RX.PA.017.CCH Intravenous Immune Globulin (IVIG), Intramuscular (IM) & Subcutaneous Immune Globulin (SCIG)

The purpose of this policy is to define the prior authorization process for all commercially available, formulary IVIG and SCIG products.

DEFINITIONS

INCAT (Inflammatory Neuropathy Cause and Treatment Scale) – is used to assess functional disability of both upper and lower extremities in chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)

Grade	Arm Disability
0	No upper limb problems
1	Symptoms, in one or both arms, not affecting the ability to perform any of the following functions: doing all zips and buttons; washing or brushing hair; using a knife and fork together; and handling small coins
2	Symptoms, in one arm or both arms, affecting but not preventing any of the above-mentioned functions
3	Symptoms, in one arm or both arms, preventing one or two of the above-mentioned functions
4	Symptoms, in one arm or both arms, preventing three or all of the functions listed, but some purposeful movements still possible
5	Inability to use either arm for any purposeful movement
Grade	**Leg Disability**
0	Walking not affected
1	Walking affected, but walks independently outdoors
2	Usually uses unilateral support (stick, single crutch, one arm) to walk outdoors
3	Usually uses bilateral support (stick, crutches, frame, two arms) to walk outdoors
4	Usually uses wheelchair to travel outdoors, but able to stand and walk a few steps with help
5	Restricted to wheelchair, unable to stand and walk a few steps with help

Inflammatory Rasch-built Overall Disability Scale (I-RODS) – intended to specifically assess activity and social participation limitations in patients with inflammatory neuropathies

Overall Disability Sum Score (ODSS) / Overall Neuropathy Limitations Scale (ONLS) – focuses on upper and lower limb functions and consists of a checklist for interviewing patients. The ODSS was the first scale designed to assess the limitations of patients with immune-mediated peripheral neuropathies. To reduce a possible ceiling

effect, the ODSS was modified to include climbing stairs and running. The new measure is called the ONLS.

Grade	Arm Disability
0	Normal
1	Minor symptoms in one or both arms but not affecting any of the functions listed
2	Disability in one or both arms affecting but not preventing any of the functions listed
3	Disability in one or both arms preventing at least one but not all functions listed
4	Disability in both arms preventing all functions listed but purposeful movement still possible
5	Disability in both arms preventing all purposeful movements
Grade	**Leg Disability**
0	Walking/climbing stairs/running not affected
1	Walking/climbing stairs/running is affected, but gait does not look abnormal
2	Walks independently but gait looks abnormal
3	Requires unilateral support to walk 10 metres (stick, single crutch, one arm)
4	Requires bilateral support to walk 10 metres (sticks, crutches, crutch and arm, frame)
5	Requires wheelchair to travel 10 metres but able to stand and walk 1 metre with the help of one person
6	Restricted to wheelchair, unable to stand and walk 1 metre with the help of one person, but able to make some purposeful leg movements
7	Restricted to wheelchair or bed most of the day, unable to make any purposeful movements of the legs

Refractory Myasthenia Gravis Disease – unchanged or worse disease after corticosteroids and at least 2 other immunosuppressants, used in adequate doses for an adequate duration, with persistent symptoms or side effects that limit functioning, as defined by patient and physician

POLICY

It is the policy of the Health Plan to maintain a prior authorization process that promotes appropriate utilization of specific drugs with potential for misuse or limited indications. This process involves a review using Food and Drug Administration (FDA) criteria to make a determination of Medical Necessity and approval by the Medical Policy Committee.

The drugs, intravenous immune globulin (IVIG) and subcutaneous immune globulin (SCIG), are subject to the prior authorization process.

PROCEDURE

IVIG and SCIG are used to increase circulating levels of gamma globulin in certain immunoglobulin deficiency states and in treatment of a limited number of specified diseases.

Must meet all the criteria listed below:

ALL REQUESTS:

- Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling
    - Ideal body weight (IBW) should be used to calculate the dose for members that are not obese
        - Actual body weight should be used if actual body weight is less than ideal body weight (IBW)
    - For obese members (BMI > 30 kg/m^2), adjusted Body Weight (ABW) must be used to calculate the dose instead of actual or ideal body weight
- Body weight calculator available at: https://www.mdcalc.com/ideal-body-weight-adjusted-body-weight
- **DOSING NOTES** are included below for several indications in which clinical guidelines recommend a specific regimen. For all dosing requests that exceed the recommended dose by either guidelines, compendia, or the manufacturer's dosing guidelines – review using off-label dosing criteria

FDA Approved Indications

1. **Primary Immunodeficiency**
    - Syndromes may include:
        - Common Variable Immunodeficiency (Hypogammaglobulinemia)
        - Congenital Agammaglobulinemia
        - Bruton's or X-linked Agammaglobulinemia
        - Severe Combined Immunodeficiency (SCID)
        - X-linked Hyper-IgM Syndrome
        - Wiskott-Aldrich Syndrome
        - Hypergammaglobulinemia Types
    - Must be prescribed by or in consultation with an immunologist or hematologist
    - Must have deficient antibody production, as evidenced through a documented IgG level ≤500mg/dL
        - Requests with IgG levels >500mg/dL require chart documentation that provides clinical rationale for the use of IVIG or SCIG (**NOTE:** Several primary immunodeficiencies do have normal levels of IgG with documented specific antibody deficiency)[149]
    - Must have history of at least 1 infection directly attributable to this deficiency
    - Initial approval is 1 year
    - Reauthorizations are granted for 1-year intervals based upon documentation from the prescriber of an updated IgG level and that the member's condition has improved as a result of treatment

2. Immune Thrombocytopenia [Idiopathic Thrombocytopenic Purpura (ITP)]

- Must be prescribed by a hematologist or oncologist
- For children with ITP:
  - Must have ONE of the following (platelet counts expressed per mm^3):
    - Active bleeding AND platelet count <30,000
    - Upcoming invasive surgery AND either platelet level below threshold designated for procedure (threshold must be provided with request) OR blood loss is expected
    - Non-life-threatening mucosal bleeding and/or diminished quality of life AND documented previous inadequate response or intolerance to corticosteroids
- For adults with ITP:
  - Must have ONE of the following:
    - Active bleeding AND platelet count <30,000
    - Upcoming invasive surgery AND platelet level below threshold designated for procedure (threshold must be provided with request)
    - Platelet count <30,000 AND documented previous inadequate response or intolerance to corticosteroids
- For pregnant women with ITP:
  - Must be pregnant and have ONE of the following:
    - Platelet count <50,000
    - Upcoming invasive surgery/procedure
    - History of splenectomy
    - Previously delivered infants with autoimmune thrombocytopenia
- Initial approval is 1 month
- Reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing the previous response and clinical rationale for re-treatment
- **DOSING NOTE:** Clinical guidelines recommend up to 1,000mg/kg/dose as a 1-time dose; dosage may be repeated if necessary[151]

