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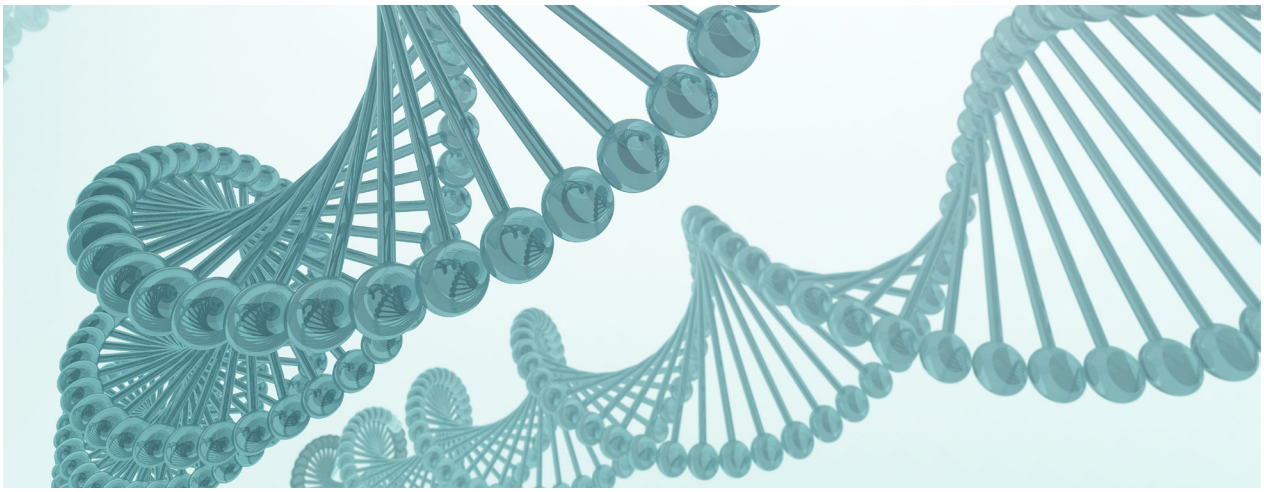
Understanding and Managing Lynch Syndrome



For people with Lynch syndrome and their family members

Read this pamphlet to learn:

- Which cancers are linked to Lynch syndrome
- What causes Lynch syndrome and who is at risk
- How to screen (test) for cancer if you have Lynch syndrome
- Symptoms (signs) of gynecologic cancers
- What supports you can get

Patient Education

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Is cancer hereditary

Everyone has a chance of getting cancer in their lifetime. As you get older, the cells in and around your organs can be damaged by:

- things in your environment (like diet, lifestyle)
- the normal aging process

Over time, this damage adds up and can lead to cancer. This may explain most cases of cancer, which we call sporadic (happens by chance).

A smaller number of cancers are caused by hereditary factors (passed down from parent to child), which can increase your risk of getting cancer. Lynch syndrome is a common hereditary condition.

Which cancers are related to Lynch syndrome?

The most common cancers related to Lynch syndrome are:

- **Colorectal cancer (CRC).** This is a cancer that starts in either the colon or the rectum. People with Lynch syndrome have a higher risk of getting colorectal cancer than people who do not have Lynch syndrome.
- **Endometrial cancer (EC).** This is a cancer that begins in the lining of the uterus (endometrium). People with Lynch syndrome have a higher risk of getting endometrial cancer than people who do not have Lynch syndrome.
- **Ovarian cancer.** This is a cancer that begins in the ovaries. People with Lynch syndrome have a higher risk of getting ovarian cancer than people who do not have Lynch syndrome.

