

MYH Associated Polyposis Syndrome (MAP)

Key Points

- MAP is a rare recessively inherited condition associated with abnormalities in the MYH gene. A recessive condition means that both inherited copies of the MYH gene (one copy is inherited from each parent) has an abnormality or mutation. The mutation in both MYH genes means that the MYH gene cannot do its job properly.
- MAP is characterised by the finding of multiple bowel polyps or outgrowths (adenomas) on the lining of the large bowel.
- **Bowel cancer** develops from adenomatous bowel polyps over a period of time .
- Adenomas in the general population do not usually occur until the age 40 – 50 years and then only in small numbers.
- People with MAP generally develop greater than 20 adenomatous bowel polyps at a younger age and the majority of people with MAP will develop bowel cancer by age 60 years.
- Surgery to remove the majority of the large bowel prevents individuals with MAP from developing bowel cancer. This surgery is normally recommended before 60 years of age.
- Yearly bowel screening after surgery is required for the part of the large bowel that remains to help prevent cancers developing.
- Siblings (brothers and sisters) of people with MAP have a 1 in 4 risk of inheriting two abnormal copies of the MYH gene. If you have two abnormal copies of the MYH gene you will have MAP. Genetic testing is recommended for siblings of people with MAP.
- Children of people with MAP will inevitably each carry one abnormal copy of the MYH gene. However, carrying one abnormal copy of the MYH gene does not clearly increase a person's risk of developing bowel cancer. Therefore, genetic testing and bowel screening by colonoscopy are not routinely recommended or available for children of people with MAP.
- The aim of the NZFGICS is to reduce the number of cancers occurring in families by facilitating the required bowel screening for registered families.

What is MAP?

MAP is an inherited condition and patients with MAP have an increased risk of developing polyps in their colon, rectum and duodenum. If the polyps are not removed there is a risk of developing cancer as bowel cancer develops from adenomatous bowel polyps over a period of time.

What is the risk of developing bowel cancer in MAP?

The majority of individuals with MAP will develop bowel cancer by 60 years of age.

What are the symptoms of MAP?

There are usually no symptoms to alert you that you have bowel polyps but sometimes you may develop rectal bleeding or persistent abdominal pain. The major concern is that the polyps will become cancerous.

What causes MAP?

In MAP, both copies of a gene called MYH are abnormal. Abnormalities in the MYH gene affect the ability of cells to correct mistakes when cells divide, allowing multiple polyps to develop.

How is MAP inherited?

It is a recessively inherited condition which means that you need to inherit one abnormal copy from your mother and one abnormal copy from your father. When you have MAP each of your brothers and sisters also has a 1 in 4 (or 25%) chance of inheriting MAP – this means that there is a 25% chance of inheriting 2 normal copies and a 50% chance of inheriting one altered copy and one normal copy. People who inherit only one abnormal copy are called carriers and do not develop the multiple polyps associated with MAP syndrome. People with no abnormal copies are also unaffected.

What does the genetic test result mean for my family?

The gene involved in MAP (called MYH) is like a very long instruction sentence. A spelling mistake or alteration in any part of this instruction can cause MAP. Each family with MAP will have a different alteration but the individuals within the family will have the same alteration which is called a mutation. We can look in a blood sample from someone who has MAP to try to find the mutation in a family. In a small number of people with MAP genetic testing may not be able to detect a mutation or abnormality within the MYH gene.

If we do find a genetic mutation we can test other family members to see if they have inherited it. Testing is not offered to children of those with MAP who will be carriers as there is currently no clear evidence of an increased risk in bowel cancer and therefore no recommended surveillance. However, if they develop bowel symptoms then like anybody else they need to see their GP.

If an individual has inherited MAP it is important that they have bowel checks every year starting from the age of 18- 20 years.

What if no spelling mistake is found?

If a gene alteration is not found we won't be able to offer a genetic test to other relatives but they will still need regular colonoscopies. We will discuss the best screening plan for your family with you.

Do I need other checks?

A small proportion of patients with MAP may have polyps (adenomas) in the first part of the small bowel (duodenum) and may require regular tube test examinations of the stomach and first part of the small bowel , that is gastroscopy.

What is the treatment?

Once large bowel polyps are found individuals with MAP are normally advised to have surgery to remove the large bowel. In some situations regular colonoscopy with removal of the polyps is possible and appropriate.

There are 2 main types of operation:

- Colectomy and ileo-rectal anastomosis (IRA) involves removing the large bowel and attaching the small bowel to the rectum or bottom part of the large bowel. The remaining rectum can still develop polyps so this must be examined every twelve months to identify and remove these polyps in order to prevent a cancer starting there. An individual who has had a colectomy and IRA may need further surgery if an unmanageable number of rectal polyps develop.
- Sometimes at the age when the initial surgery is being planned more extensive polyps have already developed in the rectum and colon and therefore the planned surgery involves removing the whole rectum and colon. This is called a pan procto-colectomy. This surgery involves removing the remaining bottom part of the bowel and creating a new "rectum" as described below.
- A new "rectum" or ileo anal pouch is created. The small bowel is used to fashion a new rectum called a pouch and connected to the anus. This mean you can pass a motion normally through your anus. Following pouch surgery regular endoscope checks for polyps are still required.
- A pan procto-colectomy and ileo anal pouch may, in certain circumstances, be planned as the initial surgery.
- An ileostomy is when the large bowel is removed and the end of the small bowel is brought to the surface and the waste material is collected in a disposable bag. Quite often after pouch surgery you do need an ileostomy for a short time only while the pouch heals. Your surgeon is best able to advise you.

Does diet matter?

At the moment there is no known effect of diet on those with MAP. We recommend a healthy diet and lifestyle including maintaining a healthy weight. Healthy foods include fruit, vegetables, cereals, bread (whole meal), pasta, rice, olive oil and low-fat dairy food such as yoghurt. Possible harmful foods are excessive amounts of animal fat, processed meats (ham, bacon, sausage, hot dogs etc.), large quantities of red meat and alcohol . The World Health Organization recommends that the general population eat less than 50grams of processed meat and less than 100grams of red meat daily to decrease the risk of bowel cancer. There is also convincing evidence that drinking alcohol increases the risk of some cancers including bowel cancer. The recommendation is to avoid binge drinking, stick to less than 2 standard drinks a day, and have at least two alcohol free days a week. We would also recommend you not smoke.

For more information visit <https://www.healthed.govt.nz/resource/eating-healthy-adult-new-zealandersng%C4%81-kai-t%C5%8Dtika-ma-te-hunga-pakeke-o-aotearoa>

Is there medication available?

There is currently no conclusive evidence that medication is helpful in preventing the progression of polyp growth in MAP.

In the general population there is some evidence that taking daily low dose aspirin can decrease the risk of developing bowel cancer. Long-term aspirin therapy does have potential risks in some people and any decision to take Aspirin long-term should be done in conjunction with your family doctor.

