

Specialist Working Group for Immunology

Proposed changes to the *Criteria for the clinical use of intravenous immunoglobulin in Australia, Second Edition*

ITEM	CRITERIA FOR THE CLINICAL USE OF INTRAVENOUS IMMUNOGLOBULIN IN AUSTRALIA, SECOND EDITION (CRITERIA)	PROPOSED REVISIONS TO THE CRITERIA (INCLUDING ADAPTATION TO THE IG SYSTEM)	SWG RATIONALE FOR PROPOSED CHANGE (A) Administrative (B) Progressive (C) Programmed
Condition Name	Specific antibody deficiency	Specific antibody deficiency	
Specialty	Immunology	Immunology	
Chapter	6	6	
Specific Conditions	Specific antibody deficiency Existing IgG subclass	Specific antibody deficiency Existing IgG subclass	
Level of Evidence	Small case studies only, insufficient data (Category 4a).	Small case studies only, insufficient data (Category 4a).	Unchanged
Description and Diagnostic Criteria	<p>The term ‘specific antibody deficiency’ describes failure of specific antibody response to an antigen challenge, and is most often used in the more restrictive sense of applying to polysaccharide antibody responses only.</p> <p>Patients who have normal total IgG levels but impaired production of specific antibodies, including those with isolated deficient responses to numerous polysaccharide antigens after vaccination, can present a diagnostic challenge. IgG replacement therapy should be provided when there is well-</p>	<p>The term ‘specific antibody deficiency’ describes failure to produce a normal immunoglobulin G (IgG) antibody response following antigen challenge (post vaccination) in an individual with a normal serum total IgG level. It is most often used in the context of impaired IgG or IgG2 antibody response to pooled pneumococcal polysaccharides (PcPs) or serotype-specific PcPs, after a single vaccination with unconjugated pneumococcal polysaccharides. (Boyle et al 2006).</p> <p>The definition of an abnormal low IgG antibody response following vaccination with pneumococcal polysaccharides is still debated in</p>	<p>This section has been revised and updated with supporting references. Further improvements to describe the approach to diagnosis and testing requirements for approval of Ig therapy. (A)</p>

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	<p>documented severe polysaccharide non-responsiveness and evidence of recurrent infections with a documented requirement for antibiotic therapy and ongoing recurrent infections despite antibiotic prophylaxis (Orange et al 2006).</p> <p>It is now generally agreed that IgG subclass level estimation in serum is relatively poorly predictive of infectious risk and is of limited value in the definition of those patients most likely to benefit from IVIg therapy.</p> <p>Further research investigating clinical and laboratory features of this disorder is required.</p>	<p>development, but data are available to provide a working definition (Boyle et al 2006, Hare et al 2009, Borgers et al 2012). <u>For approval of Ig therapy, the patient should exhibit an inadequate serum IgG or IgG2 antibody response to pooled pneumococcal polysaccharides (PcPs), or serotype-specific PcPs, 4 weeks after a single vaccination with unconjugated pneumococcal polysaccharides.</u></p> <p>Rarely, patients with an increased susceptibility to respiratory tract infections exhibit impaired antibody responses to protein antigens, such as tetanus toxoid. A definition of an abnormal antibody response is not clearly established, but failure to produce a 'protective' serum level of IgG antibodies against tetanus toxoid following at least two vaccinations should be considered abnormal. Assessment of IgG antibody responses following vaccination with <i>Haemophilus influenzae</i> type B (Hib) may also be undertaken.</p> <p>Immunoglobulin therapy should be provided when there is evidence of abnormally recurrent or persistent bacterial infections, or bronchiectasis and/or suppurative lung disease consistent with the position statement of the Thoracic Society of Australia and New Zealand and the Australian Lung Foundation (Chang et al 2010), with a documented requirement for antibiotic therapy, associated with a low IgG antibody response following vaccination with pneumococcal polysaccharides</p>	

