Reference number(s)
3997-A

JURISDICTION SPECIFIC MEDICARE PART B

Intravenous Immune Globulin (IVIG):

Asceniv, Bivigam, Carimune NF, Flebogamma DIF, Gammagard Liquid, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Panzyga and Privigen

POLICY

I. COVERED USES

The indications below are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. Primary humoral immunodeficiencies
B. Idiopathic thrombocytopenic purpura (ITP)
C. Chronic lymphocytic leukemia (CLL)
D. Human immunodeficiency virus (HIV) infection
E. Chronic inflammatory demyelinating polyneuritis (CIDP)
F. Multifocal motor neuropathy
G. Dermatomyositis
H. Polymyositis
I. Pemphigus vulgaris
J. Pemphigus foliaceus
K. Bullous pemphigoid
L. Mucous membrane pemphigoid
M. Epidermolysis bullosa acquisita
N. Autoimmune hemolytic anemia
O. Multiple sclerosis
P. Systemic lupus erythematosus
Q. Scleromyxedema
R. Myasthenia gravis
S. Stevens-Johnson syndrome/toxic epidermal necrolysis (TEN)
T. Systemic capillary leak syndrome (SCLS) or Clarkson's disease
U. Kawasaki disease
V. Bone marrow transplantation

All other indications will be assessed on an individual basis. Submissions for indications other than those enumerated in this policy should be accompanied by supporting evidence from Medicare approved compendia.

II. DOCUMENTATION

The following documentation must be available in a legible format with patient identification information (e.g., complete name and dates of service) and signature of physician or non-physician practitioner responsible for and providing care to the member, upon request, for all submissions:

A. The information contained in the medical record should include all relevant diagnostic laboratory studies, prior history of bleeding, infection, disease progression, prior medical/surgical therapies and any other

information essential in establishing that the patient meets the coverage indicators set forth in the NCD and LCD.

B. An accurate weight in kilograms should be documented prior to the infusion since the dosage is based on mg/kg dosage.
C. Indications for administration of immune globulin must be fully documented in the patient's medical record.
D. For scleromyxedema, if the therapy extends longer than 6 months, a review of the medical records might be requested to assess overall improvement and verify the provider is using the least amount of IVIG to maintain the changes.
E. For myasthenia gravis, documentation of prior treatments used.

Physicians or other providers filing Medicare claims for administration of immune globulin therapy at the request of another provider assume full responsibility as to the medical necessity for immune globulin under terms and conditions of NCD and LCD. These providers must also be able to meet documentation requirements given above, either directly through their own medical records or indirectly through records obtained from the referring physician.

III. CRITERIA FOR APPROVAL

A. Primary Humoral Immunodeficiencies

Authorization of 6 months may be granted for replacement therapy in members with primary humoral immunodeficiencies when all of the following criteria are met:

1. The member has a severe impairment of antibody capacity in one of the following conditions:
    i. Congenital agammaglobulinemia
    ii. Common variable immunodeficiency
    iii. Wiskott-Aldrich syndrome
    iv. X-linked immunodeficiency with hyper-IgM
    v. Severe combined immunodeficiencies
    vi. Deficient qualitative or quantitative antibody production
2. Member has at least one bacterial infection directly attributable to this deficiency

B. Acute Idiopathic Thrombocytopenic Purpura (ITP)

Authorization of 3 months may be granted for treatment of acute ITP when any of the following criteria is met:

1. IVIG will be used to manage acute bleeding due to severe thrombocytopenia (platelet count usually less than 30,000/microliter).
2. IVIG will be used to increase platelet counts prior to invasive surgical procedures (e.g., splenectomy).
3. IVIG will be used to treat severe thrombocytopenia (platelet counts less than 20,000/microliter) considered to be at risk for intracerebral hemorrhage.

C. Chronic Refractory Idiopathic Thrombocytopenic Purpura (ITP)

Authorization of 6 months may be granted for treatment of chronic refractory ITP when all of the following criteria are met:

1. The member has received prior treatment with corticosteroids and splenectomy, except if treatment with corticosteroids or splenectomy is contraindicated.
2. The duration of the illness is longer than 6 months.
3. The member does not have a concurrent illness or disease that explains the thrombocytopenia.
4. The member's platelet count is persistently at or below 20,000/microliter.

