



Immune Globulins

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

Note: This fax may contain medical information that is privileged and confidential and is solely for the use of individuals named above. If you are not the intended recipient you hereby are advised that any dissemination, distribution, or copying of this communication is prohibited. If you have received the fax in error, please immediately notify the sender by telephone and destroy the original fax message. Immune Globulins w/PANDAS SGM SOC -10/2020.

CVS Caremark Specialty Pharmacy • 2211 Sanders Road NBT-6 • Northbrook, IL 60062
Phone: 1-888-877-0518 • Fax: 1-855-330-1720 • www.caremark.com

Site of Service Questions: *If using Ig subcutaneously, please skip to Criteria Questions.*

- A. Where will this drug be administered?
- | | |
|---|--|
| <input type="checkbox"/> Ambulatory surgical, <i>skip to Clinical Questions</i> | <input type="checkbox"/> Home, <i>skip to Clinical Questions</i> |
| <input type="checkbox"/> Off-campus Outpatient Hospital | <input type="checkbox"/> On-campus Outpatient Hospital |
| <input type="checkbox"/> Physician office, <i>skip to Clinical Questions</i> | <input type="checkbox"/> Pharmacy, <i>skip to Clinical Questions</i> |
- B. Is the patient less than 21 years old or 65 years of age or older?
- Yes – less than 21 years old, *skip to Criteria Questions*
- Yes – age 65 years or older, *skip to Criteria Questions*
- No
- C. Is the request to continue previously established treatment with the requested medication?
- Yes - This is a continuation of an existing treatment
- Yes - This is a continuation request, however a gap in therapy of greater than 8 weeks has occurred, *skip to Criteria Questions*
- No - This is a new therapy request (patient has not received requested medication in the last 6 months), *skip to Criteria Questions*
- No – This is a request for a different brand immune globulin product that the patient has not received previously, *skip to Criteria Questions*
- D. Has the patient experienced an adverse event with the requested product that has not responded to conventional intervention (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 Criteria Questions* No
- E. Does the patient have laboratory confirmed autoantibodies to immunoglobulin A?
- ACTION REQUIRED: Attach supporting clinical documentation.** Yes, *skip to Criteria Questions* No
- F. Is the patient medically unstable which may include respiratory, cardiovascular, or renal conditions that may 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 Criteria Questions* No
- G. Does the patient have severe venous access issues that require the use of special interventions only available in the outpatient hospital setting?
- ACTION REQUIRED: Attach supporting clinical documentation.** Yes, *skip to Criteria Questions* No
- H. 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

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

Note: This fax may contain medical information that is privileged and confidential and is solely for the use of individuals named above. If you are not the intended recipient you hereby are advised that any dissemination, distribution, or copying of this communication is prohibited. If you have received the fax in error, please immediately notify the sender by telephone and destroy the original fax message. Immune Globulins w/PANDAS SGM SOC –10/2020.

CVS Caremark Specialty Pharmacy • 2211 Sanders Road NBT-6 • Northbrook, IL 60062
Phone: 1-888-877-0518 • Fax: 1-855-330-1720 • www.caremark.com

What drug is being prescribed?

- Asceniv Bivigam Flebogamma DIF Gammagard Liquid Gammagard S/D Gammaked
 Gammaplex Gamunex-C Octagam Panzyga Privigen Other _____

Criteria Questions:

