**Indication**

**Kawasaki Disease (KD)**
- IVIG is the treatment of choice for KD. All children with KD should be treated as early as possible & at any time if they have evidence of active disease.
- **Dose:** 2 g/kg given once. About 10% of children fail to respond to initial IVIG therapy & may be treated a 2nd time with the same dose, at least 24 hrs after the 1st dose. (A picture of aseptic meningitis with fever, vomiting, & headache may occur in children with inflammatory autoimmune disorders. This reaction lasts for about 12 hrs & should be distinguished from the recurrence of KD signs & symptoms). Failure to respond to a 2nd dose (provided the recurrence of symptoms & signs does not reflect a reaction to IVIG) should prompt reconsideration of the diagnosis & referral to a specialist with expertise in the management of KD. Specialist referral should also occur for KD in the very young infant, particularly in the presence of myocarditis, where IVIG treatment may compromise cardiac function & result in congestive heart failure.

**Juvenile Dermatomyositis**
- IVIG is an adjunctive therapy for children with juvenile dermatomyositis (JDM), and may be considered in children who are resistant, dependant, or toxic to treatment with corticosteroids and other second-line agents. In some circumstances, it may be considered as part of early therapy in the critically ill child with JDM. Normally, the use of IVIG in JDM would only be undertaken by physicians with extensive experience in the management of children with this rare illness. **Dose:** 2 g/kg/day given every 2-4 wks for a variable period of time, usually months to years.

**Juvenile Idiopathic Arthritis**
- IVIG should not be used in the primary management of polyarticular or systemic juvenile idiopathic arthritis. However, in children who are resistant to other forms of therapy consideration should be given to the use of IVIG, under the supervision of a pediatric rheumatologist. **Dose:** 1-2 g/kg once every 2-4 wks for a variable period of time, usually months to years.

**Systemic Lupus Erythematosus**
- IVIG should not be used as first-line therapy in pediatric systemic lupus erythematosus (SLE). However, specific patients, particularly those with autoimmune cytopenias, may benefit from its use. The use of IVIG in children with pediatric SLE should be carefully monitored by physicians with expertise in the management of pediatric SLE and the use of IVIG. **Dose:** 1-2 g/kg, frequency and duration variable.

**Idiopathic Thrombocytopenic Purpura (ITP)**
- There must be a diagnosis of ITP and one of the following 2 criteria: 1) surgery required or the presence of major bleeding and a platelet count less than 50 x 10^9/L. OR 2) a platelet count less than 20 x 10^9/L and treatment clinically indicated. **Dose:** One dose of 0.8 to 1 g/kg, with a second dose given within 48 hours if the platelet count has not increased to above 20 x 10^9/L. For life-threatening bleeding, concomitant treatment with platelet transfusion is recommended.

**Neonates of Mothers with ITP**
- In newborns without evidence of intracranial hemorrhage (ICH) or other serious bleeding, treatment with IVIG may be appropriate if platelets are less than 50 x 10^9/L. Newborns with imaging evidence of ICH or other serious bleeding should be treated with IVIG & platelet transfusion. A **Dose** of 1 g/kg daily for 2 days is recommended with the second 1 g/kg dose to be given only if platelets are less than 30 x 10^9/L or in the presence of clinically significant bleeding.

**Fetal Autoimmune Thrombocytopenia (FAIT)**
- IVIG is recommended as the standard first-line antenatal treatment of FAIT. Given the complexity of this disorder & its management, it is strongly recommended that treatment be under the direction of a maternal fetal medicine centre with specialized expertise in the treatment of FAIT. Candidates for treatment include: (i) Pregnant women with a previously affected pregnancy. IVIG should be initiated at a time in the pregnancy that corresponds to a gestational age in the present fetus that precedes by some weeks the time at which bleeding was thought or known to occur in the first pregnancy. (ii) Pregnant women with a familial history of FNAIT or those found on screening to have platelet autoantibodies. Expert opinion suggests IVIG should be initiated around 20 wks and no later than 30 wks. **Dose** in either case: 1 g/kg every week.