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		(see above) or tetanus toxoid. (see above) Further research investigating clinical and laboratory features of this group of disorders is required.			
Justification for Evidence Category					No update to this section
Diagnosis is required	A clinical immunologist must be consulted to confirm the diagnosis;	Yes	Which Speciality	Clinical immunologist	For SAD, respiratory physicians or clinical immunologists may be the treating specialist, but Clinical immunologist must make the diagnosis. (A)
Diagnosis must be verified		Yes	Which Speciality	Clinical immunologist if a clinical immunologist did not make the diagnosis	For existing IgG Subclass patients, confirmation of the diagnosis is required by a second immunologist. (A)
Exclusion Criteria	<ol style="list-style-type: none"> 1. Isolated IgG subclass deficiency in the absence of evidence of specific antibody deficiency. 2. Low total IgG. This should be considered under primary or secondary immunodeficiency. 	<p>Isolated IgG subclass deficiency in the absence of evidence of specific antibody deficiency.</p> <p>Low serum total IgG. This should be considered under primary or secondary immunodeficiency.</p>			Unchanged
Indications	Prevention of infections in individuals with frequent infections who have demonstrated failure to mount protective IgG antibody responses to vaccine antigen challenge	History of abnormally frequent and/or persistent bacterial infections associated with a demonstrated failure to mount a 'normal' IgG antibody response to pneumococcal polysaccharides or protein antigens, such as			Three indications are recommended. The second indication is required for re-entry of confirmed SAD patients that have a life threatening infection while on trial off therapy. A re-entry path is needed as

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	despite normal total serum IgG levels.	<p>tetanus toxoid, after vaccine challenge in a patient with a normal serum total IgG concentration.</p> <p>Life-threatening infection or a series of serious infections following trial-off Ig therapy in patients with proven specific antibody deficiency.</p> <p>Patients currently receiving immunoglobulin therapy for a diagnosis of IgG subclass deficiency are eligible for continued therapy, but a diagnosis of specific antibody deficiency should be confirmed following cessation of immunoglobulin therapy.</p>	<p>revaccination is contraindicated. The third indication is to cover existing IgG subclass patients to enable reassessment and requalification. (A)</p> <p>Some states have reasonable numbers of IgG subclass patients that have not been re-evaluated under earlier changes to the Criteria - this will be required during transfer to the Ig system unless there is presence of active suppurative lung disease (which is now required to comply with the position statement of the Thoracic Society of Australia and New Zealand (TSANZ). In 2013/14, there were 107 IgG subclass patients of which 80 were existing IgG without lung disease and 27 patients with suppurative lung disease. All IgG subclass patients will be required to requalify for ongoing access to Ig therapy. (A)</p>
Qualifying Criteria	<p>Specific Antibody deficiency</p> <p>To access IVIg for a period of 12 months, the following qualifying criteria must be met:</p> <ol style="list-style-type: none"> 1. A clinical immunologist must be consulted to confirm the diagnosis; 	<p>History of abnormally frequent and/or persistent bacterial infections associated with a demonstrated failure to mount a 'normal' IgG antibody response to pneumococcal polysaccharides or protein antigens, such as tetanus toxoid, after vaccine challenge in a patient with a normal serum total IgG concentration.</p> <ul style="list-style-type: none"> • History of abnormally frequent and/or persistent and significant bacterial infections 	<p>Evidence items will request the severity and number of recurrent bacterial infections in the last 12 months and record the type of vaccine challenge (Unconjugated pneumococcal polysaccharides, Conjugated pneumococcal polysaccharides, Tetanus</p>

