



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

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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
Grade	Description															
0	Healthy															
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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>														

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

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>0</td> <td>Healthy</td> </tr> <tr> <td>1</td> <td>Minor signs or symptoms, able to run</td> </tr> <tr> <td>2</td> <td>Able to walk 5 m independently</td> </tr> <tr> <td>3</td> <td>Able to walk 5 m with a walker, stick or one-person support</td> </tr> <tr> <td>4</td> <td>Bed or chair bound</td> </tr> <tr> <td>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															
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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															

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
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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>

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	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

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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

Adult 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

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	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

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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 <i>AND/OR</i> 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

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	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 AND 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 AND 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

4. References

Adamski, H. et al (2011) Solar Urticaria Treated with Intravenous Immunoglobulin's, *Journal of American Academy of Dermatology* 2011 Aug;65(2):336-40. doi: 10.1016/j.jaad.2010.05.040

Ahmed, M. et al (2020) Multisystem Inflammatory Syndrome in Children: A Systematic Review, *EClinicalMedicine* (2020) <https://doi.org/10.1016/j.eclinm.2020.100527>

Anderson, D. et al (2007) Guidelines on the Use of Intravenous Immune Globulin for Hematologic Conditions, *Transfusion Medicine Reviews*, Vol 21, No 2, Suppl 1 (April), 2007: pp S9-S56

ACR MIS-C and COVID-19 Related Hyperinflammation Task Force (2020) Clinical Guidance for Pediatric Patients with Multisystem Inflammatory Syndrome in Children (MIS-C) Associated with SARS-CoV-2 and Hyperinflammation in COVID-19, *Arthritis & Rheumatology*, November 2020

Arginelli, F. et al (2016) Long-term Efficacy of High Doses of Intravenous Immunoglobulins in generalized Scleromyxoedema: Case Report. *Journal of Internal Medicine*. 2016 Sep; 44(1 suppl): 109-112 doi: 10.1177/0300060515593259

Asano, Y. et al (2006) High-dose Intravenous Immunoglobulin Infusion in Polyarteritis Nodosa: Report on One Case and Review of the Literature, *Clinical Rheumatology* 2006 May;25(3):396-8. doi: 10.1007/s10067-005-0015-2

Basharat P *Curr Rheumatol Rep* 2015 Dec; 17(12): 72

Bidier, M. et al (2012) Scleromyxoederm: Clinical Follow-up After Successful Treatment with High-dose Immunoglobulins Reveals Different Long-term Outcomes, *Acta Dermatology and Venereology* 2012 July;92(4):408-9. doi: 10.2340/00015555-1299

Bohmig GA, et al. Immunoabsorption in severe c4d-positive acute kidney allograft rejection: a randomized controlled trial. *Am J Transplant* 2007;7:117-121.

Bounfour, T. et al (2013) Intravenous Immunoglobulins in Difficult-to-treat Ulcerated Livedoid Vasculopathy; Five Cases and a Literature Review, *International Journal of Dermatology* 2013 Sep;52(9):1135-9. doi: 10.1111/j.1365-4632.2012.05826.x.

Cakmak, S. et al (2013) Intravenous Immunoglobulin Therapy in Dermatology: An Update, *Inflamm Allergy Drug Targets* 2013 Apr;12(2):132-46.

Canavan, T. et al (2015) Mycoplasma Pneumoniae-induced Rash and Mucositis as a Syndrome Distinct from Stevens-Johnson Syndrome and Erythema Multiforme: A Systematic Review, *Journal of Academy of Dermatology* 2015 Feb; 72(2): 239-45/ doi: 10.1016/j.jaad.2014.06.026

Carapetis et al, (2014) Effectiveness of Clindamycin and Intravenous Immunoglobulin, and Risk of Disease in Contacts, in Invasive Group A Streptococcal Infections, *Clinical Infectious Diseases* 2014; 59(3): 358-65. doi: 10.1093/cid/ciu304

Chiarello, F. et al (2017) An Expert Opinion on PANDAS/PANS: Highlights and Controversies, *International Journal of Psychiatry in Clinical Practice* 2017, vol. 21, No. 2, 91-98
<https://doi.org/10.1080/13651501.2017.1285941>

Clin Rev Allergy Immunol 2016 Jan 14 – Statin-induced necrotizing myositis

Cohen-Buca A, et al. Advances in BK virus complications in organ transplantation and beyond. *Kidney Med* 2020;2(1):771-786.

