Policy Title:	Medical Policy - Immune globulins		
Policy Number:	000581	**Department:**	PHA
Effective Date:	12/13/2017		
Review Date:	09/12/2018		
Revision Date:	09/12/2018		

Purpose: To support appropriate use of Immune globulins

Scope: Medicaid, Exchange and MMP

Policy Statement:

IVIG will be covered under the medical benefit if used within the following guidelines. Use outside of these guidelines may result in non-payment unless approved under an exception process. This policy applies to immune globulin therapies including, but not limited to, the following:

Bivigam (IVIG), Carimune NF (IVIG), Flebogamma DIF (IVIG), Gammagard S/D (IVIG), Gammagard Liquid (IVIG and SCIG), Gammaked (IVIG and SCIG), Gammaplex (IVIG), GamaSTAN S/D (IMIG), Gamunex –C (IVIG and SCIG), Hizentra (SCIG), Octagam (IVIG), Privigen (IVIG), Hyqvia (SCIG),

Procedure:

Coverage of immune globulins will be reviewed prospectively via the prior authorization process based on criteria below. The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

Coverage Criteria:

1. **FDA approved indications:** :
    a. Primary immunodeficiency
    b. Idiopathic Thrombocytopenia purpura (ITP),
    c. Kawasaki syndrome
    d. Chronic B- cell lymphocytic leukemia (CLL)
    e. Multifocal motor neuropathy
    f. Chronic inflammatory demyelinating polyneuropathy

2. **Compendial Uses:**
    a. Prophylaxis of bacterial infections in pediatric human immunodeficiency virus (HIV) infection
    b. Prophylaxis of bacterial infections in bone marrow transplant (BMT)/hematopoietic stem cell transplant (HSCT) recipients
    c. Dermatomyositis
    d. Polymyositis
    e. Myasthenia gravis
    f. Guillain-Barre syndrome
    g. Lambert-Eaton myasthenic syndrome
    h. Fetal/neonatal alloimmune thrombocytopenia
    i. Parvovirus B19-induced pure red cell aplasia
    j. Stiff-person syndrome

All other indications are considered experimental/investigational and are not a covered benefit. Neighborhood does not provide coverage for drugs when used for investigational purses.

3. Administered immune globulin is administered for a confirmed diagnosis that is a medically accepted indication defined by one of the following sources: the Food and Drug Administration (FDA), Drugdex, American Hospital Formulary Service (AHFS), United States Pharmacopeia Drug Information for the Healthcare Professional (USP DI), and the Drug Package Insert.
4. **Investigational use:** Immune globulins are considered for investigational use when used at a dose and/or for a condition other than those that are recognized as medically accepted indications as defined in one of the above listed resources. Neighborhood does not provide coverage for drugs when used for investigational purses.
5. Diagnosis-specific criteria are provided below along with accepted durations.

Coverage Criteria:

Required Documentation:

6. Primary immunodeficiency(Congenital agammaglobulinemia, Hypogammaglobulinemia, Common Variable Immunodeficiency, Severe combined immunodeficiency , Wiskott-Aldrich syndrome, X-linked agammaglobulinemia or Bruton's Hypergammaglobulinemia, and X-linked Hyper IgM syndrome):
7. Diagnostic test results (when applicable) are required:
    i. Copy of laboratory with serum immunoglobulin levels: IgG, IgA, IgM, and IgG subclasses
    ii. Vaccine response to pneumococcal polysaccharide vaccine (post-vaccination Streptococcus pneumoniae antibody titers)
    iii. Pertinent genetic or molecular testing in members with a known genetic disorder
    iv. Copy of laboratory report with lymphocyte subset enumeration by flow cytometry
8. IgG trough level for those continuing with IVIG therapy
9. Secondary hypogammaglobulinemia (CLL, HIV, BMT/HSCT recipients)
    a. Copy of laboratory report with pre-treatment serum IgG level (when applicable)
10. Chronic inflammatory demyelinating polyneuropathy (CIDP) and multifocal motor neuropathy (MMN)
    a. Pre-treatment electrodiagnostic studies (electromyography [EMG] or nerve conduction studies [NCS])
    b. For CIDP, pre-treatment cerebrospinal fluid (CSF) analysis (when available)
11. Dermatomyositis and polymyositis
    a. Pre-treatment electrodiagnostic studies (EMG/NCS)
    b. Pre-treatment muscle biopsy report (when available)

