indications for the use of Intravenous Immune Globulin (IVIG) and Subcutaneous Immune Globulin (SCIG) (listed in order of appearance within the Policy section). Please see below for the specific medical necessity criteria. (NOTE: Other sections such as Experimental/Investigational and Noncovered sections below must also be reviewed).

SUBCUTANEOUS IMMUNE GLOBULIN (SCIG)

Neurological and Musculoskeletal Disorders

- Chronic Inflammatory Demyelinating Polyneuritis (CIDP)

Primary Humoral Immunodeficiencies

INTRAVENOUS IMMUNE GLOBULIN (IVIG)

Dermatologic

Autoimmune Mucocutaneous Blistering Diseases
Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis
Scleromyxedema

Hematologic

Idiopathic Thrombocytopenic Purpura (ITP)
Autoimmune Hemolytic Anemia
Evans Syndrome
Systemic Capillary Leak Syndrome (SCLS)

Immunodeficiency Syndromes, Primary and Secondary

Acquired von Willebrand disease

Antibody-Mediated Rejection (AMR)
 Chimeric Antigen Receptor (CAR) T-Cell–Related Toxicities
 Chronic Lymphocytic Leukemia (CLL)
 Hematopoietic Stem Cell Transplant (HSCT)
 Human Immunodeficiency Virus (HIV) Infection
 Immune Checkpoint Inhibitor–Related Toxicities
 Lymphomas utilizing B-cell–depleting therapies
 Kawasaki disease
 Multiple Myeloma
 Preterm and/or low-birth-weight neonates as prevention for serious infections with severe hypogammaglobulinemia
 Primary Humoral Immunodeficiencies
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Neurological and Musculoskeletal Disorders

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 CIDP
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 Immune-Mediated Necrotizing Myopathy
 Inclusion Body Myositis
 Lambert-Eaton Myasthenic Syndrome
 Multifocal Motor Neuropathy
 Multiple Sclerosis (MS)
 Myasthenia Gravis Syndrome
 Myasthenic Crisis
 Neuromyelitis optica (NMO) (Devic Syndrome)
 Overlap Syndrome with Myositis including Anti-Synthetase Syndrome
 Polymyositis
 Stiff-Person Syndrome
 Susac Syndrome
 Systemic Lupus Erythematosus (SLE)

MEDICALLY NECESSARY

SUBCUTANEOUS IMMUNE GLOBULIN (SCIG)

Use of subcutaneous immune globulin (SCIG) (Cutaquig, Cuvitru, Hizentra, HyQvia, Xembify) or subcutaneous administration of certain intravenous immune globulin (IVIG) (e.g., Gammagard Liquid, Gammaked, Gamunex-C) therapy is considered medically necessary and, therefore, covered for the following indications when the dosing and frequency requirements listed in Attachment B and the following criteria are met:

Neurological and Musculoskeletal Disorders

Chronic Inflammatory Demyelinating Polyneuritis (CIDP)

SCIG is considered medically necessary and, therefore, covered for CIDP when the individual has a documented diagnosis of CIDP that has responded to IVIG treatment.

Primary Humoral Immunodeficiencies

SCIG is considered medically necessary and, therefore, covered when used as a replacement therapy in individuals with primary immunodeficiencies, in whom severe impairment of antibody capacity is present in the following conditions*:

- Activated Phosphoinositide 3-kinase Delta Syndrome (APDS)
- Antibody deficiency with near-normal immunoglobulins or with hyperimmunoglobulinemia
- Cerebellar ataxia with defective DNA repair
- Combined immunodeficiencies
- Common variable immunodeficiency
- Common variable immunodeficiency with autoantibodies to B- or T-cells

- Common variable immunodeficiency with predominant abnormalities of B-cell numbers and function
- Common variable immunodeficiency with predominant immunoregulatory T-cell disorders
- Congenital agammaglobulinemia
- DiGeorge syndrome
- Hereditary hypogammaglobulinemia
- Hyperimmunoglobulin E [IgE] syndrome
- Immunodeficiency with increased immunoglobulin M (IgM)
- Major histocompatibility complex class I or II deficiency
- Nezelof Syndrome
- Nonfamilial hypogammaglobulinemia
- PNP deficiency
- Selective deficiency of immunoglobulin A (IgA)
- Selective deficiency of IgM
- Selective deficiency of IgG subclasses
- Severe combined immunodeficiencies (SCIDs)
- SCID due to adenosine deaminase deficiency
- Transient hypogammaglobulinemia of infancy
- Wiskott-Aldrich syndrome
- X-linked immunodeficiency with hyper-IgM

*Coverage for primary immunodeficiencies not included in this list may be reviewed for coverage through applicable Part D benefits. Individual benefits must be verified.

INTRAVENOUS IMMUNE GLOBULIN (IVIG)

Use of IVIG therapy (e.g., Alyglo, Asceniv, Bivigam, Flebogamma, Gammagard Liquid, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Panzyga, Privigen) is considered medically necessary and, therefore, covered for the following indications when the dosing and frequency requirements listed in Attachment B and the following criteria are met:

Dermatologic

Autoimmune Mucocutaneous Blistering Diseases

IVIG is considered medically necessary and, therefore, covered for the treatment of any of the following biopsy-proven conditions (i.e., pemphigus vulgaris, pemphigus foliaceus, bullous pemphigoid, mucous membrane pemphigoid [a.k.a. cicatricial pemphigoid], epidermolysis bullosa acquisita), and meets at least one of the following criteria:

- Failed or intolerant of conventional therapy
- Conventional therapy is otherwise contraindicated
- Rapidly progressive disease in which a clinical response could not be effected quickly enough using conventional agents. In such situations, IVIG therapy would be given along with conventional treatment(s), and the IVIG would be used only until the conventional therapy could take effect.

In addition, IVIG for the treatment of autoimmune mucocutaneous blistering diseases must be used only for short-term therapy and not as a maintenance therapy

Stevens-Johnson Syndrome or Toxic Epidermal Necrolysis

IVIG is considered medically necessary and, therefore, covered for the one-time treatment of Stevens-Johnson syndrome or toxic epidermal necrolysis.

Scleromyxedema

IVIG is considered medically necessary and, therefore, covered for the treatment of severe scleromyxedema.

Hematologic

Idiopathic Thrombocytopenic Purpura (ITP)

For acute ITP, IVIG is considered medically necessary and, therefore, covered for the following indications:

- Management of acute bleeding due to severe thrombocytopenia (platelet counts usually <30,000/uL)
- To increase platelet counts prior to invasive surgical procedures, e.g., splenectomy
- Severe thrombocytopenia (platelet counts less than 20,000/uL) considered to be at risk for intracerebral hemorrhage
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Chronic refractory ITP is considered medically necessary and, therefore, covered for individuals meeting **all** of the following conditions:

- Prior treatment with corticosteroids and splenectomy, except when contraindicated
- Duration of illness >6 months
- No concurrent illness/disease explaining thrombocytopenia
- Platelet counts persistently at or below 20,000/uL

Autoimmune Hemolytic Anemia (AIHA)

IVIG is considered medically necessary and, therefore, covered for individuals with AIHA when other treatment approaches have failed.

Evans syndrome

IVIG is considered medically necessary and, therefore, covered for individuals with Evans syndrome, with or without corticosteroids.

Systemic Capillary Leak Syndrome (SCLS)

IVIG is considered medically necessary and, therefore, covered for individuals with systemic capillary leak syndrome (SCLS).

Immunodeficiency Syndromes, Primary and Secondary

Acquired von Willebrand disease

IVIG is considered medically necessary and, therefore, covered for an individual with acquired von Willebrand disease associated with monoclonal gammopathy of undetermined significance of the IgG Class (IgG-MGUS) or antibody-mediated acquired von Willebrand disease

Antibody-Mediated Rejection (AMR)

IVIG is considered medically necessary and, therefore, covered for the treatment of antibody-mediated solid organ transplant rejection in combination with rituximab and plasma exchange (PE).

Chimeric Antigen Receptor (CAR) T-Cell–Related Toxicities

IVIG is considered medically necessary and, therefore, covered after CAR T-cell (anti-CD19) therapy (e.g., axicabtagene ciloleucel [Yescarta], brexucabtagene autoleucel [Tecartus], lisocabtagene maraleucel [Breyanzi], tisagenlecleucel [Kymriah®]) for one of the following:

- Hypogammaglobulinemia when the individual has a serum IgG level <400–600 mg/dL and serious or recurrent infections (particularly bacterial) until serum IgG levels normalize and infections resolve
- Management of Grade 4 cytokine release syndrome that is refractory to high-dose corticosteroids and anti-IL-6 therapy

Chronic Lymphocytic Leukemia (CLL)

IVIG is considered medically necessary and, therefore, covered when used to prevent recurrent bacterial infections in individuals with B-cell CLL who meet **all** of the following criteria:

- Documented diagnosis of CLL
- An IgG level <600 mg/dL
- Recent history of serious bacterial infection(s) requiring either oral or parenteral antibiotic therapy

Hematopoietic Stem Cell Transplant (HSCT)

- IVIG is considered medically necessary and, therefore, covered for recipients of HSCT with SCID or other primary immunodeficiencies who are functionally agammaglobulinemic because of weak B-cell engraftment.
- IVIG is considered medically necessary and, therefore, covered for recipients of allogeneic HSCT with chronic graft-versus-host disease (GVHD), recurring bacterial infections, and subprotective antibody levels following immunization against diphtheria, tetanus, or pneumococcal infection.

Human Immunodeficiency Virus (HIV) Infection

IVIG is considered medically necessary and, therefore, covered to reduce significant bacterial infection for individuals infected with HIV who meet all of the following conditions:

- Individual is younger than 14 years of age
- Evidence of either qualitative or quantitative humoral immunologic defects
- Current bacterial infections, despite appropriate antimicrobial prophylaxis

Immune Checkpoint Inhibitor–Related Toxicities

IVIG is considered medically necessary and, therefore, covered for the management of the following immunotherapy-related toxicities (Note: examples of immune checkpoint inhibitors include, but are not limited to, ipilimumab [Yervoy], PD-1/PD-L1 inhibitors [e.g., atezolizumab [Tecentriq], nivolumab [Opdivo], pembrolizumab [Keytruda])

- Myocarditis if no improvement within 24 to 48 hours of starting pulse-dose methylprednisolone
- Adjunct to rituximab for severe (Grade 3) or life-threatening (Grade 4) bullous dermatitis
- Stevens-Johnson syndrome, or toxic epidermal necrolysis
- Moderate or 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
- Severe (Grade 3-4) myasthenia gravis
- Moderate (Grade 2) or severe (Grade 3-4) Guillain-Barré syndrome or severe (Grade 3-4) peripheral neuropathy in combination with pulse-dose methylprednisolone
- Encephalitis in combination with pulse-dose methylprednisolone if severe or progressing symptoms, or if oligoclonal bands present (strongly consider if progressing over 24 hours)
- Demyelinating disease (optic neuritis, transverse myelitis, acute demyelinating encephalomyelitis)
- Moderate (Grade 3) pneumonitis if no improvement after 48 to 72 hours of corticosteroids or severe (Grades 3-4) pneumonitis if no improvement after 48 hours of methylprednisolone
- Grade 4 cytokine release syndrome that is refractory to high-dose corticosteroids and anti-IL-6 therapy

Lymphomas utilizing B-cell–depleting therapies

IVIG is considered medically necessary and, therefore, covered following treatment of lymphoma utilizing B-cell–depleting therapies for recurrent infections with hypogammaglobulinemia and subprotective antibody levels following immunization against diphtheria, tetanus, or pneumococcal infection.

Kawasaki disease

IVIG is considered medically necessary and, therefore, covered for an individual with a documented diagnosis of Kawasaki disease.

Multiple Myeloma

IVIG is considered medically necessary and, therefore, covered for an individual with multiple myeloma for recurrent infections with hypogammaglobulinemia and subprotective antibody levels following immunization against diphtheria, tetanus, or pneumococcal infection.

Preterm and/or low-birth-weight neonates as prevention for serious infections

IVIG is considered medically necessary and, therefore, covered for preterm and/or low-birth-weight neonates as prevention for serious infections (including sepsis, urinary tract infection, soft-tissue infections or cellulitis) when

severe hypogammaglobulinemia (IgG $\leq$ 400 mg/dL) is present.

