**An American Perspective** reprinted from: **Mar 99**
Title: **Splenectomy in ITP: Immediate and Long-Term Complications**
Authors: **James N. George, M.D.,** Oklahoma City,
**George R. Buchanan, M.D.,** Dallas

In patients with ITP, the spleen is an important source of anti-platelet antibody as well as the major site of destruction of antibody-coated platelets. Therefore, surgical removal of the spleen (splenectomy) is often effective for children and adults with ITP. Splenectomy is appropriate when the disease is causing significant bleeding that is unresponsive to treatment, or when the treatment has excessive toxicities. Splenectomy is performed by either an "open" technique, by which an incision several inches long is made in the left upper part of the abdomen, or by the newer "laparoscopic" technique. By this method the spleen can be removed through one of several tiny incisions, with the actual procedure facilitated by a video camera inserted into the abdominal cavity.

The risks and side effects of splenectomy are immediate and long-term. The immediate risks include those of any surgical procedure, such as complications of general anesthesia. The major immediate risk for a patient with ITP is bleeding, either from the incision or internally during or following the splenectomy. For this reason treatment with prednisone or occasionally IVIG is given to raise the platelet count prior to surgery. Platelet transfusions are usually unnecessary, but they are sometimes given to patients with platelet counts below 20,000 per mm3. Surprisingly, serious bleeding at the time of splenectomy is unusual even when the platelet count is low.

There is only one important long-term complication of splenectomy, and that is a small but real risk of septicemia or so called "blood poisoning". Septicemia results from a bacteria (or rarely a protozoan) that enters the bloodstream (most commonly from the mouth or respiratory tract). Such bacteria are normally filtered and removed from the blood by the spleen. In an individual without a spleen, especially if he or she has low levels of antibody against that particular bacteria, the bacteria may rapidly multiply in the bloodstream and produce numerous toxins that damage the body's organs. The initial manifestation of septicemia (also called sepsis) is a high fever with shaking chills. Without treatment, septicemia is rapidly fatal, often within twelve to twenty-four hours. The most common bacteria causing septicemia is Streptococcus pneumoniae, or the pneumococcus, which is also the most common cause of bacterial pneumonia in persons of all ages.

How common is septicemia following splenectomy? Fortunately, it is very rare, but when it occurs it is a terrible catastrophe. The analogy that we make with our patients is that it is like an airplane crash - i.e., not likely to occur but potentially fatal. The risk of septicemia following splenectomy is greater in very young children (especially those under four or five years of age). One study of 226 patients followed for up to forty-five years after splenectomy documented four deaths due to sepsis. But all deaths occurred before 1967, before pneumococcal vaccine (described subsequently) was available. Neither of us, with a combined fifty years of practice, has seen septicemia after splenectomy for ITP in any of our patients.

How does one prevent septicemia after splenectomy? Three measures can be taken. First, all individuals having splenectomy should receive (ideally before the operation, but afterwards is also acceptable) a dose of polyvalent pneumococcal vaccine. Some doctors recommend two other vaccines, the HIB vaccine (against Hemophilus influenzae) and meningococcal vaccine. Some experts recommend revaccination three to five years later with the pneumococcal vaccine, but there are no data that support this. The second measure is use of penicillin. Most pediatric hematologists prescribe penicillin twice daily for at least two or three years following the splenectomy, and some recommend it for life. Given the lower risk of septicemia in adults, many adult hematologists do not feel that regular penicillin is required. However, everyone would agree that an asplenic person (the term used to describe someone who has had a splenectomy) should always see a doctor immediately in the event of a fever, especially when it is associated with chills and headache. They should tell the doctor (who may not be aware of the problem of post-splenectomy septicemia) that they have had a splenectomy and need antibiotics. Hospitalization, a blood culture test, and intravenous antibiotics are usually necessary. If there is going to be any delay before seeing the doctor, taking a dose or two or penicillin by mouth is recommended. One of us asks our patients who have had a splenectomy to always have a bottle of penicillin on hand.

Overall, the risk of septicemia following splenectomy is probably less than the risk of hemorrhage in the brain due to a low platelet count in the ITP patient who hasn't had a splenectomy, and we know that risk is very small.

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Title: **Splenectomy and ITP**
Authors: **James N. George, M.D.,** Oklahoma City,
**George R. Buchanan, M.D.,** Dallas

SPLENECTOMY WAS THE FIRST effective treatment for ITP, beginning in 1913 and continued to be the only effective treatment for ITP until the discovery of steroids in the middle of the twentieth century.

