Local Coverage Determination (LCD): Immune Globulin Intravenous (IVIg) (L34074)

Links in PDF documents are not guaranteed to work. To follow a web link, please use the MCD Website.

Contractor Information

Contractor Name	Contract Type	Contract Number	Jurisdiction	State(s)
Noridian Healthcare Solutions, LLC	A and B MAC	02101 - MAC A	J - F	Alaska
Noridian Healthcare Solutions, LLC	A and B MAC	02102 - MAC B	J - F	Alaska
Noridian Healthcare Solutions, LLC	A and B MAC	02201 - MAC A	J - F	Idaho
Noridian Healthcare Solutions, LLC	A and B MAC	02202 - MAC B	J - F	Idaho
Noridian Healthcare Solutions, LLC	A and B MAC	02301 - MAC A	J - F	Oregon
Noridian Healthcare Solutions, LLC	A and B MAC	02302 - MAC B	J - F	Oregon
Noridian Healthcare Solutions, LLC	A and B MAC	02401 - MAC A	J - F	Washington
Noridian Healthcare Solutions, LLC	A and B MAC	02402 - MAC B	J - F	Washington
Noridian Healthcare Solutions, LLC	A and B MAC	03101 - MAC A	J - F	Arizona
Noridian Healthcare Solutions, LLC	A and B MAC	03102 - MAC B	J - F	Arizona
Noridian Healthcare Solutions, LLC	A and B MAC	03201 - MAC A	J - F	Montana
Noridian Healthcare Solutions, LLC	A and B MAC	03202 - MAC B	J - F	Montana
Noridian Healthcare Solutions, LLC	A and B MAC	03301 - MAC A	J - F	North Dakota
Noridian Healthcare Solutions, LLC	A and B MAC	03302 - MAC B	J - F	North Dakota
Noridian Healthcare Solutions, LLC	A and B MAC	03401 - MAC A	J - F	South Dakota
Noridian Healthcare Solutions, LLC	A and B MAC	03402 - MAC B	J - F	South Dakota
Noridian Healthcare Solutions, LLC	A and B MAC	03501 - MAC A	J - F	Utah
Noridian Healthcare Solutions, LLC	A and B MAC	03502 - MAC B	J - F	Utah
Noridian Healthcare Solutions, LLC	A and B MAC	03601 - MAC A	J - F	Wyoming
Noridian Healthcare Solutions, LLC	A and B MAC	03602 - MAC B	J - F	Wyoming

Back to Top

LCD Information

Document Information

Original Effective Date
For services performed on or after 10/01/2015

Revision Effective Date

For services performed on or after 07/01/2018

Revision Ending Date
N/A

LCD ID
L34074

Retirement Date
N/A

Original ICD-9 LCD ID
L32846

Notice Period Start Date
09/21/2015

Notice Period End Date
11/06/2015

LCD Title
Immune Globulin Intravenous (IVIg)

Proposed LCD in Comment Period
N/A

Source Proposed LCD
N/A

CMS National Coverage Policy
Title XVIII of the Social security Act; Section 1862(a)(1)(A) section allows coverage and payment for only those services that are considered to be reasonable and necessary.

Title XVIII of the Social Security Act; Section 1833(e). This section prohibits Medicare payment for any claim, which lacks the necessary information to process the claim.

CMS Manual, Pub.100-2, Chapter 15, section 50.4 and section 50.4.2. This section addresses coverage of drugs and biologicals.

CMS Manual, Pub.100-3, Chapter 1, Section 250.3. This section describes coverage for IVIg for treatment of Autoimmune Mucocutaneous Blistering Diseases.

"*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.*

Effective October 1, 2002, IVIg is covered for the treatment of biopsy-proven: (1) Pemphigus Vulgaris, (2) Pemphigus Foliaceus, (3) Bullous Pemphigoid, (4) Mucous Membrane Pemphigoid (a.k.a., Cicatricial Pemphigoid), and (5) Epidermolysis Bullosa Acquisita for the following patient subpopulations:

• Patients who have failed conventional therapy. Contractors have the discretion to define what constitutes failure of conventional therapy;
• Patients in whom conventional therapy is otherwise contraindicated. Contractors have the discretion to define what constitutes contraindications to conventional therapy; or
• Patients with rapidly progressive disease in whom a clinical response could not be affected 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. Contractors have the discretion to decide what constitutes short-term therapy."

See attached article for ICD-10 diagnosis codes.

CMS Manual, Pub. 100-2, Chapter 15, Section 50.6. This section describes coverage of IVIg for the treatment of Primary Autoimmune Deficiency Disease in the home.
"Beginning for dates of service on or after January 1, 2004, The Medicare Prescription Drug, Improvement, and Modernization Act of 2003 provides coverage of intravenous immune globulin (IVIg) for the treatment of primary immune deficiency diseases (ICD-10 diagnosis codes D80.0, D80.5, D81.0, D81.1, D81.2, D81.6, D81.7, D81.89, D81.9, D82.0, D83.0, D83.2, D83.8, D83.9) in the home. The Act defines "intravenous immune globulin" as an approved pooled plasma derivative for the treatment of primary immune deficiency disease. It is covered under this benefit when the patient has a diagnosed primary immune deficiency disease, it is administered in the home of a patient with a diagnosed primary immune deficiency disease, and the physician determines that administration of the derivative in the patient's home is medically appropriate. The benefit does not include coverage for items or services related to the administration of the derivative. For coverage of IVIg under this benefit, it is not necessary for the derivative to be administered through a piece of durable medical equipment."

