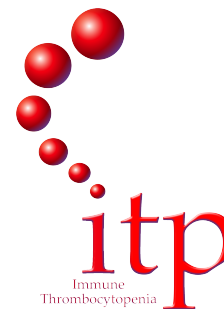


No. 63 - Splenectomy for Children with ITP



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Title: Splenectomy for Children with ITP - Similarities and Differences with Adults

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We are fortunate that most children will have resolution of their ITP by 6 months and very few children go on to have ITP that lasts longer than 1 year. This makes children with ITP different from adults who are more likely to have the disease for a long time and fail many treatments. This realization that children are not simply “little adults” becomes very important when we think about how to treat ITP.

A big area of difference is with splenectomy, however some aspects of splenectomy are similar between adults and children. In children, just like in adults, splenectomy remains very good treatment with about 75-90% children having a sustained normal platelet count following splenectomy. For a long time it was also the only treatment we had for children with ITP who were not responding to first-line treatments like steroids and IVIg. Now we have additional treatments like rituximab and the thrombopoietin- receptor agonists (TPO-RAs), but often times, splenectomy remains the best option. The major side effect of splenectomy, increased risk for infections, is also no different between adults and children. The spleen is an important infection fighter and so splenectomy removes that defense and places the child at risk for infection. Just like adults, children who undergo splenectomy will need to be on life-long antibiotics, receive special vaccinations and seek medical attention each time they have a fever. Perhaps, children may also have less side effects with splenectomy because their bodies are healthier overall when they have the surgery. Lastly, just like in adults, splenectomy should always be urgently considered with life-threatening bleeding. Splenectomy provides the quickest and most reliable response in platelet count and therefore is always considered when bleeding needs to be stopped immediately.

So how are children and adults different? First, the chance that most children are likely to have their ITP go away even months to years after the diagnosis makes doctors want to wait longer before having a child undergo an irreversible procedure such as splenectomy. Secondly, when we discuss side effects of splenectomy to a parent of a two-year old, talking about things that will occur life-long can seem very overwhelming. I have also found that sometimes parents find it very difficult to make a decision about surgery, which may be considered elective, on behalf of their child. Lastly, we now have additional medications that can be used in order to delay or avoid splenectomy, such as rituximab or the thrombopoietin receptor agonists. These have given us options and many parents wish to try these before their child has surgery.

So when should splenectomy be considered in children with ITP? The 2011 American Society of Hematology (ASH) guidelines recommended splenectomy for children with ITP of at least 3 months duration with significant bleeding, lack of responsiveness to other therapies and/or need for improved quality of life. It was, however, suggested that given the high likelihood of remission in children that splenectomy be delayed for 12 months unless the child needs more urgent treatment because of bleeding. When talking to a child or parent of a child who has failed to respond to first-line therapy the following factors should be determined: 1) How much bleeding is the child having 2) What is the impact of ITP on the child and family’s quality of life 3) what other treatments has

the child had and how did they with them 4) what is the family's biggest concern 5) what are the overall goals of treatment. This allows the physician to then discuss each treatment option, including splenectomy, with respect to each of these factors. This allows for determination of a treatment option that might best align with the needs and goals of the family. If the discussion favors splenectomy, then more time should be spent evaluating if this is truly the best option for the individual child. Physicians should consider the age of the child, how long they have had ITP, the family's experience of living with the condition, and the goals of treatment to make sure that splenectomy is still the most reasonable treatment option.

Splenectomy remains very good treatment with ITP, especially in the setting of significant bleeding, parent/child who desire "curative" treatment with long lasting results, and children with long-standing disease. There is no one right treatment for children who have ongoing ITP, but understanding the options and the goals of the family can assist with ensuring the best option is chosen. Sometimes this is splenectomy!

I have seen many (approximately 50 children) with ITP during my last 3 years at Columbia University Hospital. Fortunately most have recovered very quickly from their ITP. For those that have not responded a handful have needed additional treatment. Two such children underwent splenectomy (one for severe bleeding and one for desired remission). All responded with normal platelet counts, which have remained normal. So splenectomy for children with ITP is rarely done, but it is a very effective treatment when it's needed.