INTRAVENOUS IMMUNE GLOBULIN (IVIG) FOR NEUROLOGICAL AND OTHER DISORDERS CHECKLIST
Medicare Local Coverage Determination (LCD) - L34007
Medicare National Coverage Determination (NCD) - 250.3

See a separate checklist for Intravenous Immune Globulin for Immunodeficiency Disorders

LCD-L34007 NCD 250.3	**Patient Name:**	**MR:**

Effective Date: For services performed on or after 08/13/2019

Intravenous Immune Globulin (IVIG) is a solution of human immunoglobulin specifically prepared for intravenous infusion. It contains a broad range of antibodies that specifically act against bacterial and viral antigens. The use of intravenous immune globulin should be reserved for patients with serious defects of antibody function. The goal is to provide immunoglobulin G (IgG) antibodies to those who lack them. Affected patient populations are those with neurological and other disorders **(discussed below)** and immunodeficiency disorders **(see a separate checklist).**

COVERAGE INDICATIONS/LIMITATIONS/SPECIFIC REQUIREMENTS

Intravenous Immune Globulin IS considered medically reasonable and necessary when the applicable criteria below are met:

I. Neurological Disorders (A & B)

A. General Requirements

□ Disease status: evidence of rapid progression or relapse or other forms of treatment failed.
□ Diagnosis: unequivocal and supported by clinical (history, quantitative exam), electrophysiological motor-sensory nerve conductions, electromyography (EMG), cerebrospinal fluid (CSF), and biopsy (muscle-nerve) data.
□ Treatment progress: meticulously documented including quantitative assessment (e.g., ADL measurements) when treatment continued after initial improvement with parameter changes monitored.
Note: Subjective or experiential improvement alone is insufficient.
□ Treatment reduction/stoppage: when patient improves, evidence of attempts to reduce frequency of administration and/or dosage; when improvement sustained with reduced dosage, evidence of attempts to stop IVIG infusion; when no improvement, IVIG discontinued.
□ Dosing: According to FDA drug label for specific indications, when available. Dosing regimen for off-label indications may be not established.

B. Additional Requirements Specific to Disorder

Guillain-Barre' syndrome:
□ High-dose immunoglobulin given over 5 days

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP):
□ Physician documentation in the medical record substantiates contraindications to steroids or when patient had no response to corticosteroids, as applicable.

Myasthenia Gravis:
□ Diagnosis is confirmed by a positive Tensilon test
□ Treatment results :
- Patients refractory to corticosteroids over a 6 week period or
- unable to successfully taper corticosteroids below moderately high doses or
- develop severe side effects due to steroid therapy, and
- failed at least one immunosuppressive agent (e.g., azathiprime, Methotrexate, cyclosporine, cyclophosphamide).

Polymyositis and Dermatomyositis:
□ Diagnostic studies with elevated creatine phosphokivase (CPK), abnormal electromyography (EMG), and/or abnormal muscle biopsy.
□ Treatment results :
- refractory to corticosteroids over a 6 week period or
- unable to successfully taper corticosteroids below moderately high doses or
- develop severe side effects due to steroid therapy; and
- failed at least one immunosuppressive agent (e.g., azathiprime, Methotrexate, cyclosporine).

□ The need for continuation of IVIG documented and demonstrated by continued decreased muscle strength, elevated CPKs, and/or EMG abnormalities
□ Medical records indicate the reason(s) when corticosteroid or immunosuppressive agents are not tolerated by or contraindicated for the patient.

Autoimmune optic neuropathy:
□ Unresponsive to corticosteroids; refractory to standard treatments

Multiple Sclerosis: N/A

II. Other Disorders

Autoimmune Hemolytic Anemia
□ Patients who have failed to respond to other forms of therapy and/or require rapid cessation of hemolysis due to severe or life threatening manifestations of this condition.
□ A short course (3-5 weeks) treatment.
□ Dosing varies per patient; according to recommended current literature and standard of practice; rational for departure from the standard FDA dosing documented in the medical record.

Autoimmune Neutropenia
□ An absolute neutrophil count < 800mm with recurrent bacterial infections.