Criteria:

Primary Immunodeficiency

12. Members with a diagnosis of severe combined immunodeficiency (SCID) or congenital agammaglobulinemia (eg, X-linked or autosomal recessive agammaglobulinemia):
    a. Diagnosis confirmed by genetic or molecular testing, OR
    b. Pretreatment IgG level < 200 mg/dL, OR
    c. Absence or very low number of T cells (CD3 T cells < 300/microliter) or the presence of maternal T cells in the circulation (SCID only)
13. Wiskott-Aldrich syndrome, DiGeorge syndrome, or ataxia-telangiectasia (or other non-SCID combined immunodeficiency):
    a. Diagnosis confirmed by genetic or molecular testing (if applicable), and
    b. History of recurrent bacterial infections (eg, pneumonia, otitis media, sinusitis, sepsis,

gastrointestinal), and

c. Impaired antibody response to pneumococcal polysaccharide vaccine

14. Common variable immunodeficiency (CVID):
    a. . Age 4 years or older
    b. Other causes of immune deficiency have been excluded (eg, drug induced, genetic disorders, infectious diseases such as HIV, malignancy)
    c. Pretreatment IgG level < 500 mg/dL or ≥ 2 SD below the mean for age
    d. History of recurrent bacterial infections
    e. Impaired antibody response to pneumococcal polysaccharide vaccine
15. Hypogammaglobulinemia (unspecified), IgG subclass deficiency, selective IgA deficiency, selective IgM deficiency, or specific antibody deficiency:
    a. History of recurrent bacterial infections
    b. Impaired antibody response to pneumococcal polysaccharide vaccine
    c. Any of the following pre-treatment laboratory findings:
        i. Hypogammaglobulinemia: IgG < 500 mg/dL or ≥ 2 SD below the mean for age
        ii. Selective IgA deficiency: IgA level < 7 mg/dL with normal IgG and IgM levels
        iii. Selective IgM deficiency: IgM level < 30 mg/dL with normal IgG and IgA levels
        iv. IgG subclass deficiency: IgG1, IgG2, or IgG3 ≥ 2 SD below mean for age assessed on at least 2 occasions; normal IgG (total) and IgM levels, normal/low IgA levels
        v. Specific antibody deficiency: normal IgG, IgA and IgM levels

Initial Approval duration and dosing:

16. Approved for one year when above criteria is met and dosing will be administered within FDA recommended guidelines.

Continuation of therapy Criteria and dosing:

17. Approved for one year when criteria below is met and dosing will be administered within FDA recommended guidelines.
    a. A reduction in frequency of bacterial inefections has been demonstrated since initiation of IVIG therapy, AND
    b. IgG trough levels are monitored at least yearly and maintained at or above the lower range of normal for age (when applicable for indication), OR
    c. The prescriber will re-evaluate the dose of IVIG and consider a dose adjustment (when appropriate).

Myasthenia Gravis

18. Members with acute exacerbation, worsening weakness or in preparation for surgery:
    a. Worsening weakness includes an increase in any of the following symptoms: diplopia, ptosis blurred vision, difficulty speaking (dysarthria), difficulty swallowing (dysphagia), difficulty chewing, impaired respiratory status, fatigue, and limb weakness.
    b. Acute exacerbations include more severe swallowing difficulties and/or respiratory failure
    c. Pre-operative management (eg, prior to thymectomy)

19. Members with refractory myasthenia gravis who have tried and failed 2 or more of standard therapies (eg, corticosteroids, azathioprine, cyclosporine, mycophenolate mofetil, rituximab).

Initial Approval duration and dosing:

20. Approved for one month for members with acute exacerbation, worsening weakness or in preparation for surgery or approved for three months for refractory myasthenia gravis when above criteria is met and dosing will be administered within FDA recommended guidelines.