Primary Humoral Immunodeficiencies

IVIG is considered medically necessary and, therefore, covered when used as a replacement therapy in individuals with primary immunodeficiencies in whom severe impairment of antibody capacity is present in the following conditions*:

- APDS
- Antibody deficiency with near-normal immunoglobulins or with hyperimmunoglobulinemia
- Cerebellar ataxia with defective DNA repair
- Combined immunodeficiencies
- Common variable immunodeficiency
- Common variable immunodeficiency with autoantibodies to B or T cells
- Common variable immunodeficiency with predominant abnormalities of B-cell numbers and function
- Common variable immunodeficiency with predominant immunoregulatory T-cell disorders
- Congenital agammaglobulinemia
- DiGeorge syndrome
- Hereditary hypogammaglobulinemia
- IgE syndrome
- Immunodeficiency with increased IgM
- Major histocompatibility complex class I or II deficiency
- Nezelof syndrome
- Nonfamilial hypogammaglobulinemia
- Purine nucleoside phosphorylase [PNP] deficiency
- Selective deficiency of IgA
- Selective deficiency of IgM
- Selective deficiency of IgG subclasses
- SCIDs
- SCID due to adenosine deaminase deficiency
- Transient hypogammaglobulinemia of infancy
- Wiskott-Aldrich syndrome
- X-linked immunodeficiency with hyper-IgM

*Coverage for primary immunodeficiencies not included in this list may be reviewed for coverage through applicable Part D benefits. Individual benefits must be verified.

Solid Organ Transplant

IVIG is considered medically necessary and, therefore, covered as human leukocyte antigen (HLA) and ABO desensitization protocols for the prevention of acute humoral rejection in renal transplantation.

IVIG is considered medically necessary and, therefore, covered for the treatment of hypogammaglobulinemia in solid organ transplants.

Thyroid eye disease, also referred to as Graves' disease

IVIG is considered medically necessary and, therefore, covered for the treatment of thyroid eye disease, also referred to as Graves' disease, in individuals who have failed or have contraindications to treatment with teprotumumab (Teppezza).

Neurological and Musculoskeletal Disorders

Autoimmune Encephalitis

IVIG is considered medically necessary and, therefore, covered for the treatment of autoimmune encephalitis, once infection is ruled out, as an alternative in individuals who fail to respond or do not tolerate other treatments (e.g., high-dose corticosteroids, plasma exchange).

Chronic Inflammatory Demyelinating Polyneuritis (CIPD)

IVIG is considered medically necessary and, therefore, covered for CIPD when the individual has a documented diagnosis of CIPD.

Dermatomyositis

IVIG is considered medically necessary and, therefore, covered for an individual with a documented diagnosis of dermatomyositis.

Guillain-Barré Syndrome

IVIG is considered medically necessary and, therefore, covered for Guillain-Barré syndrome in adults.

Immune-Mediated Necrotizing Myopathy

IVIG is considered medically necessary and, therefore, covered for immune-mediated necrotizing myopathy resistant to treatment with glucocorticosteroids and immunosuppressants.

Inclusion Body Myositis

IVIG is considered medically necessary and, therefore, covered for severe forms of inclusion body myositis with dysphagia when individuals are otherwise treatment-resistant.

Lambert-Eaton Myasthenic Syndrome (LEMS)

IVIG is considered medically necessary and, therefore, covered in individuals with LEMS who fail to respond or do not tolerate other treatments.

Multifocal Motor Neuropathy

IVIG is considered medically necessary and, therefore, covered as initial therapy in individuals who have progressive, symptomatic, multifocal motor neuropathy that has been diagnosed on the basis of electrophysiologic findings that rule out other possible conditions that may not respond to this treatment.

Multiple Sclerosis (MS)

IVIG is considered medically necessary and, therefore, covered in individuals with relapsing-remitting MS.

Myasthenia Gravis Syndrome

IVIG is considered medically necessary and, therefore, covered for moderate to severe myasthenia gravis.

Myasthenic Crisis

IVIG is considered medically necessary and, therefore, covered for myasthenic crisis (i.e., an acute episode of respiratory muscle weakness) when there is a contraindication to plasma exchange.

Neuromyelitis optica (NMO) (Devic syndrome)

IVIG is considered medically necessary and, therefore, covered for individuals with NMO who have severe relapses not responding to corticosteroids and who are not candidates for plasma exchange.

Overlap Syndrome with Myositis including Anti-Synthetase Syndrome

IVIG is considered medically necessary and, therefore, covered for individuals with overlap syndrome with myositis including anti-synthetase syndrome resistant to treatment with glucocorticosteroids and immunosuppressants.

Polymyositis

IVIG is considered medically necessary and, therefore, covered for the treatment of severe forms of polymyositis resistant to treatment with glucocorticosteroids and immunosuppressants.

Stiff-Person Syndrome (Stiff-man Syndrome)

IVIG is considered medically necessary and, therefore, covered for individuals with Stiff-Person syndrome.

Susac Syndrome

IVIG is considered medically necessary and, therefore, covered for individuals with Susac syndrome in combination with high-dose intravenous corticosteroids.

Systemic Lupus Erythematosus (SLE)

IVIG is considered medically necessary and, therefore, covered for individuals with severe active systemic lupus erythematosus (SLE) for whom other interventions have been unsuccessful, have become intolerable, or are contraindicated.

EXPERIMENTAL/INVESTIGATIONAL (SCIG, IVIG)

All other uses for SCIG, IVIG are considered experimental/investigational and, therefore, not covered unless the indication is supported as an accepted off-label use, as defined in the Company medical policy on off-label coverage for prescription drugs and biologics.

NONCOVERED (IVIG)

IVIG is considered not medically necessary and, therefore, noncovered for the following indications:

- Routine use in the immediate peritransplantation period for the prevention of infection or GVHD following marrow or peripheral blood allogeneic transplantation
- Acute GVHD with hematopoietic stem cell transplantation (HSCT) in the immediate posttransplantation phase
- HSCT in the immediate posttransplantation phase with a history of sinusoidal obstructive syndrome
- Cord blood stem cell transplantation for children or adults
- Polyneuropathy associated with IgM monoclonal gammopathy
- Idiopathic neuropathies
- Brachial plexopathy
- Adrenoleukodystrophy
- Amyotrophic lateral sclerosis
- Critical illness polyneuropathy
- POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes)

NOT ELIGIBLE FOR REIMBURSEMENT

Carimune NF and Vivaglobin are no longer manufactured and have been withdrawn from the market; therefore, they are not eligible for reimbursement.

DOSING AND FREQUENCY REQUIREMENTS

The Company reserves the right to modify the Dosing and Frequency Requirements listed in this Policy to ensure consistency with the most recently published recommendations for the use of IVIG and SCIG. Changes to these guidelines are based on a consensus of information obtained from resources such as, but not limited to, the US Food and Drug Administration (FDA), drug manufacturer's guidelines, Company-recognized authoritative pharmacology compendia, or published peer-reviewed clinical research. The professional provider must supply supporting documentation (i.e., published peer-reviewed literature) in order to request coverage for an amount of IVIG and SCIG outside of the Dosing and Frequency Requirements listed in this policy. For a list of Company-recognized pharmacology compendia and criteria for peer-reviewed clinical research, view the policy on off-label coverage for prescription drugs and biologics.

Accurate member information is necessary for the Company to approve the requested dose and frequency of these

drugs. If the member's dose, frequency, or regimen changes (based on factors such as changes in member weight or incomplete therapeutic response), the provider **must** submit those changes to the Company for a new approval based on those changes as part of the precertification process. The Company reserves the right to conduct postpayment review and audit procedures for any claims submitted for IVIG and SCIG.

Refer to Attachment B for dosing and frequency requirements for IVIG and SCIG.

REQUIRED DOCUMENTATION

When coverage of IVIG and SCIG is requested outside of the Dosing and Frequency Guidelines listed in this policy, the prescribing professional provider must supply documentation (i.e., published peer-reviewed literature) to the Company that supports this request.

The Company may conduct reviews and audits of services to our members regardless of the participation status of the provider. Medical record documentation must be maintained on file to reflect the medical necessity of the care and services provided. These medical records may include but are not limited to: records from the professional provider's office, hospital, nursing home, home health agencies, therapies, and test reports.

STANDARD WRITTEN ORDER REQUIREMENTS

Before submitting a claim to the Company, the supplier must have on file a timely, appropriate, and complete order for each item billed that is signed and dated by the professional provider who is treating the member. Requesting a provider to sign a retrospective order at the time of an audit or after an audit for submission as an original order, reorder, or updated order will not satisfy the requirement to maintain a timely professional provider order on file.

PROOF OF DELIVERY

Medical record documentation must include a contemporaneously prepared delivery confirmation or member's receipt of supplies and equipment. The medical record documentation must include a copy of delivery confirmation if delivered by a commercial carrier and a signed copy of delivery confirmation by member/caregiver if delivered by the durable medical equipment (DME) supplier/provider. All documentation is to be prepared contemporaneous with delivery and be available to the Company upon request.

CONSUMABLE SUPPLIES (WHEN APPLICABLE)

The DME supplier must monitor the quantity of accessories and supplies an individual is actually using. Contacting the individual regarding replenishment of supplies should not be done earlier than approximately 30 days prior to the delivery/shipping date. Dated documentation of this contact with the individual is required in the individual's medical record. Delivery of the supplies should not be done earlier than approximately 10 days before the individual would exhaust their on-hand supply.

If required documentation is not available on file to support a claim at the time of an audit or record request, the DME supplier may be required to reimburse the Company for overpayments.

The maximum number of supplies that can be dispensed at one time is no more than a 1-month supply.

Guidelines

This policy is consistent with Medicare's coverage determination. The Company's payment methodology may differ from Medicare.

BLACK BOX WARNINGS

Refer to the specific manufacturer's prescribing information for any applicable Black Box Warnings.

BENEFIT APPLICATION

Subject to the terms and conditions of the applicable Evidence of Coverage, subcutaneous immune globulin (SCIG) and intravenous immune globulin (IVIG) are covered under the medical benefits of the Company's Medicare Advantage products when the medical necessity criteria, dosing and frequency requirements, and precertification/preapproval requirements listed in this medical policy are met.

Certain drugs may be available through either the member's medical benefit (Part B benefit) or pharmacy benefit

(Part D benefit), depending upon how the drug is prescribed and dispensed or administered. This medical policy only addresses instances when IVIG and SCIG may be covered under a member's medical benefit. It does not address instances when IVIG and SCIG may be covered under a member's pharmacy benefit. For individuals with primary immunodeficiency, IVIG and SCIG in the home setting may be covered under the medical benefit (Part B benefit). All other clinical indications in the home setting may be covered under the pharmacy benefit (Part D benefit). Refer to the policy entitled "Medicare Part B vs. Part D Crossover Drugs - MA08.007" for appropriate benefit application.

US FOOD AND DRUG ADMINISTRATION (FDA) STATUS

The FDA has approved numerous formulations of SCIG and IVIG.

Description

Intravenous immune globulin (IVIG) is a blood product prepared from the pooled plasma of donors. It has been used to treat a variety of autoimmune diseases, including mucocutaneous blistering diseases. It has fewer side effects than steroids or immunosuppressive agents.

IVIG can replace missing antibodies and decrease infection in primary immune deficiency and chronic lymphocytic leukemia, increase platelets in idiopathic thrombocytopenic purpura, prevent complications in Kawasaki disease, and possibly decrease morbidity in some other conditions.

IVIG is the preferred treatment method for patients who require an immediate increase in intravascular immunoglobulin antibody levels and are unable to produce sufficient amounts of immunoglobulin G (IgG) antibodies. The therapeutic effect of IVIG is immediate, well-tolerated, and less likely to produce side effects if infused at the properly indicated rate(s). Sensitivity to these reactions is usually related to the infusion rate. Caution should be exercised in the administration of IVIG; reactions may cause a rapid fall in blood pressure and clinical anaphylaxis.

The US Food and Drug Administration (FDA) has approved the subcutaneous infusion preparation of an immune globulin product (SCIG) for the prevention of serious infections in those with primary immunodeficiency disease (PIDD). SCIG is given under the skin using an infusion pump, usually on a weekly basis, in CIDP and PIDD. SCIG may be an option for individuals who have received IVIG and wish to transition to the subcutaneous infusion route. Subcutaneous infusion of immune globulin allows for an alternative route of administration in those who may have problems with chronic IV administration, and enables individuals to self-administer the product at home.

