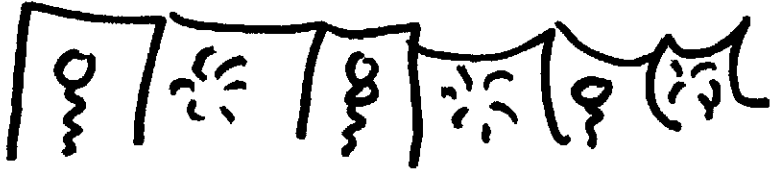


What's It Called Again?



Answers to the most commonly
asked questions about
Immune and Idiopathic
Thrombocytopenic Purpura (ITP)
in children

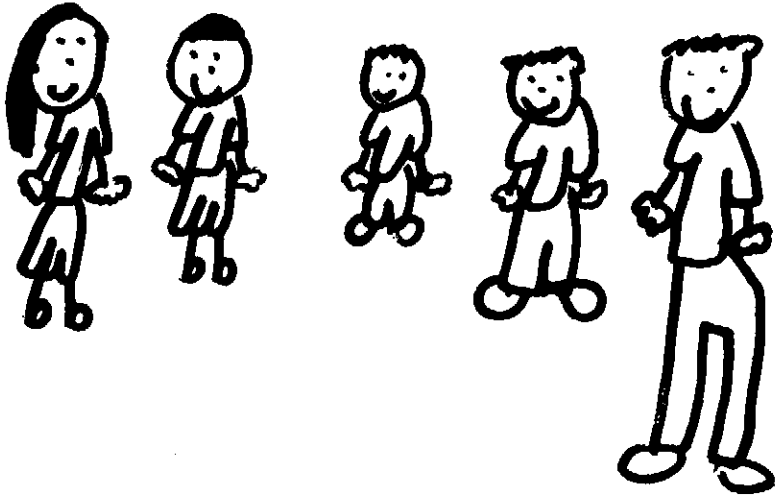


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This booklet was prepared by the parents of a child who has ITP. Their goal was to provide answers to many of the questions that come up after a child has been diagnosed with ITP. Especially because medical information is ever-changing, this booklet is not meant to replace discussions with your child's hematologist or other health professionals—simply to help you and your family cope with the ups and downs of the ITP "roller coaster". We hope you find it helpful.



What is ITP?

ITP is a disorder of the blood that involves the immune system. ITP stands for Immune Thrombocytopenic (a decrease in blood platelets) Purpura (purplish areas of the skin and mucous membranes). You may also hear it referred to as Idiopathic Thrombocytopenic Purpura which means the cause of the low platelet count is unknown. As far as experts know, ITP occurs when a person makes anti-platelet antibodies which attach to his or her own platelets. The body's immune system eliminates the platelets because of the antibodies on them, thinking that they may be bacteria. This results in a decrease in the platelet count, or thrombocytopenia.

Why are platelets important?

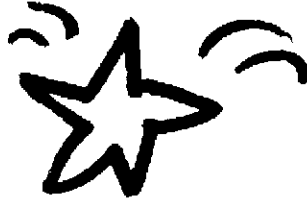
Platelets are the microscopic parts of our blood that help stop bleeding and heal bruises by promoting clotting.

What's a normal platelet count?

The normal range for a child is 150,000-400,000 platelets per microliter.

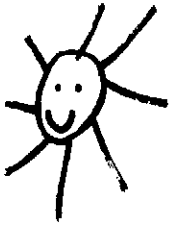
How did my child get ITP?

If your child is otherwise healthy, chances are he or she has ITP as a result of a recent viral infection. This is the situation in about one-half of the cases of ITP in children.



Some cases of ITP are the result of medication, other infections, or are associated with immune



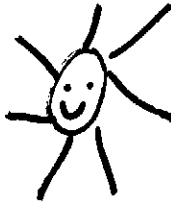


disorders such as systemic lupus erythematosus, HIV infection, or Epstein-Barr virus. Infants can develop ITP as a result of ITP in their mother during pregnancy, premature birth, and /or a number of other syndromes, but these cases go away rapidly.

How can I be sure it's ITP?



If there is any doubt about your child's diagnosis a hematologist (blood specialist) may ask that a bone marrow test be done. This involves taking a sample of the liquid part of a pelvic bone in the back. This test may also be done to rule out leukemia.



Is it contagious?

No. And most children who may have had the same viral infection will not develop ITP.



Will it go away?

In most cases, yes. The vast majority of ITP cases in children are "acute" or temporary. Statistics show that 85-90% of children with ITP recover (to normal platelet counts) within one year—most even sooner than that. In some children ITP does not go away. This would be termed "chronic" ITP. Medical professionals differ on the time frame before ITP is considered "chronic". Some call it chronic if it lasts more than six months, others after one year.