Coverage Guidance
Coverage Indications, Limitations, and/or Medical Necessity

Note: Providers should seek information related to National Coverage Determinations (NCD) and other Centers for Medicare & Medicaid Services (CMS) instructions in CMS Manuals. This LCD only pertains to the contractor's discretionary coverage related to this drug.

IVIg is a solution of human immunoglobulins specifically prepared for intravenous infusion. Immunoglobulin contains a broad range of antibodies that specifically act against bacterial and viral antigens.

There may be acceptable off-label uses for IVIg in rare patient populations or in rare individual patient clinical scenarios which are not covered by this LCD. In such instances, a request for an individual patient consideration by the Medical Director should accompany the appeal of any denied claim.

There are several off-label uses for IVIg, especially in neurological disorders. There is good scientific evidence that supports use in a few of the disorders; in others, however, the evidence is either poor or absent. This policy addresses the off-label uses of IVIg in certain neurological conditions, and idiopathic thrombocytopenic purpura (ITP) in pregnancy. It also clarifies the conditions under which certain FDA-approved uses may be covered. This policy does not address the use of IVIg in any condition covered by a National Coverage Determination (NCD) or CMS manual instruction. (See attached article)

Idiopathic Thrombocytopenic Purpura (ITP) in Pregnancy:

Pregnant women with this disease are at risk for delivering thrombocytopenic infants. Protection of the fetus becomes an important consideration in management of a pregnant woman with immune thrombocytopenic purpura. IVIg may be recommended in the following:

1. Pregnant women who have previously delivered infants with autoimmune thrombocytopenia;
2. Pregnant women who have platelet counts less than 75,000/mm3 during the current pregnancy; or
3. Pregnant women with past history of splenectomy

In the presence of one of the above indications, the use of IVIg may be covered if one of the following situations is present:

• Failure of or contraindications to other therapy; and/or

• Rapidly progressive form of the disease;

All the conditions listed below (see Neurological Disorders) for Medicare coverage are met.

Neurological Disorders:
The use of IVIg in some neurological conditions has been associated with demonstrable clinical benefit. Studies with acceptable methodological bases have shown that IVIg may halt and/or reverse disease progression in Myasthenia Gravis, Guillain-Barre Syndrome and Chronic Inflammatory Demyelinating Neuropathy (CIDP). In a few neurological conditions, such as Polymyositis, Multiple Myeloma, Multifocal Motor Neuropathy (MMN), Dermatomyositis and Lambert-Eaton myasthenic syndrome, IVIg may be of benefit.

Medicare may provide coverage for the use of IVIg use in the above disease conditions if the following requirements are met.

For Guillain-Barre, Myasthenia Gravis, Acute or Chronic Inflammatory Demyelinating Neuropathy (see CIDP below for additional criteria), Dermatomyositis, and Relapsing-Remitting Multiple Sclerosis (MS), the use of IVIg may be covered if one of the following scenarios is present:

• Failure of or contraindications to other therapy (absolute requirement for Dermatomyositis and MS); and/or

• Rapidly progressive form of the disease.

The diagnosis of the disorder must be reasonably certain, based on a thorough history and examination as well as, when necessary, electromyography (EMG), spinal fluid tests, serum tests and biopsy findings.

The clinical record **must document** the medical necessity to initiate IVIg therapy, and the ongoing need as long as treatment continues. The reasons for prescribing IVIg must be clear and include all required information. For example, previous treatment failures must be recorded.

Once treatment is initiated, documentation of progress must be meticulous. If there is initial improvement and continued treatment is necessary, then some type of quantitative assessment to monitor and document the progress is required. Quantitative monitoring may include any accepted metric assessment such as MRC scale and activities of daily living (ADL) measurements. Changes in these measures must be clearly documented. Subjective or experiential improvement alone is insufficient to either continue IVIg or to expect coverage.

Clinical monitoring takes precedence over laboratory monitoring. If significant clinical improvement is evident, then laboratory monitoring, solely to guide IVIg therapy, is not medically necessary.

When improvement has occurred, attempts to decrease/wean the dosage must be made and documented. Following dosage reduction, if improvement is sustained, an attempt to discontinue IVIg must be made. If documentable improvement does not occur with IVIg administration, then infusions should not continue.

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) and its variants (multifocal motor neuropathy, multifocal acquired demyelinating polyneuropathy, multifocal motor neuropathy, pure sensory CIDP).

CIDP is an autoimmune disorder caused by an attack on peripheral nervous system myelin. Clinically CIDP follows *a subacute onset of weakness and/or sensory loss, evolving progressively, or in a stepwise fashion, over several months. Reflexes are usually decreased or absent. Electrodiagnostic testing (EDX) reveals the classic features of demyelination, with prolonged distal latencies, conduction slowing, prolonged F-waves, conduction block, and temporal dispersion in most cases. Most patients have an elevated spinal fluid protein level.* (Jonathan S. Katz, MD, Dept. of Neurology, Stanford University)

Chronic progressive painful peripheral sensory neuropathy, which is common with diabetes mellitus or toxins, may eventually show demyelinating features on electrodiagnostic testing. Typically, these cases have progressed for more than one year prior to electrodiagnostic testing. Many patients with CIDP are not seen until several years into their illness.

