Xembify

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 1-855-330-1720**. If you have questions regarding the prior authorization, please contact CVS Caremark at **1-888-877-0518.** 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:** _____
Physician's Name: _____
Specialty: _____ **NPI#:** _____
Physician Office Telephone: _____ **Physician Office Fax:** _____

Referring Provider Info: ❑ **Same as Requesting Provider**
Name: _____ **NPI#:** _____
Fax: _____ **Phone:** _____

Rendering Provider Info: ❑ **Same as Referring Provider** ❑ **Same as Requesting Provider**
Name: _____ **NPI#:** _____
Fax: _____ **Phone:** _____

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

Required Demographic Information:

Patient Weight: _____*kg*

*Patient Height:*_____*cm*

Send completed form to: Case Review Unit CVS Caremark Specialty Programs Fax: 1-855-330-1720

Site of Service Questions:

A. Where will this drug be administered?
- ❑ Ambulatory surgical, *skip to Clinical Questions*
- ❑ Home infusion, *skip to Clinical Questions*
- ❑ Off-campus Outpatient Hospital
- ❑ On-campus Outpatient Hospital
- ❑ Physician office, *skip to Clinical Questions*
- ❑ Pharmacy, *skip to Clinical Questions*

B. Is this request to continue previously established treatment with the requested medication?
- ❑ Yes - This is a continuation of an existing treatment.
- ❑ No - This is a new therapy request (patient has not received requested medication in the last 6 months). *skip to Clinical Criteria Questions*

C. Has the patient experienced an adverse event with the requested product that has not responded to conventional interventions (eg acetaminophen, steroids, diphenhydramine, fluids or other pre-medications) or a severe adverse event (anaphylaxis, anaphylactoid reactions, myocardial infarction, thromboembolism, or seizures) during or immediately after an infusion? ***ACTION REQUIRED: Attach supporting clinical documentation.***
❑ Yes, *skip to Clinical Criteria Questions* ❑ No

D. Does the patient have laboratory confirmed anti-IgA antibodies? ***ACTION REQUIRED: Attach supporting clinical documentation.*** ❑ Yes, *skip to Clinical Criteria Questions* ❑ No

E. Is the patient medically unstable which may include respiratory, cardiovascular, or renal conditions that may limit the member's ability to tolerate a large volume or load or predispose the member to a severe adverse event that cannot be managed in an alternate setting without appropriate medical personnel and equipment? ***ACTION REQUIRED: Attach supporting clinical documentation.*** ❑ Yes, *skip to Clinical Criteria Questions* ❑ No

F. Does the patient have significant behavioral issues and/or physical or cognitive impairment that would impact the safety of the infusion therapy AND the patient does not have access to a caregiver? ***ACTION REQUIRED: Attach supporting clinical documentation.*** ❑ Yes ❑ No

Criteria Questions:

1. What is the diagnosis?
- ❑ Myasthenia gravis
- ❑ Chronic inflammatory demyelinating polyneuropathy (CIDP)
- ❑ Immune thrombocytopenic purpura (ITP)
- ❑ B-cell chronic lymphocytic leukemia (CLL)
- ❑ Stiff-person syndrome
- ❑ Dermatomyositis
- ❑ Polymyositis
- ❑ Multifocal motor neuropathy
- ❑ Human immunodeficiency virus (HIV) infection
- ❑ Guillain-Barré syndrome
- ❑ Lambert-Eaton myasthenic syndrome
- ❑ Parvovirus B19-induced pure red cell aplasia
- ❑ Fetal/neonatal alloimmune thrombocytopenia
- ❑ Immune checkpoint inhibitor related toxicity
- ❑ CAR-T therapy related hypogammaglobulinemia
- ❑ Rasmussen encephalitis
- ❑ Enteroviral meningoencephalitis
- ❑ Systemic lupus erythematosus
- ❑ Hematophagocytic lymphohistiocytosis (HLH)
- ❑ Major surgery associated secondary immunosuppression
- ❑ Major burns associated secondary immunosuppression
- ❑ Macrophage Activation Syndrome (MAS)
- ❑ Hyperimmunoglobulinemia E syndrome
- ❑ Multiple myeloma
- ❑ Opsoclonus-myoclonus
- ❑ Post-transfusion purpura
- ❑ Solid organ transplantation
- ❑ Stevens-Johnson syndrome
- ❑ Toxic necrotizing fasciitis
- ❑ Toxic epidermal necrolysis
- ❑ Toxic shock syndrome
- ❑ Kawasaki syndrome (pediatric)
- ❑ Isoimmune hemolytic disease of newborn
- ❑ Neonatal hemochromatosis
- ❑ Acquired red cell aplasia
- ❑ Acute disseminated encephalomyelitis
- ❑ Autoimmune neutropenia
- ❑ Autoimmune hemolytic anemia
- ❑ Autoimmune neutropenia
- ❑ BK virus associated nephropathy
- ❑ Churg-Strauss syndrome
- ❑ Birdshot retinochoroidopathy
- ❑ Hematologic malignancy associated secondary immunosuppression
- ❑ Collagen-vascular disease associated secondary immunosuppression
- ❑ Bone marrow transplant/hematopoietic stem cell transplant recipient

❑ Autoimmune mucocutaneous blistering disease (includes pemphigus vulgaris, pemphigus foliaceus, bullous pemphigoid, mucous membrane pemphigoid, and epidermolysis bullosa aquisita)
❑ Primary immunodeficiency (e.g., common variable immunodeficiency, X-linked agammaglobulinemia, severe combined immunodeficiency, Wiskott-Aldrich syndrome)
❑ Other _____

2. What is the ICD-10 code? _____

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

Section A: Primary Immunodeficiency

3. Is this a request for continuation of immune globulin therapy? ❑ Yes ❑ No *If No, skip to #8*

4. Has the patient experienced a reduction in the frequency of bacterial infections since starting immune globulin therapy? ❑ Yes ❑ No

5. Does the prescriber measure trough IgG levels at least once per year?
❑ Yes ❑ No ❑ Not applicable to diagnosis *If Not applicable to diagnosis, no further questions.*

6. Is the most recent trough IgG level at or above the lower range of normal for age?
ACTION REQUIRED: If Yes, attach a copy of the laboratory report with a recent IgG trough level.
If Yes or Not applicable, no further questions. ❑ Yes ❑ No ❑ Not applicable for diagnosis

7. Will the prescriber re-evaluate the dose of immune globulin and consider a dose adjustment (when clinically appropriate)? ❑ Yes ❑ No ❑ Not applicable/not clinically appropriate *No further questions.*

8. What is the specific immunodeficiency disorder?
❑ Severe combined immunodeficiency (SCID), ***specify:*** _____
❑ Congenital agammaglobulinemia (e.g., X-linked or autosomal recessive agammaglobulinemia)
❑ Other non-SCID combined immunodeficiency disorder, ***specify:*** _____
❑ Common variable immunodeficiency (CVID)
❑ Hypogammaglobulinemia (unspecified) or other predominant antibody deficiency disorder
❑ Selective IgA deficiency
❑ Selective IgM deficiency
❑ IgG subclass deficiency
❑ Specific antibody deficiency
❑ Other immunodeficiency disorder/none of the above, ***specify*****:** _____