How is ITP treated?

Treatment of ITP varies.

Medicines are stopped if they are known to be the cause. Any infection that may result in ITP would be treated as vigorously as possible.

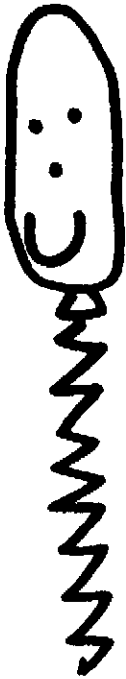


Children with no actual bleeding and a stable platelet count above 20,000 to 30,000 cells per microliter may receive no treatment. This is in hopes that the child's body will rid itself of the antibody and therefore, ITP. Children in this situation can lead an almost totally normal life.

For those children who require treatment, a hematologist will most likely prescribe one or more drugs that change or suppress the child's immune system. The thinking behind this is to "trick" the body's spleen and liver into NOT removing or destroying the platelets. Since fewer platelets are eliminated, the child's platelet count should rise.

The drugs and therapies currently used to treat ITP are:

- Corticosteroids or "steroids". High doses of these drugs over a short period of time are thought to slow the removal of platelets by the spleen and liver and temporarily lower the level of anti-platelet antibodies. This class of drugs may also stabilize the blood vessels, lowering the risk of bleeding. Steroids may be prescribed in pill form or given intravenously (through the veins).
- Intravenous Gamma Globulin. (IVIG, IgG, or GammaG) This substance may be used separately or in combination with steroids. IVIG is given intravenously over a course of 2-4 hours for a period of 1-5 days. This product is prepared from blood plasma and is thought to block the antibody that results in destruction of the platelets.
- IV-Anti-D. Recently licensed for the treatment of ITP, this form of IVIG is prepared from the plasma of a smaller, more specific group of donors. IV-Anti-D is given intravenously and functions like IVIG, acting to provide temporary increases in the platelet





count. It is effective only in people who have a positive blood type (i.e. A+, B+, O+) and have not had their spleen removed. Indications are that IV-Anti-D is particularly useful for children who are attempting to keep their platelet count at a safe level while waiting for their condition to improve.

Why treat at all?

None of these treatments cure ITP. The main reason to treat a child with ITP is to keep him or her out of danger—that is, to keep the child's platelet count in a range considered to be safe until the body cures itself. That range is considered by many experts to be greater than 20,000-30,000 cells per microliter. Platelet counts below 10,000 cells per microliter enormously increase the risk of injury-related or spontaneous internal bleeding and/or bleeding on the brain (intracranial hemorrhage). Either of these conditions can be life-threatening for a child; as a safeguard, most experts will treat children at 20,000 - 30,000 cells per microliter or below.

Are there risks or side effects to these treatments?

- Steroids, as they are used in treatment of ITP, can cause mood changes ranging from euphoria to irritability, weight gain, puffiness in the face and/or back of the neck, and irritation of the stomach. Menstrual periods may also be affected. Patients on steroids may also experience an increased risk of infection, thinning of the skin, calcium loss from bones, kidney stones, high blood pressure, pancreatitis, and other side effects. Limits on dosages and duration of steroid use make these less likely.





- Patients who receive IVIG may experience temporary headaches, nausea, light-headedness, or a slight fever. On very rare occasions, a patient can be severely allergic to IVIG. This substance is derived from human blood, but requires a "virus inactivation" step in processing and is therefore thought to be as free of blood-borne diseases as is possible today. IVIG is significantly more expensive than steroids.

- IV-Anti-D is a type of IVIG but comes from a much smaller group of donors. It can cause temporary anemia, and occasionally, fever and chills 1-2 hours after infusion. It is less expensive than IVIG and can be given in less than an hour.

- Any of these treatments involve needle-sticks and therefore pain. A topical cream can be used to temporarily numb your child's hand or arm. This often helps minimize the pain and fear of needle-sticks and IVs. This anaesthetic cream is available through your child's health care professional.

What if these don't work?

For a few children, these treatments will not cause even a temporary increase in platelet count. If this is the case, your hematologist will likely test for a rarer cause of your child's low platelet count.

For others, maintaining a safe platelet count is impossible without on-going treatments. If this is your child's situation, your hematologist will work with you to determine what the next step will be, based on the risk of serious bleeding and other factors. If serious bleeding is a risk, a splenectomy may be recommended.



What is a splenectomy?

It is the surgical removal of the spleen. Splenectomy is the only treatment known to cure ITP, and works for 70-90% of the people who undergo it. It is major surgery, and carries with it a life-long increased risk of serious infection. Splenectomy is usually not used unless the ITP has persisted for more than one year, and the child is 5 or older. It is a serious step and needs to be discussed at length with your child's health care professional.