People with Lynch syndrome are at a higher risk of getting some cancers than the average person. These cancers include:

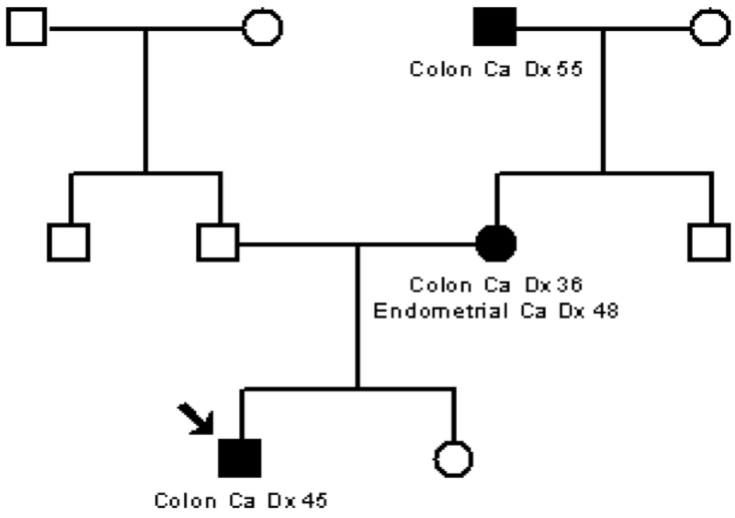
- Small bowel
- Ureter and kidney (transitional cell carcinomas)
- Stomach or gastroesophageal junction (where the esophagus connects to the stomach)
- Hepatobiliary tract (pancreas and bile duct)
- Brain
- Skin (specifically sebaceous tumours that are cancerous or non-cancerous)

If you have Lynch syndrome, the risk of getting one of these cancers is a bit higher than in someone without Lynch syndrome, but the actual chances are still quite low.

Who is at risk for Lynch syndrome?

Families with Lynch syndrome often have:

- Multiple family members with colorectal cancer, endometrial cancer or other Lynch syndrome-related cancers. These cancers are often diagnosed (found out about) at younger ages, often under age 50.
- More than one generation with these cancers. For example, grandparents and parents and siblings have these cancers.
- More than one type of Lynch syndrome cancer in the same person.



This family tree diagram shows how cancer can be passed down.
Your health care provider can explain it in more detail.

What causes Lynch syndrome?

Lynch syndrome is passed down from parent to child through genes. Genes carry instructions to help our body work properly.

There are 5 genes known to be linked to Lynch syndrome:

- MLH1
- MSH2
- MSH6
- PMS2
- EPCAM

Everyone has 2 copies (a pair) of these 5 genes, and they normally help protect us from getting cancer. One copy is passed down from each parent.

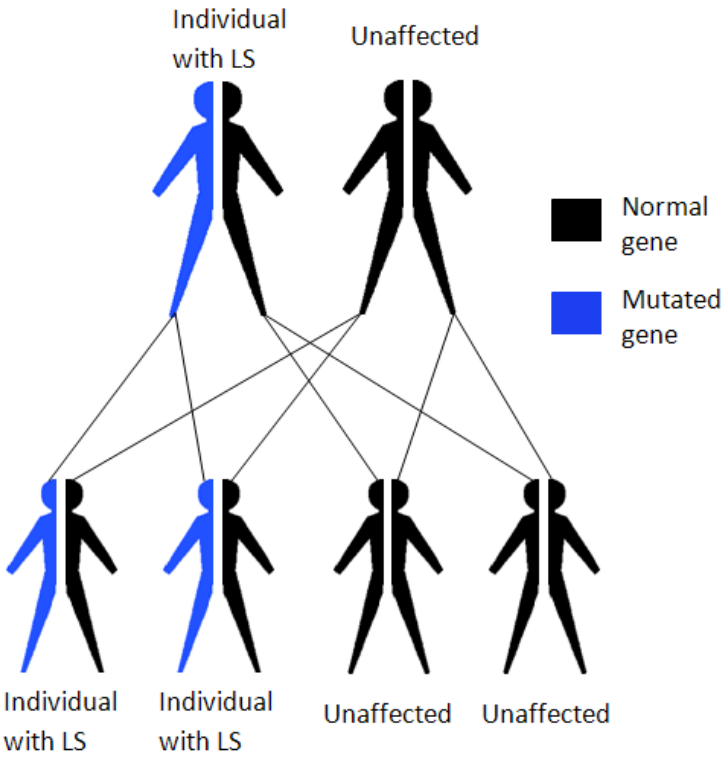
People with Lynch syndrome are born with a mutation (change) in one copy of one of these genes.

Children and siblings of someone with Lynch syndrome have a 50% (1 in 2) chance of getting that same Lynch syndrome gene mutation.

It is not the cancer itself that is passed down from parent to child, but a gene mutation that is passed down.