9. ***ACTION REQUIRED:*** Please indicate and attach a copy of the following **pre-treatment** laboratory information (where applicable):
- IgG (total) level: _____ mg/dL
  a) Is IgG (total) level within the normal reference range? ❑ Yes ❑ No
  b) If No, is the IgG level greater than or equal to (≥) 2 SD below the mean for age? ❑ Yes ❑ No
- IgG subclass levels:
  a) IgG1 _____ mg/dL; b) IgG2 _____ mg/dL; c) IgG3 _____ mg/dL;
  d) Other_____
  e) Are the IgG subclass levels within the normal reference range? ❑ Yes ❑ No
  f) If No, is the level(s) greater than or equal to (≥) 2 SD below the mean for age? ❑ Yes ❑ No
  g) Were IgG subclass levels measured on at least 2 different occasions? ❑ Yes ❑ No
- IgA level: _____ mg/dL; Is the IgA level within the normal reference range? ❑ Yes ❑ No
- IgM level: _____ mg/dL; Is the IgM level within the normal reference range? ❑ Yes ❑ No

10. *If diagnosis is severe combined immunodeficiency*, are maternal T cells present in the circulation?
If Yes, skip to #12. ❑ Yes ❑ No

11. *If diagnosis is severe combined immunodeficiency*, what is the patient's CD3 T cell count?
ACTION REQUIRED: Attach a copy of the laboratory report with lymphocyte subset enumeration by flow cytometry. _____ per microliter

12. Was the diagnosis confirmed by molecular or genetic testing? ***ACTION REQUIRED: Please attach a copy of the laboratory report or other medical record that shows the results of molecular/genetic testing.***
❑ Yes ❑ No ❑ Not applicable to diagnosis

13. *If the diagnosis is common variable immunodeficiency*, have other causes of immune deficiency been excluded (e.g., drugs, infectious disease, malignancy)? ❑ Yes ❑ No ❑ Not applicable to diagnosis

14. Does the patient have a history of recurrent bacterial infections (e.g., pneumonia, otitis media, sinusitis, sepsis, gastrointestinal infections)? ❑ Yes ❑ No

15. Was the immune globulin therapy initiated in the hospital setting? ❑ Yes ❑ No

16. Has the patient demonstrated an impaired antibody response to vaccination with a pneumococcal polysaccharide vaccine? ***ACTION REQUIRED: If Yes, please attach a copy of the laboratory report with post-vaccination titers.*** ❑ Yes ❑ No ❑ Not applicable

Section B: Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

17. Is this a request for continuation of immune globulin therapy? *If Yes, skip to #21* ❑ Yes ❑ No

18. Is the disease course progressive or relapsing/remitting for 2 months or longer? ❑ Yes ❑ No

19. Does the patient have moderate to severe functional disability? ❑ Yes ❑ No

20. Were electrodiagnostic studies (electromyography [EMG] or nerve conduction studies [NCS]) and the evaluation of cerebrospinal fluid (when available) performed to confirm the diagnosis? ***ACTION REQUIRED: If 'yes', attach a copy of the EMG or NCS test results and CSF analysis.*** ❑ Yes ❑ No *No further questions*

21. Has the patient demonstrated significant improvement in disability and maintenance of improvement since starting IG therapy? ❑ Yes ❑ No

22. Is IG being used at the lowest effective dose and frequency? ❑ Yes ❑ No

Section C: Multifocal Motor Neuropathy (MMN)

23. Is this a request for continuation of immune globulin therapy? *If Yes, skip to #26* ❑ Yes ❑ No

24. Has the patient experienced progressive, multifocal, asymmetrical weakness without objective sensory loss in 2 or more nerves for at least 1 month? ❑ Yes ❑ No

25. Were electrodiagnostic studies (electromyography [EMG] or nerve conduction studies [NCS]) performed to confirm the diagnosis? ***ACTION REQUIRED: If Yes, attach a copy of the EMG or NCS test results.***
❑ Yes ❑ No

26. Has the patient demonstrated significant improvement in disability and/or maintenance of improvement since starting IG therapy? ❑ Yes ❑ No

Section D: Dermatomyositis (DM) or Polymyositis (PM)

