CASE REPORT

BASSEN - KORNZWEIG SYNDROME

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Abstract: *Abetalipoproteinemia is a rare, autosomal recessive disease characterized by steatorrhea, acanthocytosis and hypolipidemia in infancy with a frequency <1 in 100,000. Deficiency of fat-soluble vitamins, atypical retinitis pigmentosa, coagulopathy, posterior column neuropathy and myopathy may develop by late childhood. Low fat diet and fat-soluble vitamins especially mega doses of Vitamin E are the main stays of therapy.*

Keywords: *Abetalipoproteinemia, Acanthocytes, Steatorrhea, Retinitis pigmentosa, MTTP gene*

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