Intravenous Immune Globulin

HMSA Medicare Advantage- Prior Authorization Request

CVS Caremark administers the prescription benefit plan for the patient identified. This patient's benefit plan requires prior authorization for certain medications in order for the drug to be covered. To make an appropriate determination, providing the most accurate diagnosis for the use of the prescribed medication is necessary. **Please respond below and fax this form to CVS Caremark toll-free at 1-866-237-5512**. If you have questions regarding the prior authorization, please contact CVS Caremark at **1-808-254-4414.** For inquiries or questions related to the patient's eligibility, drug copay or medication delivery; please contact the Specialty Customer Care Team: CaremarkConnect® 1-800-237-2767.

The recipient of this fax may make a request to opt-out of receiving telemarketing fax transmissions from CVS Caremark. There are numerous ways you may opt-out: The recipient may call the toll-free number at 877-265-2711, at any time, 24 hours a day/7 days a week. The recipient may also send an opt-out request via email to do_not_call@cvscaremark.com. An opt out request is only valid if it (1) identifies the number to which the request relates, and (2) if the person/entity making the request does not, subsequent to the request, provide express invitation or permission to CVS Caremark to send facsimile advertisements to such person/entity at that particular number. CVS Caremark is required by law to honor an opt-out request within thirty days of receipt.

Patient's Name: _____ **Date**: _____
Patient's ID: _____ **Patient's Date of Birth:** _____
Patient's Phone Number: _____
Physician's Name: _____
Specialty: _____ **NPI#:** _____
Physician Office Telephone: _____ **Physician Office Fax:** _____

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

Additional Demographic Information:

Patient Weight: _____*kg*

Patient Height: _____*ft* _____*inches*

Send completed form to: CVS Caremark Specialty Programs. Fax: 1-866-237-5512

Criteria Questions:

1. ***Indicate where the drug is being dispensed:***
   ❑ Office ❑ Outpatient Hospital ❑ Ambulatory Surgical ❑ Inpatient Hospital
   ❑ Off Campus Outpatient Hospital ❑ Urgent Care ❑ Emergency Room ❑ Birthing Center
   ❑ Military Facility ❑ Skilled Nursing Facility ❑ Nursing Facility ❑ Hospice
   ❑ Inpatient Psychiatric ❑ Psychiatric Residential Treatment ❑ End Stage Renal Facility
   ❑ Psychiatric Facility ❑ Pharmacy ❑ Other

2. ***Indicate where the drug is being administered:***
   ❑ Ambulatory surgical ❑ Home ❑ Inpatient Hospital
   ❑ Office ❑ Outpatient Hospital ❑ Pharmacy

3. What is the drug being prescribed?
   ❑ Asceniv
   ❑ Gammaplex
   ❑ Gamunex-C
   ❑ Octagam
   ❑ Privigen
   ❑ Panzyga
   ❑ Other_____
   ❑ Bivigam
   ❑ Carimune
   ❑ Flebogamma/Flebogamma DIF
   ❑ Gammagard Liquid
   ❑ Gammagard S/D
   ❑ Gammaked

4. Is IVIG prescribed for a patient with an ICD-10 code(s) listed in Appendix B: ICD-10 Codes That Support Medical Necessity? ❑ Yes ❑ No

