Interior Health Intravenous Immune Globulin (IVIG) Utilization Management Program Recommendations

Interior Health

This IVIG Reference is a companion document to the IH IVIG request form (826406).

IH IVIG Forms for Initial and Renewal Requests			
Form #	**Name and Link to IH Forms Library**	**Initial**	**Repeat**
826406	IVIG Request form http://insidenet.interiorhealth.ca/infoResources/forms/Documents/826406.pdf	x	x
826703	Immune Globulin (IVIG and SCIG) Outcome Questionnaire http://insidenet.interiorhealth.ca/infoResources/forms/Documents/827063.pdf		x
828101	Patient Information – What is IVIG? http://insidenet.interiorhealth.ca/infoResources/forms/Documents/828101.pdf	x	

Interior Health IVIG Utilization Program is compliant with the British Columbia Provincial Policy on Health Authority Immune Globulin Utilization Management and Ministry of Health (MoH) Directives:

1. Requests for IVIG to treat conditions on approved list must meet the specific prerequisites. The dose and duration of therapy must be in accordance with BC IVIG Utilization Management Guidelines. Most IVIG approvals are for 6 months; for patients with Primary Immune Deficiency, the approval may be 12 months.
2. IVIG use for conditions not listed here or in cases where prerequisites are not met, must be reviewed and approved at the discretion of an IH designated pathologist/hematopathologist that has IVIG expertise.
   - **Requests without appropriate evidence-based rationale may be rejected per MoH Directives.**
3. The maximum amount of IVIG administered should reflect **adjusted body weight dosing**. Actual body weight dosing will be used to determine dose for patients younger than 18 years of age, weighing less than ideal body weight, patients under 153 cm (60 inches), and during pregnancy.
4. **Assessment of the efficacy of IVIG therapy is mandatory and all renewal requests must include a completed IVIG Outcome Questionnaire.**
5. **IVIG is not recommended or is contraindicated in the following conditions:**
   - **Hematology:** aplastic anemia
   - **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, skin changes)

- **Fax all IVIG request forms and outcome questionnaires, including physician orders, and booking forms to the IH IVIG Coordinators at 250-862-4052.**
- **Once the IVIG is reviewed and approved, an approval fax is sent to the requesting physician(s). Copies are sent to clinical area transfusing IVIG and TM/Labs issuing IVIG are notified.**
- **Confidential email address for both IH IVIG Coordinators: IHLabIVIG@interiorhealth.ca**

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List of Approved Medical Conditions for IVIG Use		
Condition	**Prerequisites / Comments**	**Dose and Duration**

Primary immune deficiency (PIDD)	• Hypogammaglobulinemia (reduced total IgG or IgG subclasses) with recurrent **bacterial** infection(s)
	• Monitor IgG trough level as appropriate to achieve desired clinical outcome
	• See https://www.pbco.ca/index.php/programs/primary-immunodeficiency/diagnostic-and-treatment for more information
		• **Adults based on severity of condition:**
		  • **Less severe**: 0.2 to 0.4 g / kg **dosing** weight every 21 to 30 days
		  • **More severe**: 0.4 to 0.6 g / kg **dosing** weight every 21 to 30 days
		• **Pediatric**: 0.4 to 0.6 g / kg **actual** weight every 21 to 30 days

