Li-Fraumeni Syndrome (LFS)

Li-Fraumeni (LFS) is a rare inherited syndrome that can lead to an increased risk of several types of cancer. It is also called the sarcoma, breast, leukemia, and adrenal gland (SBLA) cancer syndrome. Li-Fraumeni syndrome is hereditary, meaning that it is caused by a change (mutation) in a gene that can be passed down in families.

People with Li-Fraumeni syndrome (LFS) have an increased risk of several types of cancer, including:

- Osteosarcoma[1]
- Soft-tissue sarcomas[2]
- Leukemia[3]
- Brain (central nervous system) tumors[4]
- Adrenal cortex[5]
- Breast[6]
- Gastrointestinal cancers
- Melanoma[7]

These cancers often develop in relatively young adults or even children.

What causes Li-Fraumeni syndrome?

This syndrome is most often caused by an inherited mutation in the *TP53* gene, which is a tumor suppressor gene[8]. A normal *TP53* gene makes a protein that helps stop

abnormal cells from growing. When a mutation occurs in the *TP53* gene, abnormal cells can divide uncontrollably and become cancer.

How is Li-Fraumeni syndrome diagnosed?

Li-Fraumeni syndrome (LFS) is often diagnosed when a person suspected of having the syndrome has genetic testing to look for an inherited *TP53* gene mutation in their cells. However, not everyone with Li-Fraumeni syndrome is found to have a *TP53* mutation.

LFS is usually considered in people who are diagnosed with a sarcoma when they are younger than 45 years old, or who have family members diagnosed with any cancer when they are younger than 45 years old. These people are often advised to get genetic counseling, and possibly genetic testing.

To learn more about genetic counseling and testing, see Genetic Testing for Cancer Risk[9].

Why is it important to know if you have Li-Fraumeni syndrome?

People with Li-Fraumeni syndrome (LFS) have a high risk of developing some cancers during their lifetime (listed above). Some people might even develop more than one type of cancer. There are several reasons it might be important to know if you have Li-Fraumeni syndrome.

It might let you take steps to lower your cancer risk or find it early

- Ask your doctor if you should start getting cancer screenings at an earlier age, if you should be screened more often than normal, and if you should get certain kinds of screening tests.
- There are two different screening schedules for people with Li-Fraumeni syndrome: One for children, and another for adults. For children, the screenings are more focused on looking for adrenocortical carcinomas, brain, soft tissue, and bone cancers. For adults, the screenings focus more on looking for breast, brain, soft tissue, bone, skin, and gastrointestinal (especially upper) cancers.
- People with Li-Fraumeni syndrome have a higher risk of getting cancer from radiation exposure, so doctors advise them not to get any imaging tests[10] or treatments[11] that include any form of radiation.
- Ask your doctor if there are other things you can do to lower your cancer risk, such as staying at a healthy weight[12], being active[13], and avoiding or limiting alcohol[14].

It might affect your family members

If you have a *TP53* mutation, some of your blood-related family members might have it, too. Talk to your close relatives (parents, siblings, and children) about getting tested for Li-Fraumeni. If they prefer to not get tested, they may want to start screening for certain cancers early or take other precautions to lower their risk of cancer.

Hyperlinks

1. www.cancer.org/cancer/types/osteosarcoma.html
2. www.cancer.org/cancer/types/soft-tissue-sarcoma.html
3. www.cancer.org/cancer/types/leukemia.html
4. www.cancer.org/cancer/types/brain-spinal-cord-tumors-adults.html
5. www.cancer.org/cancer/types/adrenal-cancer.html
6. www.cancer.org/cancer/types/breast-cancer.html
7. www.cancer.org/cancer/types/melanoma-skin-cancer.html
8. www.cancer.org/cancer/understanding-cancer/genes-and-cancer/oncogenes-tumor-suppressor-genes.html
9. www.cancer.org/cancer/risk-prevention/genetics/genetic-testing-for-cancer-risk.html
10. www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests.html
11. www.cancer.org/cancer/managing-cancer/treatment-types/radiation.html
12. www.cancer.org/cancer/risk-prevention/diet-physical-activity/body-weight-and-cancer-risk.html
13. www.cancer.org/cancer/risk-prevention/diet-physical-activity/get-active.html
14. www.cancer.org/cancer/risk-prevention/diet-physical-activity/alcohol-use-and-cancer.html

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https://www.lfsassociation.org/what-is-lfs/treatment-preventive-screening/

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