



Atlantic Clinical Indications and Criteria for Intravenous and Subcutaneous Immunoglobulin (IVIG/SCIG)

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1. Background

Since 2001, the use of intravenous/subcutaneous immunoglobulins (IVIG/SCIG) in Canada increased at a steady rate of five to ten percent each year. The increased utilization has led to concerns in the Atlantic Provinces over the appropriateness of the use of IVIG/SCIG. In 2003, the Atlantic Deputy Ministers determined that an Atlantic Collaborative, the Atlantic Blood Utilization Strategy (ABUS) Working Group be struck to assess and develop interventions to ensure appropriate IVIG/SCIG utilization. The Nova Scotia Provincial Blood Coordinating Team (NSPBCT) acts as the secretariat for ABUS.

It was agreed that ABUS would provide professional leadership in identifying, designing and implementing cost-effective IVIG/SCIG utilization management initiatives to achieve optimal patient outcomes. In 2007, The National Advisory Committee on Blood and Blood Products (NAC) developed guidelines on the use of IVIG for the most common Neurological and Hematological indications. In 2010, NAC also developed guidelines for the use of IVIG/SCIG in Solid Organ Transplant and Primary Immune Deficiencies. During 2016, the following list of indications, along with any pre-requisites/criteria required for the release of product to access publicly funded IVIG and SCIG were developed by ABUS using the NAC recommendations along with expert clinical advice from 307 Atlantic physicians in adult and pediatric hematology, neurology, immunology, rheumatology, dermatology, infectious disease, solid organ transplant, internal medicine, family medicine, obstetrics and gynecology, oncology and emergency medicine.

In the spring of 2018, the Atlantic Deputy Ministers of Health endorsed the *Atlantic Ministries of Health Common Policy for Intravenous and Subcutaneous Immunoglobulin* which refers to the *Atlantic Clinical Indications and Criteria for Intravenous and Subcutaneous Immunoglobulin (IVIG/SCIG)*. Each province developed their own provincial policy based on the Atlantic Common policy and in the fall of 2018, these policies were implemented.

The use of SCIG to treat medical conditions in addition to Inborn Errors of Immunity/Primary Immune Deficiency (IEI/PID) and Secondary Immune Deficiency (SID) has expanded. SCIG is being used nationally by patients with autoimmune neurological diseases. In 2020, the ABUS group engaged neurologists in the Atlantic Provinces and determined there was an interest in using SCIG in neurology. In April 2022, the *Atlantic Clinical Indications and Criteria for Intravenous and Subcutaneous Immunoglobulin (IVIG/SCIG)* was revised to include additional indications, pre-requisites/criteria as well as adding the use of SCIG for patients with autoimmune neurological diseases receiving long-term therapy (e.g. Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)).

2. Introduction

Intravenous/subcutaneous immunoglobulins (IVIG and SCIG) are blood products made from pooled human plasma and as such, are not risk-free to patients. In appropriately selected patients and clinical settings, IVIG/SCIG therapy can be lifesaving. However, serious adverse reactions can occur, such as: hemolysis, renal failure, aseptic meningitis, anaphylaxis and thromboembolic events. Patients must be monitored throughout their treatment to confirm efficacy of the product and that the desired clinical outcomes are achieved.

Efforts must be made to ensure that IVIG/SCIG is provided by physicians only where evidence suggests that it is the most appropriate therapy. To help limit non-evidence based use of IVIG/SCIG and to mitigate an unsustainable increase in utilization in the Atlantic Provinces, the Atlantic Deputy Ministers of Health endorsed the implementation of the *Atlantic Ministries of Health Common Policy for Intravenous and Subcutaneous Immunoglobulin* which refers to the *Atlantic Clinical Indications and Criteria for Intravenous and Subcutaneous Immunoglobulin (IVIG/SCIG)*. This strategy supports consistency in access to IVIG/SCIG across the Atlantic Provinces by building on the existing process and introducing new measures. Adherence to this strategy is intended to address issues of non-evidence-based product utilization, appropriate dosing, and appropriate duration of treatment. Each order is reviewed prior to dispense of product to ensure any pre-requisites have been met, as well to confirm that the dosing, frequency and duration of treatment meet the indications and criteria for use. In the event of an incongruity, the ordering physician will be contacted and discussion ensue regarding the discrepancy. If the discrepancy cannot be resolved after the discussion, product will not be issued until the appropriate clinical expert has been contacted for consultation and direction.

