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X-Linked Hyper-IgM Syndrome

National Cancer Institute

Source

National Cancer Institute. *X-Linked Hyper-IgM Syndrome*. NCI Thesaurus. Code C158531.

An X-linked subtype of hyper-IgM caused by mutation(s) in the CD40LG gene, encoding CD40 ligand. It is characterized by normal or elevated IgM concentrations, with markedly decreased concentrations of other immunoglobulins. The clinical course is variable; it manifests as susceptibility to bacterial and opportunistic infections, neutropenia, and increased risk of lymphoma.