

The Oklahoma ITP Registry Newsletter

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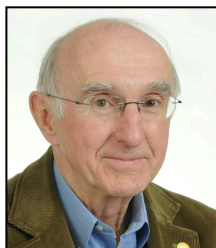
Hello!

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

The ITP Registry

The Oklahoma ITP Registry began in November 2001. Since that time we have enrolled 106 people. With your participation we hope to document the clinical course and long-term outcomes of patients with ITP

Dr. George's Perspective



What are the benefits and risks to splenectomy?

Splenectomy was the first effective treatment for ITP. Ninety-eight years after the first reports of successful treatment for ITP in 1913, splenectomy is still the most effective treatment. Two-thirds of ITP patients who have a splenectomy achieve normal platelet counts and need no further treatment for ITP. Although a few of these patients may have a recurrence of ITP some years later, most patients continue to have normal platelet counts. An additional 20% of patients will have a partial response, defined as an increased platelet count but not all the way to normal. Only 10-15% of patients do not respond to splenectomy and need additional treatments for their ITP.

Splenectomy causes a very small increased risk for infections because the spleen helps

the body's immune system to prevent infections. This can occur in anyone who has had a splenectomy for any reason (probably the most common reason is trauma causing a ruptured spleen). The risk for serious infections, described as sepsis, is extremely rare. The most serious infection is caused by the bacteria *Streptococcus pneumoniae* (called "pneumococcus") which can cause sepsis and meningitis. In my 40 years as a hematologist, I have had only one patient with a critical *Streptococcus pneumoniae* infection following splenectomy (which was not for ITP). I have learned about one other patient who had a splenectomy for ITP many years previously who had a serious *Streptococcus pneumoniae* infection causing meningitis.

What are the recommendations for prevention of infection in people who have had a splenectomy?

Recommendations for prevention of infection after splenectomy are different for children and adults. In England, daily penicillin may be recommended until age 16, or sometimes for life. In the US, daily penicillin is only routinely recommended for children until age 5, or for at least 3 years. Some *Streptococcus pneumoniae* have become resistant to penicillin, so antibiotics other than penicillin are now commonly prescribed.

I think that these 3 rules are the best and safest thing for adults to do.

1. Immunizations

Before surgery to remove the spleen, you should have received 3 immunizations:

1. Pneumococcal vaccine for *Streptococcus pneumoniae*. You should repeat the pneumococcal vaccine every 5 years.
2. *Hemophilus influenzae* type b vaccine (this is the routine vaccine for children, called Hib)
3. Meningococcus vaccine

The Hib and meningococcus vaccines only need to be given once. In addition, patients should take the annual influenza vaccine ("flu shot").

2. Keep antibiotic tablets at home and with you when you travel

The current best and cheapest antibiotic is amoxicillin-clavulanate (its brand name is Augmentin). The dose is one 875 mg tablet twice a day. I recommend that you keep 4 tablets at home or with you when you travel (a 2 day supply). Four (generic) tablets cost less than \$20.00. The label will say that they are good for a year; they should be good for at least 18 months. Then you need to refill the prescription to be sure that the antibiotic is effective. Call us if you need a prescription for the amoxicillin.

3. If you have symptoms that may be the first sign of serious infection: fever of 101° or higher, especially if you also have chills, headache, neck ache, nausea and vomiting:

- Take one amoxicillin tablet immediately
- See a doctor immediately and tell him/her that you have had your spleen removed. The 4 amoxicillin tablets you have allow for a delay in seeing a doctor, but do not delay.

These rules are for safety. My experience is that you may never need these antibiotics. But these rules are the same as other safety rules, like wearing seat belts when you drive and a helmet when you ride a bicycle.

Patient Stories: Christy's Story

The following story is from one of the patients in our registry. She was selected to tell her story because she has important messages for all patients with ITP. Her name was changed to protect her confidentiality.