Making IVIG and SCIG available for patients with medical conditions where there is evidence of clinical efficacy is a primary objective of this strategy as supply may not be able to meet demand without control points in place (e.g. the Ig Outcome Questionnaire to evaluate the effectiveness and appropriateness of treatment).

Orders deemed to be **urgent** will be dispensed immediately and the order will be reviewed after dispense. Any follow up required with the ordering physician will still occur. However, as patient safety is the main focus, the follow up will occur after the order has been dispensed. In the indications and criteria list, any indications deemed by the experts as having a possibility of urgency, are marked with an asterisk (*) and any additional criteria required is written in red.

For all indications, the dose is tailored to the lowest clinically effective dose and the shortest duration required to achieve the desired outcome, after alternative therapies have been explored.

For IEI/PID patients, treaters monitor IgG trough levels every 3 to 6 months to achieve a trough level of 7 – 10 g/L. **Clinical considerations:** The IgG trough generally stabilizes after 3 to 4 months of treatment with IVIG. After this time, regular monitoring of IgG trough levels and overall clinical picture allows adjustment of the immunoglobulin dosage to the lowest clinically effective dose.

3. Indications and Criteria

3.1 Hematology

| | Medical Condition | Pre-requisites | Dose/Frequency of Administration |
|---|--|--|---|
| Adult Hematology | Indicated Conditions | | |
| | Immune Thrombocytopenia (ITP)* | Patient must meet 1 of the following 3 criteria: 1. Major bleeding and platelets less than $50 \times 10^9/L$ OR 2. Failed to respond to steroids after 3 or more days OR 3. To produce an increase in platelet count to a level considered safe | Acute: 1 g/kg per day for 1 or 2 consecutive days depending on response Chronic: 1 to 2 g/kg no more frequently than every 2 weeks |
| | Pregnancy-Associated ITP* | Patient must meet 1 of the following 3 criteria: 1. There is major bleeding OR 2. Platelet counts fall below $10 \times 10^9/L$ anytime in the pregnancy OR 10 to $30 \times 10^9/L$ during the second or third trimester OR 3. Rapid elevation of platelets required before delivery or any invasive procedure (e.g. amniocentesis) | 1 g/kg per day for 2 consecutive days (dosing body weight is based on the pre-pregnancy weight for determining IVIG dose) No maximum dose |
| | Post-Transfusion Purpura (PTP)* | No criteria are required other than a diagnosis of PTP | 1 g/kg repeated if necessary |
| Fetal Alloimmune Thrombocytopenia (FAIT)* | Patient must meet both of the following criteria: 1. Mother has been found to have anti-platelet alloantibodies through a prior affected pregnancy or close family member (e.g. sister) with an affected pregnancy AND 2. Treatment is under the direction of a maternal fetal medicine center | 1 to 2 g/kg per week throughout the pregnancy (dosing body weight is based on the pre-pregnancy weight for determining IVIG dose; disease severity also considered) No maximum dose | |

| Possibly Indicated Conditions | | | |
|-------------------------------|---|--|--|
| Adult Hematology | Acquired Hemophilia with Factor VIII Inhibitor* | Order must be in consultation with a Hematologist | 2 g/kg divided over 2 to 5 days |
| | Factor XIII Inhibitor* | Order must be in consultation with a Hematologist | 2 g/kg divided over 2 to 5 days |
| | Secondary Immunodeficiency (SID) | Order must be in consultation with a Hematologist | * IVIG dose: 0.4 g/kg every 3 to 4 weeks * SCIG dose: 0.1 to 0.13 g/kg every week |
| | Warm Autoimmune Hemolytic Anemia | Patient must be resistant to steroids and exhibit symptomatic anemia | Up to 2 g/kg |
| | Hemophagocytic Lymphohistiocytosis (HLH)* | Order must be in consultation with a Rheumatologist, Hematologist or General Internist | 2 g/kg divided over 2 to 5 days |