In patients with sensory or sensorimotor polyneuropathies, when a CIDP diagnosis is uncertain, a response

to a therapeutic trial of prednisone (e.g. 30-60 mg/d or perhaps 50-100 mg/d for 2-4 months with a taper) should be helpful to increase the specificity of the diagnosis in order to help assure that IVIg will be effective. In the absence of other supporting information, a subjective response to a therapeutic trial of IVIg is not sufficient to validate the diagnosis of CIDP. The principal goal of the treatment is to improve motor function in most patients.

If a diagnosis of multifocal motor neuropathy is suspected, a trial of IVIg is recommended since this condition does not respond to prednisone.

Specific diagnostic criteria for CIDP should include:

• **In typical CIDP**, symmetrical muscle weakness affects proximal and distal muscles of all four limbs. Sensory loss may affect the distal limbs and usually involves large fiber modalities. The clinical evolution tends to be gradually progressive, evolving over periods of more than 8 weeks although patients typically present to clinicians within 6 months of onset. Decreased or absent reflexes in affected nerve distributions occur in nearly all CIDP presentations, and develop during the acute phase typically within 8 weeks of symptom onset. The patient should have a neurologic function assessment score of at least 3 or greater on the Rankin Scale at the time of initial therapy. However, IVIg can be used in patients with rapidly worsening weakness regardless of the Rankin score.

• **A multifocal variant of CIDP** (multifocal acquired demyelinating sensory and motor neuropathy or MADSAM) leads to sensory and motor dysfunction in multiple individual nerve distributions (for example, ulnar or median). Weakness may affect the upper or lower limbs, but it most commonly affects distal musculature and is more common in the hands. Progression tends to be step-wise with episodes of weakness compiling over time to cause gradually increasing debility.

• **Multifocal motor neuropathy (MMN)** is a purely motor syndrome that tends to affect the hands. Like MADSAM, the weakness affects the distribution of individual nerves and tends to progress in a step-wise fashion over time. Patients may have subjective sensory complaints but objective sensory findings are not present. The diagnosis is generally made using motor and sensory nerve conduction studies. MMN responds to IVIg but not to Prednisone. Therefore, Prednisone is never indicated in this condition.

• Occasionally, a patient with **CIDP** may have **only sensory symptoms**. The sensory loss may affect the upper or lower limbs and tends to be relatively symmetrical. Like more common sensory-motor CIDP presentations, patients typically seek medical attention within 6-9 months from onset. The sensory loss may begin relatively acutely and progresses in a stepwise or gradual fashion. The sensory distribution is usually not simply limited to the feet or in a stocking distribution, but takes on unusual patterns involving the trunk, arms, or proximal legs. The condition is rare compared with the relatively common purely sensory neuropathies such as distal diabetic, toxic, alcoholic, and idiopathic neuropathies. Pure sensory CIDP also must be distinguished from distal demyelinating neuropathies associated with an IgM paraprotein, which is not responsive to IVIg or prednisone.

• Laboratory evidence of CIDP includes:
o Conduction block at sites not prone to nerve compression.
o Motor nerves characteristically show segmental conduction slowing and increased distal latencies consistent with a demyelinating polyneuropathy. This is present in typical CIDP, MADSAM, MMN, and purely sensory CIDP.
o Conduction slowing from a demyelinating neuropathy should be distinguished from conduction slowing secondary to moderate to severe axonal loss.
o Cytoalbuminologic dissociation in more than 90% of cases.

• Serum tests may show:
o An IgG monoclonal gammopathy (e.g. on immunofixation electrophoresis); however, an IgM monoclonal gammopathy places the diagnosis in question.
o An elevation of a specific antibody to GM-1 increases the likelihood of MMN. Antibodies to myelin associated glycoprotein (MAG) or sulfatide may occur in patients with demyelinating neuropathies besides CIDP and place the diagnosis in question.

• No other explanation for the diagnosis, such as
o HIV disease;
o Distally predominant diabetic neuropathy;
o Diabetic amyotrophy;
o Diabetic cachectic neuropathy;

o Distal acquired demyelinating symmetric neuropathy with an IgM paraprotein; or

• No evidence of another treatable cause of the polyneuropathy;

• No evidence of hereditary demyelinating neuropathy.

Special consultation recommendations for IVIg use for CIDP, CIDP variants, and MMN:

• Before beginning the initial treatment (i.e. the induction dose) for CIDP, or, for patients currently on treatment for CIDP within 3 months of the effective date of this policy, a consultation is expected from a neurologist or rheumatologist who is an expert in the field of CIDP. This will help validate the diagnosis is correct and the IVIg treatment is reasonable and necessary. The consultation should include a comprehensive history and examination as defined in the CPT book, validate the diagnosis of CIDP, and clarify the need for IVIg treatment. The consultation should set forth the recommended treatment regimen, appropriate measures of therapeutic benefit, and any recommendation for follow-up consultation.