D. Chronic Lymphocytic Leukemia (CLL)

Authorization of 6 months may be granted for preventing recurrent bacterial infections in members with B-cell chronic lymphocytic leukemia (CLL) when all of the following criteria are met:

1. The member has unequivocally documented CLL.
2. The immunoglobulin G (IgG) level is less than 600 mg/dl.
3. The member has a recent history of serious bacterial infection(s) requiring either oral or parenteral antibiotic therapy.

E. Human Immunodeficiency Virus (HIV)

Authorization of 6 months may be granted to reduce significant bacterial infections in members infected with HIV virus when all of the following criteria are met:

1. The member is younger than 14 years of age.
2. There is evidence of either qualitative or quantitative humoral immunologic defects.
3. The member continues to have bacterial infections despite appropriate antimicrobial prophylaxis.

F. Chronic Inflammatory Demyelinating Polyneuritis (CIDP)

Authorization of 3 months may be granted for treatment of chronic inflammatory demyelinating polyneuritis when all of the following criteria are met:

1. The member is new to IVIG therapy or received less than 3 months of therapy.
2. The diagnosis of this condition is documented in the medical record and must be consistent with one of the following published diagnostic criteria:
   i. European Federation of Neurological Societies/Peripheral Nerve Society (EFNS/PNS) Guidelines
   ii. Koski Guidelines
   iii. American Academy of Neurology (AAN) Guidelines

Authorization of 6 months may be granted for treatment of chronic inflammatory demyelinating polyneuritis when all of the following criteria are met:

1. The member has received at least 3 months of IVIG therapy.
2. The member experienced significant improvement with an initial course of IVIG.
3. The member has experienced unequivocal neurological deterioration.
4. The maintenance dose will be at the lowest IVIG dose possible.
5. If the member has received 1 to 2 years of IVIG therapy, the provider should attempt to reduce the IVIG dose. Continued dosing without attempts to reduce the dosing and check responses is considered inappropriate and subject to review.

G. Multifocal Motor Neuropathy

Authorization of 6 months may be granted for treatment of multifocal motor neuropathy when all of the following criteria are met:

1. The disease has been diagnosed on the basis of electrophysiology findings that rule out other possible conditions that may not respond to IVIG treatment.
2. IVIG will be used to treat disease that is progressive and symptomatic, including as first-line treatment.

H. Dermatomyositis and Polymyositis

Authorization of 6 months may be granted for treatment of severe active dermatomyositis or polymyositis when all of the following criteria are met:

1. Other interventions have been unsuccessful, have become intolerable or are contraindicated, defined as one of the following:
   i. The disease has responded poorly or was unresponsive to high-dose steroids either alone or in combination with other immunosuppressive agents (azathioprine, cyclophosphamide, or methotrexate).
   ii. The member is responsive but intolerant to of continual high-dose steroids as reflected by severe adverse events (e.g., steroid myopathy or severe osteoporosis) in whom trials of other immunosuppressive agents, unless contraindicated have been unsuccessful in achieving significant long-term steroid dose reductions.

2. Member has biopsy-proven disease or unequivocal diagnostic features through history, exam, and electromyography (EMG) or nerve conduction studies (NCS).
3. The member has received at least a four month trial of prednisone or prednisone combination therapies.
4. There is a lack of response or poor response to therapies as reflected by persistently elevated serum creatine kinase (CK) levels or lack of improvement on muscle strength improvement scales.

I. Autoimmune Mucocutaneous Blistering Diseases

Authorization of 6 months may be granted for treatment of autoimmune mucocutaneous blistering disease when all of the following criteria are met:

1. The member has one of the following diagnoses: pemphigus vulgaris, pemphigus foliaceus, bullous pemphigoid, mucous membrane pemphigoid (cicatricial pemphigoid), or epidermolysis bullosa acquista.
2. At least one of the following is met regarding prior treatment with conventional therapy:
   i. Member has failed conventional therapy
   ii. Member has a contraindication to conventional therapy
   iii. Member has rapidly progressive disease and a clinical response could not be affected quickly enough using conventional agents, and IVIG will be given along with conventional treatment(s).
3. IVIG will be used for short-term control of the member's condition and will not be used as maintenance therapy.