* **May be considered URGENT if notified by ordering physician as such**

| | Medical Condition | Pre-requisites | Dose/Frequency of Administration |
|-----------------------------------|---|--|--|
| Pediatric Hematology | Indicated Conditions | | |
| | Post CAR-T cell therapy* | Order must be in consultation with a pediatric Hematologist | 0.4 to 0.6 g/kg every 3 to 4 weeks |
| | Neonatal Alloimmune Thrombocytopenia (NAIT)* | Treatment includes consultation with or is within a high-risk neonatal center | 1 g/kg per day x 2 days |
| | Hemolytic Disease of the Newborn (HDN)* | Total serum bilirubin (TSB) rising despite intensive phototherapy | 0.5 to 1 g/kg, with repeat dosing every 12 to 24 hours as necessary |
| | Immune Thrombocytopenia (ITP)* | Patient must meet 1 of the following 2 criteria: 1. Platelets less than $50 \times 10^9/L$ AND either the presence of major bleeding or surgery required OR 2. Platelets less than $20 \times 10^9/L$ AND treatment clinically indicated | 0.8 to 1 g/kg, with a 2 nd dose within 48 hours if the platelet count has not increased to above $20 \times 10^9/L$ |
| | Neonates of Mothers with ITP* | Patient must meet 1 of the following 2 criteria: 1. Platelets less than $50 \times 10^9/L$ OR 2. Imaging evidence of intracranial hemorrhage or other serious bleeding | 1 g/kg daily for 2 days with a second dose of 1 g/kg if platelet count is still less than $30 \times 10^9/L$ |
| | Possibly Indicated Conditions | | |
| | Hematological Malignancy* | Patient must meet criteria number 1 and either criteria number 2 or 3 1. Acquired hypogammaglobulinemia PLUS 2. History of severe invasive or recurrent sinopulmonary infections OR 3. Registered on a protocol which requires IVIG support | 0.4 to 0.6 g/kg every 3 to 4 weeks |
| Secondary Immunodeficiency (SID)* | Order must be in consultation with a pediatric Hematologist | *IVIG dose: 0.4 g/kg every 3 to 4 weeks) *SCIG dose: 0.1 to 0.13 g/kg every week | |

*** May be considered URGENT if notified by ordering physician as such**

3.2 Neurology

| | Medical Condition | Pre-requisites | Dose/Frequency of Administration | | | | | | | | | | | | | | |
|-----------------------------------|---|---|--|-------------|----------|---------|----------|--------------------------------------|----------|--------------------------------|----------|---|----------|--------------------|----------|--------------------------------|---------------------------------|
| Adult Neurology | Indicated Conditions | | | | | | | | | | | | | | | | |
| | Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) | Order must be in consultation with a Neurologist | <p>*IVIG dose: 2 g/kg divided over 2 to 5 days</p> <p>Maintenance dose: 1 g/kg every 2 to 6 weeks Tailor to the lowest dose that maintains clinical efficacy, usually 0.5 to 1g/kg q 4 to 8 weeks</p> <p>*SCIG dose: 0.2 to 0.4 g/kg every week</p> | | | | | | | | | | | | | | |
| | Guillain-Barré Syndrome (GBS)* | <p>Patient must meet both of the following criteria:</p> <ol style="list-style-type: none"> 1. IVIG is being given within 2 weeks of symptom onset <p>AND</p> <ol style="list-style-type: none"> 2. Hughes Disability score of 3 or more or less than 3 with symptoms progressing <p style="text-align: center;">Hughes Disability Scale:</p> <table border="1" style="margin-left: auto; margin-right: auto;"> <thead> <tr> <th>Grade</th> <th>Description</th> </tr> </thead> <tbody> <tr> <td style="text-align: center;">0</td> <td>Healthy</td> </tr> <tr> <td style="text-align: center;">1</td> <td>Minor signs or symptoms, able to run</td> </tr> <tr> <td style="text-align: center;">2</td> <td>Able to walk 5 m independently</td> </tr> <tr> <td style="text-align: center;">3</td> <td>Able to walk 5 m with a walker, stick or one-person support</td> </tr> <tr> <td style="text-align: center;">4</td> <td>Bed or chair bound</td> </tr> <tr> <td style="text-align: center;">5</td> <td>Requiring assisted ventilation</td> </tr> </tbody> </table> | Grade | Description | 0 | Healthy | 1 | Minor signs or symptoms, able to run | 2 | Able to walk 5 m independently | 3 | Able to walk 5 m with a walker, stick or one-person support | 4 | Bed or chair bound | 5 | Requiring assisted ventilation | 2 g/kg divided over 2 to 5 days |
| | Grade | Description | | | | | | | | | | | | | | | |
| 0 | Healthy | | | | | | | | | | | | | | | | |
| 1 | Minor signs or symptoms, able to run | | | | | | | | | | | | | | | | |
| 2 | Able to walk 5 m independently | | | | | | | | | | | | | | | | |
| 3 | Able to walk 5 m with a walker, stick or one-person support | | | | | | | | | | | | | | | | |
| 4 | Bed or chair bound | | | | | | | | | | | | | | | | |
| 5 | Requiring assisted ventilation | | | | | | | | | | | | | | | | |
| Multifocal Motor Neuropathy (MMN) | No criteria are required other than a diagnosis of MMN | <p>*IVIG dose: 2 g/kg divided over 2 to 5 days</p> <p>Maintenance dose: 1 g/kg every 2 to 6 weeks</p> <p>*SCIG dose: 0.2 to 0.4 g/kg every week</p> | | | | | | | | | | | | | | | |