Note: There are a few formulations of IVIG that may be administered via subcutaneous infusion for the indication of PIDD (e.g., Gammagard Liquid, Gamunex-C, Gammaked).

OFF-LABEL INDICATIONS

There may be additional indications contained in the Policy section of this document due to evaluation of criteria highlighted in the Company's off-label policy, and/or review of clinical guidelines issued by leading professional organizations and government entities.

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Coding

Inclusion of a code in this table does not imply reimbursement. Eligibility, benefits, limitations, exclusions, precertification/referral requirements, provider contracts, and Company policies apply.

The codes listed below are updated on a regular basis, in accordance with nationally accepted coding guidelines. Therefore, this policy applies to any and all future applicable coding changes, revisions, or updates.

In order to ensure optimal reimbursement, all health care services, devices, and pharmaceuticals should be reported using the billing codes and modifiers that most accurately represent the services rendered, unless otherwise directed by the Company.

The Coding Table lists any CPT, ICD-10, and HCPCS billing codes related only to the specific policy in which they appear.

CPT Procedure Code Number(s)

N/A

ICD - 10 Procedure Code Number(s)

N/A

ICD - 10 Diagnosis Code Number(s)

MEDICALLY NECESSARY

SUBCUTANEOUS IMMUNE GLOBULIN (SCIG)

C9399, J1555, J1558, J1559, J1561, J1569, J1575, AND J3590 are medically necessary when reported with the following diagnosis codes:

THIS IS NOT AN ALL INCLUSIVE LIST

- D80.0 Hereditary hypogammaglobulinemia
- D80.2 Selective deficiency of immunoglobulin A [IgA]
- D80.3 Selective deficiency of immunoglobulin G [IgG] subclasses
- D80.4 Selective deficiency of immunoglobulin M [IgM]
- D80.5 Immunodeficiency with increased immunoglobulin M [IgM]
- D80.6 Antibody deficiency with near-normal immunoglobulins or with hyperimmunoglobulinemia
- D80.8 Other immunodeficiencies with predominantly antibody defects
- 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
- D81.31 Severe combined immunodeficiency due to adenosine deaminase deficiency
- D81.4 Nezelof's syndrome
- D81.5 Purine nucleoside phosphorylase [PNP] deficiency
- D81.6 Major histocompatibility complex class I deficiency
- D81.7 Major histocompatibility complex class II deficiency
- D81.82 Activated Phosphoinositide 3-kinase Delta Syndrome (APDS)
- D81.89 Other combined immunodeficiencies
- D81.9 Combined immunodeficiency, unspecified
- D82.0 Wiskott-Aldrich syndrome
- D82.1 Di George's syndrome
- D82.4 Hyperimmunoglobulin E [IgE] syndrome
- D83.0 Common variable immunodeficiency with predominant abnormalities of B-cell numbers and function
- D83.1 Common variable immunodeficiency with predominant immunoregulatory T-cell disorders
- D83.2 Common variable immunodeficiency with autoantibodies to B- or T-cells
- D83.8 Other common variable immunodeficiencies
- D83.9 Common variable immunodeficiency, unspecified
- G11.3 Cerebellar ataxia with defective DNA repair
- G61.81 Chronic inflammatory demyelinating polyneuritis

INTRAVENOUS IMMUNE GLOBULIN (IVIG)

J1459, J1556, J1557, J1561, J1566, J1568, J1569, AND J1599 are medically necessary when reported with the following diagnosis codes:

THIS IS NOT AN ALL INCLUSIVE LIST

REPORT THE MOST APPROPRIATE DIAGNOSIS CODE FOR TOXICITIES RELATED TO THE MANAGEMENT OF IMMUNE CHECKPOINT INHIBITOR TOXICITIES.

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

C81.00 Nodular lymphocyte predominant Hodgkin lymphoma, unspecified site

C81.01 Nodular lymphocyte predominant Hodgkin lymphoma, lymph nodes of head, face, and neck

C81.02 Nodular lymphocyte predominant Hodgkin lymphoma, intrathoracic lymph nodes

C81.03 Nodular lymphocyte predominant Hodgkin lymphoma, intra-abdominal lymph nodes

C81.04 Nodular lymphocyte predominant Hodgkin lymphoma, lymph nodes of axilla and upper limb

C81.05 Nodular lymphocyte predominant Hodgkin lymphoma, lymph nodes of inguinal region and lower limb

C81.06 Nodular lymphocyte predominant Hodgkin lymphoma, intrapelvic lymph nodes

C81.07 Nodular lymphocyte predominant Hodgkin lymphoma, spleen

C81.08 Nodular lymphocyte predominant Hodgkin lymphoma, lymph nodes of multiple sites

C81.09 Nodular lymphocyte predominant Hodgkin lymphoma, extranodal and solid organ sites

C81.10 Nodular sclerosis Hodgkin lymphoma, unspecified site

C81.11 Nodular sclerosis Hodgkin lymphoma, lymph nodes of head, face and neck

C81.12 Nodular sclerosis Hodgkin lymphoma, intrathoracic lymph nodes

C81.13 Nodular sclerosis Hodgkin lymphoma, intra-abdominal lymph nodes

C81.14 Nodular sclerosis Hodgkin lymphoma, lymph nodes of axilla and upper limb

C81.15 Nodular sclerosis Hodgkin lymphoma, lymph nodes of inguinal region and lower limb

C81.16 Nodular sclerosis Hodgkin lymphoma, intrapelvic lymph nodes

C81.17 Nodular sclerosis Hodgkin lymphoma, spleen

C81.18 Nodular sclerosis Hodgkin lymphoma, lymph nodes of multiple sites

C81.19 Nodular sclerosis Hodgkin lymphoma, extranodal and solid organ sites

C81.20 Mixed cellularity Hodgkin lymphoma, unspecified site

C81.21 Mixed cellularity Hodgkin lymphoma, lymph nodes of head, face, and neck

C81.22 Mixed cellularity Hodgkin lymphoma, intrathoracic lymph nodes

C81.23 Mixed cellularity Hodgkin lymphoma, intra-abdominal lymph nodes

C81.24 Mixed cellularity Hodgkin lymphoma, lymph nodes of axilla and upper limb

C81.25 Mixed cellularity Hodgkin lymphoma, lymph nodes of inguinal region and lower limb

C81.26 Mixed cellularity Hodgkin lymphoma, intrapelvic lymph nodes

C81.27 Mixed cellularity Hodgkin lymphoma, spleen

C81.28 Mixed cellularity Hodgkin lymphoma, lymph nodes of multiple sites

C81.29 Mixed cellularity Hodgkin lymphoma, extranodal and solid organ sites

C81.30 Lymphocyte depleted Hodgkin lymphoma, unspecified site

C81.31 Lymphocyte depleted Hodgkin lymphoma, lymph nodes of head, face, and neck

C81.32 Lymphocyte depleted Hodgkin lymphoma, intrathoracic lymph nodes

C81.33 Lymphocyte depleted Hodgkin lymphoma, intra-abdominal lymph nodes

C81.34 Lymphocyte depleted Hodgkin lymphoma, lymph nodes of axilla and upper limb

C81.35 Lymphocyte depleted Hodgkin lymphoma, lymph nodes of inguinal region and lower limb

C81.36 Lymphocyte depleted Hodgkin lymphoma, intrapelvic lymph nodes

C81.37 Lymphocyte depleted Hodgkin lymphoma, spleen

C81.38 Lymphocyte depleted Hodgkin lymphoma, lymph nodes of multiple sites

C81.39 Lymphocyte depleted Hodgkin lymphoma, extranodal and solid organ sites

C81.40 Lymphocyte-rich Hodgkin lymphoma, unspecified site

C81.41 Lymphocyte-rich Hodgkin lymphoma, lymph nodes of head, face, and neck

C81.42 Lymphocyte-rich Hodgkin lymphoma, intrathoracic lymph nodes

C81.43 Lymphocyte-rich Hodgkin lymphoma, intra-abdominal lymph nodes

C81.44 Lymphocyte-rich Hodgkin lymphoma, lymph nodes of axilla and upper limb

C81.45 Lymphocyte-rich Hodgkin lymphoma, lymph nodes of inguinal region and lower limb

C81.46 Lymphocyte-rich Hodgkin lymphoma, intrapelvic lymph nodes

C81.47 Lymphocyte-rich Hodgkin lymphoma, spleen

C81.48 Lymphocyte-rich Hodgkin lymphoma, lymph nodes of multiple sites

C81.49 Lymphocyte-rich Hodgkin lymphoma, extranodal and solid organ sites

C81.70 Other Hodgkin lymphoma, unspecified site

C81.71 Other Hodgkin lymphoma, lymph nodes of head, face, and neck

C81.72 Other Hodgkin lymphoma, intrathoracic lymph nodes

C81.73 Other Hodgkin lymphoma, intra-abdominal lymph nodes

C81.74 Other Hodgkin lymphoma, lymph nodes of axilla and upper limb

C81.75 Other Hodgkin lymphoma, lymph nodes of inguinal region and lower limb

C81.76 Other Hodgkin lymphoma, intrapelvic lymph nodes

C81.77 Other Hodgkin lymphoma, spleen

C81.78 Other Hodgkin lymphoma, lymph nodes of multiple sites

C81.79 Other Hodgkin lymphoma, extranodal and solid organ sites

C81.90 Hodgkin lymphoma, unspecified, unspecified site

C81.91 Hodgkin lymphoma, unspecified, lymph nodes of head, face, and neck

C81.92 Hodgkin lymphoma, unspecified, intrathoracic lymph nodes

C81.93 Hodgkin lymphoma, unspecified, intra-abdominal lymph nodes

C81.94 Hodgkin lymphoma, unspecified, lymph nodes of axilla and upper limb

C81.95 Hodgkin lymphoma, unspecified, lymph nodes of inguinal region and lower limb

C81.96 Hodgkin lymphoma, unspecified, intrapelvic lymph nodes

C81.97 Hodgkin lymphoma, unspecified, spleen

C81.98 Hodgkin lymphoma, unspecified, lymph nodes of multiple sites

C81.99 Hodgkin lymphoma, unspecified, extranodal and solid organ sites

C82.00 Follicular lymphoma grade I, unspecified site

C82.01 Follicular lymphoma grade I, lymph nodes of head, face, and neck

C82.02 Follicular lymphoma grade I, intrathoracic lymph nodes

C82.03 Follicular lymphoma grade I, intra-abdominal lymph nodes

C82.04 Follicular lymphoma grade I, lymph nodes of axilla and upper limb

C82.05 Follicular lymphoma grade I, lymph nodes of inguinal region and lower limb

C82.06 Follicular lymphoma grade I, intrapelvic lymph nodes

C82.07 Follicular lymphoma grade I, spleen

C82.08 Follicular lymphoma grade I, lymph nodes of multiple sites

C82.09 Follicular lymphoma grade I, extranodal and solid organ sites

C82.10 Follicular lymphoma grade II, unspecified site

C82.11 Follicular lymphoma grade II, lymph nodes of head, face, and neck

C82.12 Follicular lymphoma grade II, intrathoracic lymph nodes

C82.13 Follicular lymphoma grade II, intra-abdominal lymph nodes

C82.14 Follicular lymphoma grade II, lymph nodes of axilla and upper limb

C82.15 Follicular lymphoma grade II, lymph nodes of inguinal region and lower limb

C82.16 Follicular lymphoma grade II, intrapelvic lymph nodes

C82.17 Follicular lymphoma grade II, spleen

C82.18 Follicular lymphoma grade II, lymph nodes of multiple sites

C82.19 Follicular lymphoma grade II, extranodal and solid organ sites

C82.20 Follicular lymphoma grade III, unspecified, unspecified site

C82.21 Follicular lymphoma grade III, unspecified, lymph nodes of head, face, and neck

C82.22 Follicular lymphoma grade III, unspecified, intrathoracic lymph nodes

C82.23 Follicular lymphoma grade III, unspecified, intra-abdominal lymph nodes

C82.24 Follicular lymphoma grade III, unspecified, lymph nodes of axilla and upper limb

C82.25 Follicular lymphoma grade III, unspecified, lymph nodes of inguinal region and lower limb

