



No.62 - Don't Forget Splenectomy

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Title: **Don't Forget Splenectomy**

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In 2010 I wrote an American Perspective titled "Don't Forget Splenectomy". As new treatments are used more often, I think it's time to say again, "Don't forget splenectomy". This American Perspective is focused on adults with ITP.

Splenectomy

In the beginning, there was only splenectomy. Splenectomy was the first and the only effective treatment for ITP until steroids began to be used in the 1950s. During this era, every patient who was diagnosed with ITP and who had severe thrombocytopenia with bleeding symptoms had a splenectomy. Almost 90% of patients responded with an increased platelet count; two-thirds of patients achieved a normal platelet count. In almost all patients who responded, the response was durable for many years. Even in this ancient era before antibiotics and modern transfusions, complications were rare. These observations provide the basis for confidence in splenectomy as an effective treatment for ITP.

Steroids

Steroids suppress the immune system, and therefore can suppress the autoantibodies that cause platelet destruction in ITP. When steroid use began, platelet counts of most patients promptly increased, but also in most patients, platelet counts fell again when steroids were stopped. The side effects of steroids soon became apparent: facial change, mood disorders, bone weakness, risk for infection, and many other issues. In 2017, the most effective steroid treatment is a high dose of dexamethasone daily for just 4 days. This may be repeated at three weeks if the platelet count does not increase. Most patients increase their platelet count and about one-third of patients maintain a safe platelet count (over 30,000) one year later. With use of steroids, splenectomy became the "second-line" treatment for adults.

Rituximab

Rituximab is a stronger immune suppressive drug and it doesn't have the side effects of steroids. Beginning 20 years ago, rituximab appeared to replace splenectomy as the "second-line" treatment. But with more experience we have learned that the response to rituximab is significantly less than with splenectomy. Approximately half of patients have an initial response, but after 5 years only 20% of patients have maintained their response. Therefore, splenectomy remained the most effective treatment for achieving a durable and complete response.

TPO drugs

These are drugs that mimic the natural hormone, thrombopoietin, which stimulates platelet production. Beginning 10 years ago, two TPO drugs were approved for treatment of ITP: romiplostim and eltrombopag. Both are impressively effective, increasing platelet counts in about 80% of patients. These drugs must be given continuously (romiplostim is a subcutaneous injection given once per week; eltrombopag is a pill taken daily). When they are stopped, their effect stops in almost all patients. In a few patients, a spontaneous recovery from TTP occurs and the platelet count remains normal when the TPO drug is stopped. The long-term, maybe lifetime, use of these

drugs can provide effective treatment, but they are expensive and the long-term risks are unknown, although they appear to be few.

Splenectomy

So what is the role for splenectomy now? After all these years, splenectomy remains the most effective treatment for ITP. Splenectomy provides the highest frequency of durable responses. The other treatments certainly have important roles in the treatment of patients with ITP, and it is appropriate to try other treatments before considering splenectomy. But when other treatments fail, or use of the TPO drugs forever isn't appropriate, splenectomy is always there. It's a treatment that can provide permanent safe platelet counts without need for other treatments.

There are tests in the UK, done by Professor Newland and his colleagues, using radioisotope-labeled platelets to determine whether the spleen or the liver is the principal organ causing platelet destruction. But even when the test identifies the liver, not the spleen, as the principal organ, 50% of patients respond to splenectomy.

Of course splenectomy has risks, as any surgical procedure has. And since the spleen is a principal organ to make antibodies and provide immunity for infection, there is an increased risk for infection. But current immunizations for pneumococcus and other bacteria are very effective, and risk for serious infections following splenectomy is much less than it was a generation ago.

The Bottom Line

Splenectomy remains the most effective treatment for patients with ITP. When ITP is a serious problem, remember splenectomy.