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

21. Moderate to severe functional disability and the diagnosis was confirmed by electrodiagnostic studies and the evaluation of cerebrospinal fluid (CSF)

Initial Approval duration and dosing:

22. Approved for three months for members with CIDP when above criteria is met and dosing will be administered within FDA recommended guidelines .

Continuation of therapy Criteria and dosing:

23. Approved for one year when criteria below is met and dosing will be administered within FDA recommended guidelines.
    a. Significant improvement in disability and maintenance of improvement since initiation of IVIG therapy
    b. IVIG is being used at the lowest effective dose and frequency

Dermatomyositis or Polymyositis

24. Diagnosis established by clinical features (eg, proximal weakness, rash), elevated muscle enzyme levels, electrodiagnostic studies, and muscle biopsy (when available); supportive diagnostic tests include autoantibody testing and muscle imaging (eg, MRI), AND
25. Standard first-line treatments (corticosteroids or immunosuppressants) have been tried but were unsuccessful or not tolerated, OR
26. Member is unable to receive standard first-line therapy because of a contraindication or other clinical reason.

Initial Approval duration and dosing:

27. Approved for three months when above criteria is met and dosing will be administered within FDA recommended guidelines

Continuation of therapy Criteria and dosing:

28. Approved for one year when criteria below is met and dosing will be administered within FDA recommended guidelines.
    a. Significant improvement in disability and maintenance of improvement since initiation of IVIG therapy

Idiopathic Thrombocytopenic Purpura (Immune Thrombocytopenia):

29. Newly diagnosed ITP (within the past 3 months) or initital therapy:
    a. Children (< 18 years of age)
        i. Significant bleeding symptoms (mucosal bleeding or other moderate/severe bleeding) OR
        ii. High risk for bleeding, OR
        iii. Rapid increase in platelets is required(eg, surgery or procedure)
    b. Adults (≥ 18 years of age)
        i. Platelet count < 30,000/mcL, OR
        ii. Platelet count < 50,000/mcL and significant bleeding symptoms, high risk for bleeding or rapid increase in platelets is required, AND
        iii. Corticosteroid therapy is contraindicated and IVIG will be used alone or IVIG will be used in combination with corticosteroid therapy
30. Chronic ITP (≥ 3 months from diagnosis) or ITP unresponsive to first-line therapy:
    a. Platelet count < 30,000/mcL, OR
    b. Platelet count < 50,000/mcL and significant bleeding symptoms, high risk for bleeding or rapid increase in platelets is required, AND
    c. Relapse after previous response to IVIG or inadequate response/intolerance/contraindication to corticosteroid or anti-D therapy
31. Adults with refractory ITP after splenectomy:
    a. Platelet count < 30,000/mcL,OR
    b. Significant bleeding symptoms

Initial Approval duration and dosing:

32. Approved for one month for initial or newly diagnosed ITP, and six months for chronic ITP or refractory ITP after splenectomy when above criteria is met and dosing will be administered within FDA recommended guidelines

. B-cell Chronic Lymphocytic Leukemia (CLL)

33. IVIG is prescribed for prophylaxis of bacterial infections.
34. Member has a history of recurrent sinopulmonary infections requiring intravenous antibiotics or hospitalization.
35. Member has a pretreatment serum IgG level

Initial Approval duration and dosing:

36. Approved for six months when above criteria is met and dosing will be administered within FDA recommended guidelines

Continuation of therapy Criteria and dosing:

37. Approved for 6 months when criteria below is met and dosing will be administered within FDA recommended guidelines.
    a. A Reduction in the frequency of bacterial infections has been demonstrated since initiation of IVIG therapy.

Prophylaxis of Bacterial Infections in HIV-Infected Pediatric Patients

38. Member is ≤ 12 years of age.
39. IVIG is prescribed for primary prophylaxis of bacterial infections and pretreatment serum IgG < 400 mg/dL, OR
40. IVIG is prescribed for secondary prophylaxis of bacterial infections for members with a history of recurrent bacterial infections (> 2 serious bacterial infections in a 1-year period)

Initial Approval duration and dosing:

41. Approved for six months when above criteria is met and dosing will be administered within FDA recommended guidelines

Continuation of therapy Criteria and dosing:

42. Approved for 6 months when criteria below is met and dosing will be administered within FDA recommended guidelines.
    a. A Reduction in the frequency of bacterial infections has been demonstrated since initiation of IVIG therapy.