• If the indication for IVIg treatment is principally for pain control in a patient with presumed CIDP predominantly affecting the sensory nerves, before beginning the initial treatment (i.e. the induction dose), the patient should have shown a measurable response to a therapeutic trial of prednisone. In addition, the consultation from a neurologist or rheumatologist who is an expert in the field of CIDP is expected. This consultation should help validate the need for IVIg treatment for pain control as opposed to other pain treatment options that do not include IVIg.

IVIg for CIDP following the initial treatment regimen:

• Once treatment is initiated, the benefit of treatment must be measured. Quantitative monitoring may use any accepted metric as MRC scale and activities of daily living (ADL) measurements.

• Subsequent treatment with IVIg will be covered only when the patient demonstrates significant improvement in clinical condition and, when relevant, a reduction in the level of sensory loss. For long-term treatment of stable patients, the dose must be periodically reduced or withdrawn, and the effects measured, in order to validate continued use.

There is no reimbursement for the use of IVIg in the treatment of the following neurological disorders: epilepsy, Amyotrophic Lateral Sclerosis (ALS), paraneoplastic neurological syndromes, undiagnosed neuropathy or weakness and malignancies with no casual link to coexisting neurological dysfunctions.

The use of IVIg should be reserved for patients with serious defects of antibody function. The goal is to provide immune globulin to those who lack it. The following are certain FDA approved indications for IVIg, which are covered by Noridian.

Acute ITP:

• Management of acute bleeding due to severe thrombocytopenia (platelet counts less than 30,000/mm3);

• To increase platelet counts prior to invasive major surgical procedures (splenectomy); or

• In patients with severe thrombocytopenia (platelet counts less than 20,000/mm3) considered to be at risk for intracerebral hemorrhage.

Chronic Refractory ITP:

First line treatment

o Pediatric ITP;
o In combination with steroids if rapid platelet response justified or to avoid splenectomy; or
o Contraindications to steroids

Second line treatment

o Following treatment with corticosteroids with splenectomy; or
o Platelet counts persistently at or below 20,000/mm 3.

Symptomatic Human Immunodeficiency Virus (HIV):

Indications for intravenous immunoglobulin would include:

1. Patients less than 13 years of age;
2. Entry CD4+ lymphocyte counts greater than or equal to 200/mm3; and
3. Clinically symptomatic or asymptomatic, but immunologically abnormal

Other Disorders:

a. Chronic Lymphocytic Leukemia with associated hypogammaglobulinemia. To initiate IVIg for this disease, the IgG level should be less than 600 mg/dl or there should be evidence of specific antibody deficiency and the presence of repeated bacterial infections.
b. Bone Marrow/Stem Cell Transplantation
o Transplantation must have been for a Medicare covered indication;
o Patients 20 years of age or older;
o Cytomegalovirus (CMV) seropositive before transplantation; or
o Cytomegalovirus (CMV) seronegative, had seropositive marrow donors, and were undergoing allogenic transplantation for hematologic neoplasms.
c. Kawasaki Disease (Mucocutaneous Lymph Node Syndrome)
d. Transplantation rejection, kidney, stem cell or heart, antibody-mediated.
e. Desensitization for a pre-kidney or pre-heart transplantation.

f. Autoimmune retinopathy (limited to three months unless there is improvement on therapy).

Immunoglobulin Deficiencies:
The principal clinical manifestations of humoral immunodeficiency are recurrent bacterial infections of the upper and lower respiratory tract. Immunoglobulin replacement therapy is required in patients with certain immunodeficiency diseases characterized by absent or deficient antibody production and, in most cases, recurrent or unusually severe infection. Individuals with agammaglobulinemia or hypogammaglobulin need lifelong antibody replacement, however, Ig replacement is unlikely to be necessary until the IgG levels fall below 200 mg/dl or if the individual has a well documented severe polysaccharide nonresponsiveness and evidence of recurrent infections with a proven requirement for antibiotic therapy.

Summary of Evidence

NA

Analysis of Evidence
(Rationale for Determination)

NA

Back to Top

Coding Information

Bill Type Codes:

Contractors may specify Bill Types to help providers identify those Bill Types typically used to report this service. Absence of a Bill Type does not guarantee that the policy does not apply to that Bill Type. Complete absence of all Bill Types indicates that coverage is not influenced by Bill Type and the policy should be assumed to apply equally to all claims.

012x Hospital Inpatient (Medicare Part B only)
013x Hospital Outpatient
022x Skilled Nursing - Inpatient (Medicare Part B only)
023x Skilled Nursing - Outpatient
085x Critical Access Hospital

Revenue Codes:

Contractors may specify Revenue Codes to help providers identify those Revenue Codes typically used to report this service. In most instances Revenue Codes are purely advisory. Unless specified in the policy, services reported under other Revenue Codes are equally subject to this coverage determination. Complete absence of all Revenue Codes indicates that coverage is not influenced by Revenue Code and the policy should be assumed to apply equally to all Revenue Codes.