For the past fifty years, steroids have been the standard initial treatment for adults with severe and symptomatic ITP, because of the assumption that the thrombocytopenia will be persistent and therefore treatment is required. For children, as has been discussed here before, the initial management decision is whether drug treatment is required at all. Splenectomy is rarely required because most children, even those with chronic ITP, gradually improve and eventually recover on their own.

Steroid treatment has severe side effects and many potential complications; treatment is generally not tolerable for more than a month or two. In those patients who require additional treatment, splenectomy has the best record of success. A review of all publications describing splenectomy for adults with ITP documents that two-thirds of patients will achieve a normal platelet count and require no further treatment. Approximately half of the remaining patients will be improved, although their platelet count does not return to normal.

In 2004, is splenectomy still the best treatment option after failure of improvement with steroids? Or are newer treatments more effective and safer than splenectomy? Probably nothing can replace initial treatment with steroids, because steroid medications are so inexpensive and, at least in the first several weeks, have tolerable side effects, although mood changes, sleeplessness, and jitters can be disturbing. But should splenectomy be replaced as the next treatment option?

No other treatment has the overall success of splenectomy, with two-thirds of patients developing normal platelet counts and requiring no further therapy. But for some patients other treatments may be preferable to surgery. Some physicians suggest intermittent 'maintenance' treatment with IVIG or anti-D. However each treatment with these medicines only temporarily increases the platelet count for several weeks; therefore repeated intravenous infusions are required. In some patients, no response occurs or the platelet count increases for only a few days. In addition, these agents are very expensive and they have side effects that we have previously discussed. In our view, splenectomy remains preferable to long-term, intermittent treatment with IVIG or anti-D.

What about rituximab? This is a novel treatment first developed as chemotherapy for patients with malignant lymphoma, but rituximab has also been effective with patients with ITP and other autoimmune diseases. There are reports of serious reactions, but these are rare. Many ITP patients receiving rituximab have improved, but probably fewer than half have recovered with normal platelet counts. Whether this recovery will last for many years cannot yet be known. Splenectomy may still be more effective than rituximab and other chemotherapeutic agents.

Do good responses following splenectomy last forever? The answer is a qualified 'yes'. Even though ITP may recur after splenectomy, most commonly within the first several years, the overall experience with splenectomy suggests that responses are durable. The longest average follow-up times for patients after splenectomy that are described in medical reports are ten to fifteen years, and the proportion of patients with normal platelet counts (two-thirds) is the same as soon after splenectomy.

What are the risks of splenectomy? There is always a risk for death with any surgery, related to operative or anesthetic complications. Recent data for splenectomy suggest that the operative mortality is 0.2% – that is 2 patients out of 1000. Although this is low for a surgical procedure, any risk of death is a critical concern for patients with ITP, whose risk for fatal bleeding is so rare. Operative complications that require prolonged hospitalization may occur in about 10% of patients following splenectomy. Some of these complications can be serious infections; others include bleeding and surgery-related blood clots. New techniques of laparoscopic (keyhole) splenectomy have fewer risks and require only a day or two of hospitalization.

Can any test predict the outcome of splenectomy? Many features of ITP have been analyzed, but only age seems to predict success, with younger patients having better responses than older persons. This is like everything else in ITP; younger patients, particularly children, do better. Some hematologists believe that radioisotope tests* to measure whether the spleen is the dominant organ that is destroying platelets (as opposed to the liver) can predict the success of splenectomy, as well as predict who will not respond. However, the results with these tests are variable in different hospitals. When reviewing all reports describing these techniques, it is not clear how accurate they are.

Therefore, at least for adults, splenectomy remains an appropriate treatment for patients with severe and symptomatic ITP. Other options are available, but their record of success and their risk of complications have not made them preferable to splenectomy. No tests consistently predict the success or failure of splenectomy. The responses to splenectomy are usually long-lasting.

Even though splenectomy remains an appropriate treatment, the frequency of splenectomy as treatment for ITP in adults seems to be decreasing. This may be because patients and their doctors appreciate that a normal platelet count is not an essential objective for treatment; an appropriate goal is only to achieve a safe platelet count. Therefore, many patients and their doctors are more comfortable with consistently abnormal platelet counts as long as there are no significant bleeding problems. There's nothing at all wrong with such an approach.

*Readers who are interested to learn more about the Indium Labelled Platelet Spleen Scan, conducted at the Royal London Hospital, should send an SAE to Shirley at HQ for a copy of our splenectomy booklet – see publications list for details – Ed. Ω