Prophylaxis of Bacterial Infections in BMT/HSCT Recipients

43. IVIG is prescribed for prophylaxis of bacterial infections.
44. Either of the following: IVIG is requested within the first 100 days post-transplant OR Member has a pretreatment serum IgG < 400 mg/dL.

Initial Approval duration and dosing:

45. Approved for six months when above criteria is met and dosing will be administered within FDA recommended guidelines

Continuation of therapy Criteria and dosing:

46. Approved for 6 months when criteria below is met and dosing will be administered within FDA recommended guidelines.
    a. A reduction in the frequency of bacterial infections has been demonstrated since initiation of IVIG therapy.

Multifocal Motor Neuropathy (MMN)

47. Weakness without objective sensory loss in 2 or more nerves
48. The diagnosis was confirmed by electrodiagnostic studies

Initial Approval duration and dosing:

49. Approved for three months when above criteria is met and dosing will be administered within FDA recommended guidelines

Continuation of therapy Criteria and dosing:

50. Approved for 12 months when criteria below is met and dosing will be administered within FDA recommended guidelines.

a. A significant improvement in disability and maintenance of improvement since initiation of IVIG therapy.

Guillain-Barre Syndrome (GBS)
Initial Approval duration and dosing:

51. Approved for two months for treatment of GBS and dosing will be administered within FDA recommended guidelines

Lambert-Eaton Myasthenic Syndrome (LEMS)
Initial Approval duration and dosing:

52. Approved for twelve months for treatment of LEMS and dosing will be administered within FDA recommended guidelines

Kawasaki Syndrome
Initial Approval duration and dosing:

53. Approved for one month for the treatment of Kawasaki syndrome in pediatric members and dosing will be administered within FDA recommended guidelines

Fetal/Neonatal Alloimmune Thrombocytopenia (F/NAIT)
Initial Approval duration and dosing:

54. Approved for six months for the treatment of F/NAITand dosing will be administered within FDA recommended guidelines

Parvovirus B19-induced Pure Red Cell Aplasia (PRCA)
Initial Approval duration and dosing:

55. Approved for six months for the treatment of PRCA and dosing will be administered within FDA recommended guidelines

Stiff-person Syndrome
Initial Approval duration and dosing:

56. Approved for six months for the treatment of stiff-person syndrome and dosing will be administered within FDA recommended guidelines

Supplier(s): Immune globulins may be accessed through the Medical benefits. All self-administered products are available through the Pharmacy benefit at In-Network preferred specialty pharmacy or pharmacies as dictated by the enrollee's line of business benefit design. Coverage of immune globulins that are not self-administered are provided through the Medical benefit under the appropriate HCPCS code.

Coding: The below are recognized HCPCS and CPT codes for products applicable to this policy. The below tables are provided for reference purposes and may not be all inclusive. Requests received with codes from tables below do not guarantee coverage. Requests must meet all criteria are provided in the procedure section.