5. What is the ICD-10 code? _____

6. What is the diagnosis?
   ❑ Primary immunodeficiency (includes congenital agammaglobulinemia, X-lined agammaglobulinemia, hypogammaglobulinemia, common variable immunodeficiency, X-linked immunodeficiency with hyper IgM syndrome, severe combined immunodeficiency, and Wiskott-Aldrich syndrome),
   ❑ Bone marrow/hematopoietic stem cell transplant recipient
   ❑ Kidney transplant recipient
   ❑ Heart transplant recipient
   ❑ Pre-kidney transplantation *No further questions*
   ❑ Pre-heart transplantation *No further questions*
   ❑ Idiopathic thrombocytopenic purpura (ITP)
   ❑ Chronic lymphocytic leukemia (CLL)
   ❑ Multiple myeloma, *No further questions*
   ❑ Kawasaki disease (mucocutaneous lymph node syndrome), *No further questions*
   ❑ HIV infection (pediatric)
   ❑ Guillain-Barre syndrome
   ❑ Myasthenia gravis
   ❑ Chronic inflammatory demyelinating polyneuropathy (CIDP) and variants excluding MMN
   ❑ Multifocal motor neuropathy
   ❑ Polymyositis, *No further questions*
   ❑ Dermatomyositis
   ❑ Relapsing-remitting multiple sclerosis
   ❑ Lambert-Eaton myasthenic syndrome, *No further questions*
   ❑ Autoimmune mucocutaneous blistering disease (eg, pemphigus vulgaris, pemphigus foliaceus, bullous pemphigoid)
   ❑ Autoimmune retinopathy
   ❑ None of the above _____

Complete the following section based on the patient's diagnosis, if applicable.

Section A: Primary Immunodeficiency

7. Is this request for a new start or continuation of therapy? ❑ New start *skip to #9* ❑ Continuation of therapy

8. Was IVIG previously authorized by HMSA/CVS for this member?
If Yes, no further questions ❑ Yes ❑ No ❑ Unknown

9. Does the member have laboratory evidence of immunoglobulin deficiency, defined as any of the following?
ACTION REQUIRED: Please attach a copy of the laboratory report. ❑ Yes ❑ No
    - Agammaglobulinemia (total IgG less than 200mg/dL) or
    - Persistent hypogammaglobulinemia (total IgG less than 400mg/dL, or at least two standard deviations below normal, on at least two occasions) or
    - Absence of B lymphocytes

10. Does the member have a documented inability to mount an adequate response to inciting antigens, defined as either of the following? ***ACTION REQUIRED: Please attach a copy of the laboratory report.***
❑ Yes ❑ No *No further questions*
    - Lack of appropriate rise in antibody titer following provocation with a polysaccharide antigen. For example, an adequate response to the pneumococcal vaccine may be defined as at least a 4-fold increase in titers for at least 50% of serotypes tested, or
    - Lack of appropriate rise in antibody titer following provocation with a protein antigen. For example, an adequate response to tetanus/diphtheria vaccine may be defined as less than a 4-fold rise in titers 3-4 weeks after vaccine administration.

Section B: Bone Marrow/Stem Cell Transplant Recipient

11. Will IVIG be used for treatment of stem cell transplantation rejection (antibody-mediated)?
If Yes, no further questions ❑ Yes ❑ No

12. Has the patient received transplantation for a Medicare-approved indication? ❑ Yes ❑ No

13. Was the patient cytomegalovirus (CMV) seropositive before transplantation?
If Yes, no further questions ❑ Yes ❑ No

14. Did the patient undergo allogeneic transplantation for a hematologic malignancy with bone marrow/stem cells from a CMV-seropositive donor? ❑ Yes ❑ No

Section C: Kidney and Heart Transplant

15. Will IVIG be used for treatment of antibody-mediated kidney or heart transplantation rejection? ❑ Yes ❑ No

Section D: Idiopathic Thrombocytopenia Purpura (ITP)

16. Is IVIG requested for a pregnant woman with idiopathic thrombocytopenic purpura (ITP)?
If No, skip to #20 ❑ Yes ❑ No

15. Has the patient previously delivered an infant with autoimmune thrombocytopenia?
If Yes, skip to # 18 ❑ Yes ❑ No

16. Does the patient have a platelet count of less than 75,000/mm3 during the current pregnancy?
If Yes, skip to # 18 ❑ Yes ❑ No

17. Does the patient have a past history of splenectomy? ❑ Yes ❑ No

18. Has other therapy failed or does the patient have a contraindication to other therapy?
If Yes, no further questions ❑ Yes ❑ No

19. Does the patient have a rapidly progressive form of the disease?
❑ Yes ❑ No *No further questions*

20. Does the patient have <u>acute</u> idiopathic thrombocytopenic purpura (ITP)? ❑ Yes ❑ No *If No, skip to #24*

21. Will IVIG be used for management of acute bleeding due to severe thrombocytopenia (platelet count less than 30,000/mm3)? *If Yes, no further questions* ❑ Yes ❑ No