1. What is the diagnosis?
- | | |
|--|---|
| <input type="checkbox"/> Myasthenia gravis | <input type="checkbox"/> Macrophage Activation Syndrome (MAS) |
| <input type="checkbox"/> Chronic inflammatory demyelinating polyneuropathy (CIDP) | <input type="checkbox"/> Hyperimmunoglobulinemia E syndrome |
| <input type="checkbox"/> Immune thrombocytopenic purpura (ITP) | <input type="checkbox"/> Multiple myeloma |
| <input type="checkbox"/> B-cell chronic lymphocytic leukemia (CLL) | <input type="checkbox"/> Opsoclonus-myoelonus |
| <input type="checkbox"/> Stiff-person syndrome | <input type="checkbox"/> Post-transfusion purpura |
| <input type="checkbox"/> Dermatomyositis | <input type="checkbox"/> Solid organ transplantation |
| <input type="checkbox"/> Polymyositis | <input type="checkbox"/> Stevens-Johnson syndrome |
| <input type="checkbox"/> Multifocal motor neuropathy | <input type="checkbox"/> Toxic necrotizing fasciitis |
| <input type="checkbox"/> Human immunodeficiency virus (HIV) infection | <input type="checkbox"/> Toxic epidermal necrolysis |
| <input type="checkbox"/> Guillain-Barré syndrome | <input type="checkbox"/> Toxic shock syndrome |
| <input type="checkbox"/> Lambert-Eaton myasthenic syndrome | <input type="checkbox"/> Kawasaki syndrome (pediatric) |
| <input type="checkbox"/> Parvovirus B19-induced pure red cell aplasia | <input type="checkbox"/> Isoimmune hemolytic disease of newborn |
| <input type="checkbox"/> Fetal/neonatal alloimmune thrombocytopenia | <input type="checkbox"/> Neonatal hemochromatosis |
| <input type="checkbox"/> Immune checkpoint inhibitor related toxicity | <input type="checkbox"/> Acquired red cell aplasia |
| <input type="checkbox"/> CAR-T therapy related hypogammaglobulinemia | <input type="checkbox"/> Acute disseminated encephalomyelitis |
| <input type="checkbox"/> Rasmussen encephalitis | <input type="checkbox"/> Autoimmune neutropenia |
| <input type="checkbox"/> Enteroviral meningoencephalitis | <input type="checkbox"/> Autoimmune hemolytic anemia |
| <input type="checkbox"/> Systemic lupus erythematosus | <input type="checkbox"/> Autoimmune neutropenia |
| <input type="checkbox"/> Hematophagocytic lymphohistiocytosis (HLH) | <input type="checkbox"/> BK virus associated nephropathy |
| <input type="checkbox"/> Major surgery associated secondary immunosuppression | <input type="checkbox"/> Churg-Strauss syndrome |
| <input type="checkbox"/> Major burns associated secondary immunosuppression | <input type="checkbox"/> Birdshot retinochoroidopathy |
| <input type="checkbox"/> Hematologic malignancy associated secondary immunosuppression | |
| <input type="checkbox"/> Collagen-vascular disease associated secondary immunosuppression | |
| <input type="checkbox"/> Bone marrow transplant/hematopoietic stem cell transplant recipient | |
| <input type="checkbox"/> Autoimmune mucocutaneous blistering disease (includes pemphigus vulgaris, pemphigus foliaceus, bullous pemphigoid, mucous membrane pemphigoid, and epidermolysis bullosa acquisita) | |
| <input type="checkbox"/> Primary immunodeficiency (eg, common variable immunodeficiency, X-linked agammaglobulinemia, severe combined immunodeficiency, Wiskott-Aldrich syndrome) | |
| <input type="checkbox"/> Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS) | |
| <input type="checkbox"/> Pediatric acute onset neuropsychiatric syndrome (PANS) | |
| <input type="checkbox"/> Other _____ | |

2. What is the ICD-10 code? _____ *If patient's diagnosis is PANDAS or PANS, no further questions*

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.*

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

Note: This fax may contain medical information that is privileged and confidential and is solely for the use of individuals named above. If you are not the intended recipient you hereby are advised that any dissemination, distribution, or copying of this communication is prohibited. If you have received the fax in error, please immediately notify the sender by telephone and destroy the original fax message. Immune Globulins w/PANDAS SGM SOC -10/2020.

CVS Caremark Specialty Pharmacy • 2211 Sanders Road NBT-6 • Northbrook, IL 60062
Phone: 1-888-877-0518 • Fax: 1-855-330-1720 • www.caremark.com

8. What is the specific immunodeficiency disorder?
- Severe combined immunodeficiency (SCID), *specify*: _____
 - Congenital agammaglobulinemia (eg, 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 (\geq) 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 (\geq) 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 (eg, drugs, infectious disease, malignancy)?* Yes No Not applicable to diagnosis
14. Does the patient have a history of recurrent bacterial infections (eg, 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

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

Note: This fax may contain medical information that is privileged and confidential and is solely for the use of individuals named above. If you are not the intended recipient you hereby are advised that any dissemination, distribution, or copying of this communication is prohibited. If you have received the fax in error, please immediately notify the sender by telephone and destroy the original fax message. Immune Globulins w/PANDAS SGM SOC -10/2020.

CVS Caremark Specialty Pharmacy • 2211 Sanders Road NBT-6 • Northbrook, IL 60062
 Phone: 1-888-877-0518 • Fax: 1-855-330-1720 • www.caremark.com

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 anti-Jo-1 (histadyl tRNA synthetase) antibody
 - 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] or nerve conduction studies [NCS]) and the muscle biopsy (when available) performed to confirm the diagnosis? **ACTION REQUIRED: If 'Yes', attach a copy of the EMG or NCS 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

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

Note: This fax may contain medical information that is privileged and confidential and is solely for the use of individuals named above. If you are not the intended recipient you hereby are advised that any dissemination, distribution, or copying of this communication is prohibited. If you have received the fax in error, please immediately notify the sender by telephone and destroy the original fax message. Immune Globulins w/PANDAS SGM SOC -10/2020.