27. Is this request for continuation of immune globulin therapy? *If Yes, skip to #32* ❑ Yes ❑ No

28. Does the patient exhibit any of the following clinical features? *Indicate ALL that apply.*
❑ Proximal muscle weakness (upper or lower extremity and trunk)
❑ Elevated serum creatine kinase (CK) or aldolase level
❑ Muscle pain on grasping or spontaneous pain
❑ Myogenic changes on EMG (short-duration, polyphasic motor unit potentials with spontaneous fibrillation potentials)
❑ Positive for anti-synthetase antibodies (e.g., anti-Jo-1, also called histadyl tRNA synthetase)
❑ Non-destructive arthritis or arthralgias
❑ Systemic inflammatory signs (fever: more than 37°C at axilla, elevated serum CRP level or accelerated ESR of more than 20 mm/h by the Westergren method
❑ Pathological findings compatible with inflammatory myositis (inflammatory infiltration of skeletal evidence of active regeneration may be seen)
❑ None of the above

29. Were electrodiagnostic studies (electromyography [EMG]) and the muscle biopsy (when available) performed to confirm the diagnosis? ***ACTION REQUIRED: If Yes, attach a copy of the EMG test results.*** ❑ Yes ❑ No

30. Were standard first-line (corticosteroids) and second-line (immunosuppressants) treatments tried but were unsuccessful or not tolerated? ***ACTION REQUIRED: If Yes, attach supporting chart note(s) describing previous treatments and no further questions.*** ❑ Yes ❑ No

31. Is the patient unable to receive standard first-line and second-line therapy because of a contraindication or other clinical reason? ***ACTION REQUIRED: If Yes, attach supporting chart note(s) describing previous treatments.*** ❑ Yes ❑ No *No further questions*

32. Has the patient demonstrated significant improvement in disability and/or maintenance of improvement since starting IG therapy? ❑ Yes ❑ No

Section E: Parvovirus B19-Induced Pure Red Cell Aplasia (PRCA)

33. Does the patient have severe, refractory anemia associated with bone marrow suppression? ❑ Yes ❑ No

34. Does the patient have parvovirus B19 viremia? ***ACTION REQUIRED: If Yes, attach test result confirming presence of parvovirus B19.*** ❑ Yes ❑ No

Section F: Myasthenia Gravis

35. What is the primary reason IG is being prescribed?
    - ❑ Refractory myasthenia gravis, *skip to #38*
    - ❑ Acute exacerbation/crisis
    - ❑ Worsening weakness, *skip to #37*
    - ❑ Pre-operative management (e.g., prior to thymectomy), *no further questions*
    - ❑ Other _____

36. Does the patient have severe swallowing difficulty and/or respiratory failure? *If Yes, no further questions.* ❑ Yes ❑ No

37. Does the patient have weakness with an increase in any of the following symptoms: diplopia, ptosis, blurred vision, difficulty speaking (dysarthria), difficulty swallowing (dysphagia), difficulty chewing, impaired respiratory status, fatigue, or limb weakness? ❑ Yes ❑ No *No further questions*

38. Has the patient tried and failed 2 or more standard therapies (e.g., corticosteroids, azathioprine, cyclosporine, mycophenolate mofetil, rituximab)? ***ACTION REQUIRED: If Yes, attach supporting chart note(s) describing previous treatments.*** ❑ Yes ❑ No

Section G: Stiff-Person Syndrome

39. Has the diagnosis been confirmed by anti-glutamic acid decarboxylase (GAD) antibody testing? ***ACTION REQUIRED: If Yes, attach GAD antibody test results.*** ❑ Yes ❑ No

40. Has the patient received first-line treatment with benzodiazepines and/or baclofen and experienced an inadequate response? ***ACTION REQUIRED: If Yes, attach supporting chart note(s) describing previous treatments.*** ❑ Yes ❑ No

Section H: Immune Thrombocytopenic Purpura (ITP)

41. Is the patient a pregnant woman? ❑ Yes ❑ No *If yes, please provide estimated date of delivery and no further questions:* _____