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	<p>AND</p> <p>2. Frequent bacterial infections despite oral antibiotic therapy consistent with best practice recommendations;</p> <p>AND</p> <p>3. Documented failure of serum antibody response to unconjugated pneumococcal or protein vaccine challenge.</p>	<p>despite oral antibiotic prophylaxis therapy</p> <p>AND</p> <ul style="list-style-type: none"> Documented failure of serum antibody response <u>after vaccination with</u> conjugated or unconjugated pneumococcal vaccine or <u>following</u> protein vaccine challenge. <p>Please note: a diagnosis of bronchiectasis and/or suppurative lung disease must be consistent with position statement of the Thoracic Society of Australia and New Zealand and the Australian Lung Foundation (Chang et al 2010).</p> <p>Life-threatening infection or a series of serious infections following trial-off Ig therapy in patients with proven specific antibody deficiency.</p> <ul style="list-style-type: none"> Patient with proven specific antibody deficiency undertaking a trial-off Ig therapy <p>AND</p> <ul style="list-style-type: none"> One life-threatening bacterial infection in the previous 12 months <p>OR</p>	<p>toxoid or Conjugated Hib polysaccharides) and Specific IgG response and details of prophylactic antibiotics given. (A)</p> <p>The purpose of the position statement is to require that the diagnosis of bronchiectasis or suppurative lung disease has been confirmed. (A)</p>

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	<p>IgG Subclass</p> <p><u>New patients</u> IVIg is not funded for new patients diagnosed with IgG subclass deficiency.</p> <p><u>Patients who were receiving IVIg for IgG subclass deficiency before initial publication of the Criteria (December 2007)</u></p> <ul style="list-style-type: none"> Without clinically active bronchiectasis or suppurative lung disease: These patients should have ceased IVIg and had their immunological status re-evaluated. Patients with a confirmed IgG deficiency have become eligible under another indication (e.g. primary immunodeficiency with antibody deficiency). Patients without a confirmed IgG deficiency have ceased IVIg therapy. With clinically active bronchiectasis or suppurative lung disease over the 	<ul style="list-style-type: none"> at least two serious infections in the last six months requiring more than standard courses of antibiotics (eg. Hospitalisation, intravenous or prolonged antibiotic therapy). <p>Please note: a diagnosis of bronchiectasis and/or suppurative lung disease must be consistent with position statement of the Thoracic Society of Australia and New Zealand and the Australian Lung Foundation (Chang AB et al. Med J Australia 2010; 193:356-65).</p> <p>Patients currently receiving immunoglobulin therapy for a diagnosis of IgG subclass deficiency are eligible for continued therapy, but a diagnosis of specific antibody deficiency should be confirmed following cessation of immunoglobulin therapy.</p> <ul style="list-style-type: none"> Existing patient on IgG therapy funded under the National Blood Arrangements in Australia <p>AND</p> <ul style="list-style-type: none"> Clinically active bronchiectasis and/or suppurative lung disease proven by radiology (see note*) 	

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	<p>previous 12 months: To be eligible to receive IVIg for a further 12 months, the following is required:</p> <p>1 Written confirmation from the treating clinical immunologist that :</p> <ul style="list-style-type: none"> • an annual review has been undertaken; • the patient has demonstrated clinical benefit; and • a trial period of cessation of IVIg for the purpose of immunological evaluation is medically contraindicated on safety grounds. <p>AND</p> <p>2 Written confirmation by a second physician that cessation of IVIg for the purpose of immunological evaluation is medically contraindicated on safety grounds.</p> <p>NOTE: The above criteria for initial and ongoing access to IVIg funded by all governments under the National Blood Arrangements will be reviewed in light of emerging evidence at the next review of the Criteria.</p>	<p>AND</p> <ul style="list-style-type: none"> • The patient has had demonstrated clinical benefit over the last 12 months and has had at least two episodes of infection requiring antibiotics <p>AND</p> <ul style="list-style-type: none"> • A trial cessation of therapy will be undertaken next September or October to undertake an immunological evaluation, unless a trial period of cessation of immunoglobulin therapy for the purpose of immunological evaluation is medically contraindicated on safety grounds (e.g. neutropenia, or other cytopenia, immunosuppressant medications or clinically active bronchiectasis and/or suppurative lung disease*) <p>AND</p> <ul style="list-style-type: none"> • A review has been undertaken by a second immunologist, with written confirmation of the eligibility for ongoing Ig treatment documenting the contra-indication reason for not trialling off Ig treatment. <p>*Please note: a diagnosis of bronchiectasis and/or</p>	

