

## Clinical Policy: Immunoglobulin for Idiopathic Dermatomyositis

Reference Number: CP.CPA.43

Effective Date: 11.16.16

Last Review Date: 11.17

Line of Business: Commercial

[Revision Log](#)

See [Important Reminder](#) at the end of this policy for important regulatory and legal information.

### Description

The following are immunoglobulins requiring prior authorization: Bivigam™, Carimune NF®, Cuvitru™, Flebogamma DIF®, Gammagard®, Gammagard S/D®, Gammaked®, Gammaplex™, Gamunex-C®, Octagam®, Privigen, Hizentra™, and Hyqvia. Immunoglobulins are sterile preparations of highly purified immunoglobulin G (IgG) derived from large pools of human plasma and administered intravenously or subcutaneously.

### FDA approved indication

Immunoglobulins are indicated:

- As replacement therapy for primary immunodeficiency (PI). This includes, but is not limited to, congenital agammaglobulinemia, common variable immunodeficiency (CVID), X-linked agammaglobulinemia, Wiskott-Aldrich syndrome, and severe combined immunodeficiencies.
- For the treatment of patients with idiopathic thrombocytopenic purpura (ITP) to raise platelet counts to prevent bleeding or to allow a patient with ITP to undergo surgery.
- As maintenance therapy to improve muscle strength and disability in adult patients with Multifocal Motor Neuropathy (MMN).
- For prevention of bacterial infections in patients with hypogammaglobulinemia and/or recurrent bacterial infections associated with B-cell chronic lymphocytic leukemia (CLL).
- For prevention of coronary artery aneurysms associated with Kawasaki syndrome.
- For the treatment of chronic inflammatory demyelinating polyneuropathy (CIDP) to improve neuromuscular disability and impairment and for maintenance therapy to prevent relapse.

### Policy/Criteria

Provider must submit documentation (which may include office chart notes and lab results) supporting that member has met all approval criteria

It is the policy of health plans affiliated with Centene Corporation® that Immunoglobulins are **medically necessary** when the following criteria are met:

#### I. Initial Approval Criteria

##### A. Dermatomyositis, Polymyositis (must meet all):

1. Diagnosis of dermatomyositis (DM) or polymyositis (PM);
2. Biopsy-proven inflammatory myopathy (dermatomyositis, polymyositis);
3. Failure of at least a 4 month trial of continual high dose corticosteroids in combination with any of the following immunosuppressive agents unless member experiences clinically significant adverse effects or has contraindication(s):

methotrexate, azathioprine, cyclophosphamide, mycophenolate mofetil, tacrolimus, cyclosporine;

4. Dose does not exceed 2 gm/kg/month for 3-6 months.

**Approval duration: 6 months or renewal date, whichever is longer**

**B. Other diagnoses/indications**

1. Refer to CP.CPA.09 if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized).

**II. Continued Therapy**

**A. Dermatomyositis, Polymyositis (must meet all):**

1. Currently receiving medication via Centene benefit or member has previously met initial approval criteria;
2. Documentation of positive response to therapy;
3. Dose does not exceed 2 gm/kg/month for 3-6 months.

**Approval duration: 6 months or renewal date, whichever is longer**

**B. Other diagnoses/indications (must meet 1 or 2):**

1. Currently receiving medication via Centene benefit and documentation supports positive response to therapy.

**Approval duration: Duration of request or 12 months (whichever is less); or**

2. Refer to CP.CPA.09 if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized).

**III. Diagnoses/Indications for which coverage is NOT authorized:**

**A.** Non-FDA approved indications, which are not addressed in this policy, unless there is sufficient documentation of efficacy and safety according to the off label use policy – CP.CPA.09 or evidence of coverage documents

**B.** A list of specific indications for which coverage is not authorized may be found in the PA guideline: CP.CPA.191 Immune Globulin Conditions Not Medically Necessary.

