

# SMALL LYMPHOCYTIC LYMPHOMA

### WHAT IS SMALL LYMPHOCYTIC LYMPHOMA?

Small lymphocytic lymphoma (SLL) is a slow-growing, chronic blood cancer involving an abnormal overgrowth of white blood cells called B-lymphocytes. It is the same disease as chronic lymphocytic leukemia (CLL), with the main difference being where the cancer cells are located within the body. CLL is found in the blood and bone marrow, whereas SLL is found in the lymph nodes and spleen.
SLL is best understood as a different stage of CLL where there are not a significant number of cancer cells present in the blood and bone marrow yet. At the time of diagnosis, more than 90% of people qualify as having CLL, not SLL. But most of those with SLL will eventually progress to CLL.

### WHAT CAUSES SMALL LYMPHOCYTIC LYMPHOMA?

The exact cause of SLL is still unknown. As with many types of cancer, advancing age plays a significant role. Researchers are continually working to understand the exact mechanisms that cause the disease. While many suspect the cause might be related to genetic mutations occurring in the DNA, specific genes that cause SLL have not yet been identified. The disease typically occurs spontaneously. SLL is generally not considered hereditary in the sense that it cannot be directly passed down to children. However, some new research suggests that in rare cases there may be a connection to a family history of blood cancers.

#### WHAT ARE SOME OF THE RISK FACTORS FOR DEVELOPING SMALL LYMPHOCYTIC LYMPHOMA?

- Age: SLL is most often diagnosed in people over 60 years old, but it can also occur in much younger adults. SLL is extremely rare in children. About 90% of people diagnosed are older than 50 years old, with the average age of diagnosis being 71 years old.
- **Gender:** Men are almost twice as likely to develop SLL compared to women.
- **Race/Ethnicity:** Individuals who are White are more frequently affected by SLL than other racial or ethnic groups, followed by Blacks, Hispanics, and Native Americans. It is rare in Asians. There is a higher disease association with Europeans of Ashkenazi Jewish descent.
- Environmental, Occupational, and Chemical Exposure: People exposed to certain chemicals and toxins seem to be at greater risk for developing SLL. For example, those living on or near a farm have increased risk, but it is not clear

if factors such as pesticides are contributing to this risk. There is some suspicion that high levels of radon exposure in homes may also increase the risk of developing SLL.

• Veterans: The U.S. Department of Veterans Affairs lists SLL as a disease associated with exposure to Agent Orange, a chemical used during the Vietnam War. An increased risk has also been associated with exposure to burn pits in the war with Iraq.

### HOW IS SMALL LYMPHOCYTIC LYMPHOMA DIAGNOSED?

The diagnosis of SLL requires the finding of one or more enlarged lymph nodes and/or an enlarged spleen, along with an absolute lymphocyte count (ALC) of less than 5 x 10<sup>9</sup>/L. Once the ALC rises above  $5.0 \times 10^9$ /L the diagnosis becomes CLL. A lymph node biopsy is usually performed to confirm the diagnosis.

### WHAT ARE THE SYMPTOMS OF SMALL LYMPHOCYTIC LYMPHOMA?

It is very common to not have any symptoms when being diagnosed with SLL. However, symptoms may include:

- Swollen, painless lymph nodes in the neck, armpit, or groin (that can grow to be unsightly and uncomfortable)
- An enlarged spleen which can cause abdominal pain or a feeling of fullness, often resulting in discomfort and a lack of appetite
- Severe fatigue that makes it difficult to work or perform the usual daily activities
- Abnormal bruising or bleeding
- Recurrent or frequent infections
- Unexplained weight loss (more than 10% of an individual's total body weight in the previous six months)



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- Fevers higher than 100.4°F for at least two weeks without evidence of infection
- Drenching night sweats (soaking the bed sheets) that occur for more than one month without evidence of infection
- An exaggerated response to insect bites (such as mosquitoes)

### WHY ARE THOSE WITH SMALL LYMPHOCYTIC LYMPHOMA CONSIDERED IMMUNOCOMPROMISED?

B-lymphocytes are an important part of the immune system that helps to fight off infections. In SLL, these cancerous B-lymphocytes are unhealthy and do not work as well, putting individuals at much higher risk of developing severe infections due to a weakened immune system. Those with SLL can also have weakened responses to vaccines putting them at an even greater risk for poorer outcomes associated with respiratory infections (such as pneumonia, COVID-19, RSV, and influenza).

## IS SMALL LYMPHOCYTIC LYMPHOMA TREATABLE?

SLL is a chronic disease that individuals can live with for a very long time, some of whom will never need to start treatment. Most people diagnosed with SLL will not need treatment upon diagnosis. Instead, they will commonly begin a period of active observation (which is also sometimes called watch and wait) in which they are regularly monitored by their healthcare provider. Research indicates that starting treatment for SLL right away does not result in improved outcomes. Chemoimmunotherapy is almost never used to treat SLL anymore due to the development of newer and better targeted oral therapies. If the disease progresses and meets criteria for treatment, there are many different drug options available to help treat the disease.

### CAN SMALL LYMPHOCYTIC LYMPHOMA BE CURED?

SLL is generally considered a chronic incurable cancer, but it can be managed very well with treatment. SLL can go into remission, but it often comes back repeatedly over time. With the recent development of new and improved therapies, life expectancy for those with SLL is now very near that of those who do not have the disease.

### **CLL SOCIETY MISSION**

CLL Society is an inclusive, patient-centric, physician-curated nonprofit organization that addresses the unmet needs of the chronic lymphocytic leukemia and small lymphocytic lymphoma (CLL/SLL) community through patient education, advocacy, support, and research.