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		suppurative lung disease must be consistent with position statement of the Thoracic Society of Australia and New Zealand and the Australian Lung Foundation (Chang et al 2010).	
Review Criteria	<p>Natural history of specific antibody deficiency remains poorly defined, although antibody production will improve for many patients over time, particularly children.</p>	<p>History of abnormally frequent and/or persistent bacterial infections associated with a demonstrated failure to mount a 'normal' IgG antibody response to pneumococcal polysaccharides or protein antigens, such as tetanus toxoid, after vaccine challenge in a patient with a normal serum total IgG concentration.</p> <p>Natural history of specific antibody deficiency remains poorly defined, although antibody production will improve for many patients over time, particularly children.</p> <p>Regular review by a Clinical Immunologist or Respiratory Physician is required, at least annually. Documentation of clinical effectiveness is necessary for continuation of Ig therapy.</p> <p>Cessation of immunoglobulin therapy should be considered at least after each 12 months of Ig therapy, extended as required to enable cessation of therapy in September or October.</p>	

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	<p>To be eligible to receive IVIg for a further 12 months, the following is required:</p> <p>Written confirmation from the treating clinical immunologist that:</p> <ul style="list-style-type: none"> • an annual review has been undertaken; • the patient had demonstrated clinical 	<p>This should particularly be considered in patients who do not have active bronchiectasis and/or suppurative lung disease. An immunoglobulin washout period of four to six months is necessary to enable an accurate assessment. Prophylactic antibiotics may be considered to cover the period of cessation of immunoglobulin therapy.</p> <p>Patients may qualify for further immunoglobulin therapy:</p> <ul style="list-style-type: none"> • under other immunodeficiency criteria (e.g. primary antibody deficiency disorder), depending on the results of subsequent immune evaluation; or • rarely under specific antibody deficiency following re-emergence of severe significant infection requiring hospitalisation. <p>In principle, immunoglobulin therapy should only be continued or renewed if there is a demonstrated clinical benefit. Note that re-vaccination with unconjugated pneumococcal polysaccharide vaccine is not recommended because of the potential for immune hyporesponsiveness induced by repeated vaccination (O'Brien et al 2007).</p> <p>On review of the authorisation period</p>	

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	<p>benefit;</p> <ul style="list-style-type: none"> a trial period of cessation of IVIg for the purpose of immunological evaluation is medically contraindicated on safety grounds. <p>Cessation of IVIg should be considered, at least after each 12 months of therapy extended as required to enable cessation of therapy in September/October.</p> <p>This should particularly be considered in patients who do not have suppurative lung disease or bronchiectasis. An immunoglobulin washout period of four to six months is necessary to enable an accurate assessment. Prophylactic antibiotics may be considered to cover the period of IVIg cessation.</p> <p>Patients may qualify for further IVIg therapy:</p> <ul style="list-style-type: none"> under other immunodeficiency criteria (e.g. common variable immunodeficiency [CVID]) depending on the results of subsequent immune 	<ul style="list-style-type: none"> The patient had demonstrated clinical benefit during the review period. <p>AND</p> <ul style="list-style-type: none"> One life-threatening bacterial infection in the previous 12 months <p>OR</p> <ul style="list-style-type: none"> At least two serious infections in the last six months requiring more than standard courses of antibiotics (eg. Hospitalisation, intravenous or prolonged antibiotic therapy). <p>OR</p> <ul style="list-style-type: none"> There is ongoing evidence of impaired antibody production in the context of infection affecting long-term function (such as persistent purulent suppurative otitis media threatening long-term hearing) <p>AND</p> <ul style="list-style-type: none"> A trial cessation of therapy will be undertaken next September or October to undertake an immunological re-evaluation, unless a trial period of cessation of intravenous immunoglobulin (IVIg) for the purpose of immunological evaluation is medically contraindicated on safety grounds (such as a patient with neutropenia, on immunosuppressant medication or with clinically active bronchiectasis and/or 	