The Section titled “Does the 'CPT 30% Rule' Apply?” needs clarification. This rule comes from the AMA (American Medical Association), the organization that holds the copyrights for all CPT codes. The rule states that if, in a given section (e.g., **surgery**) or subsection (e.g., surgery, **integumentary**) of CPT Manual, more than 30% of the codes are listed in the LCD, then the short descriptors must be used rather than the long descriptors found in the CPT Manual.
0636 Pharmacy - Drugs Requiring Detailed Coding

CPT/HCPCS Codes
Group 1 Paragraph: N/A

Group 1 Codes:

J1459	INJECTION, IMMUNE GLOBULIN (PRIVIGEN), INTRAVENOUS, NON-LYOPHILIZED (E.G., LIQUID), 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-C/GAMMAKED), NON-LYOPHILIZED (E.G., LIQUID), 500 MG
J1566	INJECTION, IMMUNE GLOBULIN, INTRAVENOUS, LYOPHILIZED (E.G., POWDER), NOT OTHERWISE SPECIFIED, 500 MG
J1568	INJECTION, IMMUNE GLOBULIN, (OCTAGAM), INTRAVENOUS, NON-LYOPHILIZED (E.G., LIQUID), 500 MG
J1569	INJECTION, IMMUNE GLOBULIN, (GAMMAGARD LIQUID), NON-LYOPHILIZED, (E.G., LIQUID), 500 MG
J1572	INJECTION, IMMUNE GLOBULIN, (FLEBOGAMMA/FLEBOGAMMA DIF), INTRAVENOUS, NON-LYOPHILIZED (E.G., LIQUID), 500 MG
J1599	INJECTION, IMMUNE GLOBULIN, INTRAVENOUS, NON-LYOPHILIZED (E.G., LIQUID), NOT OTHERWISE SPECIFIED, 500 MG

ICD-10 Codes that Support Medical Necessity
Group 1 Paragraph: N/A

Group 1 Codes:

ICD-10 Codes	Description
B20*	Human immunodeficiency virus [HIV] disease
B25.0	Cytomegaloviral pneumonitis
B25.1	Cytomegaloviral hepatitis
B25.2	Cytomegaloviral pancreatitis

ICD-10 Codes	Description
B25.8	Other cytomegaloviral diseases
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
C91.11	Chronic lymphocytic leukemia of B-cell type in remission
C91.12	Chronic lymphocytic leukemia of B-cell type in relapse
D59.0	Drug-induced autoimmune hemolytic anemia
D59.1	Other autoimmune hemolytic anemias
D61.01*	Constitutional (pure) red blood cell aplasia
D69.3	Immune thrombocytopenic purpura
D69.42	Congenital and hereditary thrombocytopenia purpura
D69.49	Other primary thrombocytopenia
D80.0	Hereditary hypogammaglobulinemia
D80.1	Nonfamilial hypogammaglobulinemia
D80.5	Immunodeficiency with increased immunoglobulin M [IgM]
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
D81.6	Major histocompatibility complex class I deficiency
D81.7	Major histocompatibility complex class II deficiency
D81.89	Other combined immunodeficiencies
D81.9	Combined immunodeficiency, unspecified
D82.0	Wiskott-Aldrich syndrome
D83.0	Common variable immunodeficiency with predominant abnormalities of B-cell numbers and function
D83.2	Common variable immunodeficiency with autoantibodies to B- or T-cells
D83.8	Other common variable immunodeficiencies
D83.9	Common variable immunodeficiency, unspecified
G25.82	Stiff-man syndrome
G35	Multiple sclerosis
G60.3	Idiopathic progressive neuropathy
G61.0	Guillain-Barre syndrome
G61.81*	Chronic inflammatory demyelinating polyneuritis
G61.82	Multifocal motor neuropathy
G65.0	Sequelae of Guillain-Barre syndrome
G70.00	Myasthenia gravis without (acute) exacerbation
G70.01	Myasthenia gravis with (acute) exacerbation
G70.81	Lambert-Eaton syndrome in disease classified elsewhere
G73.1	Lambert-Eaton syndrome in neoplastic disease
G73.3	Myasthenic syndromes in other diseases classified elsewhere
M30.3	Mucocutaneous lymph node syndrome [Kawasaki]
M31.1	Thrombotic microangiopathy
M33.00	Juvenile dermatomyositis, organ involvement unspecified
M33.01	Juvenile dermatomyositis with respiratory involvement
M33.02	Juvenile dermatomyositis with 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
M33.19	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

ICD-10 Codes	Description
M34.83	Systemic sclerosis with polyneuropathy
M36.0	Dermato(poly)myositis in neoplastic disease
T86.01	Bone marrow transplant rejection
T86.02	Bone marrow transplant failure
T86.09	Other complications of bone marrow transplant
T86.11	Kidney transplant rejection
T86.12	Kidney transplant failure
T86.19	Other complication of kidney transplant
T86.21	Heart transplant rejection
T86.22	Heart transplant failure
T86.298	Other complications of heart transplant
T86.5	Complications of stem cell transplant
Z48.21	Encounter for aftercare following heart transplant
Z48.22	Encounter for aftercare following kidney transplant
Z76.82	Awaiting organ transplant status
Z86.19	Personal history of other infectious and parasitic diseases
Z87.01	Personal history of pneumonia (recurrent)
Z94.0	Kidney transplant status
Z94.1	Heart transplant status
Z94.81	Bone marrow transplant status
Z94.84	Stem cells transplant status

Group 1 Medical Necessity ICD-10 Codes Asterisk Explanation: *B20 is only payable for children under 13 years of age.
***D61.01 is only to be used when patient has failed all first line therapies.**
***G61.81 is not payable when associated with diabetes mellitus, dysproteinemias, renal failure, or malnutrition.**

ICD-10 Codes that DO NOT Support Medical Necessity
Group 1 Paragraph: Any diagnosis codes other than those listed in the covered ICD-10-CM codes of this policy and those in the attached article will be denied as not reasonable and necessary and will be denied provider liable unless a non-coverage notice has been issued to the beneficiary prior to the test. Screening diagnoses will be denied as routine services.