22. Will IVIG be used to increase platelet counts prior to an invasive major surgical procedure (eg, splenectomy)? *If Yes, no further questions* ❑ Yes ❑ No

23. Will IVIG be used for a patient with severe thrombocytopenia (platelet count less than 20,000/mm3) considered to be at risk for intracerebral hemorrhage? ❑ Yes ❑ No *No further questions*

24. Does the patient have chronic idiopathic thrombocytopenic purpura (ITP)? ❑ Yes ❑ No

25. Will IVIG be used for first-line/initial treatment of ITP? *If No, skip to #28* ❑ Yes ❑ No

26. Is IVIG requested for a pediatric patient? *If Yes, no further questions* ❑ Yes ❑ No

27. Will IVIG be used in combination with corticosteroids (if a rapid response is required or to avoid splenectomy) or does the patient have a contraindication to corticosteroids? ❑ Yes ❑ No *No further questions*

28. Has the patient received previous treatment with corticosteroids and has had a splenectomy? *If Yes, no further questions* ❑ Yes ❑ No

29. Are the platelet counts persistently at or below 20,000/mm3? ❑ Yes ❑ No

Section E: Chronic Lymphocytic Leukemia

30. Is IVIG prescribed for a patient with chronic lymphocytic leukemia and associated hypogammaglobulinemia? ❑ Yes ❑ No

31. At the time of initiation of IVIG therapy, does/did the patient have repeated bacterial infections? ❑ Yes ❑ No

32. At the time of initiation of IVIG therapy, does/did the patient have an IgG level less than 600 mg/dL or is/was there evidence of specific antibody deficiency? ❑ Yes ❑ No

Section F: Pediatric HIV Infection

33. Does the patient meet both of the following criteria? ❑ Yes ❑ No
    - Entry CD4+ lymphocyte counts greater than or equal to 200/mm^3 and
    - Clinically symptomatic or asymptomatic, but immunologically abnormal

Section G: Guillain-Barre Syndrome or Myasthenia Gravis

34. Has other therapy failed or does the patient have a contraindication to other therapy? *If Yes, skip to #37* ❑ Yes ❑ No

35. Does the patient have a rapidly progressive form of the disease? *If Yes, skip to #37* ❑ Yes ❑ No

Section H: Dermatomyositis or Relapsing-Remitting MS

36. Has other therapy failed or does the patient have a contraindication to other therapy? *If Yes, Continue to #37* ❑ Yes ❑ No

Section I: Neurological Disorders – Continuation Criteria

37. Is the patient a new start or continuing with IVIG therapy? ❑ New Start ❑ Continuation *If Continuation, skip to #39*

38. Will the benefit of IVIG therapy be measured using quantitative monitoring tools or any accepted metric such as MRC scale and activities of daily living (ADLs)? ❑ Yes ❑ No *No further questions*

39. Has the patient demonstrated significant clinical improvement with IVIG therapy? ❑ Yes ❑ No

40. What is the total duration of treatment with IVIG?
    ❑ Less than 2 years, *No further questions*
    ❑ Greater than or equal to 2 years

41. Has IVIG dose reduction or withdrawal of treatment been periodically attempted, and the effects measured, to validate continued use? ❑ Yes ❑ No ❑ Not applicable/condition is not stable

Section J: Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

42. Does the patient have chronic inflammatory demyelinating polyneuropathy (CIDP) or other neuropathy resulting from diabetes mellitus, dysproteinemia, renal failure or malnutrition? ❑ Yes ❑ No

43. Does the patient demonstrate clinical signs and symptoms consistent with a diagnosis of CIDP? *Refer to Appendix D.* ❑ Yes ❑ No