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	<p>evaluation; or</p> <ul style="list-style-type: none"> • rarely under specific antibody deficiency following re-emergence of severe significant infection requiring hospitalisation. <p>In principle, IVIg should only be continued or renewed if there is a demonstrated clinical benefit.</p> <p>Note that re-vaccination with pneumococcal polysaccharide vaccine is not recommended because of safety concerns, and the potential for specific hyporesponsiveness induced by repeated vaccination (O'Brien et al 2007).</p>	<p>suppurative lung disease*).</p> <p>*Please note: a diagnosis of bronchiectasis and/or suppurative lung disease must be consistent with the position statement of the Thoracic Society of Australia and New Zealand and the Australian Lung Foundation (Chang et al 2010).</p> <p>Life-threatening infection or a series of significant infections following trial-off Ig therapy in patients with proven specific antibody deficiency.</p> <p>Natural history of specific antibody deficiency remains poorly defined, although antibody production will improve for many patients over time, particularly children.</p> <p>Regular review by a Clinical Immunologist or Respiratory Physician is required, at least annually. Documentation of clinical effectiveness is necessary for continuation of Ig therapy. Cessation of immunoglobulin therapy should be considered, at least after each 12 months of Ig therapy, extended as required to enable cessation of therapy in September or October.</p>	

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	<p><u>IgG subclass deficiency</u></p> <p><u><i>Patients who were receiving IVIg for IgG subclass deficiency before initial publication of the Criteria (December 2007)</i></u></p> <ul style="list-style-type: none"> • With clinically active bronchiectasis or suppurative lung disease over the previous 12 months: To be eligible to receive IVIg for a further 12 months, the following is required: <ol style="list-style-type: none"> 1. Written confirmation from the treating clinical immunologist that : <ul style="list-style-type: none"> • an annual review has been undertaken; • the patient has demonstrated clinical benefit; and • a trial period of cessation of IVIg for the purpose of immunological evaluation is medically 	<p>This should particularly be considered in patients who do not have suppurative lung disease or bronchiectasis. An immunoglobulin washout period of four to six months is necessary to enable an accurate assessment. Prophylactic antibiotics may be considered to cover the period of cessation of immunoglobulin therapy.</p> <p>Patients may qualify for further immunoglobulin therapy:</p> <ul style="list-style-type: none"> • under other immunodeficiency criteria (e.g. primary antibody deficiency disorder), depending on the results of subsequent immune evaluation; or • rarely under specific antibody deficiency following re-emergence of severe significant infection requiring hospitalisation. <p>In principle, immunoglobulin therapy should only be continued or renewed if there is a demonstrated clinical benefit. Note that re-vaccination with unconjugated pneumococcal polysaccharide vaccine is not recommended because of the potential for immune hyporesponsiveness induced by repeated vaccination (O'Brien et al 2007).</p> <p>On review of an authorisation period</p>	

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	<p>contraindicated on safety grounds.</p> <p>AND</p> <p>2. Written confirmation by a second physician that cessation of IVIg for the purpose of immunological evaluation is medically contraindicated on safety grounds.</p> <p>Cessation of IVIg should be considered, at least after each 12 months of therapy extended as required to enable cessation of therapy in September/October.</p> <p>NOTE: The above criteria for initial and ongoing access to IVIg funded by all governments under the National Blood Arrangements will be reviewed in light of emerging evidence at the next review of the Criteria.</p>	<ul style="list-style-type: none"> The patient had demonstrated clinical benefit during the review period. <p>AND</p> <ul style="list-style-type: none"> One life-threatening bacterial infection in the previous 12 months <p>OR</p> <ul style="list-style-type: none"> At least two serious infections in the last six months requiring more than than standard courses of antibiotics (eg. Hospitalisation, intravenous or prolonged antibiotic therapy). <p>OR</p> <ul style="list-style-type: none"> There is ongoing evidence of impaired antibody production in the context of persistent infection affecting long-term function (such as persistent purulent suppurative otitis media threatening long-term hearing) <p>AND</p> <ul style="list-style-type: none"> A trial cessation of therapy will be undertaken next September or October to undertake an immunological re-evaluation <p>OR</p> <ul style="list-style-type: none"> A trial period of cessation of IVIg for the purpose of immunological evaluation is medically contraindicated on safety grounds (such as a patient with neutropenia, on immunosuppressant medication or with clinically active bronchiectasis and/or 	

