Medical Policy Bulletin

Title:
Immune Globulin Intravenous (IVIG), Subcutaneous (SCIG)
Policy #:
MA08.009w

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
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Autoimmune Hemolytic Anemia
Evans Syndrome
Systemic Capillary Leak Syndrome (SCLS)

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Antibody-Mediated Rejection (AMR)

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Hematopoietic Stem Cell Transplant (HSCT)
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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 CIPD 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 corticosteriods 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 ritiximab 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 ≤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 (Tepezza).

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 (CIDP)

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

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 medial 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)

Immune Globulin IV (IVIG), SQ (SCIG).docx

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), nonlyophilized, (e.g., liquid), 500 mg

The following code is used to represent HyQvia:

J1575 Injection, immune globulin/hyaluronidase, 100 mg immuneglobulin

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

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), intravenous, non-lyophilized, (e.g. liquid), 500 mg
J1572	Injection, immune globulin, (Flebogamma/Flebogamma Dif), intravenous, non-lyophilized (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 VIVAGLOBIN WHICH IS NO LONGER MANUFACTURED AND HAS BEEN WITHDRAWN FROM THE MARKET:

J1562	Injection, immune globulin (Vivaglobin), 100 mg

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

Policy History

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

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: 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

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.

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)

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. • *Local Coverage Article (LCA)* A56786 - Billing and Coding: Intravenous Immune Globulin (IVIG). Effective date 07/01/2023. • *Local Coverage Determination (LCD)* L35093 - Intravenous Immune Globulin (IVIG). [Novitas Solutions Web site]. Original 10/01/2015, Revised 02/05/2023. 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 **I**CD-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

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

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)

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 (cutaquig), 100 mg

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 baterial infection: timeframe of dosing expanded from every 28 days to every 2-4 weeks. • CIDP policy critieria 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 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

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

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

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.

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

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: o Benign mucous membrane pemphigoid, with or without mention of ocular movement, due to Centers for Medicare and Medicaid Services (CMS) NCD 250.3. o Erythema multiforme major, due to terminology change

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

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.

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 an as example of prior therapies. The following CPT codes were removed from the policy: 90283, 90284 **Attachment B: Dosage and Frequency** **SCIG Section:** 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. **IVIG Section:** 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) Guillain-Barre syndrome Myasthenia gravis syndrome

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.

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

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 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

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.

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.

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

Revisions From MA08.009:

01/01/2015	This is a new policy.

Version Effective Date:
06/13/2025
Version Issued Date:
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Version Reissued Date:
N/A