

The Oklahoma ITP Registry Newsletter

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Hello!

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

The ITP Registry

The Oklahoma ITP Registry began in November 2001. Since that time we have enrolled 116 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



Is childbirth dangerous for me? No. Women with ITP have no greater risk with childbirth than other women.

Even if the platelet count is very low at the time of delivery, bleeding can be controlled with proper care, which may involve a few days of treatment with corticosteroids, or with IVIg, or maybe even a platelet transfusion at the time of delivery. But these treatments are rarely required. A platelet count of over 30,000 is sufficient to prevent excessive bleeding at delivery.

Is ITP likely to return in pregnancy? It is possible, but it is uncommon. Platelet counts gradually become a little bit lower in all women during the course of a pregnancy, and then return to their usual number in the weeks after delivery. In a woman who has had ITP, platelet counts may fall a little more, and may even fall enough for treatment to be considered (that is, less than 30,000). In some women, ITP is first noticed during pregnancy, and then the platelet count increases after delivery and no further treatment is needed.

Will my baby have a low platelet count? This is a very important question, and your obstetrician must be aware that you have ITP so that your baby can have her platelet count checked right when she is born. This platelet count can be done from blood taken from the umbilical cord. Your obstetrician should arrange for a pediatrician to be aware that your baby may have a low platelet count. These are the necessary precautions, but 90% of babies will have normal platelet counts and no problem. In the other 10%, the platelet count may be low because the antibody in your blood (what destroy your platelets) can cross into the baby's circulation and also destroy her platelets. We usually only see this when the mother has had very severe ITP, such as requiring a splenectomy. If your baby's platelet count is low at birth, then the platelet count needs to be repeated every day until it begins to increase toward normal. Because it is the mother's antibody that is causing the low platelet count, your baby may have a low platelet count for several days or weeks. Although as many as 10% of babies may have low platelet counts, maybe only 1% are low enough that the baby needs any treatment (such as IVIg).

Is ITP hereditary? In all of our experience, we know of one mother and daughter who both have had ITP, and both required multiple treatments including splenectomy. We believe that this is like other autoimmune disorders that seem to have increased risk within families (such as lupus, thyroid disorders, and others). Therefore the answer to this question is that ITP can be hereditary, but it is very, very rare.

Colton's Story

The day started out just as any other day. I was working, and my kids, 2 year old Cheyenne and 1 ½ year old Colton, were at daycare. I received a call from our daycare director – a call that would be the end to my son's "normal" life. She informed me that they found a "really suspicious" bruise on Colton's thigh, and due to the location and size, she had to call child protective services. My husband and I were so upset. We didn't harm our son!

I had, however, noticed a lot of little fingertip sized bruises on him over the past week or so, and had begun to get a little worried about him. The bigger bruises, oddly enough, hadn't concerned me as much; Colton is an accident prone toddler! He is known for his activeness, and rarely slows down. I knew he probably just fell over something or ran into something. So, after receiving that phone call, I made him a doctor's appointment, feeling slightly silly to make an appointment over a few bruises.

That evening, a DHS worker came to our house. They scrutinized our house and our disciplinary actions towards our children. We had to make a list of people willing to testify that my husband and I were good parents. Humbled and humiliated, I contacted friends and family to explain our situation. I will never forget that feeling of wondering "What if they *do* think I'm not a good enough mom? What if they don't *want* to vouch for me?"

The next day, Colton had his doctor's appointment. We got his blood results – his platelets were 9. Normal is 150-350! The doctor said Colton had an autoimmune disorder; a long scary name that I couldn't pronounce, but which I would later come to know very well. The doctor told me to pick Colton up, not let him walk for fear of him falling, and take him straight to a hematologist. He said this explained the bruising, and that he could have a brain hemorrhage since his blood wouldn't clot. I was terrified!

Throughout the next several months, my son was poked, prodded, and given all kinds of treatment. We went through WinRho, IVIg, and steroids. Nothing really raised his platelets very much. My poor son went through a lot of side effects, including dramatic weight gain, rage spells, and trouble sleeping. His platelets averaged between 20-50. They got as low as 3 one time. I hardly recognized my son, and the treatments weren't even working! Finally, we decided to stop all treatments and just watch him. His

platelets hovered between 25-30, but he was back to being my sweet toddler again.

Over a year later, Colton is now considered chronic. It is considered extremely rare for a child as young as him to move into the chronic stage. Though we had hoped and prayed that it would resolve, we consider ourselves incredibly blessed! His platelets now stay around 50 and we have not used any medication in about a year. He is still incredibly active, and never acts like anything is wrong. He is so used to our hematologist visits that he asks for his nurses by name. He doesn't cry when he gets his blood drawn; he just smiles and asks the nurse for his band aid and a hug. The nurses adore him.