Group 1 Codes: N/A

ICD-10 Additional Information Back to Top

General Information

Associated Information
Medical record documentation maintained by the treating physician must clearly document the medical necessity to initiate IVIg therapy and the continued need thereof. Required documentation of medical necessity could include:
• History and physical;
• Office/progress note(s);
• Test results with written interpretation; and An accurate weight in kilograms should be documented prior to the infusion since the dosage is based mg/kg/dosage.

The Section titled "Does the 'CPT 30% Rule' apply?" needs clarification. This rule comes from the AMA (American Medical Association), the organization that holds the copyrights for all CPT codes. The rule states that if, in a given section (e.g., ,b>surgery) or subsection (e.g., surgery, integumentary) of the CPT Manual, more than 30% of the codes are listed in the LCD, then the short descriptors must be used rather than the long descriptors found in the CPT Manual.

Sources of Information

N/A

Bibliography

1. Anderson D, Ali K, Blanchette V, Brouwers M, Couban S, Radmoor P, et al. Guidelines on the use of intravenous immune globulin for hematologic conditions. Transfus Med Rev. 2007;21(2 Suppl 1):S9-56.
2. Archdeacon P, Chan M, Neuland C, Velidedeoglu E, Meyer J, Tracy L, et al. Summary of FDA antibody-mediated rejection workshop. Am J Transplant. 2011;11(5):896-906.
3. Bonilla FA, Khan DA, Ballas ZK, Chinen J, Frank MM, Hsu JT, et al. Practice parameter for the diagnosis and management of primary immunodeficiency. J Allergy Clin Immunol. 2015;136(5):1186-205 e1-78.
4. Buckley RH, Schiff RI. The use of intravenous immune globulin in immunodeficiency diseases. N Engl J Med. 1991;325(2):110-7.
5. Dalakas MC. Controlled studies with high-dose intravenous immunoglobulin in the treatment of dermatomyositis, inclusion body myositis, and polymyositis. Neurology. 1998;51(6 Suppl 5):S37-45.
6. Dalakas MC. Mechanism of action of intravenous immunoglobulin and therapeutic considerations in the treatment of autoimmune neurologic diseases. Neurology. 1998;51(6 Suppl 5):S2-8.
7. Dwyer JM. Manipulating the immune system with immune globulin. N Engl J Med. 1992;326(2):107-16.
8. Glotz D, Rostaing L, Merville P, Squifflet JP, Lebranchu Y. Intravenous Immunoglobulins in the Prevention of Rejection of a Second or Third Kidney Graft. Transplant Proc. 2018;50(1):70-1.
9. Grewal DS, Fishman GA, Jampol LM. Autoimmune retinopathy and antiretinal antibodies: a review. Retina. 2014;34(5):827-45.
10. John R, Lietz K, Burke E, Ankersmit J, Mancini D, Suciu-Foca N, et al. Intravenous immunoglobulin reduces anti-HLA alloreactivity and shortens waiting time to cardiac transplantation in highly sensitized left ventricular assist device recipients. Circulation. 1999;100(19 Suppl):II229-35.
11. Jordan SC, Tyan D, Stablein D, McIntosh M, Rose S, Vo A, et al. Evaluation of intravenous immunoglobulin as an agent to lower allosensitization and improve transplantation in highly sensitized adult patients with end-stage renal disease: report of the NIH IG02 trial. J Am Soc Nephrol. 2004;15(12):3256-62.
12. Kahwaji J, Jordan SC, Najjar R, Wongsaroj P, Choi J, Peng A, et al. Six-year outcomes in broadly HLA-sensitized living donor transplant recipients desensitized with intravenous immunoglobulin and rituximab. Transpl Int. 2016;29(12):1276-85.
13. Keller T, McGrath K, Newland A, Gatenby P, Cobcroft R, Gibson J. Indications for use of intravenous immunoglobulin. Recommendations of the Australasian Society of Blood Transfusion consensus symposium. Med J Aust. 1993;159(3):204-6.
14. Kim IK, Vo A, Jordan SC. Transplantation in Highly HLA Sensitized Patients: Challenges and Solutions. Transplantation Research and Risk Management. 2014;6:99-106.
15. Kittleson, Michelle Maya M.D., FACC; Kobashigawa, Jon A M.D., FACC Management of the Highly Sensitized Patient Awaiting Heart Transplant Expert Analysis. American College of Cardiology. Jan 08, 2015.
16. Kobashigawa J, Mehra M, West L, Kerman R, George J, Rose M, et al. Report from a consensus conference on the sensitized patient awaiting heart transplantation. J Heart Lung Transplant. 2009;28(3):213-25.
17. Lee KW, Park JB, Oh DK, Na BG, Choi JY, Cho WT, et al. Short-Term Outcomes of ABO-Incompatible Living Donor Kidney Transplantation With Uniform Protocol: Significance of Baseline Anti-ABO Titer. Transplant Proc. 2016;48(3):820-6.
18. Leech SH, Lopez-Cepero M, LeFor WM, DiChiara L, Weston M, Furukawa S, et al. Management of the sensitized cardiac recipient: the use of plasmapheresis and intravenous immunoglobulin. Clin Transplant. 2006;20(4):476-84.
19. Luque Y, Anglicheau D, Rabant M, El Karoui K, Jamme M, Aubert O, et al. Renal safety of high-dose, sucrose-free intravenous immunoglobulin in kidney transplant recipients: an observational study. Transpl Int. 2016;29(11):1205-15.
20. Mofenson LM, Moye J, Jr., Bethel J, Hirschhorn R, Jordan C, Nugent R. Prophylactic intravenous immunoglobulin in HIV-infected children with CD4+ counts of 0.20 x 10(9)/L or more. Effect on viral, opportunistic, and bacterial infections. The National Institute of Child Health and Human Development Intravenous Immunoglobulin Clinical Trial Study Group. JAMA. 1992;268(4):483-8.