C82.26 Follicular lymphoma grade III, unspecified, intrapelvic lymph nodes

C82.27 Follicular lymphoma grade III, unspecified, spleen

C82.28 Follicular lymphoma grade III, unspecified, lymph nodes of multiple sites

C82.29 Follicular lymphoma grade III, unspecified, extranodal and solid organ sites

C82.30 Follicular lymphoma grade IIIa, unspecified site

C82.31 Follicular lymphoma grade IIIa, lymph nodes of head, face, and neck

C82.32 Follicular lymphoma grade IIIa, intrathoracic lymph nodes

C82.33 Follicular lymphoma grade IIIa, intra-abdominal lymph nodes

C82.34 Follicular lymphoma grade IIIa, lymph nodes of axilla and upper limb

C82.35 Follicular lymphoma grade IIIa, lymph nodes of inguinal region and lower limb

C82.36 Follicular lymphoma grade IIIa, intrapelvic lymph nodes

C82.37 Follicular lymphoma grade IIIa, spleen

C82.38 Follicular lymphoma grade IIIa, lymph nodes of multiple sites

C82.39 Follicular lymphoma grade IIIa, extranodal and solid organ sites

C82.40 Follicular lymphoma grade IIIb, unspecified site

C82.41 Follicular lymphoma grade IIIb, lymph nodes of head, face, and neck

C82.42 Follicular lymphoma grade IIIb, intrathoracic lymph nodes

C82.43 Follicular lymphoma grade IIIb, intra-abdominal lymph nodes

C82.44 Follicular lymphoma grade IIIb, lymph nodes of axilla and upper limb

C82.45 Follicular lymphoma grade IIIb, lymph nodes of inguinal region and lower limb

C82.46 Follicular lymphoma grade IIIb, intrapelvic lymph nodes

C82.47 Follicular lymphoma grade IIIb, spleen

C82.48 Follicular lymphoma grade IIIb, lymph nodes of multiple sites

C82.49 Follicular lymphoma grade IIIa, extranodal and solid organ sites

C82.50 Diffuse follicle center lymphoma, unspecified

C82.51 Diffuse follicle center lymphoma, lymph nodes of head, face, and neck

C82.52 Diffuse follicle center lymphoma, intrathoracic lymph nodes

C82.53 Diffuse follicle center lymphoma, intra-abdominal lymph nodes

C82.54 Diffuse follicle center lymphoma, lymph nodes of axilla and upper limb

C82.55 Diffuse follicle center lymphoma, lymph nodes of inguinal region and lower limb

C82.56 Diffuse follicle center lymphoma, intrapelvic lymph nodes

C82.57 Diffuse follicle center lymphoma, spleen

C82.58 Diffuse follicle center lymphoma, lymph nodes of multiple sites

C82.59 Diffuse follicle center lymphoma, extranodal and solid organ sites

C82.60 Cutaneous follicle center lymphoma, unspecified site

C82.61 Cutaneous follicle center lymphoma, lymph nodes of head, face, and neck

C82.62 Cutaneous follicle center lymphoma, intrathoracic lymph nodes

C82.63 Cutaneous follicle center lymphoma, intra-abdominal lymph nodes

C82.64 Cutaneous follicle center lymphoma, lymph nodes of axilla and upper limb

C82.65 Cutaneous follicle center lymphoma, lymph nodes of inguinal region and lower limb

C82.66 Cutaneous follicle center lymphoma, intrapelvic lymph nodes

C82.67 Cutaneous follicle center lymphoma, spleen

C82.68 Cutaneous follicle center lymphoma, lymph nodes of multiple sites

C82.69 Cutaneous follicle center lymphoma, extranodal and solid organ sites

C82.80 Other types of follicular lymphoma, unspecified site

C82.81 Other types of follicular lymphoma, lymph nodes of head, face, and neck

C82.82 Other types of follicular lymphoma, intrathoracic lymph nodes

C82.83 Other types of follicular lymphoma, intra-abdominal lymph nodes

C82.84 Other types of follicular lymphoma, lymph nodes of axilla and upper limb

C82.85 Other types of follicular lymphoma, lymph nodes of inguinal region and lower limb

C82.86 Other types of follicular lymphoma, intrapelvic lymph nodes

C82.87 Other types of follicular lymphoma, spleen

C82.88 Other types of follicular lymphoma, lymph nodes of multiple sites

C82.89 Other types of follicular lymphoma, extranodal and solid organ sites

C82.90 Follicular lymphoma, unspecified, unspecified site

C82.91 Follicular lymphoma, unspecified, lymph nodes of head, face, and neck

C82.92 Follicular lymphoma, unspecified, intrathoracic lymph nodes

C82.93 Follicular lymphoma, unspecified, intra-abdominal lymph nodes

C82.94 Follicular lymphoma, unspecified, lymph nodes of axilla and upper limb

C82.95 Follicular lymphoma, unspecified, lymph nodes of inguinal region and lower limb

C82.96 Follicular lymphoma, unspecified, intrapelvic lymph nodes

C82.97 Follicular lymphoma, unspecified, spleen

C82.98 Follicular lymphoma, unspecified, lymph nodes of multiple sites

C82.99 Follicular lymphoma, unspecified, extranodal and solid organ sites

C83.00 Small cell B-cell lymphoma, unspecified site

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

C83.10 Mantle cell lymphoma, unspecified site

C83.11 Mantle cell lymphoma, lymph nodes of head, face, and neck

C83.12 Mantle cell lymphoma, intrathoracic lymph nodes

C83.13 Mantle cell lymphoma, intra-abdominal lymph nodes

C83.14 Mantle cell lymphoma, lymph nodes of axilla and upper limb

C83.15 Mantle cell lymphoma, lymph nodes of inguinal region and lower limb

C83.16 Mantle cell lymphoma, intrapelvic lymph nodes

C83.17 Mantle cell lymphoma, spleen

C83.18 Mantle cell lymphoma, lymph nodes of multiple sites

C83.19 Mantle cell lymphoma, extranodal and solid organ sites

C83.30 Diffuse large B-cell lymphoma, unspecified site

C83.31 Diffuse large B-cell lymphoma, lymph nodes of head, face, and neck

C83.32 Diffuse large B-cell lymphoma, intrathoracic lymph nodes

C83.33 Diffuse large B-cell lymphoma, intra-abdominal lymph nodes

C83.34 Diffuse large B-cell lymphoma, lymph nodes of axilla and upper limb

C83.35 Diffuse large B-cell lymphoma, lymph nodes of inguinal region and lower limb

C83.36 Diffuse large B-cell lymphoma, intrapelvic lymph nodes

C83.37 Diffuse large B-cell lymphoma, spleen

C83.38 Diffuse large B-cell lymphoma, lymph nodes of multiple sites

C83.390 Primary central nervous system lymphoma

C83.398 Diffuse large B-cell lymphoma of other extranodal and solid organ sites

C83.3A Diffuse large B-cell lymphoma, in remission

C83.5A Lymphoblastic (diffuse) lymphoma, in remission

C83.7A Burkitt lymphoma, in remission

C83.8A Other non-follicular lymphoma, in remission

C83.9A Non-follicular (diffuse) lymphoma, unspecified, in remission

C83.51 Lymphoblastic (diffuse) lymphoma, lymph nodes of head, face, and neck

C83.52 Lymphoblastic (diffuse) lymphoma, intrathoracic lymph nodes

C83.53 Lymphoblastic (diffuse) lymphoma, intra-abdominal lymph nodes

C83.54 Lymphoblastic (diffuse) lymphoma, lymph nodes of axilla and upper limb

C83.55 Lymphoblastic (diffuse) lymphoma, lymph nodes of inguinal region and lower limb

C83.56 Lymphoblastic (diffuse) lymphoma, intrapelvic lymph nodes

C83.57 Lymphoblastic (diffuse) lymphoma, spleen

C83.58 Lymphoblastic (diffuse) lymphoma, lymph nodes of multiple sites

C83.59 Lymphoblastic (diffuse) lymphoma, extranodal and solid organ sites

C83.70 Burkitt lymphoma, unspecified site

C83.71 Burkitt lymphoma, lymph nodes of head, face, and neck

C83.72 Burkitt lymphoma, intrathoracic lymph nodes

C83.73 Burkitt lymphoma, intra-abdominal lymph nodes

C83.74 Burkitt lymphoma, lymph nodes of axilla and upper limb

C83.75 Burkitt lymphoma, lymph nodes of inguinal region and lower limb

C83.76 Burkitt lymphoma, intrapelvic lymph nodes

C83.77 Burkitt lymphoma, spleen

C83.78 Burkitt lymphoma, lymph nodes of multiple sites

C83.79 Burkitt lymphoma, extranodal and solid organ sites

C83.80 Other non-follicular lymphoma, unspecified site

C83.81 Other non-follicular lymphoma, lymph nodes of head, face, and neck

C83.82 Other non-follicular lymphoma, intrathoracic lymph nodes

C83.83 Other non-follicular lymphoma, intra-abdominal lymph nodes

C83.84 Other non-follicular lymphoma, lymph nodes of axilla and upper limb

C83.85 Other non-follicular lymphoma, lymph nodes of inguinal region and lower limb

C83.86 Other non-follicular lymphoma, intrapelvic lymph nodes

C83.87 Other non-follicular lymphoma, spleen

C83.88 Other non-follicular lymphoma, lymph nodes of multiple sites

C83.89 Other non-follicular lymphoma, extranodal and solid organ sites

C83.90 Non-follicular (diffuse) lymphoma, unspecified, unspecified site

C83.91 Non-follicular (diffuse) lymphoma, unspecified, lymph nodes of head, face, and neck

C83.92 Non-follicular (diffuse) lymphoma, unspecified, intrathoracic lymph nodes

C83.93 Non-follicular (diffuse) lymphoma, unspecified, intra-abdominal lymph nodes

C83.94 Non-follicular (diffuse) lymphoma, unspecified, lymph nodes of axilla and upper limb

C83.95 Non-follicular (diffuse) lymphoma, unspecified, lymph nodes of inguinal region and lower limb

C83.96 Non-follicular (diffuse) lymphoma, unspecified, intrapelvic lymph nodes

C83.97 Non-follicular (diffuse) lymphoma, unspecified, spleen

C83.98 Non-follicular (diffuse) lymphoma, unspecified, lymph nodes of multiple sites

C83.99 Non-follicular (diffuse) lymphoma, unspecified, extranodal and solid organ sites

C84.00 Mycosis fungoides, unspecified site

C84.01 Mycosis fungoides, lymph nodes of head, face, and neck

C84.02 Mycosis fungoides, intrathoracic lymph nodes

C84.03 Mycosis fungoides, intra-abdominal lymph nodes

C84.04 Mycosis fungoides, lymph nodes of axilla and upper limb

C84.05 Mycosis fungoides, lymph nodes of inguinal region and lower limb

C84.06 Mycosis fungoides, intrapelvic lymph nodes

C84.07 Mycosis fungoides, spleen

C84.08 Mycosis fungoides, lymph nodes of multiple sites

C84.09 Mycosis fungoides, extranodal and solid organ sites

C84.0A Mycosis fungoides, in remission

C84.10 Sezary disease, unspecified site

C84.11 Sezary disease, lymph nodes of head, face, and neck

C84.12 Sezary disease, intrathoracic lymph nodes

C84.13 Sezary disease, intra-abdominal lymph nodes

C84.14 Sezary disease, lymph nodes of axilla and upper limb

C84.15 Sezary disease, lymph nodes of inguinal region and lower limb

C84.16 Sezary disease, intrapelvic lymph nodes

C84.17 Sezary disease, spleen

C84.18 Sezary disease, lymph nodes of multiple sites

C84.19 Sezary disease, extranodal and solid organ sites

C84.1A Sezary disease, in remission

C84.40 Peripheral T-cell lymphoma, not elsewhere classified, unspecified site

C84.41 Peripheral T-cell lymphoma, not elsewhere classified, lymph nodes of head, face, and neck

C84.42 Peripheral T-cell lymphoma, not elsewhere classified, intrathoracic lymph nodes

C84.43 Peripheral T-cell lymphoma, not elsewhere classified, intra-abdominal lymph nodes

C84.44 Peripheral T-cell lymphoma, not elsewhere classified, lymph nodes of axilla and upper limb

C84.45 Peripheral T-cell lymphoma, not elsewhere classified, lymph nodes of inguinal region and lower limb

C84.46 Peripheral T-cell lymphoma, not elsewhere classified, intrapelvic lymph nodes