Secondary immune deficiency (SID) associated with Chronic lymphocytic leukemia (CLL), Memory B cell deficiency secondary to hematopoietic stem cell transplantation (HSCT), Multiple myeloma (MM), Non-Hodgkin lymphoma (NHL), or other Hematological malignancy	• Hypogammaglobulinemia (IgG less than 5 g / L excluding paraprotein) with at least 1 life-threatening bacterial infection requiring ICU admission or 2 serious bacterial infections requiring more than standard antibiotic treatment, **AND** • Infections unrelated to chemo / radiotherapy including neutropenia or mucosal / epithelial toxicity **AND** confirmed as encapsulated bacteria and / or clinically consistent with encapsulated bacteria • Monitor IgG trough level as appropriate to achieve desired clinical outcome • See www.pbco.ca IVIG Program for more information	• **Adult Initial dose**: 0.4 g / kg **dosing** weight every 30 days if IgG level is less than 4 g / L • **Adult Maintenance dose**: 0.4 to 0.6 g / kg **dosing** weight every 28 to 30 days to achieve IgG trough level of 7.0 g / L • **Pediatric**: 0.3 to 0.6 g / kg **actual** weight every 28 to 30 days
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 FAIT • In newborn with NAIT the provision of antigen-negative compatible platelets should be first-line therapy and IVIG adjunctive • Treatment should be directed by a high-risk obstetrical centre with F / NAIT expertise	• 1 to 2 g / kg **actual** weight weekly, depending on gestational age and whether risk for complications of NAIT is standard or high
Hemolytic disease of the newborn (HDN)	• Only indicated in infants with severe hyperbilirubinemia, i.e. Total Serum Bilirubin (TSB) rising despite intensive phototherapy or TSB within 34-51 micromol / L of exchange level	• 0.5 to 1 g / kg **actual** weight • If necessary, dose may be repeated in 12 hours
Idiopathic thrombocytopenic purpura (ITP) – Adult	• No treatment required if platelet count greater than 20 × 109 / L and 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 if steroids are contraindicated • **ITP with no / slow response to adequate dose steroids**: IVIG may be considered as possible adjunctive therapy	• **Acute ITP**: one dose of 1 g / kg **dosing** weight, with a second dose within 48 hours if the platelet count has not increased to above 20 × 109 / L or clinically significant bleeding persists requiring a higher platelet count

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Condition	Prerequisites / Comments	Dose and Duration
Idiopathic thrombocytopenic purpura (ITP) – Pediatric	• **Acute ITP**: IVIG may be considered initial therapy if platelet count is less than 20 × 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 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 actual weight, with a second dose within 48 hours if the platelet count has not increased to above 20 × 109 / L or clinically significant bleeding persists requiring a higher platelet count
Staphylococcal toxic shock (STS) **Invasive Group A streptococcal fasciitis with associated toxic shock (IGAS)**	• Evidence of systemic inflammation and end organ hypoperfusion with fever, tachycardia, tachypnea, and hypotension • Consult with medical microbiologist or infectious disease specialist before treatment	• **Adult**: 1 g / kg **dosing** weight on day one and 0.5 g / kg **dosing** weight per day on days two and three, **or** 0.15 g / kg **dosing** weight per day for 5 days • **Pediatric**: 1 g / kg **actual** weight on day one and 0.5 g / kg **actual** weight per day on days two and three, **or** 0.15 g / kg **actual** weight per day for 5 days
Measles post –exposure prophylaxis (MPEP)	• As ordered by Medical / Public Health Officer to prevent measles in pregnant women, infants, and immune-deficient or immunosuppressed patients who weigh more than 30 kg or unable to tolerate intramuscular (IM) injection of hyper immune globulin	• **Adult**: 0.4 g / kg dosing weight as a single dose (**actual** weight if pregnant) • **Pediatric**: 0.4 g / kg actual weight as a single dose
See https://www.pbco.ca/index.php/programs/ivig-neuromuscular for more information about the following neuromuscular conditions:		
Chronic inflammatory demyelinating polyneuropathy (CIDP)	IVIG is considered a first line treatment for initial treatment • Some patients respond fully to IVIG alone • Other patients may have a limited or incomplete response to IVIG, and then alternate treatments and immune-suppressants may be considered • All patients should be followed by a Neuromuscular specialist	• **Initial dose**: 2 g / kg **dosing** weight over 2 to 5 days • **Maintenance therapy**: tailor to the lowest dose that maintains clinical efficacy, usually 0.5 to 1 g / kg **dosing** weight every 28 to 56 days; base continued use on objective measures of sustained effectiveness
Guillain-Barré 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 • Start treatment within 2 weeks of symptom onset • Confirm variant diagnosis by GBS specialist	• **Adult**: 2 g / kg **dosing** weight over 2 to 5 days • **Pediatric**: 2 g / kg **actual** weight over 2 days
Multifocal motor neuropathy (MMN)	• Diagnosis should be made by a neuromuscular specialist, as very specific electrodiagnostic expertise is required	• **Initial dose**: 2 g / kg **dosing** weight over 2 to 5 days • **Maintenance therapy**: tailor to the lowest dose that maintains clinical efficacy, 0.5 to 1 g / kg dosing weight every 21 to 42 days
Myasthenia gravis (MG)	• 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 dose**: 2 g / kg **dosing** weight over 2 to 5 days • **If short term maintenance therapy is required**, 0.5 to 1 g / kg **dosing** weight every 21 to 30 days