3. Kawasaki Disease

- Must be receiving aspirin concomitantly
- Must be requesting treatment within the first 10 days of illness
  - If greater than 10 days after illness onset, must have persistent signs of inflammation (e.g., persistent fever without explanation, elevated ESR or CRP, coronary artery aneurysms)
- Initial approval is 1 dose
- First reauthorization is granted (approval of only 1 dose) based upon documentation showing the member failed to respond

- Additional reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing the previous response and clinical rationale for re-treatment
- **DOSING NOTE:** Clinical guidelines recommend 2 grams/kg[152]

4. Chronic B-cell Lymphocytic Leukemia

- Must be prescribed by a hematologist, oncologist, or infectious disease specialist
- Must have Hypogammaglobulinemia (IgG <500mg/dL)
- Must have previous history of serious infection (requiring antibiotics)
- Initial approval is 1 year
- Reauthorizations are granted at 1-year intervals based upon documentation from the prescriber indicating that the member's condition has improved as a result of treatment

5. HIV (Human Immunodeficiency Virus) in pediatric patients

- Must be prescribed by an immunologist or infectious disease specialist
- Must be <13 years old
- Must have CD4 count ≥200/mm^3
- Must have ONE of the following
    - Recurrent (2 or more) serious infections such as bacteremia, meningitis, or pneumonia during a 1-year period despite administration of highly active antiretroviral therapy (HAART) and prophylactic sulfamethoxazole/trimethoprim (TMP-SMZ) or other antimicrobials
    - Hypogammaglobulinemia with an IgG <400mg/dL
    - Absence of detectable antibodies to common antigens, (measles, pneumococcal, and/or *haemophilus influnzae* Type B)
    - Bronchiectasis not optimally responsive to antibiotics and pulmonary therapy
    - A need for passive immunization for measles if Intramuscular Immune Globulin (IMIG) is contraindicated. IM injection contraindicated with severe thrombocytopenia or any coagulation disorder
- Initial approval is 1 year
- Reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing previous response and clinical rationale for re-treatment
- **DOSING NOTE:** Clinical guidelines recommend 400mg/kg every 2-4 weeks[153]

6. Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

- Must be prescribed by a neurologist
- Must have a diagnosis of CIDP
- Must provide documentation of electrodiagnostic testing (an EMG report)
- Must have moderate-to-severe functional disability
- Must provide a baseline disability score (using a validated disability scale, such as I-RODs, ODSS, ONLS, or INCAT)
- Initial approval is 1 year
- Reauthorizations are granted at 1-year intervals based upon documentation from the prescriber indicating the member's condition has improved as a result of treatment, as evidenced by improvement or stability in the member's baseline disability score (using a validated disability scale, such as I-RODS, ODSS, ONLS, or INCAT).

7. Multifocal Motor Neuropathy (MMN)

- Must be prescribed by a neurologist
- Must provide chart note documentation supporting a clinical examination of the member and BOTH of the following:
  - Progressive, asymmetric limb weakness over a course of at least 1 month
  - No objective sensory abnormalities except for minor vibration sense abnormalities in the lower limbs
- Must provide documentation of electrophysiologic findings that support the diagnosis of MMN (such as nerve conduction studies showing conduction blocks and EMG report)
- Initial approval is 1 year
- Reauthorizations are granted for 1-year intervals based upon documentation from the prescriber indicating the member's condition has improved as a result of treatment
- **DOSING NOTE:** Clinical guidelines recommend up to 2g/kg given over 2-5 days[154]

Off-label Uses

1. Guillain-Barre Syndrome

- Must start immune globulin within 4 weeks of onset of neuropathic symptoms
- Must be unable to walk independently
- Initial approval is 1 month
- First reauthorization is granted at a duration of 1 additional month

- Additional reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing previous response to treatment and clinical rationale for re-treatment

2. Dermatomyositis and Polymyositis (including juvenile)

- Must have dermatomyositis and polymyositis confirmed by biopsy
- Must have tried and failed or have a contraindication to both of the following:
  - Corticosteroids for 3 months
  - Concomitant adjuvant therapy (azathioprine, methotrexate, cyclosporine)
- Initial approval is 1 year
- Reauthorizations are granted for 1-year intervals based upon documentation from the prescriber indicating the member's condition has improved as a result of treatment

3. Systemic Lupus Erythematosus (SLE)

- Must have severe active SLE
- Must have tried and failed or have contraindications to ALL of the following:
  - Corticosteroids
  - Antimalarials
  - 1 additional immunosuppressant (e.g., azathioprine, cyclophosphamide, cyclosporine, methotrexate)
- Initial approval is 1 year
- Reauthorizations are granted for 1-year intervals based upon documentation from the prescriber indicating the member's condition has improved as a result of treatment

4. Multiple Sclerosis (MS)

- Must be prescribed by a neurologist
- For acute exacerbations of MS:
  - Must have a trial and failure or have contraindications to corticosteroids or plasma exchange
  - Initial approval is 1 month
  - Reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing previous response to treatment and clinical rationale for re-treatment if immune globulin did not provide a sufficient response
- For chronic maintenance treatment of MS:
  - Must have relapsing, remitting type of MS

- Must have a trial and failure (duration of at least 3 months) or have contraindications to ALL the following:
  - At least one interferon [interferon beta-1a (Rebif®) or interferon beta-1b (Betaseron®)]
  - Glatiramer (Copaxone®)
  - Fingolimod (Gilenya®)
- No previous trials are required if:
  - Member is pregnant
  - Member is immunosuppressed or is having recurrent infections
- Initial approval is 1 year
- Reauthorizations are granted for 1-year intervals, subject to the above criteria, and require chart documentation describing previous response to treatment and clinical rationale for re-treatment

5. Autoimmune Mucocutaneous Blistering Disease (AMBD)

- Must have ONE of the following supported by biopsy:
  - Pemphigus vulgaris,
  - Pemphigus foliaceus,
  - Bullous pemphigoid,
  - Mucous membrane pemphoid (a.k.a., cictrical pemphigoid), OR
  - Epidermolysis bullosa acquisita
- Must have a trial and failure or have contraindications to corticosteroids or immunosuppressive agents
  - **EXCEPTION:** In rapidly progressive, extensive, or debilitating cases, immune globulin may be approved along with corticosteroids or immunosuppressive agents
- Initial approval is 4 months
- Reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing previous response to treatment and clinical rationale for re-treatment

6. Myasthenia Gravis Syndrome

- Must have a diagnosis of Myasthenia Gravis
- Must be prescribed by a neurologist
- For acute use:
  - Chart documentation of acute exacerbation and impaired function is required (e.g., respiratory insufficiency, inability to swallow)
  - Initial approval is 1 month