44. Does electrodiagnostic and laboratory evidence support the diagnosis of CIDP, including the following? ❑ Yes ❑ No
    - Conduction block at sites not prone to compression
    - Motor nerves show segmental conduction slowing and increased distal latencies consistent with a demyelinating polyneuropathy
    - Conduction slowing from moderate to severe axonal loss has been excluded
    - Cerebrospinal fluid (CSF) analysis shows cytoalbuminologic dissociation (occurs in >90% of cases)
    - Serum tests do NOT support an alternative diagnosis (eg, IgG or IgM monoclonal gammopathy, anti-MAG antibodies, or anti-GM1 antibodies)

45. Does the patient have any other explanation or any condition below that is associated with CIDP? ❑ Yes ❑ No
    - HIV disease
    - Distally predominant diabetic neuropathy
    - Diabetic amyotrophy
    - Diabetic cachectic neuropathy
    - Distal acquired demyelinating symmetric neuropathy with an IgM paraprotein

46. Is there evidence of another treatable cause of polyneuropathy or hereditary demyelinating neuropathy? ❑ Yes ❑ No

47. Does the patient have CIDP that predominantly affects sensory nerves and IVIG is prescribed principally for pain control? *If No, skip to #50* ❑ Yes ❑ No

48. Prior to starting IVIG therapy, did the patient experience a measurable response to a therapeutic trial of prednisone? ❑ Yes ❑ No

49. Did the patient receive a consultation from a neurologist or rheumatologist who is an expert in the field of CIDP and who validated the need for IVIG to control pain? *If Yes, skip to #51* ❑ Yes ❑ No

50. Did the patient receive a consultation from a neurologist or rheumatologist who is an expert in the field of CIDP that resulted in all of the following? ❑ Yes ❑ No
    - Comprehensive history and examination
    - Validation of correct CIDP diagnosis
    - Validation of need for IVIG treatment, recommended regimen and appropriate measures of therapeutic benefit, and follow-up

51. Has other therapy failed or does the patient have a contraindication to other therapy? *If Yes, skip to #53* ❑ Yes ❑ No

52. Does the patient have a rapidly progressive form of the disease? ❑ Yes ❑ No

53. Is the patient a new start or continuing with IVIG therapy for CIDP?
    ❑ New Start
    ❑ Continuation of therapy, *Skip to #55*

54. Will the benefit of IVIG therapy be measured using quantitative monitoring tools or any accepted metric such as MRC scale and activities of daily living (ADLs)? ❑ Yes ❑ No *No further questions.*

55. Has the patient demonstrated significant clinical improvement with IVIG therapy (and when relevant, a reduction in the level of sensory loss)? ❑ Yes ❑ No

56. What is the total duration of treatment with IVIG for CIDP?
    ❑ Less than 2 years, *No further questions*
    ❑ Greater than or equal to 2 years

57. Has IVIG dose reduction or withdrawal of treatment been periodically attempted, and the effects measured, to validate continued use? ❑ Yes ❑ No ❑ Not applicable/condition is not stable

Section K: Multifocal Motor Neuropathy (MMN)

58. Does the patient demonstrate clinical signs and symptoms consistent with a diagnosis of multifocal motor neuropathy (MMN)? Refer to Appendix E ❑ Yes ❑ No

59. Does electrodiagnostic and laboratory evidence support the diagnosis of MMN? ❑ Yes ❑ No

60. Did the patient receive a consultation from a neurologist or rheumatologist who is an expert in the field of MMN that resulted in all of the following? ❑ Yes ❑ No
    - Comprehensive history and examination
    - Validation of correct MMN diagnosis
    - Validation of need for IVIG treatment, recommended regimen and appropriate measures of therapeutic benefit, and follow-up

61. Is the patient a new start or continuing with IVIG therapy for MMN?
    ❑ New start
    ❑ Continuation of therapy, *Skip to #63*

62. Will the benefit of IVIG therapy be measured using quantitative monitoring tools or any accepted metric such as MRC scale and activities of daily living (ADLs)? ❑ Yes ❑ No *No further questions.*

63. Has the patient demonstrated significant clinical improvement with IVIG therapy (and when relevant, a reduction in the level of sensory loss)? ❑ Yes ❑ No

64. What is the total duration of treatment with IVIG for MMN?
    ❑ Less than 2 years, *No further questions*
    ❑ Greater than or equal to 2 years