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Condition	Prerequisites / Comments	Dose and Duration
Pemphigus Vulgaris (PV)	• Firm histological / immunodiagnosis needed • Consider IVIG if no response or a contraindication to corticosteroids and immunosuppressive agents	• 2 g / kg **dosing** weight over 5 days
Juvenile Dermatomyositis (JD) - Pediatric patients (18 years old or younger).	• Lack of response or contraindication to corticosteroids, Methotrexatate, and / or Azathioprine therapy	• **Initial dose:** 2 g / kg **actual** weight over 2 days • **Maintenance therapy**: use systematic approach to determine minimum effective dose; continued use should be based on objective measures of sustained effectiveness • Maximum dose per treatment course not to exceed 2 g / kg **actual** weight
Kawasaki Disease (KD)	• Validity of diagnosis must be established • Screen for COVID if KD may be related	• 2 g / kg **actual** weight × 1 day • Repeat once if no response

IVIG Utilization managed by BC Provincial Blood Coordinating Office Screening Panels and IH:

Adult Rheumatology conditions include, but are not limited to:

- Adult Still's Disease (ASD)
- Adult Dermatomyositis (AD)
- Behcet's Disease (BE)
- Non-inclusion Body Myositis (NONIBM_RH)
- Other non-specific vasculitic syndromes (NSVS)
- Overlap Syndrome (OS)
- Polyarteritis Nodosa (PAR)
- Polymyositis (PM)
- Polymyositis with connective tissue disease (PMC)
- Rheumatoid Arthritis (RA)
- Scleroderma (SC)
- Sjogren's Syndrome (SJO)
- Systemic Lupus Erythromatosus (SLE)
- Vasculitis (V)
- ANCA Vasculitis - eGPA (Churg Strauss), MPA (Microscopic Polyangitis), (WG) Wegener Granulomatosis

- **Requests for patients over 18 years of age: must be screened by PBCO Rheumatology IVIG Panel**
- Screening request form is available from PBCO website https://www.pbco.ca/images/Programs/IVIG_Provincial_Program/UM.IVIG.0002F3-TMS---IVIG-Screening-Request-Form_V2.2.pdf or IH IVIG Coordinator
- See https://www.pbco.ca/index.php/programs/ivig-rheumatology for more information

Other Neuromuscular Conditions where IVIG may be indicated:

- Atypical / Possible Chronic Inflammatory Demyelinating Polyneuropathy (ACIDP)
- Refractory Vasculitic Neuropathy (RVN)
- Lambert Eaton Syndrome (LE)
- Sensory Ganglionopathy (SG)
- Stiff Person Syndrome (SPS)
- Severe Diabetic Radiculoplexopathy (SDR)
- Voltage Gated K+ Channelopathy (VGKC)

- **Initial dose**: 2 g / kg **dosing** weight over 2 to 5 days repeated every 30 days, limited to a trial of 3 infusions
- **If maintenance therapy after 3 month trial is required, all requests are screened by PBCO Neuromuscular Panel**
- Tailor lowest dose that maintains clinical efficacy, usually 0.5 to 1 g / kg **dosing** weight every 28 to 56 days
- Base continued use on objective measures of sustained effectiveness and review every 6 months
- See https://www.pbco.ca/images/Programs/IVIG_Neuro/Diagnostic_and_Treatment_Algorithms/UM.IVIG.0036_V1.2_IVIG-for-Neuromuscular-Neurology-Conditions.pdf for more information

Reference: BC Provincial Blood Coordinating Office IVIG Provincial Program, https://www.pbco.ca/index.php/programs/ivig-provincial-program/guidelines-forms-templates July 2019 - adapted for IH IG Utilization Program 25August2020

IVIG Reference Document - Companion to IH IVIG Request form 826406