    - Reauthorizations are granted on a case-by-case basis with chart documentation describing previous response to treatment and clinical rationale for re-treatment
- For temporary use as a bridge to immunotherapy:
    - Must have a history of myasthenia gravis exacerbation
    - Must be recently started (within 3 months) on immunosuppressant therapy (e.g., azathioprine, mycophenolate, cyclosporine, or tacrolimus)
    - Chart documentation of use as bridge therapy is required
    - Initial approval is for 6 months
    - Reauthorizations are granted on a case-by-case basis with chart documentation describing previous response to treatment and clinical rationale for re-treatment
- For stabilization prior to surgery:
    - Must have a history of myasthenia gravis with current or previous difficulty with swallowing, speech, or respiratory involvement (e.g., shortness of breath or reduced force vital capacity on pre-op pulmonary function test). Chart documentation of symptoms is required.
    - IVIG infusion must be scheduled within 14 days of anticipated surgery date
    - Initial approval is for 1 month
    - Reauthorizations are granted on a case-by-case basis with chart documentation describing previous response to treatment and clinical rationale for re-treatment
- For chronic use in refractory disease:
    - Must have an adequate trial with inadequate response, significant side effects/toxicity, or have a contraindication to BOTH of the following:
        - Cholinesterase inhibitors (pyridostigmine or neostigmine)
        - Corticosteroids
    - Must have an adequate trial (of at least 3 months each) with inadequate response, significant side effects/toxicity, or have a contraindication to TWO of the following:
        - Azathioprine
        - Mycophenolate mofetil
        - Cyclosporine
        - Tacrolimus
    - Initial approval is for 1 year
    - Reauthorizations for patients with refractory disease are granted for 1-year intervals on a case-by-case basis and require chart documentation describing previous response to treatment, including improvement in symptoms that limit daily function

7. Parvovirus B19 Infection

- Must have documentation (e.g., Polymerase Chain Reaction test result) confirming presence of HPV-B19 infection
- Must have severe anemia defined as hemoglobin level <8ng/dL
- Must have low reticulocyte count defined as $<8x10^9$/L
- Must have history of immunodeficiency due to suppressive medications or HIV
- Initial approval is 1 month
- Reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing previous response to treatment and clinical rationale for re-treatment
- **DOSING NOTE:** Clinical guidelines recommend 400-500mg for 5 days[154]

8. Renal and/or Pancreatic Transplant Desensitization in Combination with Rituxan

- Must be prescribed by a transplant specialist
- Must be age 18 or older
- Must be awaiting kidney and/or pancreas transplant requiring desensitization as defined by the following criteria:
    - For deceased donor transplants:
        - Panel reactive antibody (PRA) level >30% **OR**
        - PRA <30% with previous kidney and/or pancreas transplant
    - For living donor transplants:
        - Positive crossmatch **OR**
        - Positive donor-specific antibody using Luminex® assay
- Initial approval is 1 course of treatment (2 doses)
- Reauthorizations are subject to the above criteria and are not granted until 6 months have passed since the initial treatment

9. Renal Transplant Desensitization

- Must be prescribed by a transplant specialist
- Must be awaiting kidney transplant (either from a living or deceased donor) and requiring desensitization
- Initial approval is 4 months
- Reauthorizations are subject to the above criteria and are not granted until 12 months have passed since the initial treatment

10. Renal Post-Transplant Rejection

- Must have received a renal transplant from a living donor with post-transplant rejection
- Initial approval is 1 month
- Reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing previous response to treatment and clinical rationale for re-treatment

11. Allogenic or Hematopoietic Stem Cell Transplantation (HSCT) (or Bone Marrow Transplant)

- Must have severe hypogammaglobulinemia (IgG <400 mg/dL)
- Must have history of recurrent infections
- Initial approval is 6 months
- Reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing the previous response to treatment and clinical rationale for re-treatment

12. Autoimmune Hemolytic Anemia

- Must have warm-type diagnosis
- Must have a trial and failure or have a contraindication to corticosteroids
- Initial approval is 1 month
- Reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing the previous response to treatment and clinical rationale for re-treatment

13. Stiff-Person Syndrome

- Must have a diagnosis of Stiff-Person Syndrome confirmed by electromyography (EMG) or elevated levels of glutamic acid decarboxylase (GAD)
- Must be prescribed by a neurologist
- Must have an adequate trial with inadequate responses, significant side effects/toxicity, or have contraindications to THREE of the following:
    - Corticosteroids
    - Antiepileptics
    - Benzodiazepines
    - Muscle relaxants
    - Gabapentin
- Initial approval is 4 months

- Reauthorization for an additional 2 months of treatment may be made on a case-by-case basis, is subject to the above criteria, and require chart documentation describing the previous response to treatment and clinical rationale for re-treatment. Continued use beyond 6 months of therapy is not authorized.

14. Pediatric Acute-Onset Neuropsychiatric Syndrome (PANS)

- Documented diagnosis of pediatric acute-onset neuropsychiatric syndrome (PANS) made by or in consultation with a pediatric psychiatrist or pediatric neurologist
- Must meet the following criteria consistent with PANS:
    - Abrupt, dramatic onset of obsessive-compulsive disorder or severely restricted food intake
    - Concurrent presence of additional neuropsychiatric symptoms, (with similarly severe and acute onset), from at least two of the following seven categories
        - Anxiety
        - Emotional lability and/or depression
        - Irritability, aggression, and/or severely oppositional behaviors
        - Behavioral (developmental) regression
        - Deterioration in school performance (related to attention deficit/hyperactivity disorder [ADHD]-like symptoms, memory deficits, cognitive changes)
        - Sensory or motor abnormalities
        - Somatic signs and symptoms, including sleep disturbances, enuresis, or urinary frequency
    - Symptoms are not better explained by a known neurologic or medical disorder, such as Sydenham chorea (SC)
- Initial approval is 1 month
- Reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing the previous response and clinical rationale for re-treatment

15. Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infection (PANDAS)

- Documented diagnosis of pediatric autoimmune neuropsychiatric disorders associated with streptococcal infection (PANDAS) made by or in consultation with a pediatric psychiatrist or pediatric neurologist
- Must meet the following criteria consistent with PANDAS
    - Obsessive-compulsive disorder (OCD) and/or tic disorder (such as Tourette disorder, chronic motor or vocal tic disorder)
    - Pediatric onset (between three years and onset of puberty)
    - Abrupt onset and episodic course of symptoms

  - Temporal relation between group A streptococcal (GAS) infection and onset and/or exacerbation
  - Neurologic abnormalities, such as motoric hyperactivity, choreiform movements, or tics during exacerbations
- Initial approval is 1 month
- Reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing the previous response and clinical rationale for re-treatment

16. Steven's Johnson Syndrome (SJS) or Toxic Epidermal Necrolysis (TEN)

- Initial approval is 1 week
- Reauthorizations are granted on a case-by-case basis, are subject to the above criteria, and require chart documentation describing the previous response and clinical rationale for re-treatment
- **DOSING NOTE:** Package insert/clinical guidelines recommend 1 g/kg/day in divided doses for 3-4 days[155]

17. CAR-T Therapy-Related Toxicity

- Must be prescribed by a hematologist, oncologist, or infectious disease specialist
- Must have Hypogammaglobinemia (IgG level <400mg/dL)
- Must have documentation of serious or recurrent infections
- Initial approval is 1 year
- Reauthorizations are granted at 1-year intervals based upon documentation from the prescriber indicating that the member's condition has improved as a result of treatment
- **DOSING NOTE:** Clinical guidelines recommend 400-500mg/kg monthly[155]

Limitations:

Length of Authorization (if above criteria met)	
Initial Authorization	Case-by-case bases (see criteria above)
Reauthorization	Same as initial

If the established criteria are not met, the request is referred to a Medical Director for review.