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| Adult Neurology | Myasthenia Gravis (MG)* | Patient must meet 1 of the following 3 criteria: 1. Acute exacerbation (myasthenic crisis) OR 2. Optimization prior to surgery and/or thymectomy OR 3. As maintenance therapy for moderate to severe MG in combination with immunosuppressive agents | *IVIG dose: 2 g/kg divided over 2 to 5 days every 4 to 6 weeks *SCIG dose: 0.2 to 0.4 g/kg every week |
| | Possibly Indicated Conditions | | |
| | Autoimmune Encephalitis: N-Methyl-D-Aspartate (NMDA) | Patient must meet both of the following criteria 1. Cared for in consultation with a Neurologist AND 2. Used in conjunction with immunosuppressives and/or plasmapheresis | 2 g/kg divided over 2 to 5 days |
| | Autoimmune Encephalitis: Rasmussen's Encephalitis* | IVIG is used as a short term, temporizing measure | 2 g/kg divided over 2 to 5 days |
| | Autoimmune Optic Neuropathy | Patient has failed or has contraindications to steroids | 2 g/kg divided over 2 to 5 days |
| | Lambert-Eaton Myasthenic Syndrome (LEMS) | Order must be in consultation with a Neurologist | Induction dose: 2 g/kg in 2 to 5 divided doses Maintenance dose: 0.4 to 1 g/kg every 2 to 6 weeks |
| | Multiple Sclerosis (MS) Relapsing/ Remitting Only | Patient must meet 1 of the following 2 criteria: 1. Pregnant/immediate post-partum period when other immunomodulation is contraindicated OR 2. Relapsing/remitting MS who fail or have contraindications to standard immunomodulatory therapies | 1 g/kg monthly with or without a 5-day induction of 0.4 g/kg daily |
| | Neuromyelitis Optica (NMO) | Patient has failed or has contraindications to plasma exchange and/or steroids | 1-2 g/kg in 2 to 5 divided doses |
| | Anti-myelin oligodendrocyte glycoprotein (Anti-MOG) syndromes | Patient has failed or has contraindications to immunosuppressive therapy | 2 g/kg in 2 to 5 divided doses Maintenance dose: 1 g/kg every 2 to 6 weeks |

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| Adult Neurology | Paraneoplastic Cerebellar Degeneration | Patient must meet both of the following criteria: 1. Treated within 1 month of symptom onset <i>AND</i> 2. Used in conjunction with chemotherapy treatment | 2 g/kg every 4 to 6 weeks |
| | Stiff Person Syndrome | Patient has failed or has contraindications to GABAergic medications | 2 g/kg divided over 2 to 5 days every 4 to 6 weeks |