Christy was eleven years old when she was first diagnosed with ITP. This is an update to the complete story which is posted on our website (<http://www.ouhsc.edu/platelets>).

For the first two years, after we last wrote about Christy in November 2006, she did remarkably well. She had no major bleeding episodes- only mild, scattered bruises or petechiae. Her only serious bleeding episode since we last spoke was in November and December of 2008. She describes the episode as "by far the worst bleeding that I've ever had." It started about a month before Christmas when she started having multiple episodes of heavy vaginal bleeding. She had three visits to the hospital and required six blood transfusions and three platelet transfusions during this time. Christmas came, and she spent most of her time in the house feeling too weak to get out. The day after Christmas, she was so tired of being stuck in the house that she decided to go with her husband to feed their cows. Her only excursion outside of the truck they were riding in resulted in her feeling so light-headed that it wasn't long until she went back to the truck to rest. The next thing she remembers is waking up to her frantic husband shaking her, screaming at her, and threatening to start CPR. He drove her to the hospital where her hemoglobin was extremely low at 4.8, well below the normal of 12g/dl, the result of continued vaginal bleeding. Her platelets were less than 6,000, too low for the machine to accurately count. She stayed in the hospital for 3 days where she received 6 more blood transfusions and IVIG. Even with premedication of Benadryl and steroids, she again suffered from the torturous side effects of headaches and nausea from chemically-induced meningitis that

IVIG infusions can cause. Eventually her platelet count rose to above 100,000 and her bleeding slowed to a manageable rate. To stop her bleeding completely her doctors recommended a hysterectomy. She wasn't ready for this yet so the next option was high-dose birth control pills to stop the bleeding and regular birth control pills to control future bleeding. Christy's periods have been controlled since then and she's had no more serious bleeding and no more need for blood transfusions or IVIG in the past 2 years.

Over the years, Christy has been asked to participate in various clinical trials of new drugs to treat ITP. In recent years she has declined to participate in any because she thought she would want to have another child. Recently, she and her husband have decided that they are blessed to have their daughter, and given her trouble with menstrual bleeding, they have decided to not have any more children. Therefore in January of 2010 she agreed to be part of a trial involving a drug that stimulates platelet production (romiplostim- the brand name is Nplate). At first she was very optimistic that the drug would help her because it has had such high success rates in other patients, even patients with very prolonged, severe low platelet counts like Christy has had. She was also intrigued by how few side effects patients were experiencing. She began making the trips to Oklahoma City every few weeks, with her husband along so he can take her out to eat when she is done with her appointment. Initially her platelet counts went up to as high as 62,000, but gradually, even with increasing dosage of the drug, her platelet counts have dwindled back down to around 2,000. In hindsight, she still thinks that participating in the drug trial was worth it, just on the chance that it might have worked. She would like to tell other ITP patients to do their homework when thinking about starting a new drug or participating in a drug trial. Find out the benefits and side effects. About participating in drug trials: Christy's advice is to do it because there is not a whole lot to lose and there is so much that might be gained.

Christy continues to live her life without hindrance. She is very active, even enjoying Tae Kwon Do with her nine year-old daughter, who has no signs of ITP. She has also been known to take a skiing trip with a platelet count of 7,000 (though she "forgot" to tell her doctors about the trip ahead of time). She continues to only think about ITP when she is symptomatic. She states it may be in the back of her mind at times, but she doesn't focus on it because it can "drive you crazy worrying about it." She doesn't want to let ITP control her life and refuses to live in fear of a low platelet count. Christy believes as long as she is happy and doing alright, she is content to live with ITP.