Codes:

CPT Codes / HCPCS Codes / ICD-10 Codes		
Code	Brand	Description
J1459	PRIVIGEN	INJECTION, IMMUNE GLOBULIN IV, NON-LYOPHILIZED, 500MG
J1460	GAMASTAN	INJECTION, GAMMA GLOBULIN, INTRAMUSCULAR, 1 CC
J1551	CUTAQUIG	INJECTION, IMMUNE GLOBULIN (CUTAQUIG), 100 MG
J1554	ASCENIV	INJECTION, IMMUNE GLOBULIN (ASCENIV), 500 MG
J1555	CUVITRU	INJECTION, IMMUNE GLOBULIN, 100 MG
J1556	BIVIGAM	INJECTION, IMMUNE GLOBULIN (BIVIGAM), 500 MG
J1557	GAMMAPLEX	INJECTION, IMMUNE GLOBULIN, IV, NONLYOPHILIZED, 500 MG
J1558	XEMBIFY	INJECTION, IMMUNE GLOBULIN, 100 MG
J1559	HIZENTRA	INJECTION, IMMUNE GLOBULIN (HIZENTRA), 100 MG
J1560	GAMASTAN S-D	INJECTION, GAMMA GLOBULIN, INTRAMUSCULAR, OVER 10 CC
J1561	GAMUNEX-C, GAMMAKED	INJECTION, IMMUNE GLOBULIN, NONLYOPHILIZED, 500 MG
J1566	GAMMAGARD S/D, CARIMUNE NF	INJECTION, IMMUNE GLOBULIN, INTRAVENOUS, LYOPHILIZED, NOT OTHERWISE SPECIFIED, 500 MG
J1568	OCTAGAM	INJECTION, IMMUNE GLOBULIN, INTRAVENOUS, NON-LYOPHILIZED, 500 MG
J1569	GAMMAGARD	INJECTION, IMMUNE GLOBULIN, INTRAVENOUS, NON-LYOPHILIZED, 500 MG
J1572	FLEBOGAMMA, FLEBOGAMMA DIF	INJECTION, IMMUNE GLOBULIN, INTRAVENOUS, NON-LYOPHILIZED, 500 MG
J1575	HYQVIA	INJECTION, IMMUNE GLOBULIN/HYALURONIDASE 100 MG
J1576	Panzyga	INJECTION, IMMUNE GLOBULIN (PANZYGA), INTRAVENOUS, NON-LYOPHILIZED (E.G., LIQUID), 500 MG

REFERENCES

1. Immune globulin intravenous (human) In: Drug information for the health care professional. USPDI—Volume I. 25rd ed. Greenwood Village, CO: 2005.; Thomson Reuters Micromedex; 2012.
2. Knezevic-Maramica I, Kruskall MS. Intravenous immune globulins: an update for clinicians. *Transfusion.* 2003;43:1460-1480.
3. Carimune NF [package insert]. Kankakee, IL: CSL Behring LLC; January 2013.

4. Flebogamma 5% solution [package insert]. Los Angeles, CA: Grifols USA, Inc (manufactured by Instituto Grifols, SA, Barcelona, Spain); January 2004.
5. Gammagard Liquid [package insert]. Westlake Village, CA: Baxter Healthcare Corporation; July 2012.
6. Gamunex-C [package insert]. Research Triangle Park, NC: Grifols Therapeutics Inc. ; December 2015.
7. Octagam [package insert]. Vienna, Austria: Octapharma Pharmazeutika Produktionsges.m.b.H); June 2021.
8. Buckley RH, Schiff RI. The use of intravenous immune globulin in immunodeficiency diseases. *NEJM*. 1991;325:110-117.
9. Neunert et al. ASH 2011 Evidence Based Pratice Guideline for Idiopathic thrombocytopenia purpura. Blood April 2011. 4191-4204.
10. Cines DB, Blanchette VS. Immune thrombocytopenic purpura. *NEJM.* 2002 Mar 28;346:995-1008.
11. British Committee for Standards in Haematology General Haematology Task Force. Guidelines for the investigation and management of idiopathic thrombocytopenic purpura in adults, children and in pregnancy. *Br J Haematol.* 2003;120:574-596.
12. Vesely SK, Perdue JJ, Rizvi MA, et al. Management of adult patients with persistent idiopathic thrombocytopenic purpura following splenectomy: a systematic review. *Ann Intern Med.* 2004;140:112-120.
13. Buchanan GR, de Alarcon PA, Feig SA, et al. Acute idiopathic thrombocytopenic purpura – management in childhood. *Blood.* 1997;89:1464-1465.
14. Immune globulin (Drugdex® Drug Evaluation). Micromedex® Healthcare Series: Thomson Micromedex, Greenwood Village, Colorado. Accessed:January 2012
15. Newburger JW, Takahashi M, Gerber MA, et al; Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease; Council on Cardiovascular Disease in the Young; American Heart Association; American Academy of Pediatrics. Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. *Circulation.* 2004;110:2747-2771.
16. Ratko TA, Burnett DA, Foulke GE, et al. Recommendations for off-label use of intravenously administered immunoglobulin preparations*. JAMA*. 1995;273:1865-1870.
17. Ballow M. Intravenous immunoglobulins: clinical experience and viral safety. *J Am Pharm Assoc*. 2002;42:449-459.
18. British Columbia Blood Coordinating Office. *IVIG utilization management handbook.* 1st Ed. British Columbia, Canada: Provincial Blood Coordinating Office; April 2002. http://www.bloodlink.bc.ca/ivig.html Accessed 03/14/2005.
19. [No authors listed]. Intravenous immunoglobulin for the prevention of infection in chronic lymphocytic leukemia. A randomized, controlled clinical trial. Cooperative Group for the Study of Immunoglobulin in Chronic Lymphocytic Leukemia. *N Engl J Med.* 1988;319:902-907.
20. Gamm H, Huber C, Chapel H, et al. Intravenous immune globulin in chronic lymphocytic leukaemia. *Clin Exp Immunol.* 1994;97 Suppl 1:17-20.
21. Centers for Disease Control and Prevention; Infectious Disease Society of America; American Society of Blood and Marrow Transplantation. Guidelines for preventing opportunistic infections among hematopoietic stem cell transplant recipients. *MMWR Recomm Rep.* 2000 Oct;49(RR-10):1-125, CE1-7.
22. Sullivan KM, Storek J, Kopecky KJ, et al. A controlled trial of long-term administration of intravenous immunoglobulin to prevent late infection and chronic graft-vs.-host disease after marrow transplantation: clinical outcome and effect on subsequent immune recovery. *Biol Blood Marrow Transplant.* 1996;2:44-53.
23. Mouthon L, Lortholary O. Intravenous immunoglobulins in infectious diseases: where do we stand? *Clin Microbiol Infect.* 2003;9:333-338.