*** May be considered URGENT if notified by ordering physician as such**

| | Medical Condition | Pre-requisites | Dose/Frequency of Administration | | | | | | | | | | | | | | |
|---|---|--|----------------------------------|-------------|----------|---------|----------|--------------------------------------|----------|--------------------------------|----------|---|----------|--------------------|----------|--------------------------------|---------------------------------|
| Pediatric Neurology | Indicated Conditions | | | | | | | | | | | | | | | | |
| | Guillain-Barré Syndrome (GBS)* | Patient must meet both of the following criteria: 1. IVIG is being given within 2 weeks of symptom onset AND 2. Hughes Disability score of 3 or more or less than 3 with symptoms progressing Hughes Disability Scale: <table border="1" style="margin-left: auto; margin-right: auto;"> <thead> <tr> <th>Grade</th> <th>Description</th> </tr> </thead> <tbody> <tr> <td style="text-align: center;">0</td> <td>Healthy</td> </tr> <tr> <td style="text-align: center;">1</td> <td>Minor signs or symptoms, able to run</td> </tr> <tr> <td style="text-align: center;">2</td> <td>Able to walk 5 m independently</td> </tr> <tr> <td style="text-align: center;">3</td> <td>Able to walk 5 m with a walker, stick or one-person support</td> </tr> <tr> <td style="text-align: center;">4</td> <td>Bed or chair bound</td> </tr> <tr> <td style="text-align: center;">5</td> <td>Requiring assisted ventilation</td> </tr> </tbody> </table> | Grade | Description | 0 | Healthy | 1 | Minor signs or symptoms, able to run | 2 | Able to walk 5 m independently | 3 | Able to walk 5 m with a walker, stick or one-person support | 4 | Bed or chair bound | 5 | Requiring assisted ventilation | 2 g/kg divided over 2 to 5 days |
| | Grade | Description | | | | | | | | | | | | | | | |
| | 0 | Healthy | | | | | | | | | | | | | | | |
| | 1 | Minor signs or symptoms, able to run | | | | | | | | | | | | | | | |
| | 2 | Able to walk 5 m independently | | | | | | | | | | | | | | | |
| 3 | Able to walk 5 m with a walker, stick or one-person support | | | | | | | | | | | | | | | | |
| 4 | Bed or chair bound | | | | | | | | | | | | | | | | |
| 5 | Requiring assisted ventilation | | | | | | | | | | | | | | | | |
| Myasthenia Gravis (MG)* | Patient must meet 1 of the following 3 criteria: 1. Acute exacerbation (myasthenic crisis) OR 2. Optimization prior to surgery and/or thymectomy OR 3. As maintenance therapy for moderate to severe MG in combination with immunosuppressive agents | 2 g/kg divided over 2 to 5 days | | | | | | | | | | | | | | | |
| Possibly Indicated Conditions | | | | | | | | | | | | | | | | | |
| Acute Disseminated Encephalomyelitis (ADEM)* | Patient failed to respond to or has contraindications to corticosteroids | 1 g/kg daily for 2 days every 4 to 6 weeks | | | | | | | | | | | | | | | |
| Autoimmune Encephalitis: N-Methyl-D-Aspartate (NMDA)* | Patient must meet both of the following criteria 1. Cared for in consultation with a pediatric Neurologist AND 2. Used in conjunction with immunosuppressives and/or plasmapheresis | 1 g/kg daily for 2 days | | | | | | | | | | | | | | | |
| Autoimmune Encephalitis: Rasmussen's Encephalitis | IVIG is used as a short term, temporizing measure | 2 g/kg daily for 2 days | | | | | | | | | | | | | | | |

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|---------------------|--|--|-----------------------|
| Pediatric Neurology | Post-streptococcal Autoimmune Disorders: Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections (PANDAS), Pediatric Acute-onset Neuropsychiatric Syndrome (PANS) and Sydenham's Chorea | Order must be in consultation with a pediatric Neurologist | 1 to 2 g/kg per month |
|---------------------|--|--|-----------------------|

*** May be considered URGENT if notified by ordering physician as such**

3.3 Immunology

| | Medical Condition | Pre-requisites | Dose/Frequency of Administration |
|------------------|---|---|--|
| Adult Immunology | Indicated Conditions | | |
| | Inborn Errors of Immunity (IEI) also known as Primary Immunodeficiency (PID)* | <p>Order must be in consultation with an Immunologist, Hematologist, General Internist or Infectious Disease Specialist</p> <p>Monitor IgG trough level every 3 to 6 months to maintain 7 – 10g/L in most patients</p> <p>May be considered urgent if acute/severe infection</p> | <p>*IVIG dose: 0.4 to 0.7 g/kg every 3 to 4 weeks</p> <p>*SCIG dose: 0.1 to 0.23 g/kg every week</p> |
| | Secondary Immunodeficiency (SID)* | <p>Patient has/had recent life-threatening or recurrent clinically significant infection(s) related to low levels of polyclonal immunoglobulin</p> <p>May be considered urgent if acute/severe infection</p> | <p>*IVIG dose: 0.4 to 0.7 g/kg every 3 to 4 weeks</p> <p>*SCIG dose: 0.1 to 0.23 g/kg every week</p> |
| | Possibly Indicated Conditions | | |
| | Chronic Idiopathic Urticaria | <p>Patient must meet both of the following criteria</p> <ol style="list-style-type: none"> Has failed to respond or has contraindications to high dose antihistamines <p>AND</p> <ol style="list-style-type: none"> Failed to respond or has contraindications to Omalizumab (if covered). | <p>Induction dose: 1 g/kg/day for 3 days</p> <p>Maintenance dose: 1 g/kg every 4 weeks</p> |

