

- 1) For immune deficiency conditions, serum IgG levels must be clinically assessed to ensure optimum dosing.
- 2) For all other conditions, IVIG should be used only when other, less expensive, equally safe and efficacious alternative therapy has failed. The use of IVIG should be the exception, rather than the rule.
- 3) There must be regular clinical outcome assessment.

List of Approved Medical Conditions for IVIG Use

Specialty	Medical Condition	Prerequisites / Comments	Dose and Duration
Immunology	Primary immune deficiency	<ul style="list-style-type: none"> - Hypogammaglobulinemia (reduced total IgG or IgG subclasses) with recurrent bacterial infection 	<p>Adult: 0.4-0.6 g/kg every 3-4 weeks. Pediatric: 0.3-0.6 g/kg every 4 weeks. Monitor IgG trough level to maintain low normal range.</p>
	Secondary immune deficiency		
Hematology	Fetal-Neonatal alloimmune thrombocytopenia (F/NAIT)	<ul style="list-style-type: none"> - Previous affected pregnancy or family history of F/NAIT or mother found on screening to have platelet alloantibodies. IVIG is first-line treatment of FAIT. - In newborn with NAIT the provision of antigen-negative compatible platelets should be first-line therapy and IVIG adjunctive. - Treatment should be under the direction of a high-risk obstetrical centre with expertise in F/NAIT. 	1 g/kg every week
	Hemolytic disease of the newborn (HDN)	<ul style="list-style-type: none"> - IVIG is indicated only in HDN infants with severe hyperbilirubinemia; i.e. TSB rising despite intensive phototherapy or TSB level within 34-51 micromol/L of the exchange level (TSB = total serum bilirubin). - IVIG is indicated only in HDN infants with severe hyperbilirubinemia; i.e. TSB rising despite intensive phototherapy or TSB level within 34-51 micromol/L of the exchange level (TSB = total serum bilirubin) 	0.5-1.0 g/kg over 2 hours. If necessary, dose can be repeated in 12 hours.
	Idiopathic thrombocytopenic purpura (ITP) – PEDIATRIC	<ul style="list-style-type: none"> - Acute ITP: IVIG may be considered initial therapy if the platelet count is <20 x 10⁹/L. Consultation with a pediatric hematologist is advised. IVIG is recommended as part of multimodality therapy (with platelet transfusions and bolus intravenous MP) when the patient has life-threatening bleeding. IVIG is not indicated if only mild bleeding (petechiae, bruises or asymptomatic). - Chronic ITP: IVIG may be considered. 	Acute or chronic ITP: one dose of 0.8 to 1 g/kg, with a second dose within 48 hours if the platelet count has not increased to above 20 x 10 ⁹ /L Acute ITP with life-threatening bleeding: 1 g/kg daily for 2 days
	Idiopathic thrombocytopenic purpura (ITP) – ADULT	<ul style="list-style-type: none"> - No treatment is required if the platelet count >20 x 10⁹/L and there is no active bleeding Acute ITP with bleeding: IVIG is recommended as part of multimodality therapy for major or life-threatening bleeding complications and/or clinically important mucocutaneous bleeding. Acute ITP with severe thrombocytopenia but no bleeding: IVIG is not considered first-line therapy, except for patients with contraindications to steroids. ITP with no/slow response to adequate dose steroids: IVIG may be considered as possible adjunctive therapy. Chronic ITP post-splenectomy: IVIG may be considered possible adjunctive therapy as a steroid-sparing measure. 	Acute ITP: 1 g/kg daily for 2 days Chronic ITP post-splenectomy: 0.5 g/kg every 4 weeks; gradually decrease to the minimum effective dose at maximum intervals to maintain safe platelet levels. Re-evaluate every 3-6 months. Consider alternative therapies for patients who do not achieve a durable response for a minimum of 2-3 weeks.
Infectious Diseases	Staphylococcal toxic shock Invasive Group A streptococcal fasciitis with associated toxic shock	<ul style="list-style-type: none"> - Evidence of systemic inflammation and end organ hypoperfusion with fever, tachycardia, tachypnea and hypotension - Consult with medical microbiologist or infectious disease specialist before treatment 	1 g/kg on day one and 0.5 g/kg per day on days two and three, OR 0.15 g/kg per day over 5 days

