

IMMUNOLOGY

INNATE IMMUNE DEFECTS

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Abstract: *The innate immune system is a phylogenetically ancient, germline-encoded system that enables eukaryotes to defend themselves against infections. It is “inborn” and does not require a developmental phase and forms the first line of defense against foreign material. It has an immediate or near immediate effect. Innate immune defects can be broadly classified as predisposition to invasive bacterial infections, predisposition to parasitic and fungal infections, Mendelian susceptibility to mycobacterial disease and predominant susceptibility to viral infections. Each defect has a narrow spectrum of infections and knowledge of the specific causative organism in each case helps in early diagnosis and therapeutic decision making.*

Keywords: *Inborn errors of immunity, Innate immunity, Toll-like receptor pathway, Mendelian susceptibility to mycobacterial disease.*

Points to Remember

- *Innate immune defects can be broadly classified as predisposition to invasive bacterial infections, predisposition to parasitic and fungal infections, Mendelian susceptibility to mycobacterial disease and predominant susceptibility to viral infections.*
- *Interleukin 1 Receptor Associated Kinase 4 (IRAK4), Myeloid differentiation factor 88 (MyD88) and toll-interleukin 1 receptor (TIR) domain containing adaptor protein (TIRAP) deficiencies are associated with invasive bacterial infections.*
- *Chronic mucocutaneous candidiasis is caused by defects in the IL-17 pathway, whereas defects in phagocytic defects and CARD9 deficiency cause invasive fungal infections.*
- *Inborn errors in IFN- γ cause MSMD.*
- *Errors in TLR3 signaling pathway and Type-1 Interferons lead to predisposition to viral infections and Type-1 Interferon pathway defects are also associated with severe COVID-19.*

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