Cummins, D. et al (2007) Treatment of Pyoderma Gangrenosum with Intravenous Immunoglobulin, *British Journal of Dermatology* 2007 Dec;157(6):1234-9 Epub 2007 Oct 4. doi: 10.1111/j.1365-2133.2007.08217.x.

Czernik, A. (2014) Intravenous Immunoglobulin G in the Treatment of Autoimmune Bullous Disease, *Clinical Exp Immunol*. 2014 Dec;178 Suppl 1:118-9. doi: 10.1111/cei.12535

Dalakas et al (1993) A Controlled Trial of High-dose Intravenous Immune Globulin Infusions as Treatment for Dermatomyositis, *The New England Journal of Medicine* 1993 Vol. 329 No, 27

Dawn, G. et al (2003) Effect of High-dose Intravenous Immunoglobulin in Delayed Pressure Urticaria, *British Journal of Dermatology* 2003 Oct;149(4):836-40

De Zwaan, S. et al (2009) Treatment of Refractory Pyoderma Gangrenosum with Intravenous Immunoglobulin, *Australia Journal of Dermatology*. 2009 Feb;50(1):56-9. doi: 10.1111/j.1440-0960.2008.00506.x. doi: 10.1093/rheumatology/keq021

Dourmishev, L. (2016) Intravenous Immunoglobulins: Mode of Action and Indications in Autoimmune and Inflammatory Dermatoses, *International Journal of Inflammation* 2016;2016:3523057 doi: 10.1155/2016/3523057

Eastham, A. et al (2014) Paraproteinemia-associated Scleredema Treated Successfully with Intravenous Immunoglobulin, *JAMA Dermatology* 2014 Jul;150(7):788-9. doi: 10.1001/jamadermatol.2013.8835

Enk, A. (2009) Guidelines on the Use of High-dose Intravenous Immunoglobulin in Dermatology, *European Journal of Dermatology* 2009 Jan-Feb;19(1):90-8. doi: 10.1684/ejd.2008.0580

Enk, A. et al (2016) European Guidelines (s1) on the Use of High-dose Intravenous Immunoglobulin in Dermatology, *Journal of European Academy of Dermatology and Venereology*. 2016 July 13; doi: 10.1111/jvd.13725 [Epub ahead of print]

Feasby T., Banwell B., Benstead T. et coll. « Guidelines on the Use of Intravenous Immune Globulin for Neurologic Conditions », *Transfusion Medicine Reviews*, vol. 21, no 2, suppl. 1 (avril 2007), p. S57-S107

Fernandez, A. and Kerdel, F. (2007) The Use of I.V. IG Therapy in Dermatology, *Dermatology Ther.* 2007 Jul-Aug;20(4):288-305. doi: 10.1111/j.1529-8019.2007.00142.x.

Frankovich, J. et al (2017) Clinical Management of Pediatric Acute-Onset Neuropsychiatric Syndrome: Part II- Use of Immunomodulatory Therapies, *Journal of Child and Adolescent Psychopharmacology*. Vol. 27, no. 7, p. 574-593, doi: 10.1089/cap.2016.0148

Ghazal, P. and Dissemond, J. (2015) Therapy of Pyoderma Gangrenosum in Germany: results of a Survey Among Wound Experts, *Deutsche Dermatologische Gesellschaft (DDG)* 2015 apr; 13(4): 317-24. doi: 10.1111/ddg.12585

Goodarzi, H. et al (2012) Effective Strategies for the Management of Pyoderma Gangrenosum, *Advance Wound Care (New Rochelle)*. 2012 Oct;1(5):194-199. doi: 10.1089/wound.2011.0339

Goodfield, M. et al (2004) Intravenous Immunoglobulin (IVIg) for Therapy-Resistant Cutaneous Lupus Erythematosus (LE), *Journal of Dermatology Treatment* 2004 Jan;15(1):46-50. doi: 10.1080/0951440042000269

Gourgiotou, K. et al (2002) Epidermolysis Bullosa Acquisita: Treatment with Intravenous Immunoglobulins, *European Journal of Dermatology and Venereology* 2002 Jan;16(1):77-80

Gubensek J, et al. Plasma exchange and intravenous immunoglobulin in the treatment of antibody-mediated rejection after kidney transplantation: a single-center historic cohort study. *Transplant Proc* 2013; 25: 1524-1527.