21. National Institute of Child H, Human Developments Intravenous Immunoglobulin Study G. Intravenous immune globulin for the prevention of bacterial infections in children with symptomatic human immunodeficiency virus infection. N Engl J Med. 1991;325(2):73-80.
22. NIH consensus conference. Intravenous immunoglobulin. Prevention and treatment of disease. JAMA. 1990;264(24):3189-93.
23. Orange JS, Ballow M, Stiehm ER, Ballas ZK, Chinen J, De La Morena M, et al. Use and interpretation of diagnostic vaccination in primary immunodeficiency: a working group report of the Basic and Clinical Immunology Interest Section of the American Academy of Allergy, Asthma & Immunology. J Allergy Clin Immunol. 2012;130(3 Suppl):S1-24.
24. Orange JS, Grossman WJ, Navickis RJ, Wilkes MM. Impact of trough IgG on pneumonia incidence in primary immunodeficiency: A meta-analysis of clinical studies. Clin Immunol. 2010;137(1):21-30.
25. Perez EE, Orange JS, Bonilla F, Chinen J, Chinn IK, Dorsey M, et al. Update on the use of immunoglobulin in human disease: A review of evidence. J Allergy Clin Immunol. 2017;139(3S):S1-S46.
26. Pisani BA, Mullen GM, Malinowska K, Lawless CE, Mendez J, Silver MA, et al. Plasmapheresis with intravenous immunoglobulin G is effective in patients with elevated panel reactive antibody prior to cardiac transplantation. J Heart Lung Transplant. 1999;18(7):701-6.
27. Ratko TA, Burnett DA, Foulke GE, Matuszewski KA, Sacher RA. Recommendations for off-label use of intravenously administered immunoglobulin preparations. University Hospital Consortium Expert Panel for Off-Label Use of Polyvalent Intravenously Administered Immunoglobulin Preparations. JAMA. 1995;273(23):1865-70.
28. Reinsmoen NL, Lai CH, Vo A, Jordan SC. Evolving paradigms for desensitization in managing broadly HLA sensitized transplant candidates. Discov Med. 2012;13(71):267-73.
29. Shaffer D, Feurer ID, Crowe D, Schaefer H. Early and Sustained Reduction in Donor-Specific Antibodies in Desensitized Living Donor Kidney Transplant Recipients: A 3-Year Prospective Study. Transplant Direct. 2016;2(2):e62.
30. Sharma A, King A, Kumar D, Behnke M, McDougan F, Kimball PM. Perioperative Desensitization Improves Outcomes Among Crossmatch Positive Recipients of Deceased Donor Renal Transplants. Prog Transplant. 2016;26(2):157-61.
31. Subhadra C, Dudek AZ, Rath PP, Lee MS. Improvement in visual fields in a patient with melanoma-associated retinopathy treated with intravenous immunoglobulin. J Neuroophthalmol. 2008;28(1):23-6.
32. Sullivan KM, Kopecky KJ, Jocom J, Fisher L, Buckner CD, Meyers JD, et al. Immunomodulatory and antimicrobial efficacy of intravenous immunoglobulin in bone marrow transplantation. N Engl J Med. 1990;323(11):705-12.
33. Ten RM. Primary immunodeficiencies. Mayo Clin Proc. 1998;73(9):865-72.
34. Toyoda M, Shin BH, Ge S, Mirocha J, Thomas D, Chu M, et al. Impact of Desensitization on Antiviral Immunity in HLA-Sensitized Kidney Transplant Recipients. J Immunol Res. 2017;2017:5672523.
35. Velidedeoglu E, Cavaille-Coll MW, Bala S, Belen OA, Wang Y, Albrecht R. Summary of 2017 Fda Public Workshop: Antibody Mediated Rejection in Kidney Transplantation. Transplantation. 2018.
36. Vo AA, Jordan SC. Benefits, efficacy, cost-effectiveness and infectious complications in transplant patients desensitized with intravenous immunoglobulin and anti-CD20 therapy. Clin Exp Immunol. 2014;178 Suppl 1:48-51.
37. Vo AA, Lukovsky M, Toyoda M, Wang J, Reinsmoen NL, Lai CH, et al. Rituximab and intravenous immune globulin for desensitization during renal transplantation. N Engl J Med. 2008;359(3):242-51.
38. Yu Z, Lennon VA. Mechanism of intravenous immune globulin therapy in antibody-mediated autoimmune diseases. N Engl J Med. 1999;340(3):227-8.

