

are developing an infection (fever, chills, sore throats, unexplained coughs, listlessness, muscle pain, etc.) and report them to your doctor right away.

- Sometimes a splenectomy can lead to an exceptionally high platelet count, which can in turn lead to blood clotting. Your doctor should monitor your platelet count on a regular basis and may ask you to take baby aspirin daily.

This sounds kind of frightening. Is a splenectomy really a wise choice?

That's a decision that must be made in each individual case. A doctor with significant experience with thalassemia is going to be in the best position to offer advice about this; however, most people who are splenectomized are able to manage the challenges it presents with relatively little difficulty.

Of course, it's best to avoid any circumstances that can lead to the need for a splenectomy in the first place. For a person with thalassemia, this means following a transfusion regimen that keeps hemoglobin levels above 9 or 10gm/dL. Such a regimen not only helps protect the spleen but can have other benefits as well.



If you have questions about the spleen and thalassemia, please contact your treating physician, or the Cooley's Anemia Foundation at (800) 522-7222 or info@cooleysanemia.org. If we cannot answer your question, we will be happy to put you in touch with one of the Thalassemia Treatment Centers.

Published by the Cooley's Anemia Foundation, 330 Seventh Ave., #900, New York, NY 10001 www.cooleysanemia.org (800) 522-7222. This publication is made possible by an unrestricted educational grant from Novartis Pharmaceuticals.

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Living with Thalassemia

Issues in
Thalassemia Care

Thalassemia and the Spleen

by Marie B. Martin, RN, and Craig Butler



What is the spleen?

The spleen is a small organ (normally about the size of a fist) that lies in the upper left part of the abdomen, near the stomach and below the ribcage.

What does it do?

The spleen has a number of functions, the most important of which are **filtering blood** and creating **lymphocytes**. It also acts as a "reservoir" of blood, keeping a certain amount on hand for use in emergencies.

In its filtering capacity, the spleen is able to remove large foreign particles from the blood. In addition, when red blood cells come to the end of their lifecycle (after about 30-60 days in thalassemia), the spleen breaks the cells down, removing the globin and iron from these cells so that they can be reused.

Lymphocytes are small white blood cells that are part of the body's immune system and help defend the body against disease. These lymphocytes help the spleen play an "early warning signal" role. When blood passes into the spleen, the lymphocytes react to anything suspicious in the blood and produce antibodies to fight the "invader." Thus, the spleen plays a very important role in fighting infections in the body.

How does thalassemia affect the spleen?

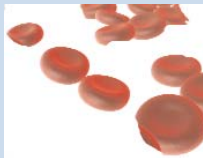
Thalassemia can affect the spleen in a couple of ways.

- When the spleen breaks down a red blood cell in a person with thalassemia, that iron often stays inside the spleen rather than being reused. Thalassemic red blood cells often also get stuck in the spleen because they're small and misshapen. Both of these factors can cause the spleen to become enlarged.

- In a person with thalassemia intermedia, hemoglobin levels may drop too low, causing his/her body to try to create more and more red blood cells. This is also the case in thalassemia major if a patient is not being adequately transfused. These cells are typically created in the bone marrow; but in thalassemia, the body demands more cells than the bone marrow can produce, and so other organs, including the spleen, may start creating them. (This is called **extramedullary erythropoiesis**.) This can also cause the spleen to become enlarged. It may also cause the spleen to become hyperactive, a condition called **hypersplenism**.

Is hypersplenism something to be concerned about?

Yes. In hypersplenism, the spleen starts operating at a faster rate - which means that it starts destroying more and more red blood cells, thereby lowering hemoglobin levels. This may result in a patient with thalassemia intermedia beginning transfusion therapy or in a patient with thalassemia major needing blood more often. More frequent transfusions in turn mean that more iron is being introduced into the body that must be removed through chelation therapy.



How is hypersplenism treated?

Hypersplenism usually can't be reversed, so it is important to prevent this condition from developing, which means ensuring that the patient is being transfused at an appropriate rate. The spleen size can be reduced once the patient is transfused at a rate high enough to turn off the need for increased production.

Sometimes it may be necessary to treat hypersplenism with a **splenectomy** (the removal of the spleen.)

Is splenectomy dangerous?

There is always a risk in surgery and anesthesia, but many people have their spleens removed due to various circumstances. However, because of the role that the spleen plays in getting rid of bacteria, microbes, etc., those who undergo a splenectomy are at increased risk of infection for the rest of their lives.

This risk is especially high for the first 1-4 years following the splenectomy, after which it remains a significant but lesser concern. Risks are higher with young children, so a splenectomy is frequently avoided if possible in children under the age of 5.

There is increasing evidence that splenectomy may increase the risk of pulmonary hypertension (PAH) by thrombosis and other mechanisms; therefore, patients who have a splenectomy should be monitored for PAH.

Sometimes hypersplenism can be treated by a **partial splenectomy**, in which only a portion of the spleen is removed. This reduces the risk of serious infection in the patient. However, a partial splenectomy is more complicated than a full splenectomy and should be considered only in specific situations.

What can be done to help reduce the risk of infection in a person with a splenectomy?

A number of treatments can help reduce the risk of infection. These include:

- A series of vaccinations both before and after the splenectomy. These include the pneumococcal hemophilus influenza (often called Hib), pneumovax, and meningococcal vaccines, which protect against meningitis, pneumonia, and infections of the blood, bones, and joints.
- Administration of penicillin or other antibiotics, prophylactically, for life after a splenectomy; some doctors will limit the period of time for antibiotic use.

Other things to consider include:

- If traveling to another country, find out if there are diseases there (such as malaria) which might pose a special threat and for which you would need to be prepared (via vaccination, medication, etc.)
- Pay attention to any warning signs that may indicate you