65. Has IVIG dose reduction or withdrawal of treatment been periodically attempted, and the effects measured, to validate continued use? ❑ Yes ❑ No ❑ Not applicable/condition is not stable

Section L: Autoimmune Mucocutaneous Blistering Diseases

66. Has treatment with conventional therapy failed or is conventional therapy contraindicated?
    If Yes, skip to #68 ❑ Yes ❑ No
67. Does the patient have rapidly progressive disease in which a clinical response could not be affected quickly enough using conventional agents? ❑ Yes ❑ No

68. Will IVIG be used for short-term therapy (NOT maintenance therapy)? ❑ Yes ❑ No

Section M: Autoimmune Retinopathy

69. How long has the patient received treatment with IVIG for autoimmune retinopathy?
    ❑ Less than 3 months, *No further questions*
    ❑ Greater than or equal to 3 months

70. Has the condition improved with IVIG therapy? ❑ Yes ❑ No

I attest that this information is accurate and true, and that documentation supporting this information is available for review if requested by CVS Caremark or the benefit plan sponsor.

X_____

Prescriber or Authorized Signature **Date (mm/dd/yy)**

APPENDICES

Appendix A: IVIG CPT/HCPCS Codes

J1459	INJECTION, IMMUNE GLOBULIN (PRIVIGEN), INTRAVENOUS, NONLYOPHILIZED (E.G. LIQUID), 500 MG
J1556	INJECTION, IMMUNE GLOBULIN (BIVIGAM), 500 MG
J1557	INJECTION, IMMUNE GLOBULIN, (GAMMAPLEX), INTRAVENOUS, NONLYOPHILIZED (E.G. LIQUID), 500 MG
J1561	INJECTION, IMMUNE GLOBULIN, (GAMUNEX-C/ GAMMAKED), NONLYOPHILIZED (E.G. LIQUID), 500 MG
J1566	INJECTION, IMMUNE GLOBULIN, INTRAVENOUS, LYOPHILIZED (E.G. POWDER), NOT OTHERWISE SPECIFIED, 500 MG
J1568	INJECTION, IMMUNE GLOBULIN, (OCTAGAM), INTRAVENOUS, NONLYOPHILIZED (E.G. LIQUID), 500 MG
J1569	INJECTION, IMMUNE GLOBULIN, (GAMMAGARD LIQUID), NONLYOPHILIZED, (E.G. LIQUID), 500 MG
J1572	INJECTION, IMMUNE GLOBULIN, (FLEBOGAMMA/FLEBOGAMMA DIF), INTRAVENOUS, NONLYOPHILIZED (E.G. LIQUID), 500 MG
J1599	INJECTION, IMMUNE GLOBULIN, INTRAVENOUS, NONLYOPHILIZED (E.G. LIQUID), NOT OTHERWISE SPECIFIED, 500 MG

Appendix B: ICD-10 Codes That Support Medical Necessity

ICD-10 Code	Description
B20*	Human immunodeficiency virus [HIV] disease
B25.0	Cytomegaloviral pneumonitis
B25.1	Cytomegaloviral hepatitis
B25.2	Cytomegaloviral pancreatitis
B25.8	Other cytomegaloviral diseases
C90.00	Multiple myeloma not having achieved remission
C90.02	Multiple myeloma in relapse
C91.10	Chronic lymphocytic leukemia of B-cell type not having achieved remission
C91.11	Chronic lymphocytic leukemia of B-cell type in remission
C91.12	Chronic lymphocytic leukemia of B-cell type in relapse
D59.0	Drug-induced autoimmune hemolytic anemia
D59.1	Other autoimmune hemolytic anemias
D61.01*	Constitutional (pure) red blood cell aplasia
D69.3	Immune thrombocytopenic purpura
D69.42	Congenital and hereditary thrombocytopenia purpura
D69.49	Other primary thrombocytopenia
D80.0	Hereditary hypogammaglobulinemia
D80.1	Nonfamilial hypogammaglobulinemia
D80.2	Selective deficiency of immunoglobulin A [IgA]
D80.3	Selective deficiency of immunoglobulin G [IgG] subclasses
D80.4	Selective deficiency of immunoglobulin M [IgM]