24. Centers for Disease Control and Prevention. Guidelines for preventing opportunistic infections among HIV-infected persons – 2002 recommendations of the U.S. Public Health Service and the Infectious Diseases Society of America. *MMWR*. 2002;51(No. RR-8):1-52.
25. American Academy of Pediatrics. **Human Immunodeficiency Virus Infection**. In: Pickering LK, ed. *Red Book: 2003 Report of the Committee on Infectious Diseases*. 26th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2003:360-382.
26. [No authors listed]. Antiretroviral therapy and medical management of pediatric HIV infection and 1997 USPHS/IDSA report on the prevention of opportunistic infections in persons infected with human immunodeficiency virus. *Pediatrics.* 1998;102(4 Pt 2):999-1085.
27. Mofenson LM, Oleske J, Serchuck L, et al; CDC; National Institutes of Health; Infectious Diseases Society of America. Treating opportunistic infections among HIV-exposed and infected children: recommendations from CDC, the National Institutes of Health, and the Infectious Diseases Society of America. *MMWR Recomm Rep.* 2004 Dec 3;53(RR-14):1-92.
28. Spector SA, Gelber RD, McGrath N, et al. A controlled trial of intravenous immune globulin for the prevention of serious bacterial infections in children receiving zidovudine for advanced human immunodeficiency virus infection. Pediatric AIDS Clinical Trials Group. *N Engl J Med.* 1994;331:1181-1187.
29. Ahmed AR, Dahl MV. Consensus statement on the use of intravenous immunoglobulin therapy in the treatment of autoimmune mucocutaneous blistering diseases. *Arch Dermatol.* 2003;139:1051-1059.
30. Ruetter A, Luger TA. Efficacy and safety of intravenous immunoglobulin for immune-mediated skin disease. Current view. *Am J Clin Dermatol.* 2004;5:153-160.
31. Dalakas MC. Intravenous immunoglobulin in autoimmune neuromuscular diseases. *JAMA.* 2004;291:2367-2375.
32. Hadden RD, Hughes RA. Management of inflammatory neuropathies. *J Neurol Neurosurg Psychiatry*. 2003;74 Suppl 2:ii9-ii14.
33. Danieli MG, Cappelli M, Malcangi G, et al. Long term effectiveness of intravenous immunoglobulin in Churg-Strauss syndrome. *Ann Rheum Dis.* 2004;63:1649-1654.
34. Tsurikisawa N, Taniguchi M, Saito H, et al. Treatment of Churg-Strauss syndrome with high-dose intravenous immunoglobulin. *Ann Allergy Asthma Immunol.* 2004;92:80-87.
35. Ljungman P, Reusser P, de la Camara R, et al; for the Infectious Diseases Working Party of the European Group for Blood and Marrow Transplantation. Management of CMV infections: recommendations from the infectious diseases working party of the EBMT. *Bone Marrow Transplant*. 2004;33:1075-1081.
36. Sokos DR, Berger M, Lazarus HM. Intravenous immunoglobulin: appropriate indications and uses in hematopoietic stem cell transplantation. *Biol Blood Marrow Transplant*. 2002;8:117-130.
37. John R, Lietz K, Burke E, et al. Intravenous immunoglobulin reduces anti-HLA alloreactivity and shortens waiting time to cardiac transplantation in highly sensitized left ventricular assist device recipients. *Circulation.* 1999;100[suppl II]:II-229-II235.
38. Itescu S, Burke E, Lietz K, et al. Intravenous pulse administration of cyclophosphamide is an effective and safe treatment for sensitized cardiac allograft recipients. *Circulation.* 2002;105:1214-1219.
39. Jordan SC, Tyan D, Stablein D, et al. Evaluation of intravenous immunoglobulin as an agent to lower allosensitization and improve transplantation in highly sensitized adult patients with end-stage renal disease: report of the NIH IG02 trial. *J Am Soc Nephrol.* 2004;15:3256-3262.
40. Mackay MT, Weiss SK, Adams-Webber T, et al; American Academy of Neurology; Child Neurology Society. Practice parameter: medical treatment of infantile spasms: report of the American Academy of Neurology and the Child Neurology Society. *Neurology.* 2004;62:1668-1681.
41. Dykes AC, Walker ID, Lowe GD, et al. Combined prednisolone and intravenous immunoglobulin treatment for acquired factor VIII inhibitors: a 2-year review. *Haemophilia.* 2001;7:160-163.
42. Collins PW. Management of acquired haemophilia A – more questions than answers. *Blood Coagul Fibrinolysis.* 2003;14 Suppl 1:S23-27.
43. Rubinger M, Rivard GE, Teitel J, et al, the Inhibitor Subcommittee of the Association of Hemophilia Clinic Directors of Canada (AHCDC). Suggestions for the management of hemophiliacs and non-

hemophiliacs with factor VIII inhibitors. 3rd ed. May 14, 1999. http://www.ahcdc.ca/Suggestions.htm Accessed 3/10/2005.
44. Baschieri L, Antonelli A, Nardi S, et al. Intravenous immunoglobulin versus corticosteroid in treatment of Graves' ophthalmopathy. *Thyroid.* 1997;7:579-585.
45. Kahaly G, Pitz S, Muller-Forell W, et al. Randomized trial of intravenous immunoglobulins versus prednisolone in Graves' ophthalmopathy. *Clin Exp Immunol.* 1996;106:197-202.
46. Hughes RA, Wijdicks EF, Barohn R, et al; Quality Standards Subcommittee of the American Academy of Neurology. Practice parameter: immunotherapy for Guillain-Barre syndrome: report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology.* 2003;61:736-740.
47. Jahnke L, Applebaum S, Sherman LA, et al. An evaluation of intravenous immunoglobulin in the treatment of human immunodeficiency virus-associated thrombocytopenia. *Transfusion.* 1994;34:759-764.
48. Majluf-Cruz A, Luna-Castanos G, Huitron S, et al. Usefulness of a low-dose intravenous immunoglobulin regimen for the treatment of thrombocytopenia associated with AIDS. *Am J Hematol.* 1998;59:127-132.
49. WinRho SDF® [package insert]. Westlake Village, CA (manufactured by Cangene Corporation, Winnipeg, Canada): October 2004.
50. Comi G, Roveri L, Swan A, et al; Inflammatory Neuropathy Cause and Treatment Group. A randomised controlled trial of intravenous immunoglobulin in IgM paraprotein associated demyelinating neuropathy. *J Neurol.* 2002;249:1370-1377.
51. Cherin P, Pelletier S, Teixeira A, et al. Intravenous immunoglobulin for dysphagia of inclusion body myositis. *Neurology.* 2002;58:326.
52. Jordan SC, Vo AA, Nast CC, et al. Use of high-dose human intravenous immunoglobulin therapy in sensitized patients awaiting transplantation: the Cedars-Sinai experience. *Clin Transpl.* 2003;:193-198.
53. Jordan S, Cunningham-Rundles C, McEwan R. Utility of intravenous immune globulin in kidney transplantation: efficacy, safety, and cost implications. *Am J Transplant.* 2003;3:653-664.
54. Maddison P, Newsom-Davis J. Treatment for Lambert-Eaton myasthenic syndrome. *Cochrane Database Syst Rev.* 2003;(2):CD003279.
55. Achiron A, Kishner I, Sarova-Pinhas I, et al. Intravenous immunoglobulin treatment following the first demyelinating event suggestive of multiple sclerosis: a randomized, double-blind, placebo-controlled trial. *Arch Neurol.* 2004;61:1515-1520.
56. Gray O, McDonnell GV, Forbes RB. Intravenous immunoglobulins for multiple sclerosis. *Cochrane Database Syst Rev.* 2003;(4):CD002936.
57. Goodin DS, Frohman EM, Garmany GP Jr, et al; Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology and the MS Council for Clinical Practice Guidelines. Disease modifying therapies in multiple sclerosis: report of the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology and the MS Council for Clinical Practice Guidelines. *Neurology.* 2002;58:169-178.
58. Achiron A, Kishner I, Dolev M, et al. Effect of intravenous immunoglobulin treatment on pregnancy and postpartum-related relapses in multiple sclerosis. *J Neurol.* 2004;251:1133-1137.
59. Bux J, Behrens G, Jaeger G, et al. Diagnosis and clinical course of autoimmune neutropenia in infancy: analysis of 240 cases. *Blood.* 1998;91:181-186.
60. Herrod HG. Management of the patient with IgG subclass deficiency and/or selective antibody deficiency. *Ann Allergy.* 1993;70:3-8.
61. Lawton AR. IgG subclass deficiency and the day-care generation. *Pediatr Infect Dis J.* 1999;18:462-466.
62. Buckley RH. Immunoglobulin G subclass deficiency: fact or fancy? *Curr Allergy Asthma Reports.* 2002;2:356-360.