CPT Codes	
90281	Immune globulin (Ig), human, for intramuscular use [when specified for disease treatment as described in this document]
90283	Immune globulin, (IgIV), human, for intravenous use
90284	Immune globulin, (SCIg), human, for use in subcutaneous infusions, 100 mg each
S9338	Home infusion therapy; immunotherapy, administrative services, professional pharmacy services, care coordination, all necessary supplies and equipment, per diem
96365	Intravenous infusion, for therapy, prophylaxis, or diagnosis(specify substance or drug), initial, up to 1 hour
96366	Intravenous infusion ,Each additional hour
96372	Therapeutic, prophylaxis, or diagnostic injection(specify substance or drug); subcutaneous or intramuscular
96369	Subcutaneous infusion, for therapy, prophylaxis, or diagnosis(specify substance or drug), initial, up to 1 hour, including pump set up
96370	Subcutaneous infusion, each additional hour
96371	Additional pump set up, with establishment of new subcutaneous infusion site
HCPCS Codes	
J0850	Injection, cytomegalovirus immune globulin
J1459	Injection, immune globulin (Privigen), intravenous, non-lyophilized (e.g., liquid), 500 mg
J1460	Injection, gamma globulin, intramuscular, 1 cc
J1555	Injection, human, for use in subcutaneous infusions, 100mg, each
J1556	Injection, immune globulin (Bivigam), 500 mg
J1557	Injection, immune globulin, (Gammaplex), intravenous, non-lyophilized (e.g., liquid), 500 mg
J1559	Injection, immune globulin (Hizentra), 100 mg
J1560	Injection, gamma globulin, intramuscular, over 10 cc
J1561	Injection, immune globulin, (Gamunex-C/Gammaked), non-lyophilized (e.g., liquid), 500 mg
J1566	Injection, immune globulin, intravenous lyophilized (e.g., powder), not otherwise specified, 500 mg [Carimune, Gammagard S/D]
J1568	Injection, immune globulin, (Octagam), intravenous, non-lyophilized (e.g., liquid), 500 mg
J1569	Injection, immune globulin, (Gammagard Liquid), non-lyophilized (e.g., liquid), 500 mg
J1572	Injection, immune globulin, (Flebogamma/Flebogamma DIF), intravenous, non-lyophilized (e.g., liquid); 500 mg
J1575	Injection, immune globulin/hyaluronidase, (HyQvia), 100 mg immune globulin
J1599	Injection, immune globulin, intravenous, nonlyophilized (e.g., liquid), not otherwise specified, 500 mg

References:

1. Bivigam [package insert]. Boca Raton, FL: Biotest Pharmaceuticals Corporation; April 2012. Accessed 6 August 2014.
2. Carimune NF [package insert]. Kankakee, IL: CSL Behring LLC; October 2010. Revised June 2012. Accessed 6 August 2014.
3. Flebogamma 5% DIF [package insert]. Los Angeles, CA: Grifols Biologicals, Inc.; December 2011. Revised September 2013. Accessed 6 August 2014.
4. Flebogamma 10% DIF [package insert]. Los Angeles, CA: Grifols Biologicals, Inc.; December 2011. Revised September 2013. Accessed 6 August 2014.
5. Gammagard Liquid [package insert]. Westlake Village, CA: Baxter Healthcare Corporation; July 2012. Revised September 2013. Accessed 6 August 2014.
6. Gammagard S/D [package insert]. Westlake Village, CA: Baxter Healthcare Corporation; December 2011. Revised September 2013. Accessed 6 August 2014.
7. Gammaked [package insert]. Research Triangle Park, NC: Talecris Biotherapeutics, Inc.; June 2011. Revised September 2013. Accessed 6 August 2014.