Comments by Dr. George about Christy's story:

Christy is a wonderful person. I think she knows as much about ITP as anyone, hematologists included. She and I recently gave a talk on ITP at her hospital. My part was the usual doctor material on ITP; Christy's part was terrific. For true education about a chronic health problem, there is no substitute for learning directly from a patient. The important messages in Christy's story are that you can have a happy, successful life, even with severe thrombocytopenia and even with occasional episodes of severe bleeding. Christy is calm and sensible, and I assume she was also calm and sensible as a teenager, years before I first met her. Her determination to have a normal active life is only normal. What are the alternatives? Only worse. Christy's calmness comes through in her description of interactions with doctors who were unfamiliar with ITP and alarmed by her severe thrombocytopenia. Most patients with ITP experience this sometimes. Her calmness was also apparent in the story of her participation as the first human subject to be treated in a phase I clinical trial of a new agent for the treatment of ITP. (This agent was effective in a few other patients with ITP, but its development was discontinued.) Participation in clinical trials is very important for patients with a chronic illness that has no effective treatment, such as ITP. The patient may not benefit, but the process is critical for progress to effective management. The process can also be very educational and supportive for the participating patients. This has been Christy's rationale for her participation in the current clinical trial with romiplostim. Although Christy's platelet count has not increased with romiplostim treatment during this clinical trial, I still think her participation in the study is important for her, as I think participation in clinical trials are important for all patients. Christy has had severe bleeding episodes in the past and even though her platelet count has not increased with romiplostim, I think that it is probable that the increased platelet production that is caused by romiplostim provides more platelets whenever there is some trauma or risk for bleeding – even though the platelets don't remain in the circulation and get counted. Also the continuous evaluations over months and years provide a very clear picture of Christy's ITP. This can be important information whenever future treatments may be considered.

Of course Christy was selected to tell her story because hers is a happy one – but it also includes some scary episodes of severe bleeding. Some patients with ITP suffer much more, but too often the severe problems receive the most attention. Christy's experience is common. Her most important message is optimism. Her story is even more optimistic than when we first recorded it in 2001. Although she has times when her platelet count was higher with an apparent spontaneous remission, and now her platelet count is very low again, Christy is perfectly comfortable with her life. People around her would never

know that she has a significant health problem. Christy's story may seem exceptional but I think it could also be a typical story of a young person with ITP.

ITP² – Initial Treatment of Patients with ITP

There are many clinical trials for patients with ITP and we just wanted to tell you about a study that our office is conducting.

The Initial Treatment of Patients with Immune Thrombocytopenic purpura is called ITP² (ITP squared) for short. It is a large clinical trial that is conducted at many hospitals across the United States including OU Health Sciences Center. A clinical trial, in this case, is a study where patients who have ITP are assigned to be in one of two treatment groups. The patients do not get to choose which group they are in because they are assigned to their group randomly (like flipping a coin). They also will not know which group they are in.

Depending on what group the patient is in, they will either receive [1] the normal treatment for ITP, which is a course of steroids (prednisone) or [2] a new treatment, which is a course of high dose steroids (dexamethasone). Researchers want to compare these two treatments because it is thought that patients who take high dose steroids might recover from ITP faster and might have a longer period before their symptoms of ITP come back (if the symptoms ever do come back). Also, high dose steroids might have more side effects including how unpleasant a patient feels. Therefore, the researchers want to compare the amount and severity of the side effects between the two groups.

In order to be in this study the patient:

1. Must be 15 years old or older.
2. Must have had a platelet count of less than or equal to 30,000/ μ L at some point since their diagnosis of ITP.
3. Must have had a diagnosed ITP for less than 30 days.

If anyone you know has recently been diagnosed with ITP (less than 30 days ago) then please feel free to tell them about this study and tell them to contact our office for more information regarding this study (contact information is on page 7).

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 Jessica Reese at **Jessica-Reese@ouhsc.edu** or calling at **(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

Contact Information

Phone: (405) 271-4222

Mailing address: James George, MD
Attention: ITP Registry
OU Health Sciences Center
Hematology-Oncology Section
P.O. Box 26901 CHB #237
Oklahoma City, OK 73126

Website: <http://www.ouhsc.edu/platelets>

