UT Southwestern
Harold C. Simmons
Comprehensive Cancer Center

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## What you should know about MYH-associated polyposis (MAP) syndrome

MAP syndrome is form of inherited colorectal cancer. Individuals with MAP syndrome develop multiple polyps (adenomas) in the colon. Individuals with MAP syndrome usually have between 15 and 100 polyps, but MAP syndrome can occur in individuals with less than 15 polyps or greater than 100 polyps. Aside from colon polyps, individuals with MAP syndrome can develop tumors in the upper gastrointestinal system, CHRPE (multiple areas of pigmentation in the retina of the eye), osteomas (benign bone tumors) of the jaw, impacted teeth, extra teeth, and benign tumors of the hair follicle.

## The risk for cancer associated with MAP

If left untreated, the polyps in the colon will develop in to cancer. Individuals with MAP syndrome caused by two mutations in the MYH gene have up to an 80% risk to develop colon cancer in their lifetime and a 5% risk to develop cancer in the duodenum. Individuals who have one mutation in the MYH gene also seem to have an increased risk for colon cancer, although the risk is not known.

## The risks to family members

MAP syndrome is caused by mutations in the MYH gene. MAP syndrome is inherited in an autosomal recessive fashion, meaning that a person must inherit a mutation in the MYH gene from both of their parents to have MAP syndrome. Brothers and sisters of a person with MAP have a 25% (1 in 4) risk to inherit MAP syndrome, a 50% (1 in 2) risk to have one MYH mutation, and a 25% (1 in 4) chance that they will not have a MYH mutation. Approximately 1-2% of the population has a MYH mutation, so it is important to know that individuals with one MYH mutation have an increased risk for having a child with MAP syndrome, and their spouse should be offered testing to see if they also have a MYH mutation.

## Managing the Risk

Non-steroidal anti-inflammatory drugs (NSAIDS) have been shown to reduce the number and progression of adenomas. Colonoscopy every 2-3 years, beginning at age 25-30, and upper endoscopy every 3-5 years, beginning at age 30-35 years are recommended for individuals with MAP syndrome. Once severe polyposis or cancer is detected, it is recommended that the colon be removed. After colon surgery, annual surveillance of the rectum is recommended.

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