IMPORTANT NOTE ABOUT THIS REIMBURSEMENT POLICY

This policy is applicable to UnitedHealthcare Medicare Advantage Plans offered by UnitedHealthcare and its affiliates.

You are responsible for submission of accurate claims. This reimbursement policy is intended to ensure that you are reimbursed based on the code or codes that correctly describe the health care services provided. UnitedHealthcare reimbursement policies use Current Procedural Terminology (CPT®), Centers for Medicare and Medicaid Services (CMS), or other coding guidelines. References to CPT or other sources are for definitional purposes only and do not imply any right to reimbursement.

This reimbursement policy applies to all health care services billed on CMS 1500 forms and, when specified, to those billed on UB04 forms (CMS 1450). Coding methodology, industry-standard reimbursement logic, regulatory requirements, benefits design and other factors are considered in developing reimbursement policy. This information is intended to serve only as a general resource regarding UnitedHealthcare’s reimbursement policy for the services described and is not intended to address every aspect of a reimbursement situation. Accordingly, UnitedHealthcare may use reasonable discretion in interpreting and applying this policy to health care services provided in a particular case. Further, the policy does not address all issues related to reimbursement for health care services provided to UnitedHealthcare enrollees. Other factors affecting reimbursement may supplement, modify or, in some cases, supersede this policy. These factors may include, but are not limited to: legislative mandates, the physician or other provider contracts, and/or the enrollee’s benefit coverage documents. Finally, this policy may not be implemented exactly the same way on the different electronic claims processing systems used by UnitedHealthcare due to programming or other constraints; however, UnitedHealthcare strives to minimize these variations.

UnitedHealthcare may modify this reimbursement policy at any time by publishing a new version of the policy on this Website. However, the information presented in this policy is accurate and current as of the date of publication.

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Intravenous Immune Globulin (IVIg)

electronic equivalent or its successor form. This policy applies to all products, all network and non-network physicians, and other health care professionals.
The HCPCS/CPT code(s) may be subject to Correct Coding Initiative (CCI) edits. This policy does not take precedence over CCI edits. Please refer to the CCI for correct coding guidelines and specific applicable code combinations prior to billing UnitedHealthcare. It is not enough to link the procedure code to a correct, payable ICD-9-CM diagnosis code. The diagnosis must be present for the procedure to be paid. Compliance with the provisions in this policy is subject to monitoring by pre-payment review and/or post-payment data analysis and subsequent medical review. The effective date of changes/additions/deletions to this policy is the committee meeting date unless otherwise indicated. CPT codes and descriptions are copyright 2010 American Medical Association (or such other date of publication of CPT). All rights reserved. CPT is a registered trademark of the American Medical Association. Applicable FARS/DFARS restrictions apply to Government use. Fee schedules, relative value units, conversion factors, and/or related components are not assigned by the AMA, are not part of CPT, and the AMA is not recommending their use. The AMA does not directly or indirectly practice medicine or dispense medical services. The AMA assumes no liability for data contained or not contained herein. Current Dental Terminology (CDT), including procedure codes, nomenclature, descriptors, and other data contained therein, is copyright by the American Dental Association, 2002, 2004. All rights reserved. CDT is a registered trademark of the American Dental Association. Applicable FARS/DFARS apply.

Summary

Overview
The use of intravenous immune globulin should be reserved for patients with serious defects of antibody function. The goal is to provide immune globulin to those who lack it. Medicare will provide coverage for intravenous immune globulin when it is used in treatment of the following conditions:

* Primary immunodeficiency.
* Immune-mediated Thrombocytopenia (ITP).
* Kawasaki disease.
* Human Immunodeficiency Virus (HIV) (for pediatric use only).
* Bone marrow transplantation.
* Chronic B-cell lymphocytic leukemia.

Intravenous Immune Globulin (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 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 intravenous immune globulin; reactions may cause a rapid fall in blood pressure and clinical anaphylaxis.

IVIG is covered for treatment of the following biopsy-proven conditions:

* Pemphigus vulgaris.
* Pemphigus foliaceus.
* Bullous pemphigoid.
* Mucous membrane pemphigoid (aka, cicatricial pemphigoid), benign mucous membrane pemphigoid, with or without mention of ocular movement.
* Epidermolysis bullosa acquisita.

Patients must meet at least one of the following criteria:

* Failed conventional therapy. Contractors have the discretion to define what constitutes failure of conventional therapy.
* Conventional therapy is contraindicated. Contractors have the discretion to define what constitutes contraindications to conventional therapy.
* Have rapidly progressive disease in which a clinical response could not be affected quickly enough using conventional agents. In these situations, IVIG therapy would be given along with conventional treatment(s) and the IVIG would be used only until conventional therapy could take effect.
Intravenous Immune Globulin (IVIg)

Note: In addition, IVIG for the treatment of autoimmune mucocutaneous blistering disease must be used only for short-term therapy and not as a maintenance therapy (see NCD 250.3 for details).