**C.** Inclusion-body myositis (IBM).

**IV. Appendices/General Information**

*Appendix A: Abbreviation/Acronym Key*

IgG: Immune globulin G

PI: Primary immunodeficiency

ITP: Idiopathic thrombocytopenic purpura

MMN: Multifocal Motor Neuropathy

CLL: Chronic Lymphocytic Leukemia

CIDP: Chronic inflammatory

demyelinating polyneuropathy

IGSC: Subcutaneous immune globulin

CVID: Common variable immunodeficiency

DM: Dermatomyositis

PM: Polymyositis

IBM: Inclusion-body myositis

CK: Creatine kinase

IVIG: Intravenous immune globulin

*Appendix B: General Information*

- IVIG may be medically necessary after less than 4 months trial of prednisone or prednisone combination therapies if the patient has profound, rapidly progressive and/or potentially life threatening muscular weakness (e.g., life-threatening aggressive disease with involvement of respiratory musculature, possibly requiring hospitalization, elective intubation and mechanical ventilatory support) and is refractory to or intolerant of previous therapy.
- Failure or clinically significant adverse effects to continual high dose steroids in combination with other immunosuppressive agents is defined as the patient being unresponsive or poorly responsive to therapy (persistently elevated serum creatine kinase (CK) levels and/or lack of improvement on muscle strength improvement scales) or intolerant of therapy (i.e., steroid myopathy or severe osteoporosis).
- IBM is classified as one of the idiopathic inflammatory myopathies. However, despite some histologic similarities, the clinical manifestations, treatment and prognosis are different from DM and PM. IBM is relatively resistant to standard immunosuppressive therapy.

*Appendix C: Therapeutic Alternatives*

Drug	Dosing Regimen	Dose Limit/ Maximum Dose
systemic glucocorticoid (Various - <u>prednisone</u> , <u>prednisolone</u> , or <u>methylprednisolone</u> )	An equivalent dose of <u>prednisone</u> 1 mg/kg per day PO, to a maximum daily dose of 80 mg. Begin slow taper after 4-6 weeks. Pulse <u>methylprednisolone</u> at 1000 mg IV QD for three days may be used for patients who are severely ill.	2 mg/kg/day
azathioprine (Imuran®)	50 mg PO QD for two weeks, then increase the daily dose by 25-50 mg each week to 1.5 mg/kg/day. In patients with an inadequate response after three months of therapy, increase the dose as tolerated up to as high as 2.5 mg/kg/day	2.5 mg/kg/day
methotrexate (Rheumatrex®)	10-15 mg/week PO/IV, increasing slowly by 2.5 mg increments to 25 mg/week if there is inadequate response to the lower dose after two to three months.	50 mg/week. Leucovorin rescue is required at doses greater than 25 mg/week.
cyclophosphamide (Cytoxan®)	300 to 800 mg/m <sup>2</sup> IV every four weeks for at least six courses OR begin at 50-75 mg/day PO, working up to 1.5-2 mg/k/day	This section intentionally left blank
cyclosporine (Various brand names)	3.5 -4 mg/kg PO QD for six months OR begin at 50 mg PO BID, increase up to 100-150 mg BID	This section intentionally left blank

Drug	Dosing Regimen	Dose Limit/ Maximum Dose
tacrolimus (Prograf®)	0.075mg/kg/day PO BID OR begin at 1 mg PO BID, increase to reach trough of 5-10 ng/ml	This section intentionally left blank
mycophenolate mofetil (Cellcept®)	250-500 mg PO BID, increasing by 250-500 mg increments every 1-2 weeks to a target dose of 1500-3000 mg/day	3000 mg/day

**V. Dosage and Administration**

Drug Name	Indication	Dosing Regimen	Maximum Dose
Various Brand Names IVIG	Dermatomyositis, Polymyositis	1gm/kg body weight IV QD for two days every four weeks or 400mg/kg body weight for five days every four weeks in patients intolerant of high dose therapy	2 gm/kg/month for 3-6 months
IGSC (Gammaked, Gamunex-C, Gammagard)	Dermatomyositis, Polymyositis	100-200mg/kg SC per week OR initial dose can be calculated by multiplying the previous IVIG dose by 1.37, then dividing this result into weekly doses based on previous IVIG interval	Not available
IGSC (Hizentra)	Dermatomyositis, Polymyositis	SC at regular intervals from QD up to q2weeks.  Calculate initial weekly dose by multiplying the previous IVIG dose in grams by 1.37, then divide this result by number of weeks between IVIG intervals.	Not available
IGSC (HyQvia)	Dermatomyositis, Polymyositis	Infuse SC the two components of HyQvia sequentially, beginning with the hyaluronidase. Initiate the IGSC within 10 minutes.  Increase dose and frequency from a 1 week dose to a 3 or 4 week dose (see ramp-up schedule below)  For patients previous on another IG treatment, administer the first dose approximately one week after the last infusion of their previous treatment	Not available