42. Is the patient an adult with refractory ITP after splenectomy? ❑ Yes ❑ No *If No, skip to #45*

43. What is the current pre-treatment platelet count? ***ACTION REQUIRED: Attach lab report with platelet count.*** _____ per mcL *If less than 30,000/mcL, no further questions.*

44. Does the patient have significant bleeding symptoms (e.g., mucosal bleeding or other moderate to severe bleeding)? ❑ Yes ❑ No *No further questions*

45. Is the patient at high risk for bleeding or does the patient require a rapid increase in platelets? *If yes, please indicate the risk factors for bleeding or reason for a rapid increase in platelets. List continues on following page.*
    - ❑ Undergoing a medical or dental procedure where blood loss is anticipated
    - ❑ Comorbidity (e.g., peptic ulcer disease or hypertension)
    - ❑ Mandated anticoagulation therapy
    - ❑ Profession or lifestyle predisposes the patient to trauma (e.g., construction worker, fireman, professional athlete)
    - ❑ Other _____
    - ❑ No, not at high risk or does not require rapid increase in platelets

46. What is the current pre-treatment platelet count? ***ACTION REQUIRED: Attach lab report with platelet count.*** _____ mcL

47. Does the patient have significant bleeding symptoms (e.g., mucosal bleeding or other moderate to severe bleeding)? ❑ Yes ❑ No

48. What is the classification of ITP?
    - ❑ Newly-diagnosed ITP (diagnosed within the past 3 months), *no further questions if patient is less than 18 years*
    - ❑ Previously untreated ITP (initial therapy), *no further questions if patient is less than 18 years old*
    - ❑ Chronic or persistent ITP (greater than or equal to 3 months from diagnosis), *skip to #51*
    - ❑ ITP unresponsive to first-line treatment, *skip to #51*
    - ❑ Other _____

49. Please indicate the prescribed regimen.
    - ❑ IG monotherapy
    - ❑ IG in combination with corticosteroid, *no further questions*
    - ❑ Other _____

50. Is corticosteroid therapy contraindicated? ❑ Yes ❑ No *No further questions*

51. Does the patient have relapsed ITP after a previous response to IG therapy? *If Yes, no further questions.* ❑ Yes ❑ No

52. Does the patient have a history of inadequate response, intolerance or a contraindication to corticosteroid or anti-D therapy? ***ACTION REQUIRED: If Yes, attach supporting chart note(s) describing previous treatments or contraindication.*** ❑ Yes ❑ No

<u>Section I: B-Cell Chronic Lymphocytic Leukemia (CLL), Bone Marrow Transplant/Hematopoietic Stem Cell Transplant Recipient</u>

53. Is this request for continuation of immune globulin therapy? *If Yes, skip to #58* ❑ Yes ❑ No

54. Is IG prescribed for prophylaxis of bacterial infections? ❑ Yes ❑ No

55. What is the patient's pre-treatment IgG level? ***ACTION REQUIRED: If IgG is less than 500 mg/dL, attach a copy of the laboratory report with the pre-treatment IgG level.*** _____ mg/dL

56. *If diagnosis is B-cell chronic lymphocytic leukemia,* does the patient have a history of recurrent sinopulmonary infections requiring intravenous antibiotics or hospitalization? ❑ Yes ❑ No *No further questions*

57. *If diagnosis is bone marrow transplant/hematopoietic stem cell transplant recipient,* has the patient received a bone marrow/hematopoietic stem cell transplant within the past 100 days? ❑ Yes ❑ No *No further questions*

58. Has the patient experienced a reduction in the frequency of bacterial infections since starting IG therapy? ❑ Yes ❑ No

<u>Section J: HIV Infection: Prophylaxis or Thrombocytopenia</u>

59. Is the requested drug being prescribed for prophylaxis of bacterial infections in a pediatric patient? *If Yes, skip to #70* ❑ Yes ❑ No