Back to Top

Revision History Information

Revision History Date	Revision History Number	Revision History Explanation	Reason(s) for Change
07/01/2018	R7		

Revision History Date	Revision History Number	Revision History Explanation	Reason(s) for Change
		Coverage for immunodeficiency is clarified and expanded with additional diagnosis codes (Z86.19, Z87.01) due to provider reconsideration request. Revisions were made to d. and e. under Other Disorders in Indications and Limitations of Coverage and the following diagnoses were added: T86.21, T86.22, T86.298, Z48.21, Z86.19, Z87.01, Z94.1. References were updated. 05/24/18 At this time 21st Century Cures Act will apply to new and revised LCDs that restrict coverage which requires comment and notice. This revision is not a restriction to the coverage determination; and, therefore not all the fields included on the LCD are applicable as noted in this policy.	• Creation of Uniform LCDs Within a MAC Jurisdiction
07/17/2017	R6	Indications and Limitations of Coverage, under Other Disorders, the letter e. is added: Desensitization for a pre-kidney transplantation in patients with a panel reactive antibody (PRA) of 80% or below. Use in patients with a PRA of 81% or higher is considered to be experimental/investigational by this Contractor and is therefore not covered. Post transplantation to prevent rejection remains covered without regard to antibody levels. LCD is revised to add Z76.82. Sources of Information is updated to include multiple recently reviewed sources and moved to the Bibliography. 12/11/2017 At this time 21st Century Cures Act will apply to new and revised LCDs that restrict coverage which requires comment and notice. This revision is not a restriction to the coverage determination; and, therefore not all the fields included on the LCD are applicable as noted in this policy.	• Creation of Uniform LCDs With Other MAC Jurisdiction
10/01/2016	R5	LCD is revised to add diagnosis G61.82, effective 10/1/2016 per the 2016/2017 annual ICD-10 update. The JFA (L34092) LCD is retired and is combined into the JFB (L34074) LCD so that both JFA and JFB contract numbers will have the same final MCD LCD number.	• Creation of Uniform LCDs Within a MAC Jurisdiction • Revisions Due To ICD-10-CM Code Changes
11/07/2015	R4	The LCD is revised to add diagnoses M33.00, M33.01, M33.02, M33.09, M33.10, M33.11, M33.12, M33.20, M33.90, M33.91, M33.92 and M33.99 effective 11/07/2015.	• Creation of Uniform LCDs Within a MAC Jurisdiction
11/07/2015	R3	The LCD is revised to add diagnoses G61.0 and G65.0 effective for dates of service on/after 10/1/15.	• Creation of Uniform LCDs Within a MAC Jurisdiction
11/07/2015	R2	This LCD is revised to add ICD-10 D83.9 effective for dates of service on/after 10/1/2015. Due to technical restrictions in the Medicare Coverage Database, the effective date cannot be a date previous to the last revision effective date. In this case that date is 11/7/2015. Noridian will, however, pay diagnosis D83.9 effective 10/1/2015.	• Revisions Due To ICD-10-CM Code Changes
11/07/2015	R1		

Revision History Date	Revision History Number	Revision History Explanation	Reason(s) for Change
		This version of the LCD is the result of ICD-9 draft LCD DL32846 finalizing in ICD-10.	• Creation of Uniform LCDs Within a MAC Jurisdiction

Back to Top

Associated Documents

Attachments N/A

Related Local Coverage Documents Article(s) A54643 - Intravenous Immune Globulin (IVIg)-NCD 250.3 A54662 - Coverage of Intravenous Immune Globulin for Treatment of Primary Immune Deficiency Diseases in the Home – Medicare Benefit Policy Manual, Chapter 15, 50.6 A54647 - Response to Comments: Immune Globulin Intravenous (IVIg)

Related National Coverage Documents NCD(s) 250.3 - Intravenous Immune Globulin for the Treatment of Autoimmune Mucocutaneous Blistering Diseases

Public Version(s) Updated on 05/25/2018 with effective dates 07/01/2018 - N/A Updated on 12/13/2017 with effective dates 07/17/2017 - 06/30/2018 Updated on 11/12/2016 with effective dates 10/01/2016 - 07/16/2017 Some older versions have been archived. Please visit the MCD Archive Site to retrieve them. Back to Top

Keywords

- Immune
- Globulin
- Intravenous
- IVIg
- J1459
- J1556
- J1557
- J1561
- J1566
- J1568
- J1569
- J1572
- J1599

Read the **LCD Disclaimer** Back to Top