MYH-Associated Polyposis [1]

What is MYH-associated polyposis?

MYH-associated polyposis (MAP) is a hereditary condition. People with MAP tend to develop multiple adenomatous colon polyps during their lifetime and will have an increased risk of colorectal cancer [2] if they are not monitored closely with colonoscopies [3]. An adenomatous polyp is an area where the normal cells that line the inside of the colon begin to form a mass. At first, a polyp is benign, meaning it is noncancerous and will not spread. However, some polyps can eventually turn malignant, meaning cancerous, and cancers can spread to other parts of the body. It is likely that people with MAP will develop multiple polyps, and therefore their risk for colorectal cancer may be increased if these polyps cannot be removed.

In some people, MAP is associated with developing 100s of polyps, and appears to be similar to other hereditary conditions of familial adenomatous polyposis [4] (FAP) and attenuated familial adenomatous polyposis [5] (AFAP). In other cases, MAP can be diagnosed with fewer polyps (less than 20) and/or colorectal cancer at a young age.

Some people with MAP have an increased risk of developing polyps in the upper gastrointestinal tract, such as the stomach and small intestine. The risk of thyroid cancer [6] may also be increased in individuals with MAP.

What causes MAP?

MAP is a genetic condition. This means that the risk of colon polyps and colorectal cancer can be passed from generation to generation in a family. A mutation is an alteration in a gene. Mutations in the *MYH* gene, also known as the *MUTYH* gene, are known to cause MAP.

How is MAP inherited?

Normally, every cell has 2 copies of each gene: 1 inherited from the mother and 1 inherited from the father. MAP follows an autosomal recessive inheritance pattern, in which a mutation needs to be present in both copies of the *MYH* gene in order for a person to have an increased risk of getting that disease. This means that each parent must pass on a gene mutation for a child to be affected. A person who has only 1 copy of the gene mutation is called a carrier. When both parents are carriers of a recessive gene mutation, there is a 25% chance that a child will inherit 2 mutations and be affected. Individuals who carry only 1 copy of a *MYH* gene mutation do not develop MAP.

How common is MAP?

Most colorectal cancer is sporadic, meaning it occurs by chance with no known cause. The percentage of colorectal cancer that can be attributed to MAP is unknown. It is estimated that as many as 1 in every 100 people may carry a single mutation in the *MYH* gene.

How is MAP diagnosed?

MAP is considered as a possible diagnosis when a person has multiple adenomatous colon polyps but does not have a mutation in the *APC* gene associated with FAP and AFAP. It may also be considered if someone has brothers or sisters with multiple colon polyps, but there is no history of colon problems in previous generations. MAP is diagnosed when a person is found to have 2 mutations in the *MYH* gene. Although most individuals with MAP carry at least 1 of the 2 most common mutations in *MYH* (*Y165C* and *G382D),* there are additional *MYH* mutations that can be detected by complete gene panel testing.

What are the estimated cancer risks associated with MAP?

In addition to the risk of colorectal cancer, the specific cancer risks associated with MAP have not been determined. The risk of colorectal cancer [2] is considered to be significantly increased, and there may be an increased risk of other cancers of the gastrointestinal tract and thyroid gland as well.

What are the screening options for MAP?

ASCO recommends the following screening for people with MAP. It is important to discuss these options with your doctor, as each individual is different:

- Colonoscopy [3] every 1 to 2 years, beginning at age 18 to 20.
- Yearly colonoscopy once a person develops polyps, with the goal of removing all large polyps

- If an individual with MAP develops colorectal cancer or if colon polyps are too numerous to be moved during a colonoscopy, surgery may be considered. Colectomy is the surgical removal of part or all of the colon. This may be considered if polyps cannot be managed with regular colonoscopies because there are too many.

- Upper endoscopy [7] (esophagogastroduodenoscopy or EGD) at age 25 to 30 or once colorectal polyps are detected, whichever occurs first

- Ultrasound [8] of the thyroid may be considered, starting at age 25 to 30

Screening options may change over time as new technologies are developed and more is learned about MAP. It is important to talk with your doctor about appropriate screening tests.

Learn more about what to expect when having common tests, procedures, and scans [9], and read more about these screening recommendations at www.asco.org/endorsements/HereditaryCRC [10].

Questions to ask the doctor

If you are concerned about your risk of developing cancer, talk with your doctor. Consider asking the following questions of your doctor:

- What is my risk of colorectal cancer [2]?

- How many colon polyps have I had in total?

- What type of colon polyps have I had? The two most common types are hyperplastic and adenomatous.

- What can I do to reduce my risk of cancer?

- What are my options for cancer screening?

If you are concerned about your family history and think your family may have MAP, consider asking the following questions:

- Does my family history increase my risk of colorectal cancer?

- Should I meet with a genetic counselor?

- Should I consider genetic testing [11]?

More Information

The Genetics of Cancer [12]

Genetic Testing [11]

What to Expect When You Meet With a Genetic Counselor [13]

Collecting Your Family Cancer History [14]

Sharing Genetic Test Results with Your Family [15]

Additional Resources

Colon Cancer Alliance
www.ccalliance.org [16]

Colorectal Cancer Coalition (C3)
www.fightcolorectalcancer.org [17]

To find a genetic counselor in your area, ask your doctor or visit the following websites:

National Society of Genetic Counselors
www.nsgc.org [18]

National Cancer Institute: Cancer Genetics Services Directory
www.cancer.gov/cancertopics/ [19]

Links
[1] http://www.cancer.net/cancer-types/myh-associated-polyposis
[2] http://www.cancer.net/node/31317
[3] http://www.cancer.net/node/24481
[4] http://www.cancer.net/node/18852
[5] http://www.cancer.net/node/18503
[6] http://www.cancer.net/node/31262
[7] http://www.cancer.net/node/24731
[8] http://www.cancer.net/node/24714
[9] http://www.cancer.net/node/24959
[10] http://www.instituteforquality.org/hereditary-colorectal-cancer-syndromes-american-society-clinical-oncology-clinica

l-practice
[11] http://www.cancer.net/node/24895
[12] http://www.cancer.net/node/24897
[13] http://www.cancer.net/node/24907
[14] http://www.cancer.net/node/30761
[15] http://www.cancer.net/node/24906
[16] http://www.ccalliance.org/
[17] http://www.fightcolorectalcancer.org/
[18] http://www.nsgc.org/
[19] http://www.cancer.gov/cancertopics/genetics/directory