The Oklahoma ITP Registry Newsletter

September 2014

Hello!

We would like to welcome you to our eleventh Oklahoma ITP Registry newsletter.

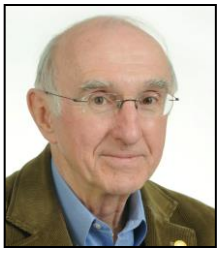
The ITP Registry

The Oklahoma ITP Registry began in November 2001. Since that time we have enrolled 118 people. With your participation we hope to document the clinical course and long-term outcomes of patients with ITP.

Follow-Up Reminder

Thank you to all of you who have returned your follow-up surveys! We send out surveys once a year so that we can document your general health. We just wanted to remind those who have not returned their follow-up surveys to please do so. Please send it back even if ITP is no longer a problem for you. In fact, if ITP is no longer a problem for you it's important for us to document that good news! If you need a new survey or would like to do your survey over the phone, please contact us by emailing Dee Terrell at **Dee-Terrell@ouhsc.edu** or Kaelyn Lu at **Kaelyn-Lu@ouhsc.edu** or by calling **(405) 271-8001** extension **48386**.

Dr. George's Perspective



What is a safe platelet count?

Platelet counts are measured in blood samples and reported as

the number per microliter (one one-thousandth of a liter). Normal individuals have platelet counts of 150,000 to 350,000. A platelet count over 50,000 appears to be safe to prevent bleeding from any type of injury or surgery. A platelet count between 20,000-30,000 is sufficient to prevent the appearance of petechiae, bruising or other spontaneous bleeding. Critical spontaneous bleeding, such as internal bleeding (intestinal bleeding, bleeding in the brain) does not occur unless the platelet count is extremely low, less than 5,000.

The goal of treatment for ITP is only to maintain a safe platelet count-to prevent bleeding. The goal is **not** to achieve a normal platelet count. Because many patients (especially children) recover spontaneously within several weeks or months, they are often observed carefully without treatment, even if their platelet count is very low. However, because the course of ITP in adults is often prolonged, some treatment is appropriate if the platelet count is very low and bleeding symptoms are present. When an adult patient is first diagnosed with ITP and a platelet count less than 30,000, some treatment is appropriate. In these adults with counts less than 30,000, the treatment goal is simply to achieve platelet counts greater than 50,000, because that will provide a safe margin for prevention of bleeding with accidents or injuries. Adults without bleeding symptoms, and who have platelet counts over 30,000, may be safely observed without any treatment.

What is a safe platelet count for medical procedures?

For medical procedures, you should consult with your hematologist for a safe platelet count range. Generally these guidelines exist:

Dentist: A trip to the dentist's office is perfectly safe, as long as the platelet count is in a safe range. Since a safe platelet count for dental procedures is 30,000-50,000, patients may need to take some medicine, such as steroids, to get their platelet count up before having teeth removed.

Pregnancy: Platelet counts often are lower during pregnancy and then recover after delivery. This rarely poses a problem during the pregnancy and the development of the fetus is normal in spite of the low platelet counts. However, potential risk for complications occurs at delivery. Doctors typically want the platelet count to be increased to 50,000 to 100,000 to allow for epidural anesthesia for delivery. This is done to prevent any risk of

bleeding around the spinal nerves when the epidural needle is inserted for the anesthesia and to prevent increased bleeding after delivery.
For other surgeries and procedures, it is recommended that you discuss your ITP and symptoms with all the physicians involved in your care.

Chronic versus Acute

"Chronic ITP" is typically considered a course of ITP that lasts more than 12 months. ITP varies from person to person, but can last from several years to a lifetime after diagnosis for someone diagnosed as a late teen or an adult. Because the course of ITP in adults is often prolonged, treatment is appropriate if the platelet count is very low and bleeding symptoms are present.

Most adult patients are treated first with steroid medicines that suppress the immune system and therefore prevent the increased platelet destruction. If the ITP does not go away then additional treatments are explored.

Children are usually considered to have "acute ITP" which means it usually goes away spontaneously within 12 months. Childhood ITP usually occurs before age 6 and symptoms such as bruising and bleeding can occur abruptly. Children are usually monitored closely by their hematologists without any treatment or intervention. Most children recover without any reoccurring problems.

Courtney's Story

In the fall of 2004, I began noticing bruises on my body. Mostly, just small bruises on my legs that I had no idea where they came from. For several months, I was having heavier periods than normal, but didn't think too much about it because I had recently stopped using birth control pills and attributed the change to that. One evening, I cut myself shaving (it was a really bad cut) and I bled through several large band-aids. The bleeding didn't completely stop for a couple of hours. I thought this was odd, but still didn't think too much about it.

A couple of days later, I noticed petechiae in several places on my body. That was when it hit me that something was actually wrong. I happen to work in a hospital laboratory and already had a pretty good idea what I was dealing with. As soon as my first CBC was done (platelet count of 7,500), I

was pretty sure I would be diagnosed with ITP. I was able to see a hematologist that day (December 1, 2004) and he confirmed my suspicion.

I was immediately started on high doses of Dexamethasone, followed by Prednisone. I had to check my platelet count every few days. Many lab tests were performed to help rule out any other causes of my lack of platelets. Everything was normal except for my platelet count. For the month of December, I was on a high dose of Prednisone. Within a few weeks, I knew there had to be better options.