ICD-10 Code	Description
D80.5	Immunodeficiency with increased immunoglobulin M [IgM]
D80.6	Antibody deficiency with near-normal immunoglobulins or with hyperimmunoglobulinemia
D80.7	Transient hypogammaglobulinemia of infancy
D81.0	Severe combined immunodeficiency [SCID] with reticular dysgenesis
D81.1	Severe combined immunodeficiency [SCID] with low T- and B-cell numbers
D81.2	Severe combined immunodeficiency [SCID] with low or normal B-cell numbers
D81.5	Purine nucleoside phosphorylase [PNP] deficiency
D81.6	Major histocompatibility complex class I deficiency
D81.7	Major histocompatibility complex class II deficiency
D81.89	Other combined immunodeficiencies
D81.9	Combined immunodeficiency, unspecified
D82.0	Wiskott-Aldrich syndrome
D82.1	Di George's syndrome
D82.4	Hyperimmunoglobulin E [IgE] syndrome
D83.0	Common variable immunodeficiency with predominant abnormalities of B-cell numbers and function
D83.1	Common variable immunodeficiency with predominant immunoregulatory T-cell disorders
D83.2	Common variable immunodeficiency with autoantibodies to B- or T-cells
D83.8	Other common variable immunodeficiencies
D83.9	Common variable immunodeficiency, unspecified
G11.3	Cerebellar ataxia with defective DNA repair
G25.82	Stiff-man syndrome
G35	Multiple sclerosis
G60.3	Idiopathic progressive neuropathy
G61.0	Guillain-Barre syndrome
G61.81*	Chronic inflammatory demyelinating polyneuritis
G61.82	Multifocal motor neuropathy
G65.0	Sequelae of Guillain-Barre syndrome
G70.00	Myasthenia gravis without (acute) exacerbation
G70.01	Myasthenia gravis with (acute) exacerbation
G70.81	Lambert-Eaton syndrome in disease classified elsewhere
G73.1	Lambert-Eaton syndrome in neoplastic disease
G73.3	Myasthenic syndromes in other diseases classified elsewhere
L10.0**	Pemphigus vulgaris
L10.1**	Pemphigus vegetans
L10.2**	Pemphigus foliaceous
L10.3**	Brazilian pemphigus [fogo selvagem]

ICD-10 Code	Description
L10.4**	Pemphigus erythematosus
L10.5**	Drug-induced pemphigus
L10.81**	Paraneoplastic pemphigus
L10.89**	Other pemphigus
L10.9**	Pemphigus, unspecified
L12.0**	Bullous pemphigoid
L12.1**	Cicatricial pemphigoid
L12.8**	Other pemphigoid
L12.9**	Pemphigoid, unspecified
L13.8**	Other specified bullous disorders
M30.3	Mucocutaneous lymph node syndrome [Kawasaki]
M31.1	Thrombotic microangiopathy
M33.00	Juvenile dermatopolymyositis, organ involvement unspecified
M33.01	Juvenile dermatopolymyositis with respiratory involvement
M33.02	Juvenile dermatopolymyositis with myopathy
M33.09	Juvenile dermatopolymyositis with other organ involvement
M33.10	Other dermatopolymyositis, organ involvement unspecified
M33.11	Other dermatopolymyositis with respiratory involvement
M33.12	Other dermatopolymyositis with myopathy
M33.19	Other dermatopolymyositis with other organ involvement
M33.20	Polymyositis, organ involvement unspecified
M33.21	Polymyositis with respiratory involvement
M33.22	Polymyositis with myopathy
M33.29	Polymyositis with other organ involvement
M33.90	Dermatopolymyositis, unspecified, organ involvement unspecified
M33.91	Dermatopolymyositis, unspecified with respiratory involvement
M33.92	Dermatopolymyositis, unspecified with myopathy
M33.99	Dermatopolymyositis, unspecified with other organ involvement
M34.83	Systemic sclerosis with polyneuropathy
M36.0	Dermato(poly)myositis in neoplastic disease
T86.01	Bone marrow transplant rejection
T86.02	Bone marrow transplant failure
T86.09	Other complications of bone marrow transplant
T86.11	Kidney transplant rejection
T86.12	Kidney transplant failure
T86.19	Other complication of kidney transplant
T86.21	Heart transplantation rejection
T86.22	Heart transplant failure