C84.47 Peripheral T-cell lymphoma, not elsewhere classified, spleen

C84.48 Peripheral T-cell lymphoma, not elsewhere classified, lymph nodes of multiple sites

C84.49 Peripheral T-cell lymphoma, not elsewhere classified, extranodal and solid organ sites

C84.4A Peripheral T-cell lymphoma, not elsewhere classified, in remission

C84.60 Anaplastic large cell lymphoma, ALK-positive, unspecified site

C84.61 Anaplastic large cell lymphoma, ALK-positive, lymph nodes of head, face, and neck

C84.62 Anaplastic large cell lymphoma, ALK-positive, intrathoracic lymph nodes

C84.63 Anaplastic large cell lymphoma, ALK-positive, intra-abdominal lymph nodes

C84.64 Anaplastic large cell lymphoma, ALK-positive, lymph nodes of axilla and upper limb

C84.65 Anaplastic large cell lymphoma, ALK-positive, lymph nodes of inguinal region and lower limb

C84.66 Anaplastic large cell lymphoma, ALK-positive, intrapelvic lymph nodes

C84.67 Anaplastic large cell lymphoma, ALK-positive, spleen

C84.68 Anaplastic large cell lymphoma, ALK-positive, lymph nodes of multiple sites

C84.69 Anaplastic large cell lymphoma, ALK-positive, extranodal and solid organ sites

C84.6A Anaplastic large cell lymphoma, ALK-positive, in remission

C84.70 Anaplastic large cell lymphoma, ALK-negative, unspecified site

C84.71 Anaplastic large cell lymphoma, ALK-negative, lymph nodes of head, face, and neck

C84.72 Anaplastic large cell lymphoma, ALK-negative, intrathoracic lymph nodes

C84.73 Anaplastic large cell lymphoma, ALK-negative, intra-abdominal lymph nodes

C84.74 Anaplastic large cell lymphoma, ALK-negative, lymph nodes of axilla and upper limb

C84.75 Anaplastic large cell lymphoma, ALK-negative, lymph nodes of inguinal region and lower limb

C84.76 Anaplastic large cell lymphoma, ALK-negative, intrapelvic lymph nodes

C84.77 Anaplastic large cell lymphoma, ALK-negative, spleen

C84.78 Anaplastic large cell lymphoma, ALK-negative, lymph nodes of multiple sites

C84.79 Anaplastic large cell lymphoma, ALK-negative, extranodal and solid organ sites

C84.7A Anaplastic large cell lymphoma, ALK-negative, breast

C84.7B Anaplastic large cell lymphoma, ALK-negative, in remission

C84.A0 Cutaneous T-cell lymphoma, unspecified, unspecified site

C84.A1 Cutaneous T-cell lymphoma, unspecified, lymph nodes of head, face, and neck

C84.A2 Cutaneous T-cell lymphoma, unspecified, intrathoracic lymph nodes

C84.A3 Cutaneous T-cell lymphoma, unspecified, intra-abdominal lymph nodes

C84.A4 Cutaneous T-cell lymphoma, unspecified, lymph nodes of axilla and upper limb

C84.A5 Cutaneous T-cell lymphoma, unspecified, lymph nodes of inguinal region and lower limb

C84.A6 Cutaneous T-cell lymphoma, unspecified, intrapelvic lymph nodes

C84.A7 Cutaneous T-cell lymphoma, unspecified, spleen

C84.A8 Cutaneous T-cell lymphoma, unspecified, lymph nodes of multiple sites

C84.A9 Cutaneous T-cell lymphoma, unspecified, extranodal and solid organ sites

C84.Z0 Other mature T/NK-cell lymphomas, unspecified site

C84.Z1 Other mature T/NK-cell lymphomas, lymph nodes of head, face, and neck

C84.Z2 Other mature T/NK-cell lymphomas, intrathoracic lymph nodes

C84.Z3 Other mature T/NK-cell lymphomas, intra-abdominal lymph nodes

C84.Z4 Other mature T/NK-cell lymphomas, lymph nodes of axilla and upper limb

C84.Z5 Other mature T/NK-cell lymphomas, lymph nodes of inguinal region and lower limb

C84.Z6 Other mature T/NK-cell lymphomas, intrapelvic lymph nodes

C84.Z7 Other mature T/NK-cell lymphomas, spleen

C84.Z8 Other mature T/NK-cell lymphomas, lymph nodes of multiple sites

C84.Z9 Other mature T/NK-cell lymphomas, extranodal and solid organ sites

C84.90 Mature T/NK-cell lymphomas, unspecified, unspecified site

C84.91 Mature T/NK-cell lymphomas, unspecified, lymph nodes of head, face, and neck

C84.92 Mature T/NK-cell lymphomas, unspecified, intrathoracic lymph nodes

C84.93 Mature T/NK-cell lymphomas, unspecified, intra-abdominal lymph nodes

C84.94 Mature T/NK-cell lymphomas, unspecified, lymph nodes of axilla and upper limb

C84.95 Mature T/NK-cell lymphomas, unspecified, lymph nodes of inguinal region and lower limb

C84.96 Mature T/NK-cell lymphomas, unspecified, intrapelvic lymph nodes

C84.97 Mature T/NK-cell lymphomas, unspecified, spleen

C84.98 Mature T/NK-cell lymphomas, unspecified, lymph nodes of multiple sites

C84.99 Mature T/NK-cell lymphomas, unspecified, extranodal and solid organ sites

C84.9A Mature T/NK-cell lymphomas, unspecified, in remission

C84.AA Cutaneous T-cell lymphoma, unspecified, in remission

C84.ZA Other mature T/NK-cell lymphomas, in remission

C85.10 Unspecified B-cell lymphoma, unspecified site

C85.11 Unspecified B-cell lymphoma, lymph nodes of head, face, and neck

C85.12 Unspecified B-cell lymphoma, intrathoracic lymph nodes

C85.13 Unspecified B-cell lymphoma, intra-abdominal lymph nodes

C85.14 Unspecified B-cell lymphoma, lymph nodes of axilla and upper limb

C85.15 Unspecified B-cell lymphoma, lymph nodes of inguinal region and lower limb

C85.16 Unspecified B-cell lymphoma, intrapelvic lymph nodes

C85.17 Unspecified B-cell lymphoma, spleen

C85.18 Unspecified B-cell lymphoma, lymph nodes of multiple sites

C85.19 Unspecified B-cell lymphoma, extranodal and solid organ sites

C85.1A Unspecified B-cell lymphoma, in remission

C85.20 Mediastinal (thymic) large B-cell lymphoma, unspecified site

C85.21 Mediastinal (thymic) large B-cell lymphoma, lymph nodes of head, face, and neck

C85.22 Mediastinal (thymic) large B-cell lymphoma, intrathoracic lymph nodes

C85.23 Mediastinal (thymic) large B-cell lymphoma, intra-abdominal lymph nodes

C85.24 Mediastinal (thymic) large B-cell lymphoma, lymph nodes of axilla and upper limb

C85.25 Mediastinal (thymic) large B-cell lymphoma, lymph nodes of inguinal region and lower limb

C85.26 Mediastinal (thymic) large B-cell lymphoma, intrapelvic lymph nodes

C85.27 Mediastinal (thymic) large B-cell lymphoma, spleen

C85.28 Mediastinal (thymic) large B-cell lymphoma, lymph nodes of multiple sites

C85.29 Mediastinal (thymic) large B-cell lymphoma, extranodal and solid organ sites

C85.2A Mediastinal (thymic) large B-cell lymphoma, in remission

C85.80 Other specified types of non-Hodgkin lymphoma, unspecified site

C85.81 Other specified types of non-Hodgkin lymphoma, lymph nodes of head, face, and neck

C85.82 Other specified types of non-Hodgkin lymphoma, intrathoracic lymph nodes

C85.83 Other specified types of non-Hodgkin lymphoma, intra-abdominal lymph nodes

C85.84 Other specified types of non-Hodgkin lymphoma, lymph nodes of axilla and upper limb

C85.85 Other specified types of non-Hodgkin lymphoma, lymph nodes of inguinal region and lower limb

C85.86 Other specified types of non-Hodgkin lymphoma, intrapelvic lymph nodes

C85.87 Other specified types of non-Hodgkin lymphoma, spleen

C85.88 Other specified types of non-Hodgkin lymphoma, lymph nodes of multiple sites

C85.89 Other specified types of non-Hodgkin lymphoma, extranodal and solid organ sites

C85.8A Other specified types of non-Hodgkin lymphoma, in remission

C85.90 Non-Hodgkin lymphoma, unspecified, unspecified site

C85.91 Non-Hodgkin lymphoma, unspecified, lymph nodes of head, face, and neck

C85.92 Non-Hodgkin lymphoma, unspecified, intrathoracic lymph nodes

C85.93 Non-Hodgkin lymphoma, unspecified, intra-abdominal lymph nodes

C85.94 Non-Hodgkin lymphoma, unspecified, lymph nodes of axilla and upper limb

C85.95 Non-Hodgkin lymphoma, unspecified, lymph nodes of inguinal region and lower limb

C85.96 Non-Hodgkin lymphoma, unspecified, intrapelvic lymph nodes

C85.97 Non-Hodgkin lymphoma, unspecified, spleen

C85.98 Non-Hodgkin lymphoma, unspecified, lymph nodes of multiple sites

C85.99 Non-Hodgkin lymphoma, unspecified, extranodal and solid organ sites

C85.9A Non-Hodgkin lymphoma, unspecified, in remission

C86.00 Extranodal NK/T-cell lymphoma, nasal type not having achieved remission

C86.01 Extranodal NK/T-cell lymphoma, nasal type, in remission

C86.10 Hepatosplenic T-cell lymphoma not having achieved remission

C86.11 Hepatosplenic T-cell lymphoma, in remission

C86.20 Enteropathy-type (intestinal) T-cell lymphoma not having achieved remission

C86.21 Enteropathy-type (intestinal) T-cell lymphoma, in remission

C86.30 Subcutaneous panniculitis-like T-cell lymphoma not having achieved remission

C86.31 Subcutaneous panniculitis-like T-cell lymphoma, in remission

C86.40 Blastic NK-cell lymphoma not having achieved remission

C86.41 Blastic NK-cell lymphoma, in remission

C86.50 Angioimmunoblastic T-cell lymphoma not having achieved remission

C86.51 Angioimmunoblastic T-cell lymphoma, in remission

C86.60 Primary cutaneous CD30-positive T-cell proliferations not having achieved remission

C86.61 Primary cutaneous CD30-positive T-cell proliferations, in remission

C88.00 Waldenstrom macroglobulinemia not having achieved remission

C88.01 Waldenstrom macroglobulinemia, in remission

C88.20 Heavy chain disease not having achieved remission

C88.21 Heavy chain disease, in remission

C88.30 Immunoproliferative small intestinal disease not having achieved remission

C88.31 Immunoproliferative small intestinal disease, in remission

C88.40 Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma] not having achieved remission

C88.41 Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma], in remission

C88.80 Other malignant immunoproliferative diseases not having achieved remission

C88.81 Other malignant immunoproliferative diseases, in remission

C88.90 Malignant immunoproliferative disease, unspecified not having achieved remission

C88.91 Malignant immunoproliferative disease, unspecified, in remission

C90.00 Multiple myeloma not having achieved remission

C90.02 Multiple myeloma in relapse

C91.10 Chronic lymphocytic leukemia of B-cell type not having achieved remission

C91.12 Chronic lymphocytic leukemia of B-cell type in relapse

D47.2 Monoclonal gammopathy

D59.0 Drug-induced autoimmune hemolytic anemia

D59.11 Warm autoimmune hemolytic anemia

D59.12 Cold autoimmune hemolytic anemia

D59.13 Mixed type autoimmune hemolytic anemia

D59.19 Immune thrombocytopenic anemia

D59.9 Acquired hemolytic anemia, unspecified

D61.01 Constitutional (pure) red blood cell aplasia

D68.00 Von Willebrand's disease, unspecified

D68.01 Von Willebrand disease, type 1

D68.020 Von Willebrand disease, type 2A

D68.021 Von Willebrand disease, type 2B

D68.022 Von Willebrand disease, type 2M

D68.023 Von Willebrand disease, type 2N

D68.029 Von Willebrand disease, type 2, unspecified

D68.03 Von Willebrand disease, type 3

D68.04 Acquired Von Willebrand disease

D68.09 Other Von Willebrand disease

D68.311 Acquired hemophilia

D68.312 Antiphospholipid antibody with hemorrhagic disorder

D68.318 Other hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors

D69.3 Immune thrombocytopenic purpura

D69.41 Evans syndrome

D69.42 Congenital and hereditary thrombocytopenia purpura

D69.49 Other primary thrombocytopenia

D80.0 Hereditary hypogammaglobulinemia

D80.2 Selective deficiency of immunoglobulin A [IgA]

