CLL SOCIETY
Resource Library

# IMMUNE THROMBOCYTOPENIC PURPURA

## WHAT IS THROMBOCYTOPENIC PURPURA?

Immune thrombocytopenic purpura (ITP) is a rare but serious complication that happens in an estimated 1-5% of those living with chronic lymphocytic leukemia (CLL) and small lymphocytic lymphoma (SLL). ITP is a blood disorder that occurs when the body mistakenly attacks and destroys its own healthy platelets, which are cells in the blood that help stop bleeding and are essential for healthy blood clotting. When ITP occurs, it can cause a large and sometimes rapid decrease in the platelet count which results in an increased risk of bleeding.

## WHAT ARE THE SYMPTOMS OF IMMUNE THROMBOCYTOPENIC PURPURA?

The symptoms of ITP can vary widely and may closely resemble other medical problems. Always consult your healthcare provider for a diagnosis. Common signs and symptoms may include:

- Easy bruising, including very large areas of purple bruising under the skin (called ecchymosis)
- Small red or purple spots on the skin that may resemble a rash, but the color will not change if you press on them
- Frequent nosebleeds that may take a long time to stop
- Heavy and/or prolonged menstrual periods in females
- Bleeding gums, especially during brushing or flossing
- Blood in vomit, urine, or stool
- Unusual fatigue and weakness

## WHY DOES IMMUNE THROMBOCYTOPENIC PURPURA OCCUR?

The connection to CLL and SLL is not yet fully understood, but it is known that the dysfunctional immune system of those with the disease has an increased tendency to attack healthy blood cells (such as platelets). It is suspected that dysfunctional T-cells, which help regulate the immune response, likely play a role.

When the immune system attacks other healthy cells within the body, the condition is classified as an autoimmune disease. ITP can rarely occur alongside another autoimmune disease known as autoimmune hemolytic anemia (AIHA). When these two conditions occur together (or when combined with another rare autoimmune disease that also causes a decrease in a type of white blood cells called neutrophils), it is called Evans Syndrome.

## HOW IS IMMUNE THROMBOCYTOPENIC PURPURA DIAGNOSED?

There is no one specific test to definitively diagnose ITP. It is diagnosed by excluding all other causes, meaning once a low platelet count has been discovered, more tests are used to rule out any other causes. A low platelet count will be compared to previous platelet levels to determine how quickly the platelets have been decreasing. In ITP, the drop in platelets can occur suddenly, while a low platelet count that occurs due to worsening of the cancer usually happens more gradually.

Additional blood tests or a bone marrow evaluation may be necessary to determine if the low platelet count is due to ITP or some other cause (such as progression of the cancer, medication side effects, infection, a massively enlarged spleen, or other blood disorders). When a low platelet count is discovered, it is critical that autoimmune disease be ruled out as the cause before considering what the treatment should be. This is because both progression of the cancer and autoimmune disease can result in low platelet counts, and treatments for each can be very different.

In severe cases of ITP (where platelet counts drop to critical levels of less than 10 x $10^9$/L), treatment may be necessary before the suspected diagnosis can be confirmed with a bone marrow biopsy or other tests. How the body responds to an ITP-specific treatment prior to testing may help to either confirm or rule out the diagnosis of ITP.

## WHEN IS TREATMENT NEEDED FOR IMMUNE THROMBOCYTOPENIC PURPURA?

ITP does not always require treatment, especially when there are no significant signs of bleeding. Treatment may be necessary when the platelet count drops too rapidly, when the platelet count drops too low (below the level of 30 x $10^9$/L), when there is significant internal bleeding present, or when a higher platelet count is needed for other reasons (such as an upcoming surgical procedure or when blood thinning medications are needed to treat other health conditions). There is no specific platelet count that is universally considered safe or not safe. But the lower the platelet count, the higher the risk of developing a serious bleeding problem.

# IMMUNE THROMBOCYTOPENIC PURPURA (continued)

## HOW IS IMMUNE THROMBOCYTOPENIC PURPURA TREATED?

Treatment options include a variety of medications that reduce the destruction of platelets or increase their production. Some options include:

- **Steroids:** Initial treatment for ITP usually includes steroids taken by mouth to help control the immune system's overactivity. However, high-dose steroids can also be administered intravenously through a vein. Steroids help to prevent bleeding by reducing the rate of platelet destruction. If effective, steroids will result in an increase in platelet counts within 2 to 3 weeks.
- **Rituximab:** When steroids are not enough to help, a medication called rituximab may be given intravenously through a vein to further calm down the immune system and keep it from attacking the healthy platelets. Rituximab works by preventing the overproduction of antibodies (made by the dysfunctional immune system) that destroy healthy platelets. It also may help treat the underlying CLL or SLL.
- **Intravenous Immunoglobulin:** If the platelet count drops dangerously low or if there is significant internal bleeding present, a treatment called intravenous immune globulin (IVIG) may be given through a vein to slow the destruction of platelets and provide rapid improvement (within 24-48 hours). IVIG is also an available option for those who do not respond to steroid therapy. A rapid increase in the platelet count after being treated with IVIG may confirm the diagnosis of ITP.
- **Platelet Transfusion:** When counts are very low or when severe internal bleeding is present, a transfusion of platelets into a vein may be necessary to quickly increase the platelet count.
- **Splenectomy:** In some severe cases, surgical removal of the spleen may be necessary when other treatments do not help.
- **Other Treatment Options:** Thrombopoietin receptor agonists (TPO-RA) can be a successful treatment option that stimulates the production of new platelets, rather than slowing their destruction as some of the other common treatments do. Other medications given less often to treat ITP can include cyclosporine and very rarely cyclophosphamide. It is common to combine different medications to increase the platelet count when the disease is not well controlled using just one medication alone. An example of this treatment approach for ITP is combining steroids and IVIG. Another successful approach can be to treat the underlying CLL or SLL in order to better control the ITP.

## DOES IMMUNE THROMBOCYTOPENIC PURPURA IMPACT SURVIVAL IN THOSE WITH CLL AND SLL?

If left untreated, ITP can cause severe and life-threatening bleeding conditions. However, most studies have not found a reduction in overall survival of those living with CLL and SLL. ITP does tend to recur and can become a chronic condition that will need to be to be treated promptly if it does come back. Fortunately, ITP can usually be well-controlled with appropriate treatment and careful monitoring.

## 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.