This gives someone a higher chance of getting cancer. It does not mean that they will get cancer for sure.

Inheritance (passing down) of Lynch syndrome



How to screen for colorectal cancer

Colorectal cancer screening:

- For people with MLH1, MSH2, EPCAM mutation screening is done with a colonoscopy (a test to see inside the colon and rectum) **every 1 to 2 years, starting at age 20 to 25.**
- For people with MSH6, PMS2 mutation screening is done with colonoscopy **every 1 to 3 years starting at age 30 to 35.**
- Some people may be advised to start screening at an earlier age depending on family history.
- Research shows that frequent colonoscopy lowers the risk of getting colorectal cancer in people with Lynch syndrome.

How to screen for endometrial cancer

- There are no known benefits of screening for endometrial cancer in people with Lynch syndrome.
- The best way to identify endometrial cancer is to screen for symptoms.
- Symptoms include postmenopausal bleeding (bleeding after someone has reached the menopause) or abnormal periods in premenopausal people (heavy vaginal bleeding, prolonged periods or bleeding between periods).
- Your doctor can check the cause of abnormal bleeding with an endometrial biopsy (a test that takes a small sample of tissue from the inner lining of the uterus). This is done by inserting a thin, flexible tube into the cervix and uterus. The tissue sample is then looked at under a microscope for pre-cancerous or cancer cells. This test is done in the office.
- Your doctor **may** recommend you do a yearly endometrial biopsy starting at age 30 to 35 until the end of childbearing or cancer preventing surgery (removal of uterus).

How to screen for ovarian cancer

- There is no effective screening to detect ovarian cancer in people Lynch syndrome.
- People with Lynch syndrome at a higher risk of ovarian cancer should be aware of symptoms of ovarian cancer (see below for ovarian cancer symptoms) and tell their doctor.

Screening for additional Lynch syndrome cancers:

At this time, routine screening for other Lynch syndrome-related cancers has not been shown to be useful.

Talk to your doctor if you have questions or concerns about your screening.

Talk to your doctor if you have questions
or concerns about your screening.

How to reduce gynecological cancer risks if you have Lynch syndrome

If you have Lynch syndrome and have a uterus and ovaries, there are things you can do that may help to reduce the risk of getting endometrial and ovarian cancer.

Some of these things include:

- Oral contraceptives (birth control pills) may reduce the risk of endometrial and ovarian cancer.
- Progestin-releasing IUD (birth control device) may reduce the risk of endometrial cancer.
- At the age of 40, or when you are done having children, you may be offered risk-reducing surgery (surgery to prevent cancer).
- Exact timing of surgery depends on many factors including:
  - whether child-bearing is complete
  - other medical or surgical issues you may have
  - the type of gene causing your Lynch syndrome (each gene has different risks of developing cancer), and
  - family history
- Surgery to prevent cancer includes removal of uterus, fallopian tubes and in most cases removal of your ovaries.
- If surgery is done before menopause, estrogen replacement therapy would be offered in most cases.

Endometrial and ovarian cancer symptoms

Contact your family doctor or your gynecologist right away, if you have:

Endometrial cancer symptoms:

- Vaginal bleeding between periods
- Heavy or prolonged periods
- Vaginal bleeding after reaching menopause

Ovarian cancer symptoms:

- Feeling bloated
- Pain in pelvis or abdomen
- Trouble eating or feeling full fast
- Need to urinate (pee) often or urgently

It is important to know there can be other causes for these symptoms other than cancer.

What support can you get?

The Familial Gastrointestinal Cancer Registry (FGICR) is part of Sinai Health System and works together with the University Health Network Division of Gynecological Oncology.

FGICR's goal is to give education and support to families with Lynch syndrome and other types of hereditary cancer. The FGICR works on research with other international groups that also specialize in hereditary cancers.

The FGICR uses research to learn more about possible:

- cancer causes
- cancer treatments
- cancer screening (testing)
- cancer prevention
- the genetics of colorectal and other cancers linked to Lynch syndrome

This booklet is meant to give you an overview of Lynch syndrome and not to replace a full genetics assessment.

If you have any questions about the booklet or its content, please contact us or your local genetics clinic, or speak to your doctor.

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Notes

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