Regional Pre-Printed Orders for
INTRAVENOUS IMMUNE GLOBULIN (IVIg)

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The IVIg Utilization Management Program is a provincial program that incorporates the BC Ministry of Health Immune Globulin Utilization Management Directives to ensure enough IVIg is available for patients likely to benefit from the treatment. Directives include:

1. A definite diagnosis must be established.
2. Dose with the adjusted body weight calculator.
3. IgG levels must be assessed to ensure optimal dosing in PID/SID.
4. There must be regular clinical outcome assessment.

Approved Medical Conditions	Dose and Duration
Immunology	
Primary and secondary immune deficiency conditions (PID and SID) • Hypogammaglobulinemia (reduced total IgG or IgG subclasses and/or inadequate response to immunization) with recurrent bacterial infection. • Monitor IgG trough levels to maintain low normal range.	**PID: Adults:** Less severe: 0.2 - 0.4 g/kg every 3-4 weeks More severe: 0.4 - 0.6 g/kg every 3-4 weeks **Pediatric:** 0.4 - 0.6 g/kg every 3-4 weeks **SID: Adult:** 0.4 - 0.6 g/kg every 3-4 weeks **Pediatric:** 0.3 - 0.6 g/kg every 4 weeks
Hematology	
Fetal-Neonatal alloimmune thrombocytopenia (F/NAIT) • Previous affected pregnancy or family history of F/NAIT or mother found on screening to have platelet alloantibodies. IVIg is first line treatment of F/NAIT. • In newborn with NAIT, antigen-negative platelets should be first-line therapy and IVIg adjunctive. • Treatment should be under the direction of a high-risk obstetrician with expertise in F/NAIT.	Maternal: 1-2 g/kg weekly, depending on gestational age and whether risk for complications of NAIT is standard or high Neonate: 1 g/kg (see FH NICU Blood Component/Product PPO)
Hemolytic disease of the newborn (HDN) • Indicated only in HDN infants with severe hyperbilirubinemia	0.5 - 1.0 g/kg If necessary, dose can be repeated in 12 hours
Immune thrombocytopenia [ITP] -Pediatric • **Acute ITP:** IVIg may be considered initial therapy if the platelet count is <20 x 10^9/L. Consultation with a pediatric hematologist is advised. IVIg is recommended as part of multimodal 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 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 **or** clinically significant bleeding persists requiring a higher platelet count.
Immune thrombocytopenia [ITP] -Adult • No treatment is required if the platelet count >20 x 10^9/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 adjunctive therapy	**Acute ITP:** one dose of 1 g/kg, with a second dose within 48 hours if the platelet count has not increased to above 20 x 10^9/L **or** clinically significant bleeding persists requiring a higher platelet count.
Infectious Diseases	
Staphylococcal toxic shock or invasive Group A streptococcal fasciitis with toxic shock • Evidence of 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
Measles - post exposure prophylaxis • To prevent post-exposure measles disease in pregnant women infants and immunosuppressed patients in whom the use of an IM preparation of hyper immune globulin is not tolerated or available.	0.4 g/kg as a single dose
Neurology	
Guillain-Barre syndrome (GBS), including Miller-Fisher syndrome and other variants • Symptoms of grade 3 severity (able to walk with aid) or greater or symptoms less than grade 3 severity that are progressing. Diagnosis of GBS variants should be made by a specialist. • Treatment should be given within 2 weeks of symptom onset.	**Adult:** 2 g/kg over 2 - 5 days **Pediatric**: 2 g/kg over 2 days
Chronic inflammatory demyelinating polyneuropathy (CIDP) • IVIg is considered a first line treatment for initial treatment of CIDP. • Alternate treatments and immunosuppressants may be considered if limited or incomplete response. • Patients receiving IVIg for 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 - 1 g/kg every 4 - 8 weeks. Continued use should be based on objective measures of sustained effectiveness
Multifocal motor neuropathy (MMN) • Diagnosis should be made by a neuromuscular specialist.	**Initial treatment:** 2 g/kg over 2 - 5 days **Maintenance therapy:** use lowest dose that maintains clinical efficacy, 0.5 - 1 g/kg every 3 - 6 weeks
Myasthenia gravis (MG) • Severe exacerbations of MG or myasthenic crisis, or to stabilize patients before surgery. • IVIg 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 every 3 - 4 weeks
Dermatology	
Pemphigus vulgaris • Firm histological and immune-diagnosis is needed. • Consider IVIg if no response or contraindication to corticosteroids and immunosuppressants.	2 g/kg over 5 days
Rheumatology *IVIg use for patients over 18 years of age must be approved by the Provincial Rheumatology Panel. On-call support and information available at* www.pbco.ca	
Juvenile dermatomyositis (JD) • Lack of response or contraindication to corticosteroids, Methotrexate and/or Azathioprine therapy.	**Initial treatment:** 2 g/kg over 2 days **Maintenance therapy:** Continued use should be based on objective measures effectiveness. Maximum dose not exceed 2 g/kg
Kawasaki disease (KD) • Validity of diagnosis must be established.	2 g/kg x 1 day. Second dose may be given for patients who fail to respond the first time

Possibly Indicated Neuromuscular Conditions	Dose and Duration
Atypical CIDP, Refractory vasculitic neuropathy, Lambert-Eaton syndrome, Sensory ganglionopathy, Stiff-Person syndrome, Severe diabetic radiculoplexopathy, Voltage-gated K+ channelopathy	2 g/kg over 2 -5 days. Initial treatment limited to 3 months. The PBCO Neuromuscular Panel will contact physician if further treatment required.

Not Indicated Conditions: IVIg is Not Recommended or Contraindicated
Hematology: aplastic anemia **Neurology:** adrenoleukodystrophy, amyotropic lateral sclerosis, autism, critical illness polyneuropathy, inclusion body myositis, intractable childhood epilepsy, paraproteinemic neuropathy (IgM variant), POEMS

Please note: Patients with blood groups A, B, or AB receiving a dose of 1g/kg or more are at an increased risk of IVIg associated hemolysis. Careful clinical follow up 1-2 weeks post-IVIg infusion is recommended. Specifically jaundice, fever and falling hemoglobin may indicate such a reaction. Completion of a transfusion reaction report form and appropriate laboratory investigations (e.g., Hb, reticulocyte, DAT, Tbili, LDH) are recommended for confirmation if there is any clinical suspicion of hemolysis.