*** May be considered URGENT if notified by ordering physician as such**

| | Medical Condition | Pre-requisites | Dose/Frequency of Administration |
|----------------------|---|---|---|
| Pediatric Immunology | Indicated Conditions | | |
| | Inborn Errors of Immunity (IEI) also known as Primary Immunodeficiency (PID)* | Order must be in consultation with an Immunologist <i>May be considered urgent if acute/severe infection</i> | *IVIG dose: 0.4 to 0.7 g/kg every 3 to 4 weeks *SCIG dose: 0.1 to 0.23 g/kg every week |
| | Secondary Immunodeficiency (SID)* | Order must be in consultation with an Immunologist or a Hematologist <i>May be considered urgent if acute/severe infection</i> | *IVIG dose: 0.4 to 0.7 g/kg every 3 to 4 weeks *SCIG dose: 0.1 to 0.23 g/kg every week |

*** May be considered URGENT if notified by ordering physician as such**

3.4 Dermatology

| | Medical Condition | Pre-requisites | Dose/Frequency of Administration |
|-------------------|--|--|---|
| Adult Dermatology | Indicated Conditions | | |
| | Scleromyxedema | Patient failed to respond or has contraindications to corticosteroids | 0.4 g/kg/day for 5 consecutive days every 4 weeks |
| | Systemic Vasculitic Syndromes including Polyarteritis Nodosa and Livedoid Vasculopathy | Order must be in consultation with a Dermatologist | 2 g/kg every 4 weeks |
| | Possibly Indicated Conditions | | |
| | Chronic Idiopathic Urticaria | Patient must meet both of the following criteria 1. Has failed to respond or has contraindications to high dose antihistamines AND 2. Failed to respond or has contraindications to Omalizumab (if covered). | Induction dose: 1 g/kg/day for 3 days Maintenance dose: 1 g/kg every 4 weeks |
| | Dermatomyositis* | Patient must meet both of the following criteria 1. Has significant muscle weakness AND 2. Failed to respond or has contraindications to corticosteroids Treatment is prescribed by a Dermatologist | 2 g/kg divided over 2 to 5 days |
| | Necrobiotic Xanthogranuloma | Patient failed to respond or has contraindications to corticosteroids | 2 g/kg every 4 weeks |
| | Pyoderma Gangrenosum | Patient must meet both of the following criteria 1. Cared for in consultation with a Dermatologist AND 2. Failed to respond or has contraindications to systemic steroids | 2 g/kg every 4 weeks |

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|-------------------|---|--|----------------------|
| Adult Dermatology | Severe Forms of Autoimmune Blistering Diseases (Pemphigus vulgaris, Pemphigus foliaceus, Pemphigoid, Cicatricial Pemphigoid, Linear IgA disease, Epidermolysis bullosa acquisita, Pemphigoid gestationis) | <p>Patient must meet both of the following criteria</p> <ol style="list-style-type: none"> 1. Disease is rapidly progressing <p>AND</p> <ol style="list-style-type: none"> 2. Failed to respond or has contraindications to systemic steroids <p>Treatment is prescribed by a Dermatologist</p> | 2 g/kg every 4 weeks |
| | Severe Lupus Erythematosus | Patient failed to respond or has contraindications to corticosteroids | 2 g/kg every 4 weeks |

*** May be considered URGENT if notified by ordering physician as such**

| | Medical Condition | Pre-requisites | Dose/Frequency of Administration |
|-----------------------|--|---|---|
| Pediatric Dermatology | Indicated Conditions | | |
| | Scleromyxedema | Patient failed to respond or has contraindications to corticosteroids | 0.4 g/kg/day for 5 consecutive days every 4 weeks |
| | Systemic Vasculitic Syndromes including Polyarteritis Nodosa and Livedoid Vasculopathy | Order must be in consultation with a Dermatologist | 2 g/kg every 4 weeks |
| | Possibly Indicated Conditions | | |
| | Chronic Idiopathic Urticaria | Patient must meet both of the following criteria 1. Has failed to respond or has contraindications to high dose antihistamines AND 2. Failed to respond or has contraindications to Omalizumab (if covered) | Induction dose: 1 g/kg/day for 3 days Maintenance dose: 1 g/kg every 4 weeks |
| | Necrobiotic Xanthogranuloma | Patient failed to respond or has contraindications to corticosteroids | 2 g/kg every 4 weeks |
| | Pediatric Atopic Dermatitis | Patient must meet both of the following criteria 1. Treatment is at the direction of a Dermatologist AND 2. Patient failed to respond or has contraindications to topical steroids and calcineurin inhibitors | 2 g/kg every 4 weeks |
| | Pyoderma Gangrenosum | Patient must meet both of the following criteria 1. Is cared for in consultation with a Dermatologist AND 2. Failed to respond or has contraindications to systemic steroids | 2 g/kg every 4 weeks |