Hirsh HH et al. BK polyomavirus in solid organ transplantation-guidelines from the American society of transplantation infectious diseases community of practice. *Clin Transplant* 2019;33(9): e13528.

Hughes, R. et al (2009) Solar urticaria Successfully Treated with Intravenous Immunoglobulin, *Clinical Exp Dermatology*. 2009 Dec;34(8):e660-2. doi: 10.1111/j.1365-2230.2009.03374.x.

Hwang SD, et al. High-dose intravenous immunoglobulin treatment of polyomavirus nephropathy developing after t cell-mediated rejection treatment: a case report. *Transplant Proc* 2018;50:2575-2578.

Jonat, B. et al (2020) Multisystem Inflammatory Syndrome in Children Associated with Coronavirus Disease 2019 in a Children's Hospital in New York City: Patient Characteristics and an Institutional Protocol for Evaluation, Management and Follow-Up, *Pediatric Critical Care Medicine*, March 2021, Vol 22, No. 3 doi: 10.1097/PCC.0000000000002598

Jolles, S. et al (1998) Dermatological Uses of High-Dose Intravenous Immunoglobulin, *Arch Dermatology* 1998 Jan;134(1):80-6

Kable K, et al. Clearance of BK virus nephropathy by combination antiviral therapy with intravenous immunoglobulin. *Transplant Direct* 2017;3:e142.

Kasiske BL, et al. Kidney Disease: improving global outcomes. KDIGO clinical practice guideline for the care of kidney transplant recipients: a summary. *Kidney Int* 2010;77:299-311.

Kim, E et al (2015) Pulsed Intravenous Immunoglobulin Therapy in Refractory Ulcerative Livedoid Vasculopathy: Seven Cases and a Literature Review, *Dermatology Ther.* 2015 Sep-Oct; 28(5): 287-90 doi: 10.1111/dth.12233

Kito, Y. et al (2012) High-dose Intravenous Immunoglobulin Monotherapy for Drug-induced Hypersensitivity Syndrome, *Acta Dermatology and Venereology* 2012 Jan;92(1):100.1. doi:10.2340/00015555-1168

Kovacevic, M., Grant, P., Swedo, S. (2015) Use of Intravenous Immunoglobulin in the Treatment of Twelve Youths with Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections. *Journal of Child and Adolescent Psychopharmacology*, 25-1, 65-69. doi:10.1089/cap.2014.0067

Kozel, M. and Sabroe, R. (2004) Chronic Urticaria: Aetiology, Management and Current and Future Treatment Options, *Drugs* 2004;64(22):2515-36

Lancet Neurol. 2013 February; 12(2): 157–165. doi:10.1016/S1474-4422(12)70310-1

Lappin, E. and Ferguson, A. (2009) Gram-positive Toxic Shock Syndromes, *Lancet Infectious Disease* 2009; 9: 281-90.

Laureano, A and Cardoso, J (2015) Unilateral Oral Mucous Membrane Pemphigoid: Refractory Atypical Presentation Successfully Treated with Intravenous Immunoglobulins, *Case Report Dermatology Medicine* 2015;2015:930859. doi: 10.1155/2015/930859

Lefaucheur C, et al. Comparison of combination plasmapheresis/ivig/anti-cd20 versus high-dose ivig in the treatment of antibody-mediated rejection. *Am J Transplant* 2009; 9: 1099-1107.

Levine M, et al. Treatment options and strategies for antibody mediated rejection after renal transplantation. *Semin Immunol* 2012;24(2): 136-142.