60. Is the requested drug being prescribed for treatment of thrombocytopenia associated with HIV? ❑ Yes ❑ No

61. Is the patient an adult? ❑ Yes ❑ No *If No, skip to #66*

62. Does the patient have significant bleeding? ❑ Yes ❑ No

63. What is the patient's platelet count? _____ / mcL

64. Is the patient Rh-positive? ❑ Yes ❑ No *If No, no further questions.*

65. Has the patient failed treatment with RhIG? ❑ Yes ❑ No *No further questions*

66. What is the patient's pre-treatment IgG level? ***ACTION REQUIRED: If IgG is less than 400 mg/dL, attach a copy of the laboratory report with the pre-treatment IgG level.*** _____ mg/dL

67. Has the patient had 2 or more bacterial infections in a 1-year period despite antibiotic chemoprophylaxis with TMP-SMZ or another active agent? *If Yes, no further questions.* ❑ Yes ❑ No

68. Does the patient have HIV-associated thrombocytopenia despite anti-retroviral therapy?
If Yes, no further questions. ❑ Yes ❑ No

69. What is the patient's T4 cell count? _____ / mm3 ❑ Unknown
If greater than or equal to 200/mm3, no further questions. If less than 200/mm3 or unknown, skip to #76.

70. Is this request for continuation of immune globulin therapy? *If Yes, skip to #79* ❑ Yes ❑ No

71. Please indicate whether IG will be used for primary or secondary prophylaxis.
❑ Primary prophylaxis ❑ Secondary prophylaxis, *skip to #73* ❑ Other _____, *skip to #74*

72. What is the patient's pre-treatment IgG level? ***ACTION REQUIRED: If IgG is less than 400 mg/dL, attach a copy of the laboratory report with the pre-treatment IgG level.*** _____ mg/dL
If less than 400 mg/dL, no further questions. If greater than or equal to 400 mg/dL, skip to #77

73. Does the patient have a history of recurrent bacterial infections (greater than 2 serious bacterial infections in a 1-year period)? *If Yes, no further questions.* ❑ Yes ❑ No

74. Has the patient failed to form antibodies to common antigens, such as measles, pneumococcal, and/or Haemophilus influenzae type b vaccine? *If Yes, no further questions.* ❑ Yes ❑ No

75. Is this request for a single dose of immune globulin for a patient who has been exposed to measles?
If Yes, no further questions. ❑ Yes ❑ No

76. Does the patient live in an area where measles is highly prevalent? ❑ Yes ❑ No *If No, skip to #78*

77. Has the patient failed to develop an antibody response after two doses of measles, mumps, and rubella live virus vaccine? *If Yes, no further questions.* ❑ Yes ❑ No

78. Does the patient have chronic bronchiectasis that is suboptimally responsive to antimicrobial and pulmonary therapy? ❑ Yes ❑ No *No further questions*

79. Has the patient experienced a reduction in the frequency of bacterial infections since starting IG therapy?
❑ Yes ❑ No

Section K: Lambert-Eaton Myasthenic Syndrome

80. Is this request for continuation of immune globulin therapy? *If Yes, skip to #86* ❑ Yes ❑ No

81. Has the diagnosis been confirmed by neurophysiology studies (e.g., electromyography) or a positive anti- P/Q type voltage-gated calcium channel antibody test? ***ACTION REQUIRED:***
If Yes, attach a copy of the laboratory report, neurophysiology study report or other supporting medical record(s).
❑ Yes – Neurophysiology studies
❑ Yes – Positive anti- P/Q type voltage-gated calcium channel antibody test
❑ No

82. Has the patient tried an anticholinesterase (e.g., pyridostigmine) but it was unsuccessful or not tolerated?
❑ Yes ❑ No

83. Has the patient tried amifampridine (e.g. 3,4-diaminopyridine phosphate, Firdapse) but it was unsuccessful or not tolerated? ❑ Yes ❑ No

