

The Oklahoma ITP Registry Newsletter

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Hello!

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

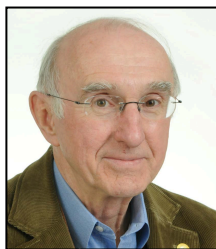
The ITP Registry

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.

Follow-Up Reminder

Thank you to all of you who have returned your follow-up surveys! We just wanted to remind those who have not returned their follow-up surveys to please do so. If you need a new survey or would like to do your survey over the phone, please contact us (contact information on page 7).

Dr. George's Perspective



Can depression be a side effect of steroids?

Yes, it can. People on prednisone commonly experience mood changes. Some have severe mood swings; others experience depression or even euphoria. Some people can take steroids and they have no effect on their mood although probably most patients who take steroids notice some mood effect. The higher the dose of steroids, the greater the risk of depression and potentially the more severe the problems. Also, the longer

the duration of steroid treatment, the greater the risk. Some people who take “pulses” of steroids – high doses for a short time, such as 3 or 4 days – may feel like they’re on stimulants while they’re taking steroids and then when the steroid dose is quickly tapered and discontinued, they may feel very depressed, blue, and tearful.

The most important things you can do about this are: [1] Appreciate that it can happen, so you don’t become alarmed by the strange feelings. [2] Tell your doctor about it, so that she can adjust the steroid dose to the minimum possible for the shortest possible time. Doctors often do not appreciate how much their patients are bothered by steroid side-effects. [3] Ask for understanding from your family. [4] Rarely is the depression so severe that another medicine has to be added to help correct the side effects of the steroid medicine. That can become very complicated.

Are people with ITP more prone to depression?

No, not that I have ever heard of. We have recently documented that patients with ITP have more problems with fatigue than the normal population, and symptoms of fatigue may seem like depression. More severe fatigue may actually limit what people can do and cause some anxiety and frustration. But I don’t think it’s appropriate to say that people with ITP are more prone to depression.

What does “being in remission” mean?

This is doctor talk. “Remission” means that there are no signs of the disease. It is often used in cancer patients, when the cancer shrinks and disappears and cannot be detected by any test. In ITP, “remission” means that the platelet count is normal. In all of these situations, the word “remission” is used because it would not be right to use the word “cure”. So “remission” is used when it seems like a cure and looks like a cure, but it’s possible that the disease may come back. In ITP, a “complete remission” means that the person has a normal platelet count and no longer needs any treatment. The longer a remission lasts, the more confident you are that it may indeed be a cure. How much time must pass before you can change the word “remission” to the much nicer word “cure”? This is not known. That’s too bad. But be happy with a remission, and at some time you’ll wake up and realize that you haven’t even thought about ITP for the past several days. That’s probably when you can begin to use the word “cure”.

Patient Stories: Rosie's Story – 2010 Update

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.

Rosie was a 46 year-old mother of three when she first started noticing her symptoms of ITP. To view Rosie's complete story, including her struggles with the treatments for ITP, please visit our website (<http://www.ouhsc.edu/platelets>).

Rosie reports that she has been doing great since our last update in 2006. She has been healthy and has not had any new medical issues arise. Rosie continues to get her platelet count annually, and her platelets have continued to stay in the normal range. Rosie states "it's hard to believe it's been close to ten years since her diagnosis of ITP, but she is thankful that she has not had any complications."

Rosie continues to get bone density testing every other year to monitor her bone density loss. With our last update in 2006, Rosie was taking bisphosphonates to help slow bone loss and increase bone mass to prevent bone fractures. Two years ago, Rosie started to have pain in her hip. Side effects of bisphosphonates can include bone, muscle, or joint pain. With this new symptom, her doctor discontinued the bisphosphonates and started her on calcium supplements to continue to increase and maintain her bone mineral density. Rosie reports her last bone density test two years ago was better than those done in the past. She is due for her next bone density test this upcoming January and hopes the results will be good!

The heavy menstrual bleeding which Rosie was dealing with at the diagnosis of ITP is officially a thing of the past! Since her endometrial ablation in January 2006, her periods have been lighter and is what Rosie believes is normal for normal women in talking to her friends. Despite her being around the age of menopause, Rosie is still having her periods every 6 weeks which she is happy for. Rosie states her doctor explained to her "as long as she is having her periods, the better her bone density status will be." For women, bone mass steadily increases during childhood and adolescence and reaches a plateau in the 20's. Women are susceptible to increased bone loss due to the decrease in estrogen levels which occur during menopause which can accelerate progressive bone loss. The steroid medications could also have contributed to bone loss while Rosie took them

10 years ago. For now, Rosie will continue on the calcium supplements and continue with the bone density checks every other year.

Overall, Rosie is doing fantastic and enjoying life. Her family has continued to grow, as her daughter recently had another baby, adding a fifth grandchild. Rosie keeps busy with her family and friends and is happy for ten years free of any ITP complications.

Comments by Dr. George about Rosie's story:

Rosie's story contains many important lessons for patients with ITP.

First, the onset of her symptoms of excessive bleeding and bruising were gradual and easily ignored. Rosie felt perfectly well; ITP causes no symptoms except for increased bleeding and bruising. The only function of blood platelets is to prevent bleeding, and having too few platelets increases the risk for bleeding. That's all. Rosie's gradual onset of bleeding symptoms is typical for adults with ITP, but different from the onset of ITP in young children. Childhood ITP usually occurs before age 6 and the beginning is usually abrupt; an active healthy child is suddenly covered by bruises and petechiae. Sometimes a day-care aide, school teacher, or nurse may suspect child abuse; this is a wrong impression that creates extreme stress. Childhood ITP is also distinct from ITP in adults because most children will recover without any treatment, within several weeks to several months. In adults, ITP is typically a long-term illness.