Do I worry about him? Do I worry about his future? Whether he will feel left out when he can't play contact sports with his friends? What kind of job he might be able to do? Yes, of course I do. But, we all have challenges in life. We can let them break us and rule over our life, or we can choose to see the good and live life to the fullest.

My son has a rare blood disorder. We don't let that shape him. He is also a sweet, loving, mischievous 3 year old that charms everyone he meets. He is full of life and love. ITP or no ITP, it does not change who he is, and we know that God has blessed us beyond measure.

Treatment Options for Childhood ITP

With this newsletter we focused on ITP in pregnancy and in children; therefore, we wanted to review some current literature on ITP in children.

Here is a short review of *Childhood immune thrombocytopenia: role of rituximab, recombinant thrombopoietin, and other therapeutics* (Journeycake JM. Hematology Am Soc Hematol Edu Program. 2012;2012:444-449.).

Childhood ITP is defined as a platelet count less than 100,000, not explained by any other cause. As in Colton's case, often children have bruising and/or bleeding from their gums. The good news is that usually childhood ITP goes away on its own without needing treatment. Serious and/or life threatening bleeding is rare. In fact, 90-95% of childhood ITP will spontaneously go into remission (defined as a normal platelet count requiring no further treatment) within 12 months. Although, new treatment options are currently being investigated to offer relief from bleeding

symptoms and to increase the quality of life, careful observation is the preferred therapy of choice for the majority of children with ITP.

The decision to treat children with ITP should be based on the patient's entire health status, striking a balance between platelet counts, bleeding symptoms, whether the child's family will be able to closely monitor them, and how treatments might affect the child's quality of life. If a child with persistent severe bleeding symptoms still fails to respond to traditional doses of steroids, IVIg, or anti-D, or is classified with chronic ITP (platelet count less than 100,000 for more than 12 months), hematologists (in combination with parents) may need to determine whether a more aggressive therapy is warranted to resolve the bleeding. Here is a summary of treatment options:

- **No treatment**-This is still an option. In fact, many children with chronic ITP will have no serious bleeding and just need an additional 12-24 months before their thrombocytopenia will go away on its own.
- **Splenectomy**- Remains the most reliable course of action to achieve a durable remission in ITP, as it removes the primary site of platelet destruction and antiplatelet antibody production. The risks involved include the unpredictability of responsiveness to the surgery. Although it is considered a relatively safe and effective treatment, it is also irreversible and places children at a higher risk for surgical and infectious complications. All vaccines should be administered prior to surgery with subsequent booster immunizations every 5 years.
- **Rituximab**- Not currently FDA approved for the treatment of ITP, but has been used as a treatment for more than 10 years. Initially about half of children respond, but in many of the children who respond, their thrombocytopenia comes back after several years. There are no current predictors (age, sex, previous therapies) of determining whether or not your child will respond to Rituximab. Risks of treatment include fever and risk of infection like pneumonia which follows administering of an immunosuppressive agent.
- **Nplate and Promacta**- have been studied and approved for treatment of chronic ITP in adults who have failed to respond to other methods of treatment (steroids, IVIg, splenectomy).

In children there has been 1 clinical trial with Nplate and 2 with Promacta. In these trials both treatments were effective however more clinical trials are currently underway. It may be that these newer agents can benefit children with ITP that are trying to delay or prevent having a splenectomy. Treatment with Nplate or Promacta can give the hematologist or parent that additional 12-24 months to see if the child will have a remission.

While childhood ITP is considered self-limiting and spontaneous remission is generally achieved, there are multiple options for those with chronic and severe bleeding. The choice to seek treatment for children should be a collaborative decision that takes into account the family's overall lifestyle and the risks and benefits that accompany each treatment option.

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.osuhsc.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

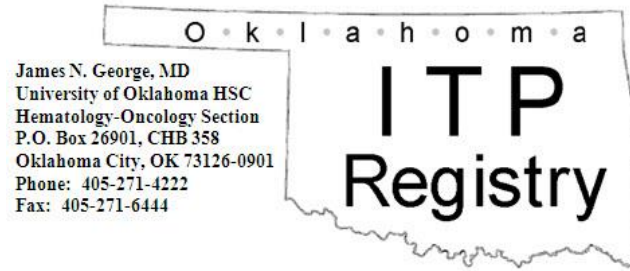
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