84. Does the patient have severe weakness? *If Yes, no further questions.* ❑ Yes ❑ No

85. Is there difficulty with venous access for plasmapheresis? ❑ Yes ❑ No *No further questions*

86. Has the patient experienced stability or improvement in symptoms relative to the natural course of LEMS? ❑ Yes ❑ No

Section L: Immune Checkpoint Inhibitor-Related Adverse Events

87. Has the patient experienced a moderate or severe adverse event to a PD-1 inhibitor (e.g., pembrolizumab, nivolumab) or PD-L1 inhibitor (e.g., atezolizumab, avelumab, durvalumab)? ❑ Yes ❑ No

88. Is the offending drug being temporarily held or has it been discontinued permanently? ❑ Yes ❑ No

89. Which of the following adverse events did the patient experience?
❑ Pneumonitis ❑ Peripheral neuropathy ❑ Transverse myelitis
❑ Myasthenia gravis ❑ Encephalitis ❑ Severe inflammatory arthritis
❑ Other _____

Section M: Hypogammaglobulinemia from CAR-T Therapy

90. Has the patient received treatment with CAR-T therapy (including but not limited to: idecabtagene vicleucel [Abecma], tisagenlecleucel [Kymriah] or axicabtagene ciloleucel [Yescarta]? ❑ Yes ❑ No

91. What is the patient's IgG level? ***ACTION REQUIRED: If IgG is less than 400 mg/dL, attach a copy of the laboratory report with the pre-treatment IgG level.*** _____ mg/dL ❑ Unknown

Section N: Guillain-Barre Syndrome (GBS)

92. Does the patient have severe disease with significant weakness (e.g., inability to stand or walk without aid, respiratory weakness)? ❑ Yes ❑ No

93. Did the onset of neurologic symptoms occur less than 4 weeks from the anticipated start of immunoglobulin therapy? ❑ Yes ❑ No

Section O: Acute Disseminated Encephalomyelitis

94. Has the patient had an insufficient response or a contraindication to intravenous corticosteroid treatment? ❑ Yes ❑ No

Section P: Autoimmune Mucocutaneous Blistering Disease (includes Pemphigus Vulgaris, Pemphigus Foliaceus, Bullous Pemphigoid, Mucous Membrane Pemphigoid, and Epidermolysis Bullosa Aquisita)

95. Has the diagnosis been proven by biopsy and confirmed by pathology report? ❑ Yes ❑ No

96. Is the condition rapidly progressing, extensive, or debilitating? ❑ Yes ❑ No

97. Has the patient failed or experienced significant complications (e.g., diabetes, steroid-induced osteoporosis) from standard treatment (corticosteroids, immunosuppressive agents)? ❑ Yes ❑ No

Section Q: Autoimmune Hemolytic Anemia

98. Which type of autoimmune hemolytic anemia does the patient have?
❑ Warm type ❑ Cold type ❑ Other _____

99. Has the patient tried corticosteroids with inadequate response? *If Yes, no further questions.* ❑ Yes ❑ No

100. Has the patient has a splenectomy with inadequate response? *If Yes, no further questions.* ❑ Yes ❑ No

101. Does the patient have a contraindication to corticosteroids or splenectomy? ❑ Yes ❑ No

Section R: Autoimmune Neutropenia

102. Is treatment with G-CSF (granulocyte colony stimulating factor) an appropriate option? Examples of G-CSF include Fulphila, Granix, Leukine, Neulasta, Neupogen, Udenyca, Zarxio. ❑ Yes ❑ No

Section S: Birdshot Retinochoroidopathy

103. Has the patient tried immunosuppressant therapy (e.g., corticosteroids, cyclosporine) with inadequate response? ❑ Yes ❑ No

Section T: Churg-Strauss Syndrome

104. Does the patient have severe, active disease? ❑ Yes ❑ No

105. Will immune globulin be used as adjunctive therapy? ❑ Yes ❑ No

106. Has the patient experienced failure, intolerance, or is contraindicated to other interventions? ❑ Yes ❑ No

Section U: Enteroviral Meningoencephalitis

107. Is the patient's condition severe? ❑ Yes ❑ No

Section V: Hematophagocytic Lymphohistiocytosis (HLH) and Macrophage Activation Syndrome (MAS)