J. Autoimmune Hemolytic Anemia

Authorization of 6 months may be granted for treatment of autoimmune hemolytic anemia when both of the following criteria are met:

1. The member has warm-type autoimmune hemolytic anemia.
2. The member meets one of the following:
   i. The disease did not respond to corticosteroids or splenectomy.
   ii. Corticosteroids and splenectomy are contraindicated.

K. Multiple Sclerosis

Authorization of 1 month may be granted as second-line therapy for treatment of an acute relapse of relapsing-remitting multiple sclerosis.

L. Systemic Lupus Erythematosus

Authorization of 6 months may be granted for treatment of severe active systemic lupus erythematosus when other interventions have been unsuccessful, have become intolerable, or are contraindicated.

M. Scleromyxedema

Authorization of 6 months may be granted for treatment of scleromyxedema when the member is new to IVIG therapy.

Authorization of 6 months may be granted for treatment of scleromyxedema when both of the following criteria are met:

1. The medical records will be provided to assess overall improvement.
2. The provider is using the least amount of IVIG to maintain the changes.

N. Myasthenia Gravis

Authorization of 1 month may be granted for treatment of acute exacerbations of myasthenia gravis when all of the following criteria are met:

1. The member is experiencing severe muscle weakness.
2. Other treatment modalities were not successful or are not available. Documentation outlining prior treatments may be requested.

3. The member is new to IVIG treatment.
4. A short course of IVIG will be used (2 gm/kg divided over up to 5 days)

Authorization of 1 month may be granted for treatment of acute exacerbations of myasthenia gravis when all of the following criteria are met:
1. The member is experiencing severe muscle weakness.
2. Other treatment modalities were not successful or are not available. Documentation outlining prior treatments may be requested.
3. The member has received IVIG for an acute episode of myasthenia gravis.
4. The member has not received IVIG in the previous 6 months.
5. A short course of IVIG will be used (2 gm/kg divided over up to 5 days)

O. Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN)
Authorization of 1 month may be granted for treatment of Stevens-Johnson syndrome or toxic epidermal necrolysis when all of the following criteria are met:
1. Member has SCORTEN level of 3 or greater
2. The member has not received IVIG treatment for SJS or TEN and the member will receive only one dose for SJS or TEN.

P. Systemic Capillary Leak Syndrome (SCLS) or Clarkson's Disease
Authorization of 3 months may be granted for treatment of systemic capillary leak syndrome or Clarkson's disease when both of the following criteria are met:
1. The disease is associated with monoclonal gammopathy.
2. IVIG will be given as monthly prophylaxis at the lowest effective dose.

Q. Kawasaki Disease
Authorization of 1 month may be granted to prevent complications in Kawasaki disease.

R. Bone Marrow Transplantation
Authorization of 6 months may be granted for bone marrow transplantation.

IV. REFERENCES

1. Intravenous Immune Globulin (IVIG) LCD (L35093) Version R17. Available at: https://www.cms.gov/medicare-coverage-database/indexes/national-and-local-indexes.aspx. Accessed April 22, 2020.
2. Billing and Coding: Intravenous Immune Globulin (IVIG) (A56786) Version R3. Available at: https://www.cms.gov/medicare-coverage-database/indexes/national-and-local-indexes.aspx. Accessed April 22, 2020.
3. National Coverage Determination (NCD) for Intravenous Immune Globulin for the Treatment of Autoimmune Mucocutaneous Blistering Diseases (250.3- Version1). Accessed at: https://www.cms.gov/medicare-coverage-database/details/ncd-details.aspx?NCDId=158&ncdver=1&SearchType=Advanced&CoverageSelection=National&NCSelection=NCA%7cCAL%7cNCD%7cMEDCAC%7cTA%7cMCD&KeyWord=Immune+Globulin&KeyWordLookUp=Title&KeyWordSearchType=Exact&kq=true&bc=IAAAACAAAAAAAA%3d%3d&. Accessed June 2, 2020.