Lynch 1 Syndrome

National Cancer Institute

Source

National Cancer Institute. Lynch 1 Syndrome. NCI Thesaurus. Code C6725.

A rare genetic neoplastic syndrome with an autosomal dominant pattern of inheritance but incomplete penetrance. It is associated with a greater than 70 % risk of developing colorectal carcinoma. It is caused by a mutation in one of the mismatch repair genes: MSH2, MLH1, MSH6 or PMS2. It usually manifests at age 50 or younger with multiple synchronous or metachronous colorectal carcinomas. Clinical course is rapidly progressive. Prognosis is variable with a high risk for the development of additional colorectal carcinomas. However, survival is significantly better than non-HNPCC carcinomas of equivalent stage.