ICD-10 Code	Description
T86.298	Other complications of heart transplant
T86.5	Complications of stem cell transplant
Z48.21	Encounter for aftercare following heart transplant
Z48.22	Encounter for aftercare following kidney transplant
Z76.82	Awaiting organ transplant status
Z86.19	Personal history of other infectious and parasitic diseases
Z87.01	Personal history of pneumonia (recurrent)
Z94.0	Kidney transplant status
Z94.1	Heart transplant status
Z94.81	Bone marrow transplant status
Z94.84	Stem cells transplant status

* Group 1 Medical Necessity ICD-10 Codes Asterisk Explanation:
B20 is only payable for children under 13 years of age.
D61.01 is only to be used when patient has failed all first line therapies.
G61.81 is not payable when associated with diabetes mellitus, dysproteinemias, renal failure, or malnutrition.
** Refer to Local Coverage Article for ICD-10 codes (A54641)

Appendix C: Conditions for Which IVIG is NOT Reasonable and Necessary

- Epilepsy
- Amyotrophic lateral sclerosis (ALS)
- Paraneoplastic neurological syndromes
- Undiagnosed neuropathy or weakness
- Malignancies with no causal link to coexisting neurological dysfunctions

Appendix D: Clinical Signs and Symptoms of Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

- Typical CIDP
  - Symmetrical muscle weakness affects proximal and distal muscles of all four limbs.
  - Sensory loss may affect the distal limbs and usually involves large fiber modalities.
  - The clinical evolution tends to be gradually progressive, evolving over periods of more than 8 weeks although patients typically present to clinicians within 6 months of onset.
  - Decreased or absent reflexes in affected nerve distributions occur in nearly all CIDP presentations, and develop during the acute phase typically within 8 weeks of symptom onset.
  - The patient should have a neurologic function assessment score of at least 3 or greater on the Rankin Scale at the time of initial therapy. However, IVIg can be used in patients with rapidly worsening weakness regardless of the Rankin score.
- A multifocal variant of CIDP (multifocal acquired demyelinating sensory and motor neuropathy or MADSAM)
  - Leads to sensory and motor dysfunction in multiple individual nerve distributions (for example, ulnar or median).
  - Weakness may affect the upper or lower limbs, but it most commonly affects distal musculature and is more common in the hands.
  - Progression tends to be step-wise with episodes of weakness compiling over time to cause gradually increasing debility.
- Predominant sensory CIDP
  - Occasionally, a patient with CIDP may have only sensory symptoms. The sensory loss may affect the upper or lower limbs and tends to be relatively symmetrical.
  - Like more common sensory-motor CIDP presentations, patients typically seek medical attention within 6-9 months from onset.

- The sensory loss may begin relatively acutely and progresses in a stepwise or gradual fashion. The sensory distribution is usually not simply limited to the feet or in a stocking distribution, but takes on unusual patterns involving the trunk, arms, or proximal legs.
- The condition is rare compared with the relatively common purely sensory neuropathies such as distal diabetic, toxic, alcoholic, and idiopathic neuropathies.
- Pure sensory CIDP also must be distinguished from distal demyelinating neuropathies associated with an IgM paraprotein, which is not responsive to IVIg or prednisone.

Appendix E: Clinical Signs and Symptoms of Multifocal Motor Neuropathy (MMN)

- MMN is a purely motor syndrome that tends to affect the hands.
- Weakness affects the distribution of individual nerves and tends to progress in a step-wise fashion over time.
- Patients may have subjective sensory complaints but objective sensory findings are not present.
- The diagnosis is generally made using motor and sensory nerve conduction studies. MMN responds to IVIg but not to prednisone. Therefore, prednisone is never indicated in this condition.