Mahadeo, K.M. et al (2019) Management guidelines for paediatric patients receiving chimeric antigen receptor T cell therapy, *Nature Reviews, Clinical Oncology* vol. 16 January 2019 pg 45-63
Makatsori M *QJM* 2014 Oct; 107(10): 821-8

Matsumura S, et al. Clinical efficacy of intravenous immunoglobulin for bk polyomavirus-associated nephropathy after living kidney transplantation. *Ther Clin Risk Man* 2020;16:947-952.

Mitzel-Kaoukhov, H. (2009) Effect of High-dose Intravenous Immunoglobulin Treatment in Therapy-resistant Chronic Spontaneous Urticaria, *Ann Allergy Asthma Immunology*. 2010 Mar;104(3):253-8. doi: 10.1016/j.anai.2009.12.007

Monshi, B. et al (2014) Efficacy of Intravenous Immunoglobulins in Livedoid Vasculopathy: Long-term Follow-up of 11 patients, *Journal of American Academy of Dermatology* 2014 Oct;71(4):738-44. doi: 10.1016/j.jaad.2014.05.039

MS Research Unit, Department of Neurology, Copenhagen University Hospital (2003) The Role of Intravenous Immunoglobulin in the Treatment of Multiple Sclerosis, *Journal of the Neurological Sciences* 206 (2013) 123-130
Mulhearn, B. and Bruce, I. (2015) Indications for IVIG in Rheumatic Diseases, *Rheumatology* 2015;54:383-391. Doi: 10.1093/rheumatology/keu429

Mydlarski, P. et al (2004) Intravenous Immunoglobulin: Use in Dermatology, *Skin Therapy Lett* 2004 May;9(5):1-6

Mydlarski, P. et al (2006) Canadian Consensus Statement on the Use of Intravenous Immunoglobulin Therapy in Dermatology, *Journal of Cutaneous Med Surg*. 2006 Sep-Oct;10(5):205-21

NHS England (2016). Evidence Review: Intravenous immunoglobulin for autoimmune encephalitis. Turnkey Clinical Evidence Review Team on behalf of NHS England Specialised Commissioning. https://www.engage.england.nhs.uk/consultation/clinical-commissioning-wave8/user_uploads/f06x05-aie-evidence-rev.pdf

Nanda, A. et al (2012) Linear Immunoglobulin a Bullous Disease of Childhood Responsive to Intravenous Immunoglobulin Monotherapy, *Pediatric Dermatology* 2012 July-Aug;29(4):529-32. Doi: 10.1111/j.1525-1470.2011.01475.x.

National Advisory Committee on Blood and Blood Products (2007) Guidelines on the Use of Intravenous Immune Globulin for Neurologic Conditions.

Nova Scotia Provincial Blood Coordinating Team (2021) *Atlantic Guideline for Subcutaneous Immune Globulin Home Administration Programs*, Halifax, NS

Nuenert, C. et al (2011) The American Society of Hematology 2011 Evidence-based Practice Guideline for Immune Thrombocytopenia, *Blood* 2011 117:4190-4207; doi: <https://doi.org/10.1182/blood-2010-08-302984>

Nguyen, T. et al (2015) Positive Clinical Outcome with IVIG as Monotherapy in Recurrent Pemphigoid Gestationis, *International Immunopharmacol.* 2015 May; 26(1): 1-3. doi: 10.1016/j.intimp.2015.02.038

O'Donnell, B. et al (1998) Intravenous Immunoglobulin in Autoimmune Chronic Urticaria, *British Journal of Dermatology.* 1998 Jan;138(1):101-6

Oktem, A. et al (2016) Long-term Results of Rituximab-intravenous Immunoglobulin Combination Therapy in Patients with Epidermolysis Bullosa Acquisita Resistant to Conventional Therapy, *Journal of Dermatology Treatment* 2016 May 10: 1-5 doi: 10.1080/09546634.2016.1179711 [Epub ahead of print]

Patel, F. et al (2015) Effective Strategies for the Management of Pyoderma Gangrenosum: A Comprehensive Review, *Academy of Dermatology and Venereology* 2015 May;95(5):525-31. doi: 10.2340/00015555-2008