63. Silk HJ, Ambrosino D, Geha RS. Effect of intravenous gammaglobulin therapy in IgG2 deficient and IgG2 sufficient children with recurrent infections and poor response to immunization with Hemophilus influenzae type b capsular polysaccharide antigen. *Ann Allergy.* 1990;64:21-25.
64. Bjorkander J, Bengtsson U, Oxelius VA, et al. Symptoms in patients with lowered levels of IgG subclasses, with or without IgA deficiency, and effects of immunoglobulin prophylaxis. *Monogr Allergy.* 1986;20:157-163.
65. Dalakas MC, Fujii M, Li M, et al. High-dose intravenous immune globulin for stiff-person syndrome. *N Engl J Med.* 2001;345:1870-1876.
66. Ioannou Y, Isenberg DA. Current concepts for the management of systemic lupus erythematosus in adults: a therapeutic challenge. *Postgrad Med J.* 2002;78:599-606.
67. O'Donnell BF, Barr RM, Kobza A, et al. Intravenous immunoglobulin in autoimmune chronic urticaria. *Br J Dermatol.* 1998;138:101-106.
68. Asero R. Are IVIG for chronic unremitting urticaria effective? *Allergy.* 2000;55:1099-1101.
69. Dawn G, Urcelay M, Ah-Weng A, et al. Effect of high-dose intravenous immunoglobulin in delayed pressure urticaria. *Br J Dermatol.* 2003;149:836-840.
70. Joint Tasks Force on Practice Parameters. The diagnosis and management of urticaria: a practice parameter part I: acute urticaria/angioedema part II: chronic urticaria/angioedema. Ann Allergy Asthma Immunol 2000;85(6 Pt 2):521-544.
71. Becker MD, Rosenbaum JT. Current and future trends in the use of immunosuppressive agents in patiens with uveitis. *Curr* Opin Ophthalmol. 2000;11:472-477.
72. LeHoang P, Cassoux N, George F, et al. Intravenous immunoglobulin (IVIg) for the treatment of birdshot retinochoroidopathy. *Ocul Immunol Inflamm.* 2000;8:49-57.
73. Jayne DR, Chapel H, Adu D, et al. Intravenous immunoglobulin for ANCA-associated systemic vasculitis with persistent disease activity. *QJM.* 2000;93:433-439.
74. Relkin N, Szabo, Adamiak B, et al. Intravenous immunoglobulin (IVIG) treatment causes dose-dependent alterations in β-amyloid (Aβ) levels and anti- Aβ antibody titers in plasma and cerebrospinal fluid (CSF) of Alzheimer's disease (AD) patients [abstract]. Presented at: American Academy of Neurology 57th Annual Meeting; April 12, 2005: Miami Beach, FL.
75. Dodel RC, Du Y, Depboylu C, et al. Intravenous immunoglobulins containing antibodies against beta-amyloid for the treatment of Alzheimer's disease. *J Neurol Neurosurg Psychiatry.* 2004;75:1472-1474.
76. Hack CE, Scheltens P. Intravenous immunoglobulins: a treatment for Alzheimer's disease? *J Neurol Neurosurg Psychiatry.* 2004;75:1374-1375.
77. Niven AS, Argyros G. Alternate treatments in asthma. *Chest.* 2003;123:1254-1265.
78. Jolles S, Sewell C, Webster D, et al. Adjunctive high-dose intravenous immunoglobulin treatment for resistant atopic dermatitis: efficacy and effects on intracellular cytokine levels and CD4 counts. *Acta Derm Venereol.* 2003;83:433-437.
79. Paul C, Lahfa M, Bachelez H, et al. A randomized controlled evaluator-blinded trial of intravenous immunoglobulin in adults with severe atopic dermatitis. *Br J Dermatol.* 2002;147:518-522.
80. Jolles S. A review of high-dose intravenous immunoglobulin treatment for atopic dermatitis. *Clin Exp Dermatol.* 2002;27:3-7.
81. Jolles S, Hughes J. Importance of trial design in studies using high-dose intravenous immunoglobulin. *Br J Dermatol.* 2003;148:1284-5; author reply 1285-6.
82. Wakim M, Alazard M, Yajima A, et al. High dose intravenous immunoglobulin in atopic dermatitis and hyper-IgE syndrome. *Ann Allergy Asthma Immunol.* 1998;81:153-158.
83. Noh G, Lee KY. Intravenous immune globulin (i.v.IG) therapy in steroid-resistant atopic dermatitis. *J Korean Med Sci.* 1999;14:63-68.
84. Hanifin JM, Cooper KD, Ho V, et al. Guidelines of care for atopic dermatitis. *J Am Acad Dermatol*. 2004;50:391-404.
85. Wolff SN, Fay JW, Herzig RH, et al. High-dose weekly intravenous immunoglobulin to prevent infections in patients undergoing autologous bone marrow transplantation or severe myelosuppressive therapy. A study of the American Bone Marrow Transplant Group. *Ann Intern Med.* 1993;118:937-942.