**Neonatal Autoimmune Thrombocytopenia (NAIT)**
- When NATI is suspected in a neonate, treatment should be commenced promptly. It is strongly recommended that treatment of newborns with NAINT be under the direction of a center with specialized expertise in the treatment of NAINT. The provision of antigen-negative compatible platelets should be considered first-line therapy for neonates with severe thrombocytopenia and/or bleeding. Antigen-negative platelets are the platelet product of choice; however, random donor platelets should be used if matched platelets are not immediately available. IVIG is used as adjunctive therapy & is effective in 75% of cases. The threshold platelet count for treatment of NAIT with platelet transfusions and IVIG is 30-50 x 10^9/L, which is derived from published studies, although data from randomized trials are lacking. **Dose:** 1g/kg/dose/day x 2. **Monitoring:** platelet counts should rise within 24 to 48 hrs after therapy & should be checked weekly. Follow up with a specialist should be done at 6-8 wks.

**Hemolytic Disease of the Newborn**
- In an effort to decrease the need for exchange transfusion, IVIG is recommended, in addition to aggressive phototherapy, for the treatment of hyperbilirubinemia due to immune hemolytic disease of the newborn (ABO, Rh or other). **Dose:** 0.5g/kg, with repeat dosing q 12-24h as necessary.

**Hematological Malignancy**
- IVIG is not recommended for routine use in children with hematologic malignancies (with or without hypogammaglobulinemia). However, it may be considered for children with hematologic malignancies with acquired hypogammaglobulinemia and either of the following criteria: a history of severe invasive infection or recurrent sinopulmonary infections, or children registered on multinational protocols which require IVIG support. **Dose:** one dose of 0.4–0.6 g/kg every 3–4 wks for prophylaxis. Reevaluation should be done every 4–6 months by a hematologist. A trough IgG level of greater than 7 g/L should be maintained, but it may be necessary to assess the effectiveness of IVIG primarily on clinical response.

**Bone Marrow Transplant**
- Routine use of IVIG for bone marrow transplant is not indicated.

**Guillain-Barré Syndrome (GBS)**
- Diagnosis of GBS variants should be made by a specialist with expertise in this area. **Acute Setting** - IVIG is recommended as a treatment option for GBS within 4 wks of symptom onset for patients with symptoms of grade 3 severity or greater (see Hughes Disability Scale below); OR patients with symptoms less than grade 3 severity whose symptoms are progressing. IVIG treatment after plasma exchange or immunoabsorption, does not produce extra benefit and is not recommended. **Relapses** - IVIG may be considered as a treatment option for patients who initially responded to IVIG & who are experiencing a relapse of symptoms within 8 wks of the initial onset of symptoms. **Acute/Relapses Dose:** 1 g/kg/day x 2 days.

<table>
<thead>
<tr>
<th>Grade</th>
<th>0</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Description</td>
<td>healthy</td>
<td>minor symptoms or signs, able to run</td>
<td>able to walk 5 m independently</td>
<td>able to walk 5 m with a walker, stick, or one-person support</td>
<td>bed- or chair-bound</td>
<td>Requiring assisted ventilation</td>
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**Acute Disseminated Encephalomyelitis**
- IVIG is recommended as an option for treatment of monophasic acute disseminated encephalomyelitis (ADEM) when first-line therapy with high-dose corticosteroids fail or when there are contraindications to steroid use. IVIG may be considered as an option for treatment of relapsing ADEM to eliminate steroid dependency or for those patients who fail to respond, or have contraindications, to steroids. **Dose:** 1 g/kg daily x 2 days. IVIG is used in the same way for other autoimmune encephalopathies (Hashimoto encephalopathy, Landau-Kleffner syndrome, & primary CNS angitis).

**Primary Immune Deficiency (PID)**
- An immunologist should be consulted for all pediatric patients with suspected immunodeficiency syndromes (including PID) prior to the administration of immunoglobulin. Patients with immunodeficiency should have their care coordinated by a comprehensive care clinic with expertise in immune deficiencies. Treatment of patients based solely on low IgG levels is not advised. In patients with PID, IVIG should be initiated at an immunology clinic. **Replacement doses** are 0.6-0.7g/kg every 3-4 weeks to achieve a minimum trough level of 7g/L. In some cases, higher doses maybe required based on clinical response. Patients & practitioners should be aware that patients with PID will usually require immunoglobulin replacement therapy indefinitely. **Re-evaluation** should be done by an immunologist every 3-6 months.

**Secondary Immune Deficiency**
- An immunologist should be consulted for all patients with suspected immunodeficiency, including secondary immune deficiency. IVIG may be required for children with hypogammaglobulinemia secondary to B-cell depletion for autoimmune disease, protein losing enteropathy, post stem cell transplantation, recurrent sinusopulmonary or invasive infections following appropriate immunologic assessment. The usual doses are as for PID.