Other preparations of IVIG are available:
- RhoD immune globulin for use in preventing postpartum Rhesus isoimmunization.
- Cytomegalovirus immune globulin for use in treating or preventing cytomegaloviral disease in transplant recipients.
- Hepatitis B immune globulin intravenous for use in treating prevention of hepatitis B recurrence following liver transplantation in hepatitis B surface antigen (HBsAG)-positive liver transplant patients. (FDA approved April 6, 2007.)

Primary Humoral Immunodeficiencies

IVIG will be covered for use as replacement therapy in patients with primary immunodeficiencies in whom severe impairment of antibody capacity is present in the following conditions:
- Congenital agammaglobulinemia.
- Common variable immunodeficiency.
- Wiskott-Aldrich syndrome
- X-linked immunodeficiency with hyper-IgM.
- Severe combined immunodeficiencies.
- Deficient qualitative and/or quantitative antibody production.
- Have at least one bacterial infection directly attributable to this deficiency.

Idiopathic Thrombocytopenic Purpura (ITP)

IVIG will be covered for both acute and chronic refractory ITP.

Acute ITP, IVIG is covered for:
- Management of acute bleeding due to severe thrombocytopenia (platelet counts usually less than 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.

Chronic refractory ITP

IVIG is covered for patients meeting all of the following conditions:
- Prior treatment with corticosteroids and splenectomy.
- Duration of illness of greater than six months.
- Age of 10 years or older.
- No concurrent illness/disease explaining thrombocytopenia.
- Platelet counts persistently at or below 20,000/ul.

Chronic Lymphocytic Leukemia (CLL)

IVIG will be covered when used to prevent recurrent bacterial infections in patients with B-cell chronic lymphocytic leukemia meeting all of the following conditions:
- Must have unequivocally documented CLL.
- An immunoglobulin G (IgG) level of less than 600 mg/dl.
- Recent history of serious bacterial infection(s) requiring either oral or parenteral antibiotic therapy.

Human Immunodeficiency Virus (HIV) Infection

IVIG will be covered for patients infected with HIV to reduce significant bacterial infection meeting all of the following conditions:
- Age younger than 14 years old.
- Evidence of either qualitative or quantitative humoral immunologic defects.
- Current bacterial infections, despite appropriate antimicrobial prophylaxis.

Chronic Inflammatory Demyelinating Polyneuritis (CIDP)

The diagnosis of this condition must be documented in the medical record and must be consistent with published diagnostic criteria for this condition.
Intravenous Immune Globulin (IVIg)


Patients responsive to an initial course of IVIG will be eligible for maintenance therapy coverage only if unequivocal neurological deterioration occurs at some future point in time. It is expected an initial trial of IVIG for CIDP to last 3 months. If no significant improvement as outlined in the above guidelines, therapy should be discontinued. Maintenance therapy should be at the lowest dose of IVIG possible. Although patients will vary in response, after a one to two year period of stable therapy, attempts to reduce should be occurring. Continued dosing without attempts to reduce the dosing and check responses would be considered inappropriate and subject to pre and post pay reviews.

Multifocal Motor Neuropathy
IVIG may be considered for first line of treatment of patients who have progressive, symptomatic multifocal motor neuropathy that has been diagnosed on the basis of electrophysiology findings that rule out other possible conditions that may not respond to this treatment.

Dermatomyositis, Polymyositis
The routine use of IVIG is not usually recommended for polymyositis or dermatomyositis. IVIG may be used in patients with severe active illness for whom other interventions have been unsuccessful, have become intolerable or are contraindicated.

Refractory myopathies
Refractory myopathies, are, by definition, diseases that are unresponsive or poorly responsive to high-dose steroids either alone or in combination with other immunosuppressive agents (azathioprine, cyclophosphamide, methotrexate). Also included in this definition are patients responsive to but intolerant of continual high-dose steroids as reflected by severe adverse side effects (e.g., steroids myopathy or severe osteoporosis) in whom trials of other immunosuppressive agents, unless contraindicated, have been unsuccessful in achieving significant long-term steroid dose reductions.

Three other coverage conditions which must all be met, in addition to the above, are:
- Biopsy-proven disease.
- At least a four- to six-month trail of prednisone or prednisone combination therapies.
- Lack of response/poor response to therapies as reflected by persistently elevated serum Creatine Kinase (CK) levels and/or lack of improvement on muscle strength improvement scales.

