The American Perspective: Revisiting Splenectomy for Treating ITP
Spero R. Cataland, M.D.

Removing the spleen has historically been a very effective treatment option for many hematologic conditions including immune thrombocytopenic (ITP). In the past 15 years there have been numerous new medications that have received regulatory approval for the treatment of ITP that work by increasing the production of platelets, (romiplostim, eltrombopag, avatrombopag) or inhibiting the destruction of platelets (fostamatinib), with even more new treatments on the way. Coupled with the other existing medical therapies available to treat ITP, it might be expected that performing a splenectomy to treat ITP might not be necessary any longer. The enthusiasm for splenectomy for the treatment of ITP seems to wax and wane over time for both providers and patients. Splenectomy for the treatment of ITP though still provides the greatest chance to alter the course of ITP and induce long-term remissions or cures of ITP. It also carries the greatest risk for patients compared to any other treatment for ITP, therefore careful consideration and discussion of the risks and benefits of undergoing a splenectomy for ITP (as well as the certainty of the diagnosis of ITP) with their physician is essential.

The spleen is the major site of removal of antibody coated platelets in patients with ITP in addition to housing lymphocytes that may be responsible for producing anti-platelet antibodies that lead to the removal of platelets. Given this it makes sense as to why removing the spleen has the best chance of modifying the disease. In one reported study that combined the results of 47 case series studies of splenectomy for ITP, 66% of patients were able to achieve a normal platelet count after splenectomy that persisted for over 10

years. Complications (discussed below) occurred in approximately 13% of patients that underwent an open, surgical splenectomy and roughly 10% of patients that underwent a laparoscopic surgical technique (uses 2-3 small surgical ports rather than the typical larger, surgical incision).

The most significant risks from the removal of the spleen stem from the spleen's role in fighting infection and the clearance of antibody-coated bacteria from the blood stream. Because of its role in fighting infection, especially from bacteria categorized as encapsulated organisms (bacterial cells covered by an outer layer that helps the bacteria to cause disease), infection is the most significant lifelong risk for patients that undergo a splenectomy. For this reason, patients are vaccinated against these encapsulated bacteria prior to surgery and receive regular booster vaccines after surgery. After splenectomy antibiotic therapy is promptly initiated for splenectomized patients that develop a febrile illness. Pediatric patients, more so than adult patients, may also be prescribed prophylactic antibiotics to prevent infections. Mortality rates for the surgery itself are also a consideration and are 1% with the open surgical procedure and 0.2% for the laparoscopic surgical approach.

Patients with ITP are also known to have a greater risk for venous thromboembolic disease compared to patients without ITP, and this risk is thought to possibly be increased in ITP patients after splenectomy. Cardiovascular complications including pulmonary hypertension are thought to be increased after splenectomy, but there is no clear evidence for this to date.

If we accept the idea that splenectomy remains a good treatment option for ITP, the next obvious question is who should undergo splenectomy for ITP? Given the potential for ITP in adults to spontaneously go into remission 20% of the time (most commonly in the first year), it is generally recommended that adults wait at least one year after diagnosis before undergoing splenectomy. It is also thought that younger patients are more likely to respond to splenectomy, but there is no clear age cutoff that predicts a decreased response to splenectomy. Studies have used various tests to attempt to predict who would be most likely to respond to splenectomy, but to date there is no reliable means to predict who will respond to splenectomy, and who will not.

The development of several new treatment options for ITP in recent years has positively impacted the lives of patients with chronic ITP, allowing them to live a normal life without persistent concerns for bleeding complications from their ITP. These treatments come in many forms (oral, subcutaneous, intravenous) also allowing patients more freedom and control over the type of treatment they choose. However, for patients who have grown intolerant of the need for ongoing treatments due to side effects, or the impact on their lives from having to make regular trips to the clinic for monitoring or treatment, splenectomy should be considered a reasonable option to treat their ITP. As with any other ITP treatment, the relative risks and benefits of a splenectomy should be discussed with their physician so that an educated and informed decision can be made by patients.