Pedrosa, A. et al (2015) Necrobiotic Xanthogranuloma With Giant Cell Hepatitis, Successfully Treated with Intravenous Immunoglobulins, *Journal of European Academy of Dermatology and Venereology* 2015 Mar-Apr; 28(2): 68-70. doi: 10.1111/dth.12211

Phuphanich S, Brock C. (2007) Neurologic Improvement After High-dose Intravenous Immunoglobulin Therapy in Patients With Paraneoplastic Cerebellar Degeneration Associated With Anti-Purkinje Cell Antibody, *Journal of Neuro-oncology* 2007 Jan;81(1):67-9

Prins, C. et al (2007) Intravenous Immunoglobulin: Properties, Mode of Action and Practical Use in Dermatology, *Acta Dermatology and Venereology* 2007;87(3):206-18. doi: 10.2340/00015555-0249

Provan, D. et al (2010) International Consensus Report on the Investigation and Management of Primary Immune Thrombocytopenia, *Blood* 2010 115:168-186 doi: 10.1182/blood-2009-06.225565

Pyrpasopoulou, A. et al (2007) Intravenous Immunoglobulins: a Valuable Asset in the Treatment of a Case of Septic Febrile Ulceronecrotic Mucha-Habermann Disease, *Dermatology* 2007;215(2):164-5 doi:10.1159/000104271

Rimensberger, P. et al (2020) Caring for Critically Ill Children with Suspected or Proven Coronavirus Disease 2019 Infection: Recommendations by the Scientific Sections' Collaborative of the European Society of Pediatric and Neonatal Intensive Care, *Pediatric Critical Care Medicine*, January 2021, Vol 22, No. 1 doi: 10.1097/PCC.0000000000002599

Rutter, A. and Luger, T. (2001) High-dose Intravenous Immunoglobulins: An approach to Treat Severe Immune-mediated and autoimmune Diseases of the Skin, *Journal of American Academy of Dermatology* 2001 Jun;44(6):1010-24. doi: 10.1067/mjd.2001.112325

Santos, G. et al (2014) Leg Ulcers in Antiphospholipid Syndrome Secondary to Systemic Lupus Erythematosus Treated with Intravenous Immunoglobulin, *Journal of Dermatology Case Report* 2014 Jun 30;8(2): 38-41. doi: 10.3315/jdcr.2014.1169