Kawasaki Disease (mucocutaneous Lymph Node Syndrome)
□ Diagnosis: fever, at least 5 day-long, and at least 4 out of the following criteria:
- cervical lymphadenopathy, often singular and unilateral
- bilateral conjunctival infection without exudate
- changes in the oropharynx such as fissured lips and strawberry tongue without discrete lesions
- polymorphic exanthem
- changes in the extremities such as edema of the hands/feet and erythema of the palms/soles

□ IVIG treatment in conjunction with aspirin.
□ Dosing: According to FDA drug label for specific FDA indications.

Chronic Lymphocytic Leukemia
□ For the prevention of recurrent bacterial infections and an immunoglobulin G (IgG) level of less than 600 mg/dl in patients with hypogammaglobulinemia associated with B-cell chronic lymphocytic leukemia (CLL) .
□ Dosing: Per FDA drug label for specific FDA indications.

Bone Marrow Transplantation (BMT)
□ To prevent the risk of acute graft-versus-host disease, associated interstitial pneumonia (infectious or idiopathic) and infections (e.g., cytomegalovirus infections [CMV], varicella-zoster virus infection, and recurrent bacterial infection); within 100 days after BMT transplant; for patients ≥ 20 years of age.
□ Dosing: Per FDA drug label for specific FDA indications. No dosing established for off-label indications.
Limitation: not indicated for patients < 20 years of age or for autologous transplants.

Autoimmune Mucocutaneous Blistering Diseases
□ One of the following is biopsy-proven: Pemphigus Vulgaris, or Pemphigus Foliaceus, or Bullous Pemphigoid, or Mucous Membrane Pemphigoid (a.k.a., Cicatricial Pemphigoid), or Epidermolysis Bullosa Acquista
□ For one of the following patient populations:
- who failed conventional therapy • for whom conventional therapy is contraindicated
- with rapidly progressive disease without quick clinical response to conventional agents to supplement until the conventional therapy takes effect.

□ Used only for short-term therapy, and not as a maintenance therapy.
□ Dosing: Per FDA drug label for specific FDA indications. No dosing established for off-label indications.

Stiff-man (stiff-person) syndrome (off-label):
□ Documentation supports the following:
1. Patient is under the care of a physician who is competent in the diagnosis of the syndrome.
2. The current defined criteria for the diagnosis are met.
3. Failed conservative treatment (such as benzodiazepines).
4. Patient's response to therapy after initial treatment (0 and 1 month).
5. Objective response for continued coverage each month or at longer intervals.

□ Dosing: Per FDA drug label for specific FDA indications. No dosing established for off-label indications.

Hypogammaglobulinemia with NNI (non neutropenic infection) induced by certain agents:
□ Documentation supports the following:
1. Recent treatment with rituximab in combination with cytotoxic chemotherapy
2. Laboratory proven hypogammaglobulinemia and an absolute neutrophil count over 1,000.
3. Acute infection requiring hospitalization or lasting > 2 weeks with antibiotics or relapsing after antibiotics end.

□ Dosing: Per FDA drug label for specific FDA indications. No dosing established for off-label indications.

GENERAL DOCUMENTATION REQUIREMENTS

Medical records must include physician's documentation of medical necessity to initiate intra-venous immunoglobulin therapy and the continued need thereof, including but not limited to:

□ History, physical & supporting rationale (within the last 12 months)	□ Physician's orders (with dose, frequency, administration route and duration; dated within 30 days prior date of service) & progress note(s) documenting the necessity for IVIG initiation/continuation.
□ Diagnosis code(s) supporting medical necessity submitted with each claim	
□ Documentation supporting the qualifying diagnoses	□ Medication administration records, and rational for departure from the standard dosing.
□ An accurate weight (in kilograms) documented prior to each infusion for dosage determination	□ Documentation indicating that conventional therapies either failed or are contraindicated
□ A copy of applicable lab and procedure test results	□ Specific documentation indicated in the section above

CODING

HCPCS Codes: J1459, J1555, J1556, J1557, J1561, J1566, J1568, J1569, J1572, J1575

ICD-10 Codes: See LCD L34007 for the list of covered codes

Checklist completed by:

Date: