

LFS AWARENESS. KNOWLEDGE IS POWER.



Empowering adolescents and young adults to drive key aspects of their healthcare and decision making.
Learn more about Li-Fraumeni syndrome (LFS).

WHAT IS LI-FRAUMENI SYNDROME AND WHAT ARE THE RISKS?

Li-Fraumeni syndrome (LFS) is a genetic condition that greatly increases the risk of developing many types of cancers. LFS affects all people regardless of sex and has been identified in families all over the world. Cancers related to this condition may occur at any age, including in children and young adults.



UNDERSTANDING WHAT CAUSES LFS

LFS is caused by a DNA change (sometimes called a variant or mutation) in the code of the *TP53* gene. When working properly, the *TP53* gene is the “guardian of our genome” which helps prevent DNA errors from occurring in our cells. Cancer is caused by accumulating lots of DNA errors in our cells so *TP53* is an important gene that protects us from developing cancer.


All of us are born with two copies of the *TP53* gene in each of our cells. For people with LFS, one of our two *TP53* genes is working and the other one has a change that causes it to not work properly. Genetic testing looks at the two *TP53* genes you were born with and looks for DNA changes that cause the gene to not work. This is usually done using a blood or saliva sample.

Most people with LFS inherit their *TP53* change from one of their parents. A small


number of people are the first person in their family to have LFS and they have a “de novo” (new) change.

When a child or young adult is found to have a *TP53* change, it is recommended that their parents and siblings (or other direct relatives) be referred to a genetic counselor to discuss testing even if no one else in the family has had cancer.

LFS is inherited, or passed down in families, in a way called “dominant inheritance.” This means that inheriting one altered *TP53* gene is enough to have LFS. For a person with LFS, there is a 50% chance that each of their children will also have LFS. There is also a 50% chance that each of their children will NOT have LFS. For this reason, it is helpful to talk with a doctor and/or a genetic counselor in the future when thinking about starting a family.



I have heard TP53 described as the 'superhero' of the genome. When it sees a cell accumulating DNA errors that cause cancer, it tries to fix the cell and, if it can't fix it, it destroys the cell. LFS occurs when one of TP53 'arms' is broken, so it has only one arm to fight with in battle.



Though the risk of developing cancer is significantly increased in people with LFS, it is important to know that not everybody with LFS will develop cancer. Some of the most common cancers seen in LFS are described below.

Cancers seen most often in children and young adults with LFS include:

- Soft tissue sarcoma (cancer of the tissues that connect, support and surround other body structures like muscles and fat)
- Osteosarcoma (cancer that starts in the bone)
- Brain tumors
- Adrenal gland cancer (the adrenal gland is a small organ located on top of each kidney which helps regulate your hormones)
- Leukemia (cancer of the blood)

Other cancers that occur more often in adults with LFS include:

- Breast cancer
- Colon cancer
- Melanoma (cancer of the skin)

The types of cancers that occur and the age when the cancers occur can vary between families and even within the same family. Some people with LFS develop two or more separate cancers over their lifetime. It is important for everyone with LFS to be monitored closely throughout their lives and to be cared for by doctors who are familiar with LFS.

WHY A **DIAGNOSIS OF LFS MATTERS**

While the diagnosis of LFS can seem overwhelming, it is important for adolescents and young adults to be aware of the condition and the steps they can take to better manage healthcare and address lifestyle behaviors (such as not smoking and using sunscreen to minimize exposure to cancer causing agents). Because people with LFS often develop cancers early, regular check-ups and cancer screenings are extremely important – the sooner a cancer is diagnosed and appropriately treated, the better the chances of a successful outcome. Coordination with individual medical providers is crucial to ensure that an LFS diagnosis is considered in the treatment of other conditions, particularly in regard to the use of radiation therapy.

GENETIC COUNSELORS AND GENETIC COUNSELING

Genetic counselors are health care professionals who specialize in talking with individuals about genetic conditions. Families often meet with a genetic counselor when LFS is diagnosed. For some young adults, this meeting may have occurred when they were children. It is important to know that genetic counselors can be a resource even after the diagnosis of LFS. Genetic counselors can help people with LFS continue to incorporate the diagnosis into their healthcare and their lives - especially at important times of transition such as adolescence, young adulthood, going off to college, and when a person is beginning to think about planning their own family. In some cases, a person may benefit from meeting with professional such as a psychologist or social worker over a longer period of time to talk about the emotional side of living with LFS. A genetic counselor can also help you identify these resources. The LFSA website has a Directory of Genetic Counselors where you can find a genetic counselor in your area.



LFS AND SCHOOL

In general, having LFS should not affect life at school on a day to day basis. Having LFS itself does not affect someone's ability to learn or to achieve academic goals in school but sometimes having symptoms when you have cancer may affect school. Because of the recommendation for frequent screening tests, there is a need (more than for other classmates) to go to extra doctor's appointments. When possible, it will be important to try to schedule these appointments at times that don't interfere with school and extracurricular activities. However most importantly, health comes first, and there may be times when a conflict cannot be avoided. There could also be times when treatment for cancer interferes with being able to attend classes in-person. Some people are able to minimize the impact of missed classes by having a tutor. And while each school's policy is different, it may be worthwhile to ask the teacher about an option to participate in class through a video conference, like Skype or FaceTime.

There may be times where thoughts related to LFS feel overwhelming and distracting. This can include a range of emotions from worry to sadness to anger. These feelings are all normal. But, if they become significant and are distracting from school or friendships, it's really important to talk about them. Identify trusted resources such as family members, a good friend or a school counselor or nurse to talk to about these feelings when they come up.

LFS AND FRIENDS

The decision to talk with friends or someone you are dating about having LFS is a personal one. People should share as much or as little information about LFS as they feel comfortable with. Good friends can be a great support system for all of life's good and bad moments including those related to LFS. It is important to remember that all people (with and without LFS) experience things in life that bring frustration, worry or feelings of being different, and that even friends without LFS can understand these kinds of feelings and provide support. Other young adults with LFS can also be a great support system since they may share similar experiences. The LFS Youth Program is a great way to meet other adolescents and young adults like you!





ABOUT THE **LFSA YOUTH PROGRAM**

The LFSA Youth Program is a program designed to provide support to teens and young adults with LFS (ages 13-25) by giving them opportunities to learn more about the condition from experts in the field in fun, engaging, and positive ways such as Youth Workshops, webinars, and quarterly Youth Newsletters. Very importantly, the Youth Program facilitates friendships between young adults living with LFS, creating a great support network amongst those who truly understand! As one member said, "I realized that I'm not alone and, more than this, I feel inspired by every teen and young adult. I can see myself in them and I see that they have fears and concerns just like me... I could see that I'm not the only one who lives with these questions, this inspires me to fight and to live one day at a time"

**To learn more about the LFSA youth program visit the website at:
www.lfsassociation.org/youth-program/**



Learn more at **LFSAssociation.org**

Legal Disclaimer: The information presented here is intended to provide general information for the LFS community. It is not intended to replace consultation with qualified medical professionals familiar with the conditions and considerations for any individual.