I met with a surgeon regarding a splenectomy, but my husband really didn't want me to jump right into surgery in hopes that a less invasive treatment would work. The first week of January, I began IV infusions of Rituximab along with IV steroids. For the 1st month, the infusions were weekly, then once a month for the next 3 months. I also started IV Immunoglobulin (IVIg) in January. I went in every 7-10 days for IVIg. I did those infusions for 5 months. The IVIg worked temporarily as expected to boost my platelets, but they would drop back down before the next treatment.

By May of that year, I had had enough of IV's and went back to the surgeon. I had a splenectomy on June 7, 2005. My platelet count immediately shot up. Within a few days it was too high (> 600,000), but within 2 months, I was back at 20,000. This is when I was introduced to Dr. George and Dr. Terrell and enrolled in the AMGEN study.

I began the N-plate injections in August. It was given once a week after a platelet count was performed. It was a double-blind study, so no one knew if I was receiving the actual drug or a placebo. Every few weeks, the dose was increased, because no change was seen in my platelet count. After about 2-2 1/2 months, my count was up around 90,000. It continued to slowly increase, which was great news. After 6 months on the study, I began injecting myself and only went in to the clinic once a month to have a platelet count done and receive supplies for the next month.

This continued for 4 years while the drug company was trying to gain FDA approval to market the drug to the public. Approval was obtained sometime in 2009. In January of 2010, I was released from the study and began seeing a hematologist again to receive the N-plate injection. My hematologist initially had me come in every 2 weeks for a platelet count and injection. We gradually increased the time between visits to 3 weeks,

then monthly. By fall of 2010, she agreed to a monthly platelet count and no shot unless I fell below 150,000.

I don't think my platelet count has been below 200,000 in 3 years except for one time when I was sick and on antibiotics. I received a N-plate injection in October of 2010 and I have not had a shot since. Even though I have considered myself cured for years, I am much more aware of any bruises that I may have and I am always on the lookout for any other symptoms. Other than that, I rarely think about the illness. I continue to see my hematologist once a year and have a platelet count performed every 3-4 months.

Dr. George's Perspective on Courtney's Story

Courtney's story tells us five important lessons about ITP. First, ITP can cause persistent low platelet counts that can cause excessive bleeding. Second, ITP can be resistant to multiple different treatments. Although, splenectomy is effective in most ITP patients, resulting in normal platelet counts that last forever, in some patients – like Courtney – even splenectomy is not effective. Third, the newer treatments for ITP, the drugs that stimulate platelet production (Nplate and Promacta) can be very effective even when all previous treatments have been ineffective. So why don't we just use these agents first? Why do we use treatments like splenectomy rituximab at all? The reason for this is that splenectomy and rituximab may stop ITP; when they are effective, no more treatment is needed. Nplate and Promacta don't stop ITP; they just keep the platelet counts up. You can think of Nplate and Promacta for ITP as similar to insulin for diabetes. Insulin doesn't do anything to stop diabetes, it only keeps the blood sugar down. And like insulin, the usual experience is that Nplate and Promacta must be taken forever. But Nplate and Promacta are essential treatments for patients like Courtney. And then the fourth lesson: Sometimes ITP just goes away. ITP may seem to last forever, and in some patients it does last forever. But in other patients, like Courtney, after several years, ITP just goes away. Finally, the last and most important lesson: Courtney had good doctors; she was a very active participant in her own care; and she was lucky. Her ITP went away.

Coming Soon

ITP and Depression

In the next few months we will be sending you a survey to ask about depressive symptoms. The physical signs of ITP can vary from person to person, along with the severity of the symptoms experienced. Both symptoms and management have an impact on the well-being of patients. This can range from emotional and functional health to work life and social activities. Using a short questionnaire, we plan to survey our patients in order to document a better understanding of their physical and mental well-being. We also plan on surveying ITP patients in the United Kingdom Support Association, much like we did with the ITP and Fatigue survey in 2009. We will send this survey out this Fall with our yearly follow-ups and would greatly appreciate the return of both! Your continued contribution to our ITP Registry is what makes our research possible.

Send Your Suggestions

Is there anything you'd like to see in the next newsletter? We'd like to hear from you! Please contact us if you have any suggestions as to what you would like to see in this newsletter in the future either by emailing Dee Terrell at **Dee-Terrell@ouhsc.edu or** Kaelyn Lu at **Kaelyn-Lu@ouhsc.edu** or by calling **(405) 271-8001** extension **48386**.

Resources for ITP Patients

Visit our website, Platelets on the Web, at http://www.ouhsc.edu/platelets.

There is also an informative website from the United Kingdom you can visit at www.itpsupport.org.uk. This site includes a support group with newsletters, publications, and information on ITP. Dr. George contributes "An American Perspective" found on this page, where you can find additional topics about ITP.
www.itpsupport.org.uk/american.htm

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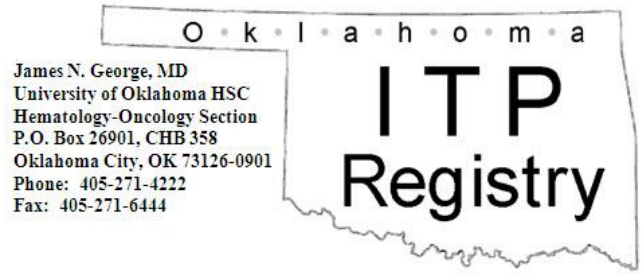

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