Clinical Review Criteria for Intravenous Immunoglobulin (IVIg) Therapy

1. Criteria for Approval

A. IVIg is covered for treatment of the following biopsy-proven conditions:

I. Pemphigus Vulgaris and Pemphigus Foliaceus [ICD9 Code: 694.4]

II. Bullous Pemphigoid [ICD9 Code: 694.5]

III. Mucous Membrane Pemphigoid (a.k.a., Cicatrical Pemphigoid) [ICD9 Codes: 694.6, 694.60, 694.61]

IV. Epidermolysis Bullosa Acquisita [ICD9 Code: 694.8]

B. Patients must meet at least one of the following criteria:

I. Failed conventional therapy

II. Conventional therapy is contraindicated

III. 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

C. 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. Short-term therapy is defined as therapy with a duration of six months or less.

D. IVIg is also covered for treatment of the following conditions:

a. Allogeneic bone marrow transplantation: IVIG is indicated to prevent the risk of acute graft-versus-host disease, associated interstitial pneumonia (infectious or idiopathic), and infections (e.g., cytomegalovirus infections, varicella-zoster virus infection, and recurrent bacterial infection) in the first 100 days after bone marrow transplantation (BMT). Note: IVIG is not indicated and thus not covered for autologous transplants, because the benefit in these cases has been shown to be slight

b. B-cell chronic lymphocytic leukemia (CLL): for patients with hypogammaglobulinemia associated with CLL

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c. Chronic inflammatory demyelinating polyneuropathies (CIDP)

d. Dermatomyositis

e. Guillain-Barré syndrome: IVIG is generally accepted as the treatment of choice for patients with Guillain-Barre syndrome, provided that they are so severely affected that they at least require aid to walk, that the disorder is diagnosed during the first 2 weeks of the illness, and that there are no contraindications to IVIG

f. HIV infected children: bacterial control or prevention

g. Hyperimmunoglobulinemia E syndrome, for treatment of severe infection

h. Immune thrombocytopenic purpura (ITP) when a rapid rise in the platelet count is required, such as prior to surgery, to control excessive bleeding, or to defer or avoid splenectomy

i. Kawasaki disease

j. Lambert-Eaton myasthenic syndrome

k. Moersch-Woltmann (Stiff-man) syndrome (unresponsive to other therapies)

l. Multifocal motor neuropathy. for patients who have progressive, symptomatic multifocal motor neuropathy that has been diagnosed on the basis of electrophysiologic findings that rule out other possible conditions that may not respond to this treatment

m. Multiple myeloma

n. Myasthenia gravis

o. Neonatal alloimmune thrombocytopenia

p. Parvovirus B19 infection, chronic, with severe anemia

q. Primary immunodeficiency diseases (such as congenital agammaglobulinemia (X-linked agammaglobulinemia), hypogammaglobulinemia, common variable immunodeficiency, X-linked immunodeficiency with hyperimmunoglobulin M, severe combined immunodeficiency, and Wiskott-Aldrich syndrome)

r. Relapsing-remitting multiple sclerosis (MS) when standard approaches (i.e., interferons) have failed, become intolerable, or are contraindicated

s. Secondary immunosuppression associated with major surgery (such as cardiac

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transplants) and certain diseases (hematologic malignancies, extensive burns, or collagen-vascular diseases)

t. Systemic lupus erythematosus (SLE), for patients with severe active SLE for whom other interventions have been unsuccessful, have become intolerable, or are contraindicated

2. What is Not Covered

A. The use of IVIG in the following clinical conditions is considered experimental and investigational and thus is not covered:

I. Alzheimer's disease

II. Autism

III. Autoimmune chronic urticaria, angioedema, and therapy for recurrent spontaneous abortion

IV. IgG subclass deficiencies

V. Inclusion body myositis

VI. Infection prevention and control in newborns

a. Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infection (PANDAS)

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Off-label Uses of IVIG for Which Use is NOT Recommended		
Acquired factor VIII inhibitors	Cystic fibrosis	Ophthalmopathy, euthyroid
Acquired von Willebrand's	Diabetes mellitus	Oral use
Acute cardiomyopathy	Diamond-Blackfan anemia	
Acute idiopathic dysautonomia	General parvovirus infection	POEMS syndrome
Acute lymphoblastic leukemia	Hemolytic transfusion reaction	Progressive lumbosacral plexopathy
Acute renal failure	Hemophagocytic syndrome	Rasmussen's syndrome
Adrenoleukodystrophy	High-risk neonates	Recurrent otitis media
Adult HIV infection	HTLV-1 associated	Recurrent miscarriage
Adults in surgery/trauma, burns	Lower motor neuron	Reiter's syndrome
Amyotrophic lateral sclerosis	Lyme radiculoneuritis	Rheumatoid arthritis
Aplastic anemia	Membranous nephropathy	Surgery/trauma
Asthma and chronic chest syndromes	Nephritic syndrome	Thrombotic thrombocytopenic purpura/ Hemolytic uremic syndrome
Autoimmune blistering disorders	Nephrotic syndrome	Uveitis
Behcet's syndrome	Nonimmune thrombocytopenia	Vogt-Koyanagi-Harada syndrome
Chronic fatigue syndrome	Congenital heart block	Opsoclonus-myoclonus

Approve for 1 year

Renewal:

1. Kidney function tests must be attached, showing normal kidney function (SCr, BUN)
2. IgG levels (quantitative) should be attached to show the drug is effective, where appropriate
3. Member has not had an excessive number (4 or less) of serious infections during the previous year.

    A. If above met, may renew for 1 year

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