Second, the diagnosis is usually straightforward, even though there are no laboratory tests that specifically document ITP and exclude other causes of a low platelet count (thrombocytopenia). In an otherwise healthy person who is taking no medicines or health food supplements, whose routine physical examination is normal (except for possibly bruises and other signs of bleeding), and whose routine laboratory data are normal except for a low platelet count, ITP is the appropriate diagnosis. No further evaluation is required. Signs of bleeding are typically on the skin and the mucous membranes of the nose and mouth and excessive menstrual bleeding - which was so dramatic in Rosie. Skin bleeding is described as "purpura" (bruises, often occurring without any remembered bumps), "ecchymoses" (also a term for extensive bruises), and "petechiae" (the tiny red hemorrhagic dots caused by leakage of blood from a small blood vessel, leaks normally sealed by platelets).

Perhaps the most important part of the laboratory evaluation is the direct examination of the blood cells. This is done by preparing a thin film of blood on a glass slide, staining the blood film with a mixture of dyes that develop specific colors for each type of blood cell, and examining the stained blood film with a microscope. In ITP, the blood should be normal except for the decreased number of platelets. The red blood cells and white blood cells are normal in number and appearance. The remaining platelets are normal in size; the presence of giant platelets suggests the possibility of an inherited problem of platelet production. Occasionally, perhaps in 1 of 1000 healthy people, platelets clump in the tube used to collect blood for routine counts, and the reported platelet count is falsely low. Without examination of the blood film to actually see the clumped platelets, this harmless occurrence can be mistaken for ITP.

If all of these observations and results are consistent with the diagnosis of ITP, a bone marrow aspiration may not be necessary. If there are unexplained abnormalities of the red blood cells or white blood cells, a bone marrow aspiration is appropriate to be certain that the production of blood cells is normal. In patients with ITP, the bone marrow examination is normal. Although an important cause of the low platelet counts in ITP is increased destruction of circulating platelets, the bone marrow production of platelets is often also insufficient. Normally the bone marrow should increase platelet production to compensate for the more rapid destruction. In patients with ITP, platelet production is often not as effective as it should be.

The third lesson is Rosie's experience with a severe headache at a time when her platelet count was very low. This is the most feared symptom for all patients with ITP. Although the ER doctors in her small community hospital had no experience with ITP, their management was exactly correct: urgent platelet transfusion and IV steroid (IVIg would also have been given if it had been available) followed by a CT scan to diagnose or exclude brain hemorrhage.

The final important lesson from Rosie's story is her experience with the treatment. The story of Rosie's treatment is familiar to most all patients with ITP. Prednisone pills or an equivalent steroid preparation are always the initial treatment for adults - inexpensive and simple to take. Most patients respond with an increased platelet count, but Rosie responded only incompletely and transiently. In most patients, thrombocytopenia recurs when the prednisone dose is decreased. The side effects of the steroids are not immediately dangerous, but they can be devastating. Rosie's

experience is common, and it is echoed in the experiences of several other patients in this series of Patient's Perspective Stories, both with ITP and TTP.

It is an important lesson for physicians that patients often feel that the treatment of ITP is worse than the disease itself. Since the goal of the treatment is not to cure ITP, but only to prevent the risk for major bleeding, patients whose platelet counts are not severely decreased may be better off with no treatment - managed only with reassurance and careful follow-up. The key phrase in this recommendation is: "...whose platelet counts are not severely decreased". How low can a platelet count be without causing risk for dangerous bleeding? In an otherwise healthy young person, platelet counts as low as 10,000 to 20,000 may be safe. Therefore it is commonly recommended that patients with ITP and platelet counts greater than 30,000 (to provide an additional margin of safety) may not require treatment. But older patients who may have other diseases, such as high blood pressure, which can increase the risk for bleeding in the brain, may need higher platelet counts to provide safety.

Rosie's experience with treatment documents (1) the emotional disturbance caused by steroids, (2) the optimum response with the first dose of anti-Rh(D) but only a brief response to the second dose, (3) the response to IVIg but also the common side effect of a severe headache and neck stiffness, symptoms which can cause alarm because they may suggest brain hemorrhage, and finally (4) what appears to be an excellent response to splenectomy. Typically excellent responses to splenectomy occur promptly after surgery, within two weeks. Rosie's delayed response caused concern, but now her platelet count is normal. We hope it will remain normal, and it most likely will, but there is always a chance that her ITP will return.

Since 2001 there are new treatments for ITP. Although the sequence of treatments Rosie received is still the most common treatment plan today, rituximab is now also used for patients who do not have a durable platelet count response to initial steroid treatment. Rituximab is a relatively safe drug that suppresses the antibody that is destroying platelets. It is not as effective as splenectomy, but perhaps one-third of patients achieve durable remissions. New medicines that increase platelet production in patients with ITP are now available: Nplate (romiplostim) and Promacta (Eltrombopag).

The most important use of these new drugs is for the patients who fail to achieve a safe platelet count with splenectomy. As well demonstrated by Rosie, splenectomy – which was the first treatment for ITP over 80 years ago – remains the most effective treatment for ITP. Although there may be

risks for infections and blood clots after splenectomy, these are rare. Most patients are just like Rosie – their platelet count recovers to normal and stays there.

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

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