UT Southwestern
Harold C. Simmons
Comprehensive Cancer Center

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What you should know about Familial Adenomatous Polyposis (FAP)

FAP is a very rare condition that accounts for about 1% of new cases of colorectal cancer. People with FAP typically develop hundreds to thousands of polyps (adenomas) in their colon and rectum by age 30-40. Polyps may also develop in the stomach and small intestine. Individuals with FAP can develop non-cancerous cysts on the skin (epidermoid cysts), especially on the scalp. Besides having an increased risk for colon polyps and cysts, individuals with FAP are also more likely to develop sebaceous cysts, osetomas (benign bone tumors) of the jaw, impacted teeth, extra teeth, CHRPE (multiple areas of pigmentation in the retina in the eye) and desmoid disease. Some individuals have milder form of FAP, called attenuated FAP (AFAP), and develop an average of 20 polyps at a later age.

The risk for cancer associated with FAP

If left untreated, the polyps in the colon and rectum will develop in to cancer, usually before age 50. Individuals with FAP also have an increased risk for stomach cancer, papillary thyroid cancer, periampullary carcinoma, hepatoblastoma (in childhood), and brain tumors.

The risks to family members

FAP is caused by mutations in the Adenomatous Polyposis Coli (APC) gene. Approximately 1/3 of people with FAP do not have family history of the disease, and thus have a new mutation. FAP is inherited in a dominant fashion. Children of a person with an APC mutation have a 50% risk to inherit the mutation. Brothers, sisters, and parents of individuals with FAP should also be checked to see if they have an APC mutation. Almost everyone who has an APC mutation will develop FAP.

Managing the Risk

Non-steroidal anti-inflammatory drugs (NSAIDS) have been shown to reduce the number and progression of adenomas. Once polyposis or cancer is detected, it is recommended that the colon be removed. After colon surgery, annual surveillance of the rectum and thyroid is recommended.at age 30-35 can be considered (depending on family history and race)

Children under the age of 5 with an APC mutation should be screened for a rare liver tumor (hepatoblastoma). Screening is usually done by measuring blood AFP levels and/or abdominal imaging studies.

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