86. Seider N, Beiran I, Scharf J, et al. Intravenous immunoglobulin therapy for resistant ocular Behcet's disease. *Br J Ophthalmol.* 2001;85:1287-1288.
87. Cordonnier C, Chevret S, Legrand M, et al; GREFIG Study Group. Should immunoglobulin therapy be used in allogeneic stem-cell transplantation? A randomized, double-blind, dose effect, placebo-controlled, multicenter trial. *Ann Intern Med.* 2003;139:8-18.
88. Gottstein R, Cooke RW. Systematic review of intravenous immunoglobulin in haemolytic disease of the newborn. *Arch Dis Child Fetal Neonatal Ed.* 2003;88:F6-10.
89. Hommes OR, Sorensen PS, Fazekas F, et al. Intravenous immunoglobulin in secondary progressive multiple sclerosis: randomised placebo-controlled trial. *Lancet.* 2004;364:1149-1156.
90. Ohlsson A, Lacy JB. Intravenous immunoglobulin for suspected or subsequently proven infection in neonates. *Cochrane Database Syst Rev.* 2004:CD001239.
91. Ohlsson A, Lacy JB. Intravenous immunoglobulin for preventing infection in preterm and/or low-birth-weight infants. *Cochrane Database Syst Rev.* 2004;(1):CD000361.
92. Fanaroff AA, Korones SB, Wright LL, et al. A controlled trial of intravenous immune globulin to reduce nosocomial infections in very-low-birth-weight infants. National Institute of Child Health and Human Development Neonatal Research Network. *N Engl J Med.* 1994;330:1107-1113.
93. American College of Obstetricians and Gynecologists (ACOG). ACOG Practice Bulletin. Management of recurrent early pregnancy loss. Washington, DC: ACOG; 2001 Feb.
94. Branch DW, Peaceman AM, Druzin M, et al. A multicenter, placebo-controlled pilot study of intravenous immune globulin treatment of antiphospholipid syndrome during pregnancy. The Pregnancy Loss Study Group. *Am J Obstet Gynecol.* 2000;182(1 Pt 1):122-127.
95. Practice Committee of the American Society for Reproductive Medicine. Intravenous immunoglobulin (IVIG) and recurrent spontaneous pregnancy loss. *Fertil Steril.* 2004;82 Suppl 1:S199-200.
96. Levy Y, Amital H, Langevitz P, et al. Intravenous immunoglobulin modulates cutaneous involvement and reduces skin fibrosis in systemic sclerosis: an open-label study. *Arthritis Rheum.* 2004;50:1005-1007.
97. White B, Bauer EA, Goldsmith LA, et al. Guidelines for clinical trials in systemic sclerosis (scleroderma). I. Disease-modifying interventions. The American College of Rheumatology Committee on Design and Outcomes in Clinical Trials in Systemic Sclerosis. *Arthritis Rheum.* 1995;38:351-360.
98. Pereyra F, Rubin RH. Prevention and treatment of cytomegalovirus infection in solid organ transplant recipients. *Curr Opin Infect Dis.* 2004;17:357-361.
99. Preiksaitis JK, Brennan DC, Fishman J, et al. Canadian Society of Transplantation consensus workshop on cytomegalovirus management in solid organ transplantation final report. *Am J Transplant.* 2005;5:218-227.
100. Hizentra™ [package insert]. Bern, Switzerland: CSL Behring; April 2021.
101. Gammaplex® [package insert]. Elstree Hertfordshire, United Kingdom: Bio Products Laboratory; July 2015.
102. Shehata N, et al. The use of immunoglobulin therapy for patients with Primary Immune Deficiency: an evidence-based practice guideline. Transfusion Medicine Reviews 2010. 24(1): S28-S50.Yellen ES, et al. Performance of 2004 American Heart Association recommendations for treatment of Kawasaki disease. Pediatrics 2010. 125(2):e234-e241.
103. Mofenson L, et al. Guidelines for the Prevention and Treatment of Opportunistic Infections Among HIV-Exposed and HIV-Infected Children: Recommendations from CDC, the National Institutes of Health, the HIV Medicine Association of the Infectious Diseases Society of America, the Pediatric Infectious Diseases Society, and the American Academy of Pediatrics. MMWR Recomm Rep. 2009;10;58(RR-4):1-207.
104. Antoine JC, the French CIDP Study Group. Recommendations on diagnostic strategies for CIDP. Journal of Neurology, Neurosurgery, Psychiatry 2008. 79: 118.
105. Van Den Bergh PYK, et al. European Federation of Neurological Societies/PNS Guidelines on management of chronic inflammatory demyelinating polyradiculoneuropathy - first revision. European Journal of Neurology 2010. 17: 356-363.

106. Bromberg MB. Review of the Evolution of Electrodiagnositic Criteria for Chronic Inflammatory Demyelinating Polyradiculoneuropathy. Issues & Opinions in Muscle & Nerve; June 2011: 780-792.
107. Hughes RA, et al. Practice parameter: immunotherapy for Gullain-Barre syndrome: report of the Quality Standards Committee of the American Academy of Neurology. August 2, 2008.
108. Winer JB. When the Guillain Barre patient fails to respond to treatment. Practice Neurology 2009. 9:227-230.
109. Marie I, Mouthon L. Therapy of Polymyositis and dermatomyositis. Autoimmunity Reviews 2011. 11: 6-13.
110. Hak AE, et al. Dermatomyositis and polymyositis: new treatment targets on the horizon. Journal of Medicine 2011. 69(10): 415-416.
111. Aggarwal R, Oddis CN. Therapeutic Approaches in myositis. Current Rheumatology Rep 2011. 13: 182-191.
112. Berger JR. Functional improvement and symptom management in multiple sclerosis: clinical efficacy of current therapies. American Journal of Managed Care 2011. 17: S146-153.
113. Elovaara I, et al. Intravenous Immunoglobulins are a therapeutic option in the treatment of multiple sclerosis relapse. Clinical Neuropharmacology 2011. 34(2):84-88.
114. Rio J, et al. Multiple Sclerosis: current treatment algorithms. Current Opinion in Neurology 2011. 24: 230-237.
115. Feasby T, et al. Guidelines for the use of IVIG for neurological conditions. Transfus Med Rev 2007. 21 (2 suppl 1): S57-107.
116. Gold R, et al. Drug Insight: the use of IVIG in neurology. Nature Clinical Practice Neurology 2007. 3:36-44.
117. McGhee SA, et al. Persistent parvovirus-associated chronic fatigue treated with high dose IVIG. Pediatric Infectious Disease Journal 2005. 24(3):272-274.
118. Tomblyn M, et al. Guidelines for Preventing Infectious Complications among Hematopoietic Cell Transplantation Recipients. Biol Blood Marrow Transplant 2009. 15:1143-1238.
119. Yildirim-Toruner C, Diamond B. Current and novel therapeutics in the treatment of systemic lupus erythematosus. Clinical reviews in allergy and immunology 2010. 303-312.
120. Bayry J et al. Intravenous immunoglobulin treatment in rheumatic disease. Nature Reviews Rheumatology 2011. 7: 351-352.
121. Petz LP. Treatment of autoimmune hemolytic anemia. Current Opinion in Hematology. 2001;8:411-416.
122. Skele GO, et al. Guidelines for the treatment of autoimmune neuromuscular transmission disorders. Euro J of Neurology 2006. 13: 691-699.
123. Fazekas et al. Randomsed placebo-controlled trial of monthly intravenous immunoglobulin therapy in relapsing-remitting multiple sclerosis. Lancet 1997; 349:589-593.
124. Achiron et al. Intrevenous immunoglobulin treatment in multiple sclerosis: effect on relapses. Neurology 1998; 50: 398-402.
125. Sorenson et al. Intravenous immunoglobulin G reduced MRI activity in relapsing multiple sclerosis. Neurology 1998 50: 1273-1281.
126. Achiron et al. Intravenous immunoglobulin treatment following the first demyelinating event suggestive of Multiple Sclerosis. Arch Neurology 2004; 61:1515-1520
127. Orange J, Hossny E, Weiler C, 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(4):S525-S553.
128. Leong H, Stachnik J, Bonk ME, Matuszewski KA. Unlabeled uses of intravenous immune globulin. Am J Health Syst Pharm. 2008;65(19)1815-1824.
129. RhoGAM® and MICRhoGAM® [package insert]. Raritan, NJ; Ortho-Clinical Diagnostics, Inc.; 2007. Available at: http://www.rhogam.com/Patient/News/Pages/default.aspx.
130. Rhophylac® [package insert]. Bern, Switzerland; CSL Behring AG; 2010. Available at: http://labeling.cslbehring.com/PI/US/Rhophylac/EN/Rhophylac-Prescribing-Information.pdf.

