New Zealand Familial GI Cancer Service

Updated: Jan 2013

# Lynch Syndrome

## Key Points

- **Lynch syndrome is a rare, inherited condition that increases your risk of developing bowel cancer and sometimes other cancers.**
- **Lynch Syndrome accounts for about 3% of all bowel cancer.**
- **Regular bowel screening by colonoscopy can prevent bowel cancers developing.**
- **Bowel Screening usually starts from the age of 25 years and is performed on a yearly basis.**
- **People with Lynch Syndrome are also at increased risk of developing certain other cancers such as uterine (womb), urinary tract, ovary and small bowel.**
- **Women should consider screening of the uterus and ovaries from the age of 30-35 years.**
- **Genetic testing is available for families with this syndrome.**
- **When you have Lynch Syndrome, each of your children have a 1 in 2 (50%) chance of inheriting it.**
- **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 Lynch Syndrome?

Lynch syndrome is a rare inherited condition that increases your risk of bowel cancer and other cancers. Lynch syndrome has historically been known as Hereditary Non Polyposis Colorectal Cancer (HNPCC). A number of inherited syndromes can increase your risk of bowel cancer, but Lynch syndrome is the most common.

### What is the risk of developing bowel cancer in Lynch Syndrome?

The average New Zealander has about a 6% chance of developing bowel cancer by the age of 75 years but in Lynch Syndrome there is up to a 50% chance of developing bowel cancer by the age of 70 years. Most bowel cancer normally occurs after the age of 50 years but in Lynch Syndrome it occurs earlier.

### What is the risk of other cancers in Lynch Syndrome?

In some families with Lynch syndrome there are other cancers, most commonly uterine or womb cancer, as well as bowel cancer.

The average New Zealand woman has less than a 2% chance of developing uterine or womb cancer but in Lynch Syndrome this can be 30% or greater.

There is also an increased risk of ovarian cancer, 3-13% by the age of 70 years compared to a risk of less than 2% for the average New Zealand women.

An increased risk of cancer at other sites namely, urinary tract, small intestine, pancreas, brain and particular types of skin cancers has also been found but by the age of 70 years the risk of developing one of these cancers is estimated to be 10% or less.

### Is the risk of cancer the same for all families with Lynch Syndrome?

It is important to remember that not all families have all cancers and that not all people develop every cancer that can occur. In recent years it has been recognised that the risk of developing both bowel and other cancers varies between families. This mostly relates to the type of genetic spelling mistake in each family. Some people with Lynch Syndrome will not develop cancer although they can still pass the gene mutation on to their children.

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**What causes Lynch Syndrome?**

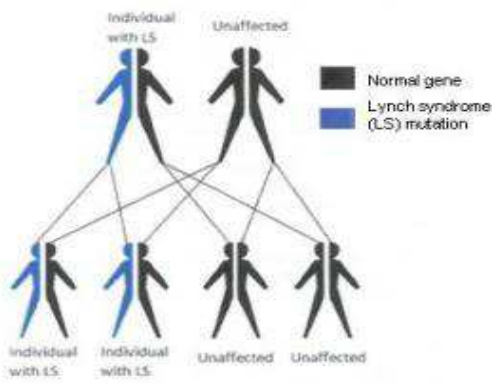
Lynch Syndrome is caused by an abnormality in one of four mismatch repair genes (MLH1, MSH2, MSH6, and PMS2). These are the genes responsible for correcting mistakes that occur in genes when body cells divide.

More recently an error in a gene called EPCAM has been identified and this also stops the MSH2 gene working properly meaning it can't fix up those "spelling" mistakes.

Nearly every cell in our bodies contains two copies of each gene and genes are the "instruction manuals" for building and running the body. DNA is the genetic material within each cell that contains instructions for every chemical process in the cells of the body. As cells grow and divide they make copies of their DNA and it is common for minor mistakes to occur.

Normally the mismatch repair genes recognize these mistakes and repair them, similar to the "spell check" function on your computer. However, people who inherit a fault in one of the four mismatch repair genes lack the ability to repair these minor mistakes. An accumulation of these mistakes may eventually lead to the development of a cancer.