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		<p>suppurative lung disease*).</p> <p>Antibiotic therapy may be indicated in addition to immunoglobulin therapy.</p> <p>*Please note: a diagnosis of bronchiectasis and/or suppurative lung disease must be consistent with position statement of the Thoracic Society of Australia and New Zealand and the Australian Lung Foundation (Chang et al 2010).</p> <p>Patients currently receiving immunoglobulin therapy for a diagnosis of IgG subclass deficiency are eligible for continued therapy, but a diagnosis of specific antibody deficiency should be confirmed following cessation of immunoglobulin therapy.</p> <p>Natural history of specific antibody deficiency remains poorly defined, although antibody production will improve for many patients over time, particularly children.</p> <p>Regular review by a Clinical Immunologist or Respiratory Physician is required, at least annually. Documentation of clinical effectiveness is necessary for continuation of Ig therapy.</p>	

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		<p>Cessation of immunoglobulin therapy should be considered, at least after each 12 months of therapy, extended as required to enable cessation of therapy in September or October.</p> <p>This should particularly be considered in patients who do not have active suppurative lung disease or bronchiectasis. An immunoglobulin washout period of four to six months is necessary to enable an accurate assessment. Prophylactic antibiotics may be considered to cover the period of cessation of immunoglobulin therapy.</p> <p>In principle, immunoglobulin therapy should only be continued or renewed if there is a demonstrated clinical benefit and ceasing therapy will compromise patient safety.</p> <p>On review of an authorisation period</p> <ul style="list-style-type: none"> • The patient had demonstrated clinical benefit during the review period and has had at least two episodes of infection requiring antibiotics <p>AND</p> <ul style="list-style-type: none"> • Clinically active bronchiectasis and/or suppurative lung disease persists (see note) 	

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		<p>AND</p> <ul style="list-style-type: none"> • A trial cessation of therapy will be undertaken next September or October to undertake an immunological re-evaluation, OR • A trial period of cessation of IVIg for the purpose of immunological evaluation is medically contraindicated on safety grounds (such as a patient with neutropenia, on immunosuppressant medication or with clinically active bronchiectasis and/or suppurative lung disease*). <p>Antibiotic therapy may be indicated in addition to immunoglobulin therapy.</p> <p>*Please note: a diagnosis of bronchiectasis and/or suppurative lung disease must be consistent with position statement of the Thoracic Society of Australia and New Zealand and the Australian Lung Foundation (Chang et al 2010).</p>	
Dose	<p>Maintenance dose: 0.4 g/kg every 4 weeks.</p> <p>Loading dose: not approved.</p>	<p>Maintenance dose - 0.4 g/kg every 4 weeks.</p> <p>Loading dose is not approved.</p> <p>Subcutaneous administration of immunoglobulin</p>	Dosing is unchanged.

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	<p>Subcutaneous administration of immunoglobulins (SCIg) is a suitable alternative to IVIg in this setting.</p> <p>Refer to the current product information sheet for further information.</p> <p>The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient.</p>	<p>(SCIg) is a suitable alternative to IVIg in this setting.</p> <p>The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient.</p> <p>Refer to the current product information sheet for further information.</p>	

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Immunodeficiency Committee of the American Academy of Allergy, Asthma and Immunology', <i>Journal of Allergy and Clinical Immunology</i> , vol. 117, no. 4, pp. S525–53.			
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