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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]. An adenomatous polyp is an area where the normal cells that line the inside of the colon begin to make mucous and form a mass. A polyp is benign, meaning it is noncancerous and will not spread. However, a polyp can eventually turn malignant, meaning cancerous, and can spread to other parts of the body. It is also likely that people with MAP will develop polyps and colorectal cancer at a relatively young age, in their 20s to 50s.

MAP has only recently been described by medical researchers, and there is much to be learned about the condition. MAP appears to be similar to other hereditary conditions of familial adenomatous polyposis [3] (FAP) and attenuated familial adenomatous polyposis [4] (AFAP). It is currently unknown if people with MAP have an increased risk of polyps or cancer in other parts of the digestive tract, such as the stomach or small intestine.

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. Mutations, meaning alterations, in the *MYH* gene are known to cause MAP.

How is MAP inherited?

Normally, every cell has two copies of each gene: one inherited from the mother and one 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 one 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 two mutations and be affected.

Options exist for couples interested in having a child when they know that one of them carries a gene mutation that increases the risk for this hereditary cancer syndrome. Preimplantation genetic diagnosis (PGD) is a medical procedure done in conjunction with in-vitro fertilization

(IVF). It allows people who carry a specific known genetic mutation to have children who do not carry the mutation. A woman?s eggs are removed and fertilized in a laboratory. When the embryos reach a certain size, one cell is removed and is tested for the hereditary condition in question. The parents can then choose to transfer embryos which do not have the mutation. PGD has been in use for over a decade, and more recently has been used for several hereditary cancer predisposition syndromes. However, this is a complex procedure with financial, physical, and emotional factors for couples to consider before starting. For more information, talk with an assisted reproduction specialist at a fertility clinic.

How common is MAP?

Most colorectal cancer [2] 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 one 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 [3] and AFAP [4]. 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 two mutations in the *MYH* gene. There are two common mutations in *MYH* called *Y165C* and *G382D*.

What are the estimated cancer risks associated with MAP?

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 digestive tract 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 [5] every one to two years, beginning at age 18 to 20.
- Yearly colonoscopy once a person develops polyps. Colectomy is the surgical removal of the entire colon. This may be considered if polyps cannot be managed with regular colonoscopies because there are too many.
- Upper endoscopy [6] (EGD) at age 25 to 30 or once colorectal polyps are detected, whichever occurs first
- Yearly ultrasound of the thyroid may be considered starting at age 25 to 30
- Computed tomography (CT) scan or magnetic resonance imaging (MRI) if a person has a family history of desmoid tumors and a mutation on the *APC* gene that is linked with these tumors

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 [7], and read more about these screening recommendations at www.asco.org/endorsements/HereditaryCRC [8].

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 [9]?

More Information

The Genetics of Cancer [10]

Genetic Testing [9]

What to Expect When You Meet With a Genetic Counselor [11]

Collecting Your Family Cancer History [12]

Sharing Genetic Test Results with Your Family [13]

Additional Resources

Colon Cancer Alliance
www.ccalliance.org [14]

Colorectal Cancer Coalition (C3)
www.fightcolorectalcancer.org [15]

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

National Society of Genetic Counselors
www.nsgc.org

[16]

National Cancer Institute: Cancer Genetics Services Directory
www.cancer.gov/cancertopics/ [17]

Links:
[1] http://www.cancer.net/cancer-types/myh-associated-polyposis
[2] http://www.cancer.net/node/31317
[3] http://www.cancer.net/node/18852
[4] http://www.cancer.net/node/18503
[5] http://www.cancer.net/node/24481
[6] http://www.cancer.net/node/24731
[7] http://www.cancer.net/node/24959
[8] http://www.asco.org/endorsements/HereditaryCRC
[9] http://www.cancer.net/node/24895
[10] http://www.cancer.net/node/24897
[11] http://www.cancer.net/node/24907
[12] http://www.cancer.net/node/30761
[13] http://www.cancer.net/node/24906
[14] http://www.ccalliance.org/
[15] http://www.fightcolorectalcancer.org/
[16] http://www.nsgc.org/
[17] http://www.cancer.gov/cancertopics/genetics/directory