- Schinstock C, et al. Recommended treatment of antibody-mediated rejection after kidney transplantation: the 2019 expert consensus from the transplantation society working group. *Transplantation* 2020;104:911-922.
- Seidling, V. et al (2013) Analysis of High-dose Intravenous Immunoglobulin Therapy in 16 Patients With Refractory Autoimmune Blistering Skin Disease: High Efficacy and No Serious Adverse Events, *Acta Dermatology and Venereology* 2013 May;93(3):346-9. doi: 10.2340/00015555-1471
- Sener A, et al. Intravenous immunoglobulin as a treatment for bk virus associated nephropathy: one-year follow-up of renal allograft recipients. *Transplantation* 2006;81:117-120.
- Shehata, N. et al (2010) The Use of Immunoglobulin Therapy for Patients Undergoing Solid Organ Transplantation: An Evidence-Based Practice Guideline, *Transfusion Medicine Reviews*, Vol 24, No 1, Suppl 1 (January), 2010: pp S7-S27
- S. Jarius, B. Wildemann (2015) Medusa Head Ataxia: The Expanding Spectrum of Purkinje Cell Antibodies in Autoimmune Cerebellar Ataxia. Part 1: Anti-mGluR1, anti-Homer-3, anti-Sj/ITPR1 and anti-CARP VIII, *Journal of Neuro-inflammation*; 12: 166. Published online 2015 September 17. doi: 10.1186/s12974-015-0356-y
- Smith, D. et al (2007) Off-Label Uses of Biologics in Dermatology: Interferon and Intravenous Immunoglobulin (part 1 of 2), *Journal of American Academy of Dermatology* 2007 Jan;56(1):e1-54. doi: 10.1016/j.jaad.2006.06.016
- Smith, S. et al (2010) The Use of Intravenous Immunoglobulin for Treatment of Dermatological Conditions in Australia: A Review, *Australia Journal of Dermatology*. 2010 Nov;51(4):227-37. doi: 10.1111/j.1440-0960.2009.00578.x.
- Snydman, D. et al (2011) Update and Review: State-of-the-Art Management of Cytomegalovirus Infection and Disease Following Thoracic Organ Transplantation, *Transplantation Proceedings*, (2011) 43, S1-S17 doi: 10.1016/j.transproceed.2011.02.069
- Spadaro G *Clin Immunol* 2016 May; 166-167 – secondary hypogammaglobulinemia
- Sroa, N. et al (2010) Intravenous Immunoglobulin Therapy of Scleromyxedema: A Case Report and Review of Literature, *J Drugs Dermatology*. 2010 Mar;9(3):263-5
- Tekin, B. and Yucelten, A. (2015) Infantile Bullous Pemphigoid Treated Using Intravenous Immunoglobulin: Case Report and Review of the Literature, *Pediatric Dermatology* 2015 Sep-Oct;32(5):723-6. Doi: 10.1111/pde.12635
- Titulaer M.J., McCracken I, Gabilondo I. et coll. « Treatment and prognostic factors for long-term outcome in patients with anti-NMDA receptor encephalitis: an observational cohort study », *The Lancet Neurology*, vol. 12, no 2 (février 2013), p. 157–165. doi :10.1016/S1474-4422(12)70310-1
- Travi, G. and Pergam, S. (2014) Cytomegalovirus Pneumonia in Hematopoietic Stem Cell Recipients, *J Intensive Care Med*. 2014; 29 (4): 200-212. doi: 10.1177/0885066613476454
- Trebst, C. et al (2013) Update on the diagnosis and treatment of neuromyelitis optica: Recommendations of the Neuromyelitis Optica Study Group (NEMOS), *Journal of Neurology* 2014; 261:1-16

Tunis, M.C et al on behalf of the National Advisory Committee on Immunization (NACI) (2018) Updated NACI recommendation for measles post-exposure prophylaxis, *CCDR*, vol. 44-9, September 6, 2018: Respiratory Infections

<https://www.canada.ca/en/public-health/services/reports-publications/canada-communicable-disease-report-ccdr/monthly-issue/2018-44/issue-9-september-6-2018/article-7-naci-recommendation-pep.html>

Turnkey Clinical Evidence Review Team on behalf of NHS England Specialised Commissioning (2016) Evidence Review: Intravenous immunoglobulin for Autoimmune Encephalitis

https://www.engage.england.nhs.uk/consultation/clinical-commissioning-wave8/user_uploads/f06x05-aie-evidence-rev.pdf

Vaitla, P.M. and McDermott, E.M. (2010) The Role of High-dose Intravenous Immunoglobulin in Rheumatology *Rheumatology* 2010; 49:1040-1048

Van Immerzell, T., van Gilst, R., Hartwig, N., (2010) Beneficial Use of Immunoglobulins in the Treatment of Sydenham Chorea. *Eur J Pediatr* (2010) 169:1151–1154 doi 10.1007/s00431-010-1172-0

VU D, et al. Efficacy of intravenous immunoglobulin in the treatment of persistent bk viremia and bk virus nephropathy in renal transplant recipients. *Transplant Proc* 2015;47:394-398.

Walker, K., Wilmshurst, J. (2010) An update on the treatment of Sydenham’s chorea: the evidence for established and evolving interventions. *Therapeutic Advances in Neurological Disorder*. 3(5) 301_309 doi: 10.1177/1756285610382063

Wang, D. et al (2012) Intravenous Immunoglobulin Therapy in Adult Patients with Polymyositis/Dermatomyositis: A Systematic Literature Review, *Clinical Rheumatology* 2012 31:801-806. doi: 10.1007/s10067-012-1940-5

Widdess-Walsh P, Tavee JO, Schuele S, Stevens GH. (2003) Response to intravenous immunoglobulin in anti-Yo associated paraneoplastic cerebellar degeneration: case report and review of the literature, *Journal of Neuro-oncology* 2003;63:187-190

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