D80.3 Selective deficiency of immunoglobulin G [IgG] subclasses

D80.4 Selective deficiency of immunoglobulin M [IgM]

D80.5 Immunodeficiency with increased immunoglobulin M [IgM]

D80.6 Antibody deficiency with near-normal immunoglobulins or with hyperimmunoglobulinemia

D80.8 Other immunodeficiencies with predominantly antibody defects

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

D81.31 Severe combined immunodeficiency due to adenosine deaminase deficiency

D81.4 Nezelof's syndrome

D81.5 Purine nucleoside phosphorylase [PNP] deficiency

D81.6 Major histocompatibility complex class I deficiency

D81.7 Major histocompatibility complex class II deficiency

D81.82 Activated Phosphoinositide 3-kinase Delta Syndrome (APDS)

D81.89 Other combined immunodeficiencies

D81.9 Combined immunodeficiency, unspecified

D82.0 Wiskott-Aldrich syndrome

D82.1 Di George's syndrome

D82.4 Hyperimmunoglobulin E [IgE] syndrome

D83.0 Common variable immunodeficiency with predominant abnormalities of B-cell numbers and function

D83.1 Common variable immunodeficiency with predominant immunoregulatory T-cell disorders

D83.2 Common variable immunodeficiency with autoantibodies to B- or T-cells

D83.8 Other common variable immunodeficiencies

D83.9 Common variable immunodeficiency, unspecified

D89.811 Chronic graft-versus-host disease

D89.812 Acute on chronic graft-versus-host disease

D89.813 Graft-versus-host disease, unspecified

D89.834 Cytokine release syndrome, grade 4

E05.00 Thyrotoxicosis with diffuse goiter without thyrotoxic crisis or storm

E05.01 Thyrotoxicosis with diffuse goiter with thyrotoxic crisis or storm

E05.80 Other thyrotoxicosis without thyrotoxic crisis or storm

E05.81 Other thyrotoxicosis with thyrotoxic crisis or storm

E05.90 Thyrotoxicosis, unspecified without thyrotoxic crisis or storm

E05.91 Thyrotoxicosis, unspecified with thyrotoxic crisis or storm

G04.81 Other encephalitis and encephalomyelitis

G04.89 Other myelitis

G04.90 Encephalitis and encephalomyelitis, unspecified

G04.91 Myelitis, unspecified

G11.3 Cerebellar ataxia with defective DNA repair

G25.82 Stiff-man syndrome

G35 Multiple sclerosis

G36.0 Neuromyelitis optica [Devic]

G37.3 Acute transverse myelitis in demyelinating disease of central nervous system

G60.3 Idiopathic progressive neuropathy

G61.0 Guillain-Barre syndrome

G61.81 Chronic inflammatory demyelinating polyneuritis

G61.82 Multifocal motor neuropathy

G61.89 Other inflammatory polyneuropathies

G61.9 Inflammatory polyneuropathy, unspecified

G62.89 Other specified polyneuropathies

G62.9 Polyneuropathy, unspecified

G64 Other disorders of peripheral nervous system

G70.00 Myasthenia gravis without (acute) exacerbation

G70.01 Myasthenia gravis with (acute) exacerbation

G70.80 Lambert-Eaton syndrome, unspecified

G70.81 Lambert-Eaton syndrome in disease classified elsewhere

G72.41 Inclusion body myositis [IBM]

G72.81 Critical illness myopathy

G72.89 Other specified myopathies

G73.1 Lambert-Eaton syndrome in neoplastic disease

G73.3 Myasthenic syndromes in other diseases classified elsewhere

G93.49 Other encephalopathy

H05.241 Constant exophthalmos, right eye

H05.242 Constant exophthalmos, left eye

H05.243 Constant exophthalmos, bilateral

H46.9 Unspecified optic neuritis

I40.8 Other acute myocarditis

I40.9 Acute myocarditis, unspecified

I77.82 Antineutrophilic cytoplasmic antibody (ANCA) vasculitis

I78.8 Other diseases of capillaries

L10.0 Pemphigus vulgaris

L10.1 Pemphigus vegetans

L10.2 Pemphigus foliaceus

L10.3 Brazilian pemphigus [fogo selvagem]

L10.4 Pemphigus erythematosis

L10.5 Drug-induced pemphigus

L10.81 Paraneoplastic pemphigus

L10.89 Other pemphigus

L10.9 Pemphigus, unspecified

L12.0 Bullous pemphigoid

L12.1 Cicatricial pemphigoid

L12.30 Acquired epidermolysis bullosa, unspecified

L12.35 Other acquired epidermolysis bullosa

L12.8 Other pemphigoid

L12.9 Pemphigoid, unspecified

L13.8 Other specified bullous disorders

L13.9 Bullous disorder, unspecified

L14 Bullous disorders in diseases classified elsewhere

L51.1 Stevens-Johnson syndrome

L51.2 Toxic epidermal necrolysis [Lyell]

L51.3 Stevens-Johnson syndrome-toxic epidermal necrolysis overlap syndrome

L90.8 Other atrophic disorders of skin

L91.8 Other hypertrophic disorders of the skin

L98.5 Mucinosis of the skin

M30.3 Mucocutaneous lymph node syndrome [Kawasaki]

M32.10 Systemic lupus erythematosus, organ or system involvement unspecified

M32.11 Endocarditis in systemic lupus erythematosus

M32.12 Pericarditis in systemic lupus erythematosus

M32.13 Lung involvement in systemic lupus erythematosus

M32.14 Glomerular disease in systemic lupus erythematosus

M32.15 Tubulo-interstitial nephropathy in systemic lupus erythematosus

M32.19 Other organ or system involvement in systemic lupus erythematosus

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

M33.99 Dermatopolymyositis, unspecified with other organ involvement

M36.0 Dermato(poly)myositis in neoplastic disease

M60.811 Other myositis, right shoulder

M60.812 Other myositis, left shoulder
M60.821 Other myositis, right upper arm
M60.822 Other myositis, left upper arm
M60.831 Other myositis, right forearm
M60.832 Other myositis, left forearm
M60.841 Other myositis, right hand
M60.842 Other myositis, left hand
M60.851 Other myositis, right thigh
M60.852 Other myositis, left thigh
M60.861 Other myositis, right lower leg
M60.862 Other myositis, left lower leg
M60.871 Other myositis, right ankle and foot
M60.872 Other myositis, left ankle and foot
M60.88 Other myositis, other site
M60.89 Other myositis, multiple sites
M60.9 Myositis, unspecified
P07.00 Extremely low birth weight newborn, unspecified weight
P07.01 Extremely low birth weight newborn, less than 500 grams
P07.02 Extremely low birth weight newborn, 500-749 grams
P07.03 Extremely low birth weight newborn, 750-999 grams
P07.10 Other low birth weight newborn, unspecified weight
P07.14 Other low birth weight newborn, 1000-1249 grams
P07.15 Other low birth weight newborn, 1250-1499 grams
T80.90XA Unspecified complication following infusion and therapeutic injection, initial encounter
T80.90XD Unspecified complication following infusion and therapeutic injection, subsequent encounter
T80.90XS Unspecified complication following infusion 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
T86.11 Kidney transplant rejection
T86.21 Heart transplant rejection
T86.31 Heart-lung transplant rejection
T86.5 Complications of stem cell transplant
T86.810 Lung transplant rejection

Z28.39 Other underimmunization status
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
Z48.298 Encounter for aftercare following other organ transplant
Z51.11 Encounter for antineoplastic chemotherapy
Z51.12 Encounter for antineoplastic immunotherapy
Z91.89 Other specified personal risk factors, not elsewhere classified
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
Z99.2 Dependence on renal dialysis

THE FOLLOWING PIDD DIAGNOSIS CODES REQUIRE FURTHER REVIEW TO ENSURE THAT THE IMMUNODEFICIENCY IS OF PRIMARY ORIGIN:

D80.1 Nonfamilial hypogammaglobulinemia
D80.7 Transient hypogammaglobulinemia of infancy

NOT COVERED

INTRAVENOUS IMMUNE GLOBULIN (IVIG)

IVIG is not covered for the following diagnosis:

E71.520 Childhood cerebral X-linked adrenoleukodystrophy
E71.521 Adolescent X-linked adrenoleukodystrophy
E71.522 Adrenomyeloneuropathy
E71.528 Other X-linked adrenoleukodystrophy
E71.529 X-linked adrenoleukodystrophy, unspecified type
D47.9 Neoplasm of uncertain behavior of lymphoid, hematopoietic and related tissue, unspecified

D89.810 Acute graft-versus-host disease
G12.21 Amyotrophic lateral sclerosis
G54.0 Brachial plexus disorders
G60.9 Hereditary and idiopathic neuropathy, unspecified
G61.89 Other inflammatory polyneuropathies
G61.9 Inflammatory polyneuropathy, unspecified
G62.81 Critical illness polyneuropathy
Z94.81 Bone marrow transplant status
Z94.84 Stem cells transplant status

HCPCS Level II Code Number(s)

MEDICALLY NECESSARY

SUBCUTANEOUS IMMUNE GLOBULIN (SCIG)

J1555 Injection, immune globulin (cuvitru), 100 mg
J1558 Injection, immune globulin (xembify), 100 mg
J1559 Injection, immune globulin (Hizentra), 100 mg
J1561 Injection, immune globulin, (Gamunex/Gamunex-C/Gammaked), nonlyophilized (e.g., liquid), 500 mg
J1569 Injection, immune globulin, (gammagard liquid/gammagard liquid erc), non-lyophilized, (e.g., liquid), 500 mg

The following code is used to represent HyQvia:

J1575 Injection, immune globulin/hyaluronidase, 100 mg immunoglobulin

The following code are used to represent Cutaquig:

J1551 Injection, immune globulin (cutaquig), 100 mg

INTRAVENOUS IMMUNE GLOBULIN (IVIG)

J1459 Injection, immune globulin (Privigen), intravenous, non-lyophilized (e.g. liquid), 500 mg
J1552 Injection, immune globulin (alyglo), 500 mg
J1553 Injection, immune globulin (yimmugo), 100 mg
J1554 Injection, immune globulin (asceniv), 500 mg
J1556 Injection, immune globulin (bivigam), 500 mg
J1557 Injection, immune globulin (Gammaplex), intravenous, non-lyophilized (e.g. liquid), 500 mg
J1561 Injection, immune globulin, (Gamunex/Gamunex-C/Gammaked), non-lyophilized (e.g. liquid), 500 mg
J1568 Injection, immune globulin, (Octagam), intravenous, non-lyophilized (e.g. liquid), 500 mg
J1569 Injection, immune globulin, (gammagard liquid/gammagard liquid erc), non-lyophilized, (e.g., liquid), 500 mg
J1572 Injection, immune globulin, (Flebogamma/Flebogamma Dif), intravenous, nonlyophilized (e.g., liquid), 500 mg

J1576 Injection, immune globulin (panzyga), intravenous, non-lyophilized (e.g., liquid), 500 mg

Q2052 Services, supplies and accessories used in the home for the administration of intravenous immune globulin (ivig)

S9338 Home infusion therapy, immunotherapy, administrative services, professional pharmacy services, care coordination, and all necessary supplies and equipment (drugs and nursing visits coded separately), per diem

The following code is used to represent GAMMAGARD S/D:

J1566 Injection, immune globulin, intravenous, lyophilized (e.g. powder), not otherwise specified, 500 mg

NOT ELIGIBLE FOR REIMBURSEMENT

THE FOLLOWING CODE REPRESENTS CARIMUNE NF WHICH IS NO LONGER MANUFACTURED AND HAS BEEN WITHDRAWN FROM THE MARKET:

J1566 Injection, immune globulin, intravenous, lyophilized (e.g. powder), not otherwise specified, 500 mg

Revenue Code Number(s)

N/A

Cross Reference



Immune Globulin IV
(IVIG), SQ (SCIG) Attac

Attachment A:

Description: ICD-10 DIAGNOSIS CODES



Immune Globulin IV
(IVIG), SQ (SCIG) Attac

Attachment B:

Description: Dosage and Frequency Requirements

Policy History

Revisions From MA08.009z:

09/18/2026	This version of the policy will become effective 09/18/2026. The following HCPCS narrative has been revised in this policy: FROM: J1569 Injection, immune globulin, (gammagard liquid), non-lyophilized, (e.g., liquid), 500 mg TO: J1569 Injection, immune globulin, (gammagard liquid/gammagard liquid erc), non-lyophilized, (e.g., liquid), 500 mg
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Revisions From MA08.009y:

06/18/2026	This version of the policy will become effective 06/18/2026. The following HCPCS code has been removed from this policy:
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	J1599 Injection, immune globulin, intravenous, nonlyophilized (e.g., liquid), not otherwise specified, 500 mg The following HCPCS code has been added to this policy: J1553 Injection, immune globulin (yimmugo), 100 mg
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Revisions From MA08.009x:

03/20/2026	This version of the policy will become effective 03/20/2026. The following HCPCS codes have been removed from this policy: J1562 Injection, immune globulin (vivaglobin), 100 mg The following HCPCS code has been added: J1572 Injection, immune globulin, (flebogamma/flebogamma dif), intravenous, non-lyophilized (e.g., liquid), 500 mg
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Revisions From MA08.009w:

06/13/2025	This version of the policy will become effective 06/13/2025. The following Healthcare Common Procedure Coding System (HCPCS) code has been added to this policy: - J1552 Injection, immune globulin (alyglo), 500 mg The following HCPCS code has been removed from this policy, which was used to represent immune globulin intravenous, human-stwk (Alyglo): - J1599 Injection, immune globulin, intravenous, nonlyophilized (e.g., liquid), not otherwise specified, 500 mg
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Revisions From MA08.009v:

12/16/2024	This version of the policy will become effective 12/16/2024. The following HCPCS codes have been termed (no longer valid codes) and removed from this policy: C83.39 Diffuse large B-cell lymphoma, extranodal and solid organ sites C86.0 Extranodal NK/T-cell lymphoma, nasal type C86.1 Hepatosplenic T-cell lymphoma C86.2 Enteropathy-type (intestinal) T-cell lymphoma C86.3 Subcutaneous panniculitis-like T-cell lymphoma C86.4 Blastic NK-cell lymphoma C86.5 Angioimmunoblastic T-cell lymphoma C86.6 Primary cutaneous CD30-positive T-cell proliferations C88.0 Waldenstrom macroglobulinemia C88.2 Heavy chain disease C88.3 Immunoproliferative small intestinal disease C88.4 Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma] C88.8 Other malignant immunoproliferative diseases The following ICD-10 CM codes have been added to this policy:
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	C83.390, C83.398, C83.3A, C83.5A, C83.7A, C83.8A, C83.9A, C84.0A, C84.1A, C84.4A, C84.6A, C84.7B, C84.9A, C84.AA, C84.ZA, C85.1A, C85.2A, C85.8A, C85.9A, C86.00, C86.01, C86.10, C86.11, C86.20, C86.21, C86.30, C86.31, C86.40, C86.41, C86.50, C86.51, C86.60, C86.61, C88.00, C88.01, C88.20, C88.21, C88.30, C88.31, C88.40, C88.41, C88.80, C88.81, C88.90, C88.91
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Revisions From MA08.009u:

09/16/2024	This version of the policy will become effective 09/16/2024. This policy has been revised to communicate the coverage position of intravenous immune globulin (IVIG) and subcutaneous immune globulin (SCIG). New product, immune globulin intravenous, human-stwk (Alyglo) was added, represented by HCPCS code: J1599 Injection, immune globulin, intravenous, nonlyophilized (e.g., liquid), not otherwise specified, 500 mg Language regarding refill requirements for supplies was added to the Required Documentation Section of the Policy, per Noridian Medicare Local Coverage Determination (LCD): Intravenous Immune Globulin (L33610) effective 01/01/2024. Attachment A: ICD-10 Diagnosis Codes The following ICD-10 Diagnosis Codes have been added to this policy for IVIG: E05.00 Thyrotoxicosis with diffuse goiter without thyrotoxic crisis or storm E05.10 Thyrotoxicosis with toxic single thyroid nodule without thyrotoxic crisis or storm E05.80 Other thyrotoxicosis without thyrotoxic crisis or storm E05.81 Other thyrotoxicosis with thyrotoxic crisis or storm E05.90 Thyrotoxicosis, unspecified without thyrotoxic crisis or storm E05.91 Thyrotoxicosis, unspecified with thyrotoxic crisis or storm Attachment B: Dosing and Frequency: Added a new reference #242 from UpToDate.
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Revisions From MA08.009t:

01/01/2024	This version of the policy will become effective 01/01/2024. The following HCPCS code has been added to this policy: Q2052 Services, supplies and accessories used in the home for the administration of intravenous immune globulin (ivig)
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Revisions From MA08.009s:

02/05/2023	This version of the policy is retro-effective to 02/05/2023. This policy has been revised to communicate the coverage position of intravenous immune globulin (IVIG) and subcutaneous immune globulin (SCIG), in accordance with Novitas Solutions, Inc. <i>Local Coverage Article (LCA)</i> A56786 - Billing and Coding: Intravenous Immune Globulin (IVIG). Effective date 07/01/2023. <i>Local Coverage Determination (LCD)</i> L35093 - Intravenous Immune Globulin (IVIG). [Novitas Solutions Web site]. Original 10/01/2015, Revised 02/05/2023.
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The following ICD-10 CM codes have been **added** to this policy as Medically Necessary for IVIG AND SCIG:

D80.8 Other immunodeficiencies with predominantly antibody defects
D81.31 Severe combined immunodeficiency due to adenosine deaminase deficiency
D81.4 Nezelof's syndrome

The following ICD-10 CM codes have been **added** to this policy as Medically Necessary for IVIG:

C81.00, C81.01, C81.02, C81.03, C81.04, C81.05, C81.06, C81.07, C81.08, C81.09, C81.10, C81.11, C81.12, C81.13, C81.14, C81.15, C81.16, C81.17, C81.18, C81.19, C81.20, C81.21, C81.22, C81.23, C81.24, C81.25, C81.26, C81.27, C81.28, C81.29, C81.30, C81.31, C81.32, C81.33, C81.34, C81.35, C81.36, C81.37, C81.38, C81.39, C81.40, C81.41, C81.42, C81.43, C81.44, C81.45, C81.46, C81.47, C81.48, C81.49, C81.70, C81.71, C81.72, C81.73, C81.74, C81.75, C81.76, C81.77, C81.78, C81.79, C81.90, C81.91, C81.92, C81.93, C81.94, C81.95, C81.96, C81.97, C81.98, C81.99, C82.00, C82.01, C82.02, C82.03, C82.04, C82.05, C82.06, C82.07, C82.08, C82.09, C82.10, C82.11, C82.12, C82.13, C82.14, C82.15, C82.16, C82.17, C82.18, C82.19, C82.20, C82.21, C82.22, C82.23, C82.24, C82.25, C82.26, C82.27, C82.28, C82.29, C82.30, C82.31, C82.32, C82.33, C82.34, C82.35, C82.36, C82.37, C82.38, C82.39, C82.40, C82.41, C82.42, C82.43, C82.44, C82.45, C82.46, C82.47, C82.48, C82.49, C82.50, C82.51, C82.52, C82.53, C82.54, C82.55, C82.56, C82.57, C82.58, C82.59, C82.60, C82.61, C82.62, C82.63, C82.64, C82.65, C82.66, C82.67, C82.68, C82.69, C82.80, C82.81, C82.82, C82.83, C82.84, C82.85, C82.86, C82.87, C82.88, C82.89, C82.90, C82.91, C82.92, C82.93, C82.94, C82.95, C82.96, C82.97, C82.98, C82.99, C83.00, C83.01, C83.02, C83.03, C83.04, C83.05, C83.06, C83.07, C83.08, C83.09, C83.10, C83.11, C83.12, C83.13, C83.14, C83.15, C83.16, C83.17, C83.18, C83.19, C83.30, C83.31, C83.32, C83.33, C83.34, C83.35, C83.36, C83.37, C83.38, C83.39, C83.50, C83.51, C83.52, C83.53, C83.54, C83.55, C83.56, C83.57, C83.58, C83.59, C83.70, C83.71, C83.72, C83.73, C83.74, C83.75, C83.76, C83.77, C83.78, C83.79, C83.80, C83.81, C83.82, C83.83, C83.84, C83.85, C83.86, C83.87, C83.88, C83.89, C83.90, C83.91, C83.92, C83.93, C83.94, C83.95, C83.96, C83.97, C83.98, C83.99, C84.00, C84.01, C84.02, C84.03, C84.04, C84.05, C84.06, C84.07, C84.08, C84.09, C84.10, C84.11, C84.12, C84.13, C84.14, C84.15, C84.16, C84.17, C84.18, C84.19, C84.40, C84.41, C84.42, C84.43, C84.44, C84.45, C84.46, C84.47, C84.48, C84.49, C84.60, C84.61, C84.62, C84.63, C84.64, C84.65, C84.66, C84.67, C84.68, C84.69, C84.70, C84.71, C84.72, C84.73, C84.74, C84.75, C84.76, C84.77, C84.78, C84.79, C84.7A, C84.A0, C84.A1, C84.A2, C84.A3, C84.A4, C84.A5, C84.A6, C84.A7, C84.A8, C84.A9, C84.Z0, C84.Z1, C84.Z2, C84.Z3, C84.Z4, C84.Z5, C84.Z6, C84.Z7, C84.Z8, C84.Z9, C84.90, C84.91, C84.92, C84.93, C84.94, C84.95, C84.96, C84.97, C84.98, C84.99, C85.11, C85.12, C85.13, C85.14, C85.15, C85.16, C85.17, C85.18, C85.19, C85.20, C85.21, C85.22, C85.23, C85.24, C85.25, C85.26, C85.27, C85.28, C85.29, C85.80, C85.81, C85.82, C85.83, C85.84, C85.85, C85.86, C85.87, C85.88, C85.89, C85.90, C85.91, C85.92, C85.93, C85.94, C85.95, C85.96, C85.97, C85.98, C85.99, C86.0, C86.1, C86.2, C86.3, C86.4, C86.5, C86.6, C88.0, C88.2, C88.3, C88.4, C88.8, D59.12, D59.13, D59.19, D89.834, G36.0, G72.81, G72.89, G93.49, H05.241, H05.242, H05.243, H46.9, I40.8, I40.9, L12.35, M32.11, M32.12, M32.13, M32.14, M32.15, M32.19, M33.03, M33.13, Z28.39, Z51.11, Z51.12, Z91.89, Z99.2

The following ICD-10 CM codes have been **removed** from this policy for IVIG:

C90.01, C91.11, D89.810, M60.80, M60.819, M60.829, M60.839, M60.849, M60.859, M60.869, M60.879, O36.0910, O36.0911, O36.0912, O36.0913, O36.0914, O36.0915, O36.0919, O36.0920, O36.0921, O36.0922, O36.0923, O36.0924, O36.0925, O36.0929, O36.0930, O36.0931, O36.0932, O36.0933, O36.0934, O36.0935, O36.0939, O36.0990, O36.0991, O36.0992, O36.0993, O36.0994, O36.0995, O36.0999

The coverage position for the following ICD-10 CM code has been **revised** from Noncovered to Medically Necessary for IVIG for:

G72.41 Inclusion body myositis [IBM]

The coverage position for the following ICD-10 CM code has been **revised** from Medically Necessary to Noncovered for IVIG for the following ICD-10 CM codes:

	G62.81 Critical illness polyneuropathy Z94.81 Bone marrow transplant status Z94.84 Stem cells transplant status The following ICD-10 CM codes have been added to this policy as Noncovered for IVIG: E71.520, E71.521, E71.522, E71.528, E71.529, D47.9, D89.810, G12.21, G54.0, G60.9, G61.89, G61.9
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Revisions From MA08.009r:

07/01/2023	This version of the policy will become effective 07/01/2023. The following NOC code has been removed from this policy and is replaced by the following HCPCS code: • Removed: J1599 Injection, immune globulin, intravenous, nonlyophilized (e.g., liquid), not otherwise specified, 500mg • Replaced With: J1576 Injection, immune globulin (panzyga), intravenous, non-lyophilized (e.g., liquid), 500 mg
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Revisions From MA08.009q:

10/01/2022	This version of the policy will become effective 10/01/2022. The following ICD-10 CM code has been termed (no longer valid codes) and removed from this policy for IVIG: D68.0 Von Willebrand's disease The following ICD-10 CM codes have been added to this policy for IVIG: D68.00 Von Willebrand disease, unspecified D68.01 Von Willebrand disease, type 1 D68.020 Von Willebrand disease, type 2A D68.021 Von Willebrand disease, type 2B D68.022 Von Willebrand disease, type 2M D68.023 Von Willebrand disease, type 2N D68.029 Von Willebrand disease, type 2, unspecified D68.03 Von Willebrand disease, type 3 D68.04 Acquired von Willebrand disease D68.09 Other von Willebrand disease I77.82 Antineutrophilic cytoplasmic antibody [ANCA] vasculitis The following ICD-10 CM code has been added to this policy for IVIG and SCIG: D81.82 Activated Phosphoinositide 3-kinase Delta Syndrome (APDS)
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Revisions From MA08.009p:

07/01/2022	This policy has been identified for the HCPCS code update, effective 07/01/2022. The following NOC codes have been removed from this policy and are replaced by the following HCPCS code: REMOVED: C9399 Unclassified drugs or biologicals J7799 NOC drugs, other than inhalation drugs, administered through DME REPLACED WITH: J1551 Injection, immune globulin (cutaquin), 100 mg
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Revisions From MA08.009o:

01/31/2022	This version of the policy will become effective 01/31/2022. This policy has been updated to communicate the coverage position for the following: • Claims for Carimune NF are not eligible for reimbursement, since it was withdrawn from the market. The HCPCS code will remain set up as eligible, since it also represents GAMMAGARD S/D. (J1566 Injection, immune globulin, intravenous, lyophilized (e.g. powder), not otherwise specified, 500 mg) • Indications added per National Comprehensive Cancer Network (NCCN) ○ NCCN Clinical Practice Guidelines in Oncology - Management of immune checkpoint inhibitor-related toxicities. V.3.2021. 05/14/2021. ○ NCCN Drugs & Biologics Compendium. Immune globulin. [NCCN Web site]. 2021. Accessed September 1, 2021. ▪ Acquired (secondary) hypogammaglobulinemia in oncologic conditions ▪ Management of Immune checkpoint inhibitor-related toxicities • HIV-infected individuals to reduce bacterial infection: timeframe of dosing expanded from every 28 days to every 2-4 weeks. • CIDP policy criteria and Dosing & Frequency were updated in alignment with Noridian Medicare and Hizentra FDA labeling <u>Attachment B: ICD-10 DIAGNOSIS CODES</u> The following Header was added: THIS IS NOT AN ALL INCLUSIVE LIST. REPORT THE MOST APPROPRIATE DIAGNOSIS CODE FOR TOXICITIES RELATED TO THE MANAGEMENT OF IMMUNE CHECKPOINT INHIBITOR TOXICITIES. The following ICD-10 CM codes have been added to this policy for IVIG as Medically Necessary: G04.81 Other encephalitis and encephalomyelitis G04.89 Other myelitis G04.90 Encephalitis and encephalomyelitis, unspecified G04.91 Myelitis, unspecified G37.3 Acute transverse myelitis in demyelinating disease of central nervous system L13.9 Bullous disorder, unspecified M60.80 - M60.89 Other myositis M60.9 Myositis, unspecified
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	T80.90XA Unspecified complication following infusion and therapeutic injection, initial encounter T80.90XD Unspecified complication following infusion and therapeutic injection, subsequent encounter T80.90XS Unspecified complication following infusion and therapeutic injection, sequela
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Revisions From MA08.009n:

04/01/2021	This policy has been identified for the HCPCS code update, effective 04/01/2021. The following HCPCS code has been termed (no longer valid code) from this policy: C9072 Injection, immune globulin (asceniv), 500 mg The following HCPCS code has been removed from this policy to represent Asceniv (but will remain in the policy to represent Panzyga): J1599 Injection, immune globulin, intravenous, nonlyophilized (e.g., liquid), not otherwise specified, 500mg The following ICD-10 CM code has been added to this policy: J1554 Injection, immune globulin (asceniv), 500 mg
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Revisions From MA08.009m:

01/01/2021	This policy has been identified for the HCPCS code update, effective 01/01/2021. The following ICD-10 CM code has been added to this policy: C9072 Injection, immune globulin (asceniv), 500 mg
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Revisions From MA08.009l:

10/12/2020	HCPCS "J3590 Unclassified biologics" has been replaced with "J7799 NOC drugs, other than inhalation drugs, administered through DME" to represent Cutaquig. This is in alignment with Billing Requirements for Medicare contractor, Noridian Healthcare Solutions.
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Revisions From MA08.009k:

10/01/2020	This policy has been identified for the ICD-10 CM code update, effective 10/01/2020. The following ICD-10 CM code has been deleted for IVIG: D59.1 Other autoimmune hemolytic anemias The following ICD-10 CM code has been added for IVIG: D59.11 Warm autoimmune hemolytic anemia
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Revisions From MA08.009j:

07/20/2020	This version of the policy will become effective 07/20/2020. This policy was updated to communicate the removal of the following indications:
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	○ Benign mucous membrane pemphigoid, with or without mention of ocular movement, due to Centers for Medicare and Medicaid Services (CMS) NCD 250.3. ○ Erythema multiforme major, due to terminology change
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Revisions From MA08.009i:

07/01/2020	This policy has been identified for the HCPCS code update, effective 07/01/2020. The following NOC codes have been removed from this policy for Xembify and are replaced by the following HCPCS code: REMOVED: C9399 Unclassified drugs or biologicals J3590 Unclassified biologics REPLACED WITH: J1558 Injection, immune globulin (xembify), 100 mg
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Revisions From MA08.009h:

10/21/2019	This policy was updated to communicate the Company's coverage criteria, including Dosing and Frequency, of the new SCIG product, Xembify. Per CMS, "MM11295 – Update to Coverage of Intravenous Immune Globulin for Treatment of Primary Immune Deficiency Diseases in the Home", additional primary immune deficiency diseases and codes were added to this policy for IVIG and SCIG.
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Revisions From MA08.009g:

06/17/2019	This Policy was revised with the following changes: • The following new SCIG product was added: Cutaquig • The following new IVIG products were added: Asceniv™, Panzyga®. • New FDA-approved indication for SCIG (e.g., Hizentra®) for CIDP was added. • New indication for IVIG, per Novitas L35093, for Systemic Capillary Leak Syndrome (SCLS) or Clarkson's Disease: IVIG is considered medically necessary and, therefore, covered in individuals with Systemic Capillary Leak Syndrome (SCLS) or Clarkson's Disease when associated with monoclonal gammopathy and used for prophylaxis to increase survival • Lambert-Eaton: plasma exchange was added as an example of prior therapies. The following CPT codes were removed from the policy: 90283, 90284 <u>Attachment B: Dosage and Frequency</u> <u>SCIG Section:</u> PIDD Induction dosing was expanded from 100-150 mg/kg/weekly to 100-200 mg/kg/weekly. PIDD: Cutaquig dosing was added. CIDP: New Indication. Hizentra dosing was added. <u>IVIG Section:</u> Dosing and Frequency were added/adjusted for the following IVIG indications: Scleromyxedema Systemic Capillary Leak Syndrome (SCLS) or Clarkson's Disease Antibody-Mediated Rejection (AMR)
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	Guillain-Barre syndrome Myasthenia gravis syndrome
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Revisions From MA08.009f:

10/22/2018	This version of the policy will become effective 10/22/2018. This policy has been updated to include coverage changes, per Novitas Solutions, Inc., to: • Include the following subtypes of Primary Humoral Immunodeficiencies: Nonfamilial hypogammaglobulinemia, Selective deficiency of IgG subclasses, and Transient hypogammaglobulinemia of infancy • Add coverage criteria for antibody-mediated rejection (AMR), multiple myeloma, and Rh and ABO incompatibility • Clarify types of solid organs included in transplant coverage.
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Revisions From MA08.009e:

01/01/2018	This policy has been identified for the HCPCS code update, effective 01/01/2018. The following NOC code has been removed from this policy and is replaced by the following HCPCS code: REMOVED: J3590 Unclassified biologics REPLACED WITH: J1555 Injection, immune globulin (cuvitru), 100 mg
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Revisions From MA08.009d:

10/01/2017	This policy has been identified for the ICD-10 CM code update, effective 10/01/2017. The following ICD-10 CM codes have been added to this policy for IVIG only: M33.03 Juvenile dermatomyositis without myopathy M33.13 Other dermatomyositis without myopathy M33.93 Dermatopolymyositis, unspecified without myopathy The following ICD-10 CM narratives have been revised in this policy for IVIG only: M33.00 FROM: Juvenile dermatopolymyositis, organ involvement unspecified TO: Juvenile dermatomyositis, organ involvement unspecified M33.01 FROM: Juvenile dermatopolymyositis with respiratory involvement TO: Juvenile dermatomyositis with respiratory involvement M33.02 FROM: Juvenile dermatopolymyositis with myopathy TO: Juvenile dermatomyositis with myopathy M33.09 FROM: Juvenile dermatopolymyositis with other organ involvement TO: Juvenile dermatomyositis with other organ involvement M33.10 FROM: Other dermatopolymyositis, organ involvement unspecified TO: Other dermatomyositis, organ involvement unspecified
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	M33.11 FROM: Other dermatopolymyositis with respiratory involvement TO: Other dermatomyositis with respiratory involvement M33.12 FROM: Other dermatopolymyositis with myopathy TO: Other dermatomyositis with myopathy M33.19 FROM: Other dermatopolymyositis with other organ involvement TO: Other dermatomyositis with other organ involvement
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Revisions From MA08.009c:

12/28/2016	This policy was updated to reflect current Medicare Policies. The new SCIG product, Cuvitru, was added to the Policy and to the Dosing and Frequency Attachment. In the Dosing and Frequency Attachment, Kawasaki disease was added to this attachment and Scleromyxedema was revised. In addition, extensive coding changes were made to reflect the Policy's covered indications; as such, the following codes have been removed from this policy: D80.3, T86.19, T86.298, T86.39, T86.49, T86.818, T86.898.
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Revisions From MA08.009b:

03/01/2016	This policy was updated to communicate the following changes: New indications with Policy and Dosing criteria were added, per Novitas LCD L32712: Erythema multiforme major (Stevens-Johnson syndrome and toxic epidermal necrolysis), Scleromyxedema, and Systemic Lupus Erythematosus (SLE). Product and Dosing criteria were added/updated per FDA labeling or peer-reviewed literature: Hizentra (SCIG), HyQvia (SCIG), Multiple sclerosis, CLL, and pediatric individuals with dermatomyositis or polymyositis. Noncovered positions were added for the following indications, per Novitas LCD L35093: Immune Modulation of highly sensitized individuals prior to transplantation and the Treatment of inclusion body myositis. Policy criteria were updated for the following indications, per Novitas LCD L32712: Multifocal Motor Neuropathy, Dermatomyositis and Polymyositis. Policy and dosing criteria were updated for the following indications, per Novitas LCD L32712: Chronic refractory ITP and Myasthenia Gravis Syndrome. Antibody-mediated rejection (AMR) was removed as a covered indication. Policy was updated for Vivaglobin, a SCIG product, that is no longer available; therefore, so longer eligible for reimbursement. The coding table has been updated to reflect above criteria. ICD-9 codes were removed from this policy since as of 10/01/2015, ICD-10 codes are effective. SCIG information was added to the Description Section. The criteria for required documentation that must be kept regarding DME and Home Infusion related supplies was updated.
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Revisions From MA08.009a:

01/01/2016	This policy has been identified for the HCPCS code update, effective 01/01/2016. The following HCPCS code has been added to this policy, as Medically Necessary: J1575 Injection, immune globulin/hyaluronidase, (hyqvia), 100 mg immunoglobulin
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Revisions From MA08.009:

01/01/2015	This is a new policy.
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Version Effective Date:

09/18/2026

Version Issued Date:

09/18/2026

Version Reissued Date:

N/A