CVS Caremark Specialty Pharmacy • 2211 Sanders Road NBT-6 • Northbrook, IL 60062
Phone: 1-888-877-0518 • Fax: 1-855-330-1720 • www.caremark.com

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 (eg, 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 (eg, 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 (eg, 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.*
 Undergoing a medical or dental procedure where blood loss is anticipated
 Comorbidity (eg, peptic ulcer disease or hypertension)
 Mandated anticoagulation therapy
 Profession or lifestyle predisposes the patient to trauma (eg, 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 (eg, mucosal bleeding or other moderate to severe bleeding)?
 Yes No

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

Note: This fax may contain medical information that is privileged and confidential and is solely for the use of individuals named above. If you are not the intended recipient you hereby are advised that any dissemination, distribution, or copying of this communication is prohibited. If you have received the fax in error, please immediately notify the sender by telephone and destroy the original fax message. Immune Globulins w/PANDAS SGM SOC -10/2020.

CVS Caremark Specialty Pharmacy • 2211 Sanders Road NBT-6 • Northbrook, IL 60062
Phone: 1-888-877-0518 • Fax: 1-855-330-1720 • www.caremark.com

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 old*
 - 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

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

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

Section J: HIV Infection: Prophylaxis or Thrombocytopenia

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? _____ / mL
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

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

Note: This fax may contain medical information that is privileged and confidential and is solely for the use of individuals named above. If you are not the intended recipient you hereby are advised that any dissemination, distribution, or copying of this communication is prohibited. If you have received the fax in error, please immediately notify the sender by telephone and destroy the original fax message. Immune Globulins w/PANDAS SGM SOC -10/2020.

CVS Caremark Specialty Pharmacy • 2211 Sanders Road NBT-6 • Northbrook, IL 60062
Phone: 1-888-877-0518 • Fax: 1-855-330-1720 • www.caremark.com

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 #74
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 (eg 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

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

Note: This fax may contain medical information that is privileged and confidential and is solely for the use of individuals named above. If you are not the intended recipient you hereby are advised that any dissemination, distribution, or copying of this communication is prohibited. If you have received the fax in error, please immediately notify the sender by telephone and destroy the original fax message. Immune Globulins w/PANDAS SGM SOC -10/2020.

CVS Caremark Specialty Pharmacy • 2211 Sanders Road NBT-6 • Northbrook, IL 60062
Phone: 1-888-877-0518 • Fax: 1-855-330-1720 • www.caremark.com

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 (e.g., 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 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 (eg 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, Neuopogen, 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

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

Note: This fax may contain medical information that is privileged and confidential and is solely for the use of individuals named above. If you are not the intended recipient you hereby are advised that any dissemination, distribution, or copying of this communication is prohibited. If you have received the fax in error, please immediately notify the sender by telephone and destroy the original fax message. Immune Globulins w/PANDAS SGM SOC -10/2020.

CVS Caremark Specialty Pharmacy • 2211 Sanders Road NBT-6 • Northbrook, IL 60062
 Phone: 1-888-877-0518 • Fax: 1-855-330-1720 • www.caremark.com

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-myooclonus-ataxia associated with neuroblastoma?
If Yes, no further questions. Yes No

115. Does the patient have refractory opsoclonus-myooclonus? 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

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

Note: This fax may contain medical information that is privileged and confidential and is solely for the use of individuals named above. If you are not the intended recipient you hereby are advised that any dissemination, distribution, or copying of this communication is prohibited. If you have received the fax in error, please immediately notify the sender by telephone and destroy the original fax message. Immune Globulins w/PANDAS SGM SOC -10/2020.

CVS Caremark Specialty Pharmacy • 2211 Sanders Road NBT-6 • Northbrook, IL 60062
Phone: 1-888-877-0518 • Fax: 1-855-330-1720 • www.caremark.com

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

Note: This fax may contain medical information that is privileged and confidential and is solely for the use of individuals named above. If you are not the intended recipient you hereby are advised that any dissemination, distribution, or copying of this communication is prohibited. If you have received the fax in error, please immediately notify the sender by telephone and destroy the original fax message. Immune Globulins w/PANDAS SGM SOC –10/2020.

**CVS Caremark Specialty Pharmacy • 2211 Sanders Road NBT-6 • Northbrook, IL 60062
Phone: 1-888-877-0518 • Fax: 1-855-330-1720 • www.caremark.com**