Reimbursement Guidelines
IVIG coverage benefits will not be extended for the treatment of: (A50875)
- Erythroblastosis,
- Secondary thrombocytopenia,
- Epilepsy,
- Amyotrophic lateral sclerosis (ALS),
- Paraneoplastic neurological syndromes,
- Undiagnosed neuropathy, or
- Weakness and malignancies with no causal link to coexisting neurological dysfunctions.

LCD Individual Consideration
Certain unusual uses of IVIG may be covered on an LCD Individual Consideration basis:
Autoimmune Hemolytic Anemia that does not respond to corticosteroids or splenectomy or those for whom the latter two treatments are contraindicated, acute relapse of Relapsing Remitting Multiple Sclerosis, severe active systemic lupus erythematosus for whom other interventions have been unsuccessful, have become intolerable or are contraindicated.

Drug Wastage
Medicare provides payment for the discarded drug/biological remaining in a single-use drug product after administering what is reasonable and necessary for the patient’s condition. If the physician has made good faith efforts to minimize the unused portion of the drug/biological in how patients are scheduled and how he ordered, accepted, stored and used the drug and made good faith efforts to minimize the unused portion of the drug in how it is supplied, then the program will cover the amount of drug discarded along with the
## Intravenous Immune Globulin (IVIg)

### CPT/HCPCS Codes

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<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<tr>
<td>J1459</td>
<td>Injection, immune globulin (Privigen), intravenous, nonlyophilized (e.g., liquid), 500 mg</td>
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<tr>
<td>J1556</td>
<td>Injection, immune globulin (bivigam), 500 mg</td>
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<td>J1557</td>
<td>Injection, immune globulin, (Gammaplex), intravenous, nonlyophilized (e.g., liquid), 500 mg</td>
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<td>Injection, immune globulin, (Gamunex/Gamunex-C/Gammaked), nonlyophilized (e.g., liquid), 500 mg</td>
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<td>Injection, immune globulin, intravenous, lyophilized (e.g., powder), not otherwise specified, 500 mg</td>
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<td>J1568</td>
<td>Injection, immune globulin, (Octagam), intravenous, nonlyophilized (e.g., liquid), 500 mg</td>
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<td>J1569</td>
<td>Injection, immune globulin, (Gammagard liquid), intravenous, nonlyophilized, (e.g., liquid), 500 mg</td>
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<td>J1599</td>
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### References Included (but not limited to):

**CMS NCD**
- NCD 250.3 Intravenous Immune Globulin for the Treatment of Autoimmune Mucocutaneous Blistering Diseases

**CMS LCD(s)**
- Numerous LCDs

**CMS Article(s)**
- Numerous Articles

**CMS Benefit Policy Manual**
- Chapter 15, § 50 Drugs and Biologicals

**CMS Claims Processing Manual**
- Chapter 17; § 80.6 – Intravenous Immune Globulin

**UnitedHealthcare Medicare Advantage Coverage Summaries**
- Medications/Drugs (Outpatient/Part B)
- Skin Treatment, Services and Procedures

**UnitedHealthcare Reimbursement Policies**
- Intravenous Immune Globulin for the Treatment of Mucocutaneous Blistering Diseases (250.3)
- Medically Unlikely Edits (MUE)

**UnitedHealthcare Medical Policies**
- Immune Globulin (IVIG and SCIG)

**MLN Matters**
- Article SE1424, Intravenous Immune Globulin (IVIG) Demonstration - Implementation

**Others**
- FDA Clinical Pharmacology Review; Trade Name Bivigam, FDA Website

### History

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<tr>
<th>Date</th>
<th>Revisions</th>
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<tr>
<td>10/08/2014</td>
<td>Annual Review for MRP Committee presentation and approval</td>
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<tr>
<td>09/25/2013</td>
<td>HCPSC Codes list includes many more codes that are not listed in this Policy</td>
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<td>MRPC approved</td>
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<tr>
<td>02/05/2013</td>
<td>Administrative updates</td>
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## Intravenous Immune Globulin (IVIg)

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<th>Date</th>
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<tr>
<td>12/19/2012</td>
<td>Policy presented to and approved by MRP Committee</td>
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<tr>
<td>10/15/2012</td>
<td>Policy re-reviewed and the following codes added to the policy: J1557, J1559, and J1562</td>
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<td>10/10/2011</td>
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<td>09/28/2011</td>
<td>PI revised</td>
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<tr>
<td>09/14/2011</td>
<td>Policy developed and presented to committee for approval</td>
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