108. What is the patient's total IgG level? ***ACTION REQUIRED: Attach a copy of the laboratory report with the pre-treatment IgG level.*** _____ mg/dL *If less than 400 mg/dL, no further questions.*

109. Is the total IgG level at least two standard deviations below the mean for age? ❑ Yes ❑ No

Section W: Hyperimmunoglobulinemia E syndrome

110. Does the patient have severe eczema? ❑ Yes ❑ No

Section X: Multiple Myeloma

111. Does the patient have recurrent, serious infections despite the use of prophylactic antibiotics? ❑ Yes ❑ No

Section Y: Neonatal Hemochromatosis

112. Is the patient currently pregnant? ❑ Yes ❑ No

113. Does the patient have a history of pregnancy ending in documented neonatal hemochromatosis? ❑ Yes ❑ No

Section Z: Opsoclonus-Myoclonus

114. Does the patient have paraneoplastic opsoclonus-myoclonus-ataxia associated with neuroblastoma? *If Yes, no further questions.* ❑ Yes ❑ No

115. Does the patient have refractory opsoclonus-myoclonus? ❑ Yes ❑ No

116. Is immune globulin being used as last-resort treatment? ❑ Yes ❑ No

Section AA: Rasmussen Encephalitis

117. Did the patient try anti-epileptic drugs with no improvement in symptoms? ❑ Yes ❑ No

118. Did the patient try corticosteroids with no improvement in symptoms? ❑ Yes ❑ No

Section BB: Solid Organ Transplantation

119. Is immune globulin being prescribed for solid organ transplantation in an allosensitized patient? *If Yes, no further questions.* ❑ Yes ❑ No

120. Is the patient undergoing renal transplantation from a live donor with ABO incompatibility or positive cross match? ❑ Yes ❑ No

Section CC: Secondary Immunosuppression Due to Surgery, Malignancy, Burns, Collagen-Vascular Diseases

121. Is immune globulin being requested to prevent or modify recurrent bacterial or viral infections? ❑ Yes ❑ No

122. What is the patient's pre-treatment IgG level? ***ACTION REQUIRED: If IgG is less than 400 mg/dL, attach a copy of the laboratory report with the pre-treatment IgG level.*** _____ mg/dL ❑ Unknown

Section DD: Toxic Epidermal Necrolysis, Stevens-Johnson Syndrome

123. Is the patient's case severe? ❑ Yes ❑ No

Section EE: Systemic Lupus Erythematosus

124. Does the patient have severe, active disease? ❑ Yes ❑ No

125. Has the patient experienced inadequate response, intolerance, or have a contraindication to first line therapy?
❑ Yes ❑ No

126. Has the patient experienced inadequate response, intolerance, or have a contraindication to second line therapy?
❑ Yes ❑ No

Section FF: Toxic Necrotizing Fasciitis

127. Does the patient have toxic necrotizing fasciitis due to invasive group A streptococcal infection?
ACTION REQUIRED: If Yes, attach documentation confirming presence of fasciitis and culture or Gram stain.
❑ Yes ❑ No

Section GG: Toxic Shock Syndrome

128. Does the patient have toxic shock syndrome due to a staphylococcal or streptococcal infection?
ACTION REQUIRED: If Yes, attach culture or Gram stain. ❑ Yes ❑ No

129. Is the infection refractory to several hours of aggressive therapy? *If Yes, no further questions.* ❑ Yes ❑ No

130. Does the patient have an undrainable focus of infection? *If Yes, no further questions.* ❑ Yes ❑ No

131. Does the patient have persistent oliguria with pulmonary edema? ❑ 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)**

Send completed form to: Case Review Unit CVS Caremark Specialty Programs Fax: 1-855-330-1720