8. Gammaplex [package insert]. Hertfordshire, United Kingdom: Bio Products Laboratory; October 2011. Revised September 2013. Acccessed 6 August 2014.
9. Gamunex-C [package insert]. Research Triangle Park, NC: Talecris Biotherapeutics, Inc.; June 2012. Revised September 2013. Accessed 6 August 2014.
10. Octagam [package insert]. Hoboken, NJ: Octapharma USA, Inc.; September 2009. Revised July 2014. Accessed 6 August 2014.
11. Privigen [package insert]. Kankakee, IL: CSL Behring LLC; May 2012. Revised September 2013. Accessed 6 August 2014.
12. Hizentra [package insert]. Kankakee, IL: CSL Behring LLC; October 2011. Revised September 2013. Accessed 6 August 2014.
13. GamaSTAN S/D [package insert]. Research Triangle Park, NC: Grifolis Therapeutics, Inc.; June 2012. Revised September 2013. Accessed 6 August 2014.
14. Hyqvia [package insert]. Westlake Village, CA: Baxter Healthcare Corporation; January 2015. Revised September 2013. Accessed 22 January 2016.
15. Cytogam [package insert]. Kankakee, IL: CSL Behring, LLC, August 2012. Accessed 27 January 2016.
16. A.J. Ammann, R.F. Ashman, R.H. Buckley, W.R. Hardie, H.J. Krantmann, J. Nelson, *et al.*Use of intravenous gamma-globulin in antibody immunodeficiency: results of a multicenter controlled trial**.** *Clin Immunol Immunopathol*, 22 (1982), pp. 60-67
17. C.M. Roifman, H.M. Lederman, S. Lavi, L.D. Stein, H. Levison, E.W. Gelfand
18. Benefit of intravenous IgG replacement in hypogammaglobulinemic patients with chronic sinopulmonary disease. Am J Med, 79 (1985), pp. 171-174
19. Orange JS, Hossny EM, Weiler CR, et al. Use of intravenous immunoglobulin in human disease: A review of evidence by members of the Primary Immunodeficiency Committee of the American Academy of Allergy, Asthma and Immunology. *J Allergy Clin Immunol* 2006;117:S525-S553
20. Chen S, Pi D, Ansari M, et al. Polyclonal intravenous immunoglobulin in patients with immune thrombocytopenic purpura: Clinical systematic review. Technology Report. HTA Issue 108. Ottawa, ON: Canadian Agency for Drugs and Technologies in Health (CADTH); 2008
21. Oates-Whitehead RM, Baumer JH, Haines L, et al. Intravenous immunoglobulin for the treatment of Kawasaki disease in children. Cochrane Database Syst Rev. 2003;(4):CD004000.
22. Liu Z, Albon E, Hyde C. The effectiveness and cost effectiveness of immunoglobulin replacement therapy for primary immunodeficiency and chronic lymphocytic leukaemia: A systematic review and economic evaluation. DPHE Report No. 54. Birmingham, UK: West Midlands Health Technology Assessment Collaboration, Department of Public Health and Epidemiology, University of Birmingham (WMHTAC); 2005
23. Raanani P, Gafter-Gvili A, Paul M, et al. Immunoglobulin prophylaxis in hematological malignancies and hematopoietic stem cell transplantation. Cochrane Database Syst Rev. 2008;(4):CD006501
24. Mofenson LM, Moye J. Intravenous immune globulin for the prevention of infections in children with symptomatic human immunodeficiency virus infection. Pediatr Res. 1993;33(1 Suppl):S80-7
25. van Schaik IN, van den Berg LH, de Haan R, Vermeulen M. Intravenous immunoglobulin for multifocal motor neuropathy. Cochrane Database Syst Rev. 2005;(2):CD004429.