General Prerequisites for IVIG Use:

List of Approved Medical Conditions for IVIG Use

Specialty	Medical Condition	Prerequisites / Comments	Dose and Duration
Neurology	Guillain-Barré syndrome (GBS), including Miller-Fisher syndrome and other variants	<ul style="list-style-type: none"> Symptoms of grade 3 severity (able to walk with aid) or greater or symptoms less than grade 3 severity that are progressing Treatment should be given within 2 weeks of symptom onset Diagnosis of GBS variants should be made by a specialist with expertise in this area 	2 g/kg over 2-5 days for adults and over 2 days for children
	Chronic inflammatory demyelinating polyneuropathy (CIDP)	<ul style="list-style-type: none"> IVIG is considered a first line treatment for initial treatment of CIDP. Some patients may respond fully to IVIG alone. Other CIDP patients may have a limited or incomplete response to IVIG and then alternate treatments and immunosuppressants may be considered. All patients receiving IVIG for chronic treatment of CIDP should be followed by a neuromuscular specialist. 	Initial treatment: 2 g/kg over 2-5 days. Maintenance therapy: tailor to the lowest dose that maintains clinical efficacy, usually 0.5-1g/kg q 4-8 weekly. Continued use should be based on objective measures of sustained effectiveness.
	Multifocal motor neuropathy (MMN)	<ul style="list-style-type: none"> Diagnosis should be made by a neuromuscular specialist, as very specific electrodiagnostic expertise is required 	Initial treatment: 2 g/kg over 2-5 days. Maintenance therapy: tailor to the lowest dose that maintains clinical efficacy, 0.5-1 g/kg q 3-6 weekly.
	Myasthenia gravis (MG)	<ul style="list-style-type: none"> Severe exacerbations of MG or myasthenic crises, or to stabilize patients before surgery IVIG is not recommended as maintenance therapy for patients with chronic MG 	Initial treatment: 2 g/kg over 2-5 days, and if short term maintenance therapy is required, 0.5-1 g/kg q 3-4 weekly
Dermatology	Pemphigus vulgaris	<ul style="list-style-type: none"> Firm histological and immuno-diagnosis is needed. Consider IVIG when there is no response or a contraindication to corticosteroids and immunosuppressive agents 	2 g/kg over 5 days
Rheumatology	IVIG use by patients over 18 years of age must be approved by the Rheumatology IVIG Consultant		
	Juvenile dermatomyositis (JD)	<ul style="list-style-type: none"> Lack of response or contraindication to corticosteroids, Methotrexate and/or Azathioprine therapy 	Initial treatment: 2 g/kg over 2 days. Maintenance therapy: a systematic approach should be taken to determine minimum effective dose. Continued use should be based on objective measures of sustained effectiveness. Maximum dose per treatment course should not exceed 2 g/kg.
	Kawasaki disease (KD)	<ul style="list-style-type: none"> Validity of diagnosis must be established 	2 g/kg x 1 day. Second dose can be given for patients who fail to respond the first time.

Use of IVIG for conditions not listed above or in cases where the prerequisites are not met must be reviewed and approved by a designated pathologist/Hematopathologist that has experience in the use of IVIG.

IVIG is not recommended or is contraindicated for use in the following conditions:

- Hematology:** aplastic anemia, heparin-induced thrombocytopenia
- Neurology:** adrenoleukodystrophy, amyotrophic lateral sclerosis, autism, critical illness polyneuropathy, inclusion body myositis, intractable childhood epilepsy, paraproteinemic neuropathy (IgM variant), POEMS (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes)

References:

- Anderson D et al. Guidelines on the use of intravenous immune globulin for hematologic conditions. Transfusion Medicine Reviews 2007(April); 21.
- Feasby T et al. Guidelines on the use of intravenous immune globulin for neurologic conditions. Transfusion Medicine Reviews 2007(April); 21(1, Suppl 1):S57-S107.
- IVIG Utilization Management Handbook: First Edition. British Columbia Provincial Blood Coordinating Office, 2002.
- Physician's Standing Order Request for IVIG – Recommended Dosing. Nova Scotia Provincial Blood Coordinating Program, 2004-05