

Abt Letterer Siwe Disease

[Langerhans cell histiocytosis](#) (LCH), previously referred to as [histiocytosis X](#), is a dendritic cell [histiocytic tumor](#) that demonstrates a variable spectrum of organ involvement.

Clinical syndromes within this entity include eosinophilic granuloma, Hand-Schuller-Christian disease, Abt-Letterer-Siwe Disease, and Hashimoto-Pritzker disease. Currently, it is classified on the basis of extent, such as unifocal, multifocal, or disseminated disease. LCH typically occurs in childhood and adolescence as solitary osteolytic lesions. When involving the central nervous system, it is usually either a result of extra-axial extension from skull vault epicenters, or is restricted to the hypothalamic-pituitary axis. Discrete intraparenchymal, intra-axial CNS lesions are rare.

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