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# Alpha Heavy Chain Disease

National Cancer Institute

## Source

National Cancer Institute. *Alpha Heavy Chain Disease*. NCI Thesaurus. Code C3132.

A clonal disorder, also known as immunoproliferative small intestinal disease or Mediterranean lymphoma, characterised by the secretion of a defective alpha heavy chain. It predominantly affects young people in the Mediterranean region. It involves the small intestine, and patients usually present with malabsorption syndrome, abdominal pain, weight loss, and fever. There is extensive villous atrophy of the small intestinal mucosa, which is heavily infiltrated by small lymphocytes and plasma cells. The small intestinal morphologic changes are consistent with a mucosa-associated lymphoid tissue lymphoma (MALT lymphoma).