

Understanding and Managing Lynch Syndrome



For women who may have Lynch syndrome and their family members

Read this pamphlet to learn:

- What 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



Is cancer hereditary (runs in your family)?

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 (something that runs in your family), which can increase your risk of getting cancer.

Lynch syndrome (LS) is a hereditary condition, and is also called Hereditary Non-Polyposis Colorectal Cancer (HNPCC).

What 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.
- **Endometrial cancer (EC).** This is a cancer that begins in the uterus. Women with Lynch syndrome have an increased risk of getting this.
- **Ovarian cancer.** This is a cancer that begins in the ovaries. Women with Lynch syndrome have an increased risk of getting this.

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

- Small bowel
- Ureter & kidney (like transitional cell carcinomas)
- Stomach
- Hepatobiliary tract (like pancreas and bile duct)
- Brain
- Skin

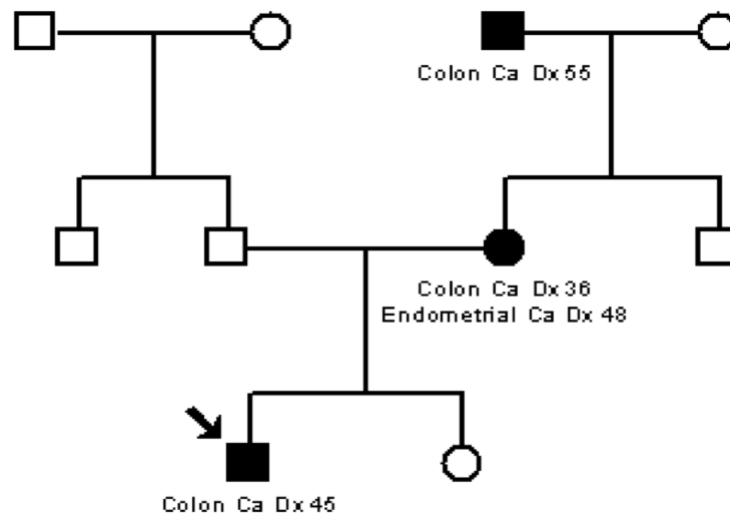
If you have Lynch syndrome, the risk of getting one of these cancers is a bit higher than in someone without Lynch syndrome, but the chances are still quite low.

Who is at risk for Lynch syndrome?

Families with sporadic (happens by chance) cancer, and families with Lynch syndrome are different.

Families with Lynch syndrome often have:

- One or more persons with colorectal cancer (CRC), endometrial cancer (EC) or other Lynch syndrome-related cancers (like ovarian cancer)
- More than one generation with these cancers
- More than one cancer in a person
- Cancers diagnosed (found out about) at younger ages (often under age 50)



What causes Lynch syndrome?

Lynch syndrome is hereditary, meaning that it can be 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 (TACSTD1)

Everyone has these 5 genes, and they normally help protect us from getting cancer.

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

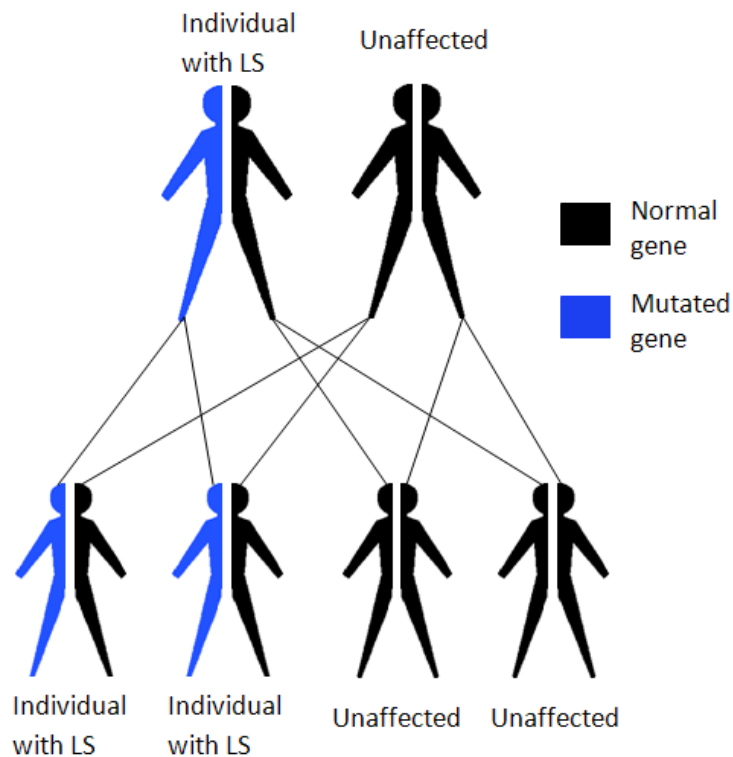
Everyone has 2 copies of every gene. One copy is passed down from each parent.

Children and siblings of someone with Lynch syndrome have a 50% (1 in 2) chance of getting that same Lynch syndrome-gene mutation. This is called **autosomal dominant inheritance**.

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



Lynch syndrome has an **autosomal dominant** pattern of inheritance.

What does this mean?

Someone with a gene mutation for LS has a 50% chance of passing that mutation on to each of their children.

How to screen for colorectal cancer

Colorectal cancer screening:

- Screening is done with a colonoscopy (a test to see inside the colon and rectum) **every 1 to 2 years, starting at age 20–25**.
- 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 ovarian cancer

Gynecological cancer screening

- Little is known about the benefits of screening for endometrial and ovarian cancers (gynecologic cancers)
- The best way to find ovarian cancer is to look for symptoms (see section on symptoms)

Ovarian Cancer Screening:

There are few options to screen for ovarian cancer. Women with Lynch syndrome can get:

1. **CA-125 Assay:** A blood test that measures your level of CA-125 (a substance in your bloodstream).

A high CA-125 may be linked to certain types of cancer. Your CA-125 can also be high for non-cancer reasons, especially in pre-menopausal women.

2. **Pelvic Exam:** An internal exam of the

- ✓ vagina
- ✓ cervix
- ✓ uterus
- ✓ fallopian tubes
- ✓ ovaries
- ✓ rectum

This is done by a doctor to feel for the size, shape and position of your ovaries and uterus.

3. **Transvaginal ultrasound:** An internal ultrasound that looks at the uterus, fallopian tubes and ovaries.

How to screen for endometrial cancer

Endometrial Cancer Screening:

Endometrial cancer grows inside your uterus, so it will not normally be found by a pap test (sample of cells from your cervix). Women with Lynch syndrome can get:

1. Endometrial biopsy: A test that takes a small sample of tissue from the inner lining of the uterus (endometrium).

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.

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.

How to reduce gynecological cancer risks for women with Lynch syndrome

If you are a woman with Lynch syndrome, 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.
 - Risk-reducing surgery is when your uterus, fallopian tubes and ovaries are removed.

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 and/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) a lot or badly

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 Mount Sinai Hospital and works together with the University Health Network Division of Gynecological Oncology.

FGICR's goal is to give education and support to families with LS and other types of hereditary (runs in your family) 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 LS

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.

Contact us

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