131. Thymoglobulin® [package insert]. Cambridge, MA; Genzyme Corporation; 2008. Available at: http://www.thymoglobulin.com/home/thymo_pdf_pi.pdf.
132. Winrho® [package insert]. Winnipeg, Manitoba; Cangene Corporation; 2010. Available at: http://www.winrho.com/pi.pdf.
133. Patwa HS, Chaudry 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;78:1009-1015
134. Leger J, Chassandre B, Musset L, et al. Intravenous immunoglobulin therapy in multifocal motor neuropathy: a double-blind, placebo-controlled study. *Brain* 2001;124:145-153
135. Federico P, Zochodne DW, Hahn AF, et al. Multifocal motor neuropathy improved by IVIG: randomized, double-blind, placebo-controlled study. *Neurology* 2000;55:1256-1262.
136. Hughes R, Bensa S, Willison H, et al. Randomized controlled trial of intravenous immunoglobulin versus oral prednisolone in chronic inflammatory demyelinating polyradiculoneuropathy. Ann Neurol 2001;50:195-201
137. Hughes RA, Donofrio P, Bril V, et al. Intravenous immune globulin (10% caprylate-chromatography purified) for the treatment of chronic inflammatory demyelinating polyradiculoneuropathy (ICE study): a randomised placebo-controlled trial. Lancet Neurol 2008;136-44
138. Mouthon L, Guillevin L, Tellier Z. Intravenous immunoglobulins in autoimmune- or parvovirus B19-mediated pure red-cell aplasia. Autoimmunity Reviews 2005;4:264-269
139. Crabol R, Terrier B, Rozenberg F, et al. Intravenous immunoglobulin therapy for pure red cell aplasia related to human parvovirus B19 infection: a retrospective study of 10 patients and review of the literature. Clinical Infectious Disease 2013;56(7):968-77
140. Umapathi T, Hughes RAC, Nobile-Orazio E, et al. Immunosuppressant and immunomodulatory treatments for multifocal motor neuropathy. Cochrane Database of Systematic Reviews 2012, Issue 4. Art. No.: CD003217. DOI: 10.1002/14651858.CD003217.pub4.
141. Karlson EW, Sudarsky L, Ruderman E, et al. Treatment of stiff-man syndrome with intravenous immune globulin. Arthritis Rheum 1994; 37:915.
142. Barker RA, Marsden CD. Successful treatment of stiff man syndrome with intravenous immunoglobulin. J Neurol Neurosurg Psychiatry 1997; 62:426.
143. Khanlou H, Eiger G. Long-term remission of refractory stiff-man syndrome after treatment with intravenous immunoglobulin. Mayo Clin Proc 1999; 74:1231.
144. Dalkas MC, Fujii M, Li M, et al. High-dose intravenous immune globulin for stiff-person syndrome. N Engl J Med 2001; 345:1870
145. International consensus guidance for management of myasthenia gravis June 2016 American Academy of Neurology.
146. Williams KA, Swedo SE, Farmer CA, et al. Randomized, Controlled Trial of intravenous Immunoglobulin for Pediatric Autoimmune Neuropsychiatric Disorders Associated With Streptococcal Infections. J AM Acad Child Adolsec Psychicatry. 2016 Oct;55(10):860-867.e2
147. Perez EE, Orange JS, Bonilla F, et al. Update on the use of immunoglobulin in human disease: A review of evidence. J Allergy Clin Immunol. 2017 Mar;139(3S):S1-S46.
148. Chang K, Frankovich J, Cooperstock M, et al. Clinical evaluation of youth with pediatric acute-onset neuropsychiatric syndrome (PANS): recommendations from the 2013 PANS Consensus Conference. J Child Adolesc Psychopharmacol. 2015 Feb;25(1):3-13.
149. Neunert C, Terrell D, Arnold DM, et al. American Society of Hematology 2019 guidelines for immune thrombocytopenia. Blood Advances. 2019 Oct; 3(23): 3829-3866
150. McCrindle BW, Rowley AH, Newburger JW. Diagnosis, Treatment and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals From the American Heart Association. Circulation. 2017; 135:927-999. doi: 10.1161/CIR.0000000000000484
151. Panel on 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 https://clinicalinfo.hiv.gov/sites/default/files/inline-files/oi_guidelines_pediatrics.pdf. Last updated Nov 2013.

152. Joint Task Force of the EFNS and the PNS. European Federation of Neurological Societies/Peripheral Nerve Society Guideline on management of multifocal motor neuropathy. Report of a Joint Task Force of the European Federation of Neurological Societies and the Peripheral Nerve Society – first revision. Journal of the Peripheral Nervous System 2010;15:295-301.
153. Narayanaswami P, Sanders DB, Wolfe G, et al. International Consensus Guidance for Management of Myasthetnia Gravis. Neurology® 2021;96:114-122. doi:10.1212/WNL.0000000000011124
154. Baden LR, et al. NCCN: Prevention and Treatment of Cancer-Related Infections. Version 1.2021.
155. Thompson, et al. NCCN: Management of Immunotherapy-Related Toxicities. Version 3.2021.
156. Poston JN, Gernsheimer TB. Management of Immune Thrombocytopenia in Pregnancy. Annals of Blood. 2021: 6.
157. Cutaquig [package insert]. Vienna, Austria; Octapharma Pharmazeutika Produktionsges.m.b.H; 2021.
158. Panzyga [package insert[. Vienna, Austria; Octapharma Pharmazeutika Produktionsges.m.b.H; 2021.

RECORD RETENTION

Records Retention for Evolent Health documents, regardless of medium, are provided within the Evolent Health records retention policy and as indicated in CORP.028.E Records Retention Policy and Procedure.

Revision History

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
New Policy	03/22
Added HCPCS code for Cutaquig; extended authorization durations for various indications to 1 year	02/23
Added HCPCS code for Panzyga	01/24

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