26. Eftimov F, Winer JB, Vermeulen M, et al. Intravenous immunoglobulin for chronic inflammatory demyelinating polyradiculoneuropathy. Cochrane Database Syst Rev. 2009
27. Hughes RA, Swan AV, Van doorn PA. Intravenous immunoglobulin for Guillain-Barré syndrome. Cochrane Database Syst Rev. 2014;(9):CD002063.
28. Perez E, Orange JS, Bonilla F, et al. Update on the use of immunoglobulin in human disease: a review of evidence by Work Group Report of the American Academy of Allergy, Asthma, and Immunology. J Allergy Clin Immunol. 2017;139:S1-46.
29. Panel o Opportunistic Infections in HIV-Exposed and HIV-Infected Children. Guidelines for the Prevention and Treatment of Opportunistic Infections in HIV-Exposed and HIV-Infected Children. Department of Health and Human Services. Available at http://aidsinfo.nih.gov/contentfiles/lvguidelines/oi_guidelines_pediatrics.pdf. Accessed June 13, 2017.
30. Tombly M, Chiller T, Einsele H, et al. Guidelines for preventing infectious complications among hematopoietic cell transplant recipients: a global perspective. Biol Blood Marrow Transplant. 2009;15(10):1143-1238.
31. FeasbyT, Banwell B, Bernstead T, et al. Guidelines on the use of intravenous immune globulin for neurologic conditions. Transfus Med Rev. 2007;21(2):S57-S107.
32. Donofro PD, Berger A, Brannagan TH 3rd, et al. Consensus statement: the use of intravenous immunoglobulin in the treatment of neuromuscular conditions report of the AANEM ad hoc committee. Muscle Nerve. 2009;40(5):890-900.
33. ElovaarI, Apostolski S, van Doorn P, et al. EFNS guidelines for the use of intravenous immunoglobulin in treatment of neurological diseases: EFNS task force on the use of intravenous immunoglobulin in treatment of neurological diseases. Eur J Neurol. 2008;15(9):893-908.
34. Patwa HS, Chaudhry V, Katzberg H, et al. Evidence-based guideline: intravenous immunoglobulin in the treatment of neuromuscular disorders: report of the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology. Neurology. 2012;78(13);1009-1015.
35. Anderson D, Kaiser A, Blanchette V, et al. Guidelines on the use of intravenous immune globulin for hematologic conditions. Transfus Med Rev. 2007;21(2):S9-S56.
36. Picard C, Al-Herz W, Bousfiha A, et al. Primary immunodeficiency diseases: an update on the classification from the International Union of Immunological Societies Expert Committee for Primary Immunodeficiency. J Clin Immunol. 2015; 35(8):696-726.
37. Bonilla FA, Khan DA, Ballas ZK, et al. Practice parameter for the diagnosis and management of primary immunodeficiency. J Allergy Clin Immunol. 2015;136(5):1186-205.e1-78
38. .Orange JS, Ballow M, Stiehm ER, et al. Use and interpretation of diagnostic vaccination in primary immunodeficiency: a working group report of the Basic and Clinical Immunology Interest section of the American Academy of Allergy, Asthma and Immunology. J Allergy Clin Immunol. 2012;130:S1-S24.
39. Ameratunga R, Woon ST, Gillis D, Koopmans W, Steele R. New diagnostic criteria for common variable immune deficiency (CVID), which may assist with decisions to treat with intravenous or subcutaneous immunoglobulin. Clin Exp Immunol. 2013;174(2):203-11.
40. Immune Deficiency Foundation. About primary immunodeficiencies. Specific disease types. http://primaryimmune.org/about-primary-immunodeficiencies/specific-disease-types/. Accessed June 13, 2017.
41. European Society for Immunodeficiencies. Diagnostic criteria for PID. http://esid.org/WorkingParties/Clinical/Resources/Diagnostic-criteria-for-PID2. Accessed July 8, 2016.
42. Immune Deficiency Foundation. Diagnostic and Clinical Care Guidelines for Primary Immunodeficiency Diseases. 3rd edition. Towson, MD: Immune Deficiency Foundation; 2015. http://primaryimmune.org/wpcontent/uploads/2015/03/2015-Diagnostic-and-Clinical-Care-Guidelines-for-PI.pdf. Accessed June 13, 2017.
43. The NCCN Clinical Practice Guidelines in Oncology® Chronic Lymphocytic Leukemia/Small Lymphocytic Lymphoma (Version 2.2017). © 2017 National Comprehensive Cancer Network, Inc. http://www.nccn.org. Accessed June 13, 2017. 33. Van den Bergh PY, Hadden RD, Bouche P, et al. European Federation of Neurological Societies/Peripheral Nerve Society guideline on management of chronic inflammatory demyelinating polyradiculoneuropathy: report of a joint task force of the European Federation of Neurological Societies and the Peripheral Nerve Society - first revision. Eur J Neurol. 2010;17(3):356-363.
44. Joint Task Force of the EFNS and the PNS. European Federation of Neurological Societies/Peripheral Nerve Societies guideline on management of multifocal motor neuropathy. J Peripher Nerv Syst. 2010;15:295-301. 35. Olney RK, Lewis RA, Putnam TD, Campellone JV. Consensus criteria for the diagnosis of multifocal motor neuropathy. Muscle Nerve. 2003;27:117-121.
45. Dalakas M. Inflammatory muscle diseases. N Engl J Med. 2015;372(18):1734-1747.

46. Neunert C, Lim W, Crowther M, et al. The American Society of Hematology 2011 evidence-based practice guideline for immune thrombocytopenia. Blood. 2011;117(16):4190-4207.
47. Provan D, Stasi R, Newland AC, et al. International consensus report on the investigation and management of primary immune thrombocytopenia. Blood. 2010;115(2):168-186.
48. Shearer WT, Dunn E, Notarangelo LD, et al. Establishing diagnostic criteria for severe combined immunodeficiency disease (SCID), leaky SCID, and Omenn syndrome: the Primary Immune Deficiency Treatment Consortium experience. J Allergy Clin Immunol. 2014;133(4):1092.