| | | | |
|-----------------------|---|--|----------------------|
| Pediatric Dermatology | Severe Forms of Autoimmune Blistering Diseases (Pemphigus vulgaris, Pemphigus foliaceus, Pemphigoid, Cicatricial Pemphigoid, Linear IgA disease, Epidermolysis bullosa acquisita, Pemphigoid gestationis) | Patient must meet both of the following criteria 1. Disease is rapidly progressing <i>AND</i> 2. Failed to respond or has contraindications to systemic steroids Treatment is prescribed by a Dermatologist | 2 g/kg every 4 weeks |
| | Severe Lupus Erythematosus | Patient failed to respond or has contraindications to corticosteroids | 2 g/kg every 4 weeks |

*** May be considered URGENT if notified by ordering physician as such**

3.5 Rheumatology

| | Medical Condition | Pre-requisites | Dose/Frequency of Administration |
|--------------------|---|---|---|
| Adult Rheumatology | Indicated Conditions | | |
| | Immune-Mediated Inflammatory Myositis* | Patient must meet 1 of the following 2 criteria 1. Failed to respond to or has contraindications to corticosteroids with/without immunosuppressive therapies AND/OR 2. The presence of life-threatening disease | Initial dose: 2 g/kg divided over 2 to 5 days every 4 to 6 weeks (Taper when disease stable) |
| | Possibly Indicated Conditions | | |
| | Catastrophic Antiphospholipid Antibody Syndrome* | Order must be in consultation with a Rheumatologist or a Hematologist | 2 g/kg divided over 2 to 5 days |
| | Adult-onset Still's Disease | Order must be in consultation with a Rheumatologist | 2 g/kg divided over 2 to 5 days |
| | Sjogren's Syndrome | Order must be in consultation with a Rheumatologist | 2 g/kg divided over 2 to 5 days |
| | Hemophagocytic Lymphohistiocytosis (HLH)* | Order must be in consultation with a Rheumatologist, Hematologist or General Internist | 2 g/kg divided over 2 to 5 days |
| | Multisystem Inflammatory Syndrome in Adults (MIS-A) | Order must be in consultation with a Rheumatologist | 2 g/kg over 1 to 2 days |

*** May be considered URGENT if notified by ordering physician as such**

| | Medical Condition | Pre-requisites | Dose/Frequency of Administration |
|------------------------|---|---|--|
| Pediatric Rheumatology | Indicated Conditions | | |
| | Juvenile Dermatomyositis* | Patient must meet both of the following criteria 1. Glucocorticoids and other 2 nd line agents are contraindicated OR IVIG is part of early therapy in a critically ill child <i>AND</i> 2. Cared for in consultation with a pediatric Rheumatologist | 2 g/kg every 2 to 4 weeks |
| | Kawasaki Syndrome* | No criteria are required other than a diagnosis of Kawasaki Syndrome | 2 g/kg given once If failure to respond to initial dose, a 2 nd dose may be given at least 24 hours after the 1 st dose |
| | Systemic Onset Juvenile Idiopathic Arthritis* | Patient must meet both of the following criteria 1. Is resistant to other forms of therapy <i>AND</i> 2. Cared for in consultation with a pediatric Rheumatologist | 1 to 2 g/kg every 2 to 4 weeks |
| | Possibly Indicated Conditions | | |
| | Multisystem Inflammatory Syndrome in Children (MIS-C) | Cared for in consultation with a pediatric Rheumatologist | 2 g/kg given once |
| | Hemophagocytic Lymphohistiocytosis /Macrophage Activation Syndrome (HLH/MAS)* | Cared for in consultation with a pediatric Rheumatologist, pediatric Hematologist or pediatric Immunologist | 2 g/kg given once |

