INTRAVENOUS IMMUNE GLOBULIN (IVIG) FOR IMMUNODEFICIENCY DISORDERS CHECKLIST
Medicare Local Coverage Determination (LCD) - L34007

See a separate checklist for Intravenous Immune Globulin for Neurological and Other Disorders.

LCD-L34007 Billing and Coding Article A56846	**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 immunodeficiency disorders **(discussed below)** and neurological and other disorders **(see a separate checklist).**

COVERAGE INDICATIONS/ LIMITATIONS/SPECIFIC REQUIREMENTS

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

PRIMARY HUMERAL IMMUNODEFICIENCY SYNDROMES:
1. Common Variable Immunodeficiency (CVID)
2. Severe combined immunodeficiency (SCID)
3. X-linked immunodeficiency with hyperimmunoglobulin M (IgM)
4. Wiskott-Aldrich syndrome
5. Congenital Agammaglobulinemia (X-linked agammaglobulinemia)

□ Dosing as follows:
- According to current standards of practice, literature, and FDA drug label for specific indications. CVID dosage regimen is based on clinical response (not standardized).
- When departing from the standard dosing and for serum trough level > 300 mg above the baseline IgG, documentation includes the rational.
- When maintenance doses exceed FDA recommended dosages, documentation reflects inadequate clinical response to FDA recommended dosage.
- IgG trough & functional antibody level testing does not exceed every 3-6 months frequency.

□ For recurrent or chronic bacterial sinusitis, documentation includes:
- diagnostic or highly suggestive findings during the first few days of, not limited to, fever, unilateral purulent nasal discharge, and unilateral facial pain. Radiographic documentation of mucosal thickening of the paranasal sinuses is not considered sufficient documentation as a sole finding.
- substantiation for prolonged/or recurrent antibiotic therapy in the treatment of infections.

Additional Requirements for Common Variable Immunodeficiency (CVID)
Documentation indicates/includes:
□ The initial IgG levels, record of vaccination with Pneumovax, pre-and post-vaccine pneumococcal antibody titers, and statement regarding antibody response to antigens.
□ Lab Results:
- IgG level < 400 mg/dl and lack of response to immunization* OR
- IgG level ≥ 400mg/dl and lack of response to immunization* with documented recurrent or chronic severe infections and antibiotic therapy.

** Lack of ability to produce an antibody response to a protein antigen (e.g., tetanus) or one of the polysaccharide antigens (e.g., Pneumococcal polysaccharide or H. Influenza type B) when two or more immunizations given in the 12 months prior to assessment of antibody response (exception applies when a severe adverse reaction occurred with the last immunization and two given over 12 month or longer interval). An adequate response includes a two to three fold increase in titers in at least 50 percent of the serotypes (4,5,6,12 or 14) tested (exception to Functional Antibody Testing applies to patients diagnosed prior to the introduction of this laboratory technology; low total IgG values to support the diagnosis of B-cell or humeral immune deficiency).*

□ Sinopulmonary disease: pneumonia, bronchiectasis, chronic bronchitis, recurrent bacterial or mycoplasma bronchitis, pneumonitis and episodes of acute bacterial sinusitis.
□ Dual Diagnosis: For hypogammaglobulinemia due to non-neutropenic infection, documentation and coding reflect:
- (D80.1) nonfamilial hypogammaglobulinemia, and
- (Z92.21) personal history of antineoplastic chemotherapy.

Limitation: IVIG is not covered for:
- treatment of viral upper respiratory infections or simple rhinosinusitis
- patients with normal humeral immunity with recurrent infections

ITP in Patients with Human Immunodeficiency Virus (HIV) Disease
□ For one of the following patient populations:
- approaching invasive surgical procedures, e.g., splenectomy, with platelet counts <20,000
- with active bleeding , with platelet counts <30,000
- pregnant women in the third trimester, with platelet counts <10,000
- pregnant women who are bleeding, with platelet counts 10,000-30,000

□ Treatment Duration: 3 to 5 days. *Note: Longer treatments are subject to medical review.*
□ Medical record reflect an actively bleeding and lab results with platelet counts < 30,000.
□ Dual Diagnosis: For patients ≥ 13 years old, documentation and coding reflect:
- primary diagnosis of (D69.6) Thrombocytopenia, unspecified, and
- secondary diagnosis of (B20) Human immunodeficiency virus [HIV] disease.

Idiopathic Thrombocytopenic Purpura (ITP)
□ Dosages according to FDA drug label for each specific indication.
□ For chronic refractory ITP when meeting all of the below criteria:
1. Prior treatment with corticosteroids and splenectomy
2. Duration of illness of greater than six months
3. Age of 10 years or older
4. No concurrent illness/disease explaining thrombocytopenia
5. Persistent platelet counts ≤ 20,000/ul

IVIG may be medically necessary for:
- management of acute bleeding due to severe thrombocytopenia with platelet count < 30,000/ul
- patients with severe thrombocytopenia (platelet count < 20,000/ul), with documented risk for intracerebral or other hemorrhage, including co-morbidities: hypertension, peptic ulcer disease.
- increasing platelet counts prior to invasive surgical procedures, e.g., splenectomy.

Limitation: IVIG is not covered for:
- children with platelet counts > 30,000/mm that are asymptomatic or have only minor purpura
- adults with a platelet count > 50,000/mm

Children with Human Immunodeficiency Virus (HIV) Disease who do not have ITP:
To reduce significant bacterial infections when:
A□ All three primary criteria are met:
1. Age < 13 years; 2. IgG level < 400 mg/dl; 3. Entry CD4+ lymphocyte count is≥ 200/mm3; ***and***

B□ One of the below secondary criteria is met:
- Two or more bacterial infections in a 1-year period despite antibiotic chemoprophylaxis with TMP-SMZ or another active agent.
- In a region with a high prevalence of measles, the child received two doses of measles vaccine.
- The child with chronic bronchiectasis whose immunological response is suboptimal to antimicrobial and pulmonary therapy.
- The child with symptomatic or asymptomatic HIV, lacks an ability to produce an antibody response to immunization with protein or carbohydrate antigens.

□ Dosages according to FDA drug label for each specific indication.
Note: No established dosing regimen for off-label indications.
□ Dual Diagnosis: documentation and coding reflect:
- (B20) Human immunodeficiency virus [HIV] disease, and
- (Z91.89) Other specified personal risk factors, not elsewhere classified or (Z78.9) Other specified health status.

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 and physical; supporting rationale (current within the last 12 months)	□	Physician's orders (with dose, frequency, administration route and duration; written within 30 days prior date of service); progress note(s) including the necessity for initiation and continuation of IVIG.
□	Documentation supporting the qualifying diagnoses		
□	A copy of applicable lab and procedure test results	□	ICD-10-CM diagnosis code(s) supporting medical necessity submitted with each claim; see above dual code requirements for certain conditions.
□	An accurate weight (in kilograms) documented prior to each infusion for dosage determination	□	Documentation indicating that conventional therapies either failed or are contraindicated
□	Medication administration records	□	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 and dual code requirements indicated above

Checklist completed by: **Date:**

The content of the checklist is an educational tool. Use of this document is not intended as a replacement for the documentation requirements published in National or Local Coverage Determinations, or the CMS's documentation guidelines, written law or regulations. Medicare policy changes frequently; Providers/Departments are reminded to review current National and Local Coverage Determination and Policy Articles for specific documentation and coding guidelines.