Pediatric Immunodefi ciencies			
DISORDER	DESCRIPTION	INFECTION RISK/TYPE	DIAGNOSIS/TREATMENT
B cell			
X-linked agamma-globulinemia (Bruton's)	A B-cell defi ciency in boys only.	Life threatening; encapsulated *Pseudomonas, Streptococcus pneumoniae,* and Haemophilus infections after six months (passive immunity through maternal antibodies wanes).	Quantitative immunoglobulin levels. If low, confi rm diagnosis with B- and T-cell subsets (absent B cells; T cells are often high); absent tonsils and other lymphoid tissue may be a clue. Treat with prophylactic antibiotics and IVIG.
Common variable immunodefi ciency	Immunoglobulin level drops in the 20s and 30s; usually a combined B- and T-cell defect.	↑ pyogenic upper and lower respiratory infections; ↑ risk of lymphoma and autoimmune disease.	Quantitative immunoglobulin levels; confirm with B- and T-cell subsets; treat with IVIG.
IgA defi ciency	Mild; the most common immunodefi ciency.	Usually asymptomatic; patients may develop recurrent respiratory or GI infections. Anaphylactic transfusion reaction due to anti-IgA antibodies is a common presentation.	Quantitative IgA levels; treat infections. Do not give immunoglobulins (can lead to the production of anti-IgA antibodies).
T cell			
Thymic aplasia	See mnemonic.	Variable risk of infection.	Absolute lymphocyte count;

(DiGeorge	Presents with tetany (2° to	↑↑↑ infections with fungi	mitogen stimulation
syndrome)	hypocalcemia) in the fi rst	and Pneumocystis jiroveci	response; delayed
	days of life.	pneumonia (formerly	hypersensitivity skin testing.
		P. carinii).	Treat with bone marrow
			transplantation and IVIG
			for antibody defi ciency;
			PCP prophylaxis. Thymus
			transplantation is an
			alternative.
Combined			
Ataxia-telangiectasia	Oculocutaneous	↑ incidence of non-Hodgkin's	No specific treatment; may
	telangiectasias and	lymphoma, leukemia, and	require IVIG depending
	progressive cerebellar	gastric carcinoma.	on the severity of the Ig
	ataxia. Caused by a DNA		defi ciency.
	repair defect.		
Severe combined	Severe lack of B and T cells.	Severe, frequent bacterial	Treat with bone marrow
immunodefi ciency		infections; chronic	transplant or stem cell
(SCID)		candidiasis; and	transplant and IVIG for
		opportunistic organisms.	antibody defi ciency. Needs
			PCP prophylaxis.