Primary immunodeficiency diseases (PID) with antibody deficiency. This excludes: specific antibody deficiency and IgG subclass deficiency.

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Condition for which IVIg has an established therapeutic role.

Specific Conditions	• Common variable immunodeficiency disease (CVID) • Severe combined immunodeficiency (SCID) • Wiskott–Aldrich syndrome • X-linked agammaglobulinaemia • Transient hypogammaglobulinaemia of infancy • Other primary immunodeficiency
Indication for IVIg Use	• Management of infection related to primary antibody deficiency.
Level of Evidence	Evidence of probable benefit – more research needed (Category 2a)
Description and Diagnostic Criteria	PID comprise a group of more than 120 separate conditions. Many of these are manifest by failure of protective antibody production. Key diagnoses include common variable immunodeficiency (CVID), severe combined immunodeficiencies, transient hypogammaglobulinaemia of infancy, Wiskott–Aldrich syndrome and X-linked agammaglobulinaemia. In certain conditions, such as Wiskott–Aldrich syndrome, antibody failure may not be manifest as hypogammaglobulinaemia, but functional antibody responses will be impaired. Some PID does not involve antibody failure, such as chronic granulomatous disease and deficiencies of complement components. In these cases, antibody replacement therapy is not justified.
Justification for Evidence Category	The Biotext (2004) review reported level 2a evidence for the use of intravenous immunoglobulin (IVIg) in the treatment of common variable immunodeficiency and primary hypogammaglobulinaemia.
Diagnosis Requirements	A diagnosis must be made by an Immunologist.
Qualifying Criteria for IVIg Therapy	• Confirmed or suspected diagnosis of primary immunodeficiency. AND • Increased susceptibility to infection. Where a clinical diagnosis has initially been suspected, confirmation will be required for access to continuing Ig therapy.
Exclusion Criteria	Miscellaneous hypogammaglobulinaemia (see secondary hypogammaglobulinaemia). - see Secondary hypogammaglobulinaemia (including iatrogenic immunodeficiency) Specific antibody deficiency. - see Specific antibody deficiency (SAD) IgG subclass deficiency. (See specific antibody deficiency) - see Specific antibody deficiency (SAD)

Review Criteria for Assessing the Effectiveness of IVIg Use	Review by a Clinical Immunologist is required within twelve months and at least annually thereafter, as mandated under the Ig Governance National Policy endorsed by all Australian governments. Documentation of clinical effectiveness is necessary for continuation of Ig therapy. The review criteria for primary immunodeficiency diseases are to ensure adequate replacement of antibody deficiency and to demonstrate clinical benefit from Ig treatment. **On review of an authorisation period** Evidence of clinical effectiveness of Ig therapy, and criteria for continued use may include: Where a diagnosis has initially been suspected, confirmation is required for access to continuing Ig therapy. • Confirmation of diagnosis. AND • Monitoring of trough or random serum IgG levels. AND • Review of any episodes of bacterial infection during the authorisation period.
Dose	• **Loading Dose** - one additional dose of 0.4 g/L in the first month of therapy is permitted if the serum IgG level is <4 g/L. • **Maintenance In chronic suppurative lung disease** - dosing to achieve IgG trough level of up to 9 g/L is permitted if chronic suppurative lung disease is not adequately controlled at an IgG trough level at the lower limit of the age-specific serum IgG reference range. A total dose of up to 1 g/kg may be given over any four week period, which might be by divided doses more frequently than monthly. • **Maintenance Dose** - 0.4 g/kg every four weeks to achieve IgG trough level of at least the lower limit of the age-specific serum IgG reference range. Subcutaneous administration of immunoglobulin can be considered as an alternative to IVIg. A suggested dose is 0.1 g/kg lean body mass every week, modified to achieve an IgG trough level of at least the lower limit of the age-specific serum IgG reference range. **The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient.** **Refer to the current product information sheet for further information.**

Bibliography

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