**How is Lynch Syndrome inherited?**




Lynch Syndrome is a condition that is passed from parent to child. Occasionally it may occur "out of the blue with no family history.

Each child of a parent with Lynch Syndrome has a 50:50 chance of inheriting the genetic spelling mistake and each of those person's children then has the 50:50 chance of inheriting it.

This is called an autosomal dominant inheritance pattern.

**Do people who have NOT inherited the spelling mistake need surveillance for cancer?**

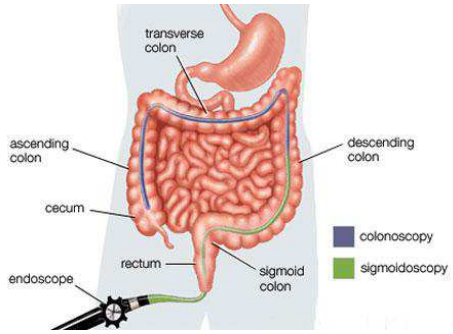
Children who do not inherit the gene cannot pass it on to their children. They have the same risk as anyone else for developing cancer and therefore do not need surveillance for the cancers linked with Lynch syndrome. They should have the screening tests normally recommended by their General Practitioners.

**Is the risk of inheriting the condition different for men and women?**

The risk of inheriting Lynch syndrome is the same whether the gene mutation carrier is the mother or father or the child is a son or daughter.

**What bowel checks are advised for individuals with Lynch syndrome?**

Most bowel cancer starts in polyps (called adenomas). In Lynch Syndrome as the DNA "spell check" isn't working and the time taken for a polyp to develop into a cancer is sped up from the 5- 10 years it takes for bowel cancer to develop in people with no family history of bowel cancer. This is the reason why we currently recommend yearly colonoscopy for people with Lynch Syndrome.




A colonoscopy is a procedure which examines the large bowel. If any polyps are detected they can be removed. Colonoscopies can also detect bowel cancer at an early stage, when they are more treatable.

You need a colonoscopy every year, usually starting from the age of 25 years.

If a relative has had a bowel cancer below the age of 35 years, we may begin screening in your family a bit earlier.

**Do I need other checks?**

For women we usually advise referral to a gynaecologist to discuss checks that can be performed to look for uterine or ovarian cancer.

Screening for cancer of the uterus and ovarian cancer can begin around the age of 30-35 years and is required every 1-2 years.

Uterine screening involves an ultrasound scan to measure the thickness of the lining of the womb. It also involves taking samples of the lining of the womb (pipelle).

Ovarian screening is usually done by an ultrasound scan.

Both of these procedures can be done as an outpatient procedure.

There is no evidence to support the effectiveness of this screening. The screening for both uterine and ovarian cancer is less reliable than bowel screening.

Some women may choose to have a hysterectomy (removal of the womb) and oophorectomy (removal of the ovaries) rather than screening if they have completed their families. We do not currently recommend this.

Check for the less common cancers in Lynch Syndrome may be recommended and we will need to discuss this with you.

**What about testing for my children?**

When you have Lynch syndrome, each of your children have a 1 in 2 (or 50%) chance of inheriting the Lynch syndrome gene.

Children can be tested for Lynch syndrome once they reach the age of 18 years.

**What about Genetic testing for family members?**

If we have found a spelling mistake in one of the mismatch repair genes that appears to be responsible for most of the cancer in your family we will then let other family members know they can have a genetic test to see if they too have inherited the gene alteration.

They need to understand the advantages and disadvantages of testing. It is a very personal decision.
Some people decide to have a genetic test so they can decide about their screening and to find out if there is a risk for their children.

Bowel screening by colonoscopy is not usually recommended in people under the age of 25y and therefore many people delay having a genetic test until this age.

People, who decide not to have genetic testing, should have regular bowel checks every 2 years.

**Does diet matter?**

At the moment we do not really know the effect diet can have on the development of cancers in Lynch Syndrome. However, we recommend that you have a healthy diet and lifestyle. We also recommend that you maintain your body weight within the healthy range recommended for your height. See the information on 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 some evidence that aspirin may reduce the chance of bowel cancer if taken regularly. This is something you may wish to consider but you would need to talk this over with your General Practitioner who knows your medical history and can help you decide if this is appropriate for you.