Cleveland Clinic
Genomic Medicine Institute

Lynch Syndrome

Lynch syndrome, also known as Hereditary Non-polyposis Colorectal Cancer (HNPCC), is one of the most common hereditary cancer syndromes, affecting as many as 1 in 400 individuals. Individuals with Lynch syndrome have a significantly increased risk of developing colorectal cancer. This risk may range from approximately 40-80% by the age of 70 years. This is much higher than the 5.5% lifetime risk of developing colorectal cancer in the general population. Colorectal cancer in Lynch syndrome is usually diagnosed at an earlier age (average age: 45 years) than sporadic colon cancer. An individual with Lynch syndrome who has had one colon cancer has an increased risk for developing a second colon cancer. This risk is 16% within ten years after the original surgery, 41% within 20 years, and 62% after 30 years.

Endometrial (uterine) cancer is the most common type of cancer reported in women with Lynch syndrome. The risk to develop endometrial cancer by age 70 is approximately 20-60% in women with Lynch syndrome which is much higher than the 2-3% risk in the general population. Women with Lynch syndrome also have up to a 20% lifetime risk of developing ovarian cancer (general population risk is 1-2%).

Stomach cancer is the third most common type of cancer associated with Lynch syndrome. Individuals with Lynch syndrome have up to a 10% risk of developing stomach cancer which is increased over the general population risk of less than 1%. Individuals with Lynch syndrome also have slightly increased risks for developing cancers of the small intestine (1-2% lifetime risk), hepatobiliary tract (1-2% lifetime risk), brain (1-8% lifetime risk), and transitional cell carcinoma of the renal pelvis (kidney), bladder, or ureter (1-12% lifetime risk).

Muir-Torre syndrome is a form of Lynch syndrome. Individuals with this condition have sebaceous neoplasms (growths) of the skin in addition to the cancers commonly associated with Lynch syndrome. The variety of skin growths associated with Muir-Torre include: sebaceous adenomas, sebaceous cysts, sebaceous carcinomas, and keratoacanthomas.

Turcot syndrome is another form of Lynch syndrome. Individuals with this condition may have brain tumors in addition to Lynch syndrome-related. Most often glioblastomas are the type of brain tumor associated with Turcot, although a variety of other tumors have been reported. Recent research suggests that Turcot syndrome is caused when a mutation is present in both copies of one of the Lynch syndrome genes (one inherited from the mother and one inherited from the father).

The Center for Personalized Genetic Healthcare