		<p>Ramp up schedule:                      Week 1: [1-week dose interval] ¼ of target dose                      Week 2: [2-week dose interval] ½ of target dose                      Week 3: no infusion                      Week 4: [3-week dose interval] ¾ of target dose                      Week 5: no infusion                      Week 6: no infusion                      Week 7: [4-week dose interval] target dose, repeat every 4 weeks</p> <p>Switching from IGIV: Administer at the same dose and frequency as the previous intravenous treatment, after the initial dose ramp-up</p> <p>Naïve to IG treatment or switching from IGSC: 300 to 600 mg/kg at 3 to 4 week intervals, after initial ramp-up</p>	
IGSC (Cuvitru)	Dermatomyositis, Polymyositis	<p>Administer SC at regular intervals from daily up to every two weeks (biweekly).</p> <p>To calculate the initial weekly dose of Cuvitru, multiply the previous IVIG dose in grams by the dose adjustment factor of 1.30; then divide this by the number of weeks between doses during the patient's IVIG treatment (i.e., 3 or 4).</p>	Not available

**VI. Product Availability**

<b>Drug</b>	<b>Availability</b>
Bivigam 10% Vial	Solution: 5gm/50ml, 10gm/100ml
Carimune NF Vial	Lyophilized Powder: 6gm, 12gm
Cuvitru 20% (200 mg/mL)	Solution: 5 mL, 10 mL, 20 mL, 40 mL vials
Flebogamma DIF 5% Vial 10% vial	Solution: 0.5gm, 2.5gm, 5gm, 10gm, 20gm Solution: 5gm, 10gm, 20gm
Gammagard 10% Vial	Solution: 1gm, 2.5gm, 5gm, 10gm, 20gm, 30gm

Gammagard S/D vial	Freeze-dried: 5gm, 10gm
Gammaked vial	Solution: 1gm, 2.5gm, 5gm, 10gm, 20gm
Gammaplex 5% vial	Solution: 2.5gm, 5gm, 10gm, 20gm
Gammunex-C vial	Solution: 1gm, 2.5gm, 5gm, 10gm, 20gm, 40gm
Octagam 5% vial	Solution: 1gm, 2.5gm, 5gm, 10gm, 25gm
Octagam 10% vial	Solution: 2gm, 5gm, 10gm, 20gm
Privigen 10% vial	Solution: 5gm, 10gm, 20gm, 40gm
Hizentra 20% vial	Solution: 1gm, 2gm, 4gm, 10gm
HyQvia 10% vial w/ Hyaluronidase recombinant	Solution: 2.5gm, 5gm, 10gm, 20gm, 30gm 160u/ml

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Reviews, Revisions, and Approvals	Date	P&T Approval Date
Converted to new template. Minor changes to verbiage and grammar. References updated.	01.19.17	11.17
Cuvitru added to criteria	02.03.17	11.17

**Important Reminder**

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. The Health Plan makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved. “Health Plan” means a health plan that has adopted this clinical policy and that is operated or administered, in whole or in part, by Centene Management Company, LLC, or any of such health plan’s affiliates, as applicable.

The purpose of this clinical policy is to provide a guide to medical necessity, which is a component of the guidelines used to assist in making coverage decisions and administering benefits. It does not constitute a contract or guarantee regarding payment or results. Coverage decisions and the administration of benefits are subject to all terms, conditions, exclusions and limitations of the coverage documents (e.g., evidence of coverage, certificate of coverage, policy, contract of insurance, etc.), as well as to state and federal requirements and applicable Health Plan-level administrative policies and procedures.

This clinical policy is effective as of the date determined by the Health Plan. The date of posting may not be the effective date of this clinical policy. This clinical policy may be subject to applicable legal and regulatory requirements relating to provider notification. If there is a discrepancy between the effective date of this clinical policy and any applicable legal or regulatory requirement, the requirements of law and regulation shall govern. The Health Plan retains the right to change, amend or withdraw this clinical policy, and additional clinical policies may be developed and adopted as needed, at any time.

This clinical policy does not constitute medical advice, medical treatment or medical care. It is not intended to dictate to providers how to practice medicine. Providers are expected to exercise professional medical judgment in providing the most appropriate care, and are solely responsible

## CLINICAL POLICY

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for the medical advice and treatment of members. This clinical policy is not intended to recommend treatment for members. Members should consult with their treating physician in connection with diagnosis and treatment decisions.

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