*** May be considered URGENT if notified by ordering physician as such**

3.6 Infectious Disease

| | Medical Condition | Pre-requisites | Dose/Frequency of Administration |
|--|---|---|--|
| Adult and Pediatric Infectious Disease | Indicated Conditions | | |
| | Group A Streptococcus (GAS) Necrotizing Fasciitis or Toxic Shock Syndrome* | Patient must be treated with a combination therapy of antibiotics and IVIG | 1 g/kg on day 1 and 0.5 g/kg per day on days 2 and 3 OR 0.15 g/kg per day for 5 days |
| | Staphylococcus Aureus Toxic Shock Syndrome (TSS)* | Patient must be treated with a combination therapy of antibiotics and IVIG | 1 g/kg on day 1 and 0.5 g/kg per day on days 2 and 3 OR 0.15 g/kg per day for 5 days |
| | Possibly Indicated Conditions | | |
| | Chronic Parvovirus Infection with Anemia | Immunocompromised patient with parvovirus B19 causing Pure Red Cell Aplasia | Initial dose: 0.4 to 1 g/kg for 5 to 10 days Maintenance dose: 0.4 g/kg every 4 weeks |
| Measles Post-Exposure Prophylaxis | <ol style="list-style-type: none"> Susceptible pregnant individuals OR immunocompromised individuals 6 months of age and older AND IVIG should only be provided within 6 days of measles exposure | 0.4g/kg given once | |

* May be considered URGENT if notified by ordering physician as such

3.7 Solid Organ Transplant

| | Medical Condition | Pre-requisites | Dose/Frequency of Administration |
|--|--|---|---|
| Adult and Pediatric Solid Organ Transplant | Indicated Conditions | | |
| | Acute Antibody Mediated Rejection* | Patient must meet the following criterion: <ul style="list-style-type: none"> • Pathology proven acute antibody mediated rejection | IVIG is commonly administered as part of a treatment protocol that includes plasmapheresis. 0.2 g/kg after each plasmapheresis session up to a total of 10 doses (i.e. 2 g/kg maximum cumulative dose) then reassess. Additional doses may be required depending on response. |
| | Possibly Indicated Conditions | | |
| | Chronic Parvovirus Infection with Anemia | Immunocompromised patient with parvovirus B19 causing Pure Red Cell Aplasia | Initial dose: 0.4 to 1 g/kg for 5 to 10 days Maintenance dose: 0.4 g/kg every 4 weeks |
| BK Polyomavirus (BKV)* | Immunocompromised patient with a pathological diagnosis of BK Polyomavirus | 0.2 g/kg per week for 5 doses (i.e. 1 g/kg maximum cumulative dose) then reassess. Additional doses may be required depending on response. | |

* May be considered URGENT if notified by ordering physician as such

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Appendix A – Atlantic Clinical Experts

| Specialty | Region | Contact Details |
|--|---------------|--|
| Hematology - Adult | Atlantic | Hematologist on call: (902) 473-2220 locating Fax if non urgent: (902) 473-3910 |
| Hematology - Pediatric | Atlantic | Pediatric Hematologist/Oncologist on call: (902) 470-8888 |
| Neurology – Adult | Atlantic | Call Dr. Ian Grant or designate in his absence Ph: (902) 473-3731 fax: (902) 473-4438 |
| Neurology – Pediatric | Atlantic | Pediatric Neurologist on call: (902) 470-8888 |
| Immunology – Adult | Atlantic | Call Dr. Gina Lacuesta or Dr. Lori Connors in Dr. Lacuesta’s absence Ph: (902) 425-3927 fax: (902) 425-3928 |
| Immunology – Pediatric | Atlantic | Pediatric Immunology Specialist on call: (902) 470-8888 |
| Rheumatology – Adult | Atlantic | Dr. Volodko Bakowsky Ph: (902) 470-7040 Fax: (902) 473-7019 In his absence Rheumatologist on call: (902) 473-2220 |
| Rheumatology – Pediatric | Atlantic | Dr. Adam Huber Ph: (902) 470-8827 fax: (902) 470-7217 |
| Infectious Disease – Adult | Atlantic | Infectious Disease Specialist on call: (902) 473-5553 |
| Infectious Disease – Pediatric | Atlantic | Pediatric Infectious Disease Specialist on call: (902) 470-8888 |
| Dermatology – Adult & Pediatric | Atlantic | Dr. Peter Hull Ph: (902) 473-7934 cell: (902) 817-6010 Dermatologist on call: 1-800-701-7774 |
| Solid Organ Transplant – Adult | Atlantic | Dr. Ken West Ph: (902) 473-2099 Pager: 2188 |
| Solid Organ Transplant - Pediatric | Atlantic | Dr. Phil Acott Ph: (902) 470-8195 Fax: (902) 470-8900 Pager: 1987 |