IMMUNOLOGY

PRIMARY ANTIBODY DEFICIENCIES

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Abstract: *Primary antibody deficiencies are a group of primary immunodeficiency disorders characterized by a marked reduction or absence of serum immunoglobulins due to intrinsic genetic defects in B-cells or impaired interaction between B-cells and T- cells. Clinical symptoms first manifest usually around 6-12 months of life when maternally acquired antibody levels are waning. The sine qua non of antibody deficiency syndromes is recurrent sino-pulmonary infections, especially with encapsulated organisms.Replacement with intravenous immunoglobulin is the mainstay of treatment in primary antibody deficiencies.*

Keywords: *Agammaglobulinemia, Primary immunodeficiency, Recurrent infections, Hypogammaglobulinemia.*

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Points to Remember

- ***Primary B-cell disorders/ primary antibody deficiency disorders (PADs) are the most common primary immunodeficiency disorders (PID) accounting for approximately 50% of all PID cases.***
- ***Recurrent infections with typical microorganisms and predilection for specific organ systems (sino-pulmonary system, gastrointestinal tract and bloodstream infections) are important clinical pointers to suspect PADs.***
- ***Absent tonsils and non-palpable lymph nodes are simple bedside clues to clinch the diagnosis of X-linked Agammaglobulinemia.***
- ***Compliance with regular intravenous immunoglobulin (IVIg) replacement and prophylactic antimicrobial agents remains the standard of care, with proven benefits in both morbidity and mortality.***
- ***Autoimmunity in the setting of underlying PADs (especially common variable immunodeficiency) has a heterogeneous spectrum of clinical manifestations and needs a high index of clinical suspicion to recognise.***
- ***Appropriate disease specific vaccination plan, genetic counselling and attempts for antenatal diagnosis for monogenic defects are crucial.***

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