If splenectomy is chosen, your child will be given medicines before, and for at least one year afterward to strengthen him or her against infections. In the years that follow, any fever over 102 degrees must be treated with antibiotics on an emergency basis as a precaution.

Why the spleen?

The spleen is one of the body's organs which works like the filter in a carburetor—removing wastes and "foreign" bodies. Normally, platelets pass through the spleen, but with ITP the spleen filters out the antibody-coated platelets "by mistake." This organ is also thought to be where some of the anti-platelet antibody is made.

What if it is chronic ITP?

ITP is considered by most to be chronic if it lasts more than six months, though some say after twelve months. In children ITP may still go away by itself after this length of time. There is no difference in the disease itself if it is chronic. The difference is in the management of the disease. The goal of the treatment becomes keeping your child's life as normal and risk-free as possible.



What does that mean?

Assuming your child is otherwise healthy, this basically means frequent blood tests to track his or her platelet count and treatments when the numbers are in the range that is risky. You can also watch for visible signs of a low platelet count, such as excessive bleeding, bruising, the small red pin-dots (petechiae), and other typical symptoms of ITP.

The actual platelet level at which you take your child for treatment is something to discuss with the hematologist or other health professional. This decision should take into consideration your child's activities and lifestyle among other factors.

Do I need to restrict my child in any way?

That depends on your child's platelet count and how ITP affects his or her bleeding and bruising.

Most experts advise restricting contact and competitive team sports at platelet counts of 30,000-50,000 cells per microliter or lower.

This is to keep the risk of internal injury and bleeding as low as possible.

However, most other sports and activities are considered okay. In general, it is important to encourage normal activity and participation. You can ask your child's health professional for more specific guidelines.



Your child should not take aspirin or aspirin related products such as ibuprofen.



Who else needs to know about ITP?

Because of the risks associated with ITP, you should make other medical professionals aware of your child's ITP. This would include your child's pediatrician, dentist, and any emergency staff who may treat your child. School and sports personnel and care-givers should be aware of the condition and any restrictions to your child's activities. The risk of excessive bleeding or bruising and internal bleeding, though rare, makes communication very important.

Does ITP make my child more susceptible to other illnesses?

Some people wonder about this since ITP involves a child's immune system. If your child is otherwise healthy, he or she is no more likely to catch colds and viruses than other children.

Does ITP affect my child's life span?

By itself, ITP is not known to change a person's life-span.

OK. So what's it like to deal with ITP?

You're likely to experience a whole range of emotions:

- Relief—that it wasn't a more serious illness
- Anger—because this has happened to your child
- Helplessness—because you can't fix it for your child
- Frustration—because of the unknowns and unpredictability of ITP and the need to be patient



- Worried, scared, and sad—for the pain and feelings your child is experiencing.

If your child's ITP goes away within a matter of days or weeks and there is no other health problem, most likely you'll feel great.

A n d i f i t g o e s o n ?

In that case, it may feel like you're on a roller coaster—going through ups and downs with your child's platelet counts and need for treatment.

If your child responds well to treatments and can carry on his or her normal life, you may begin to feel that the immediate crisis has passed. The next challenge is to "fold" ITP into your family's lifestyle as much as possible. Blood tests can be scheduled as conveniently as possible—closer to home perhaps, with results called in to the hematologist's office—and around your family's activities.

That's not to say there won't be stressful times—just before blood tests, trying to get time off for treatments, juggling other children's schedules and child care—but it can feel more "manageable".



H o w i s m y c h i l d l i k e l y t o r e a c t ?

A child with ITP will likely go through all the same emotional ups and downs you experience:

- Scared—of tests, treatments, and IV needles; of dying; of "catching" something else
- Guilt—thinking that he or she caused ITP to happen
- Embarrassed—to be "different" than other kids; by lots of bruises





- Angry—at ITP and his or her body; at parents and health professionals for “doing” this
- Frustrated—about restrictions on physical activities, changes in routine; at the lack of “control” over ITP
- Denial—of the reality of ITP and the changes to his or her lifestyle it may require (Especially true for teenagers who may be tempted to ignore the advice of parents and health care providers)

Talk about these feelings often. Listen carefully and let your child know that these are normal feelings. Help your child understand about ITP by answering questions as honestly and simply as possible. Allow your child to ask questions at, and have some say about the scheduling of, his or her appointments. Be sure your child understands that he or she did NOT cause ITP or any other illness that might develop as a result. Offer your child a chance to express his or her feelings through drawing, pretend play, and talks. Try to give your child some sense of “control” over the management of his or her ITP.

What about my other children?

Other children in the family will also be affected. They may feel:

- Stress—from the change in routine
- Worry—for their sister or brother who has ITP; for their own health and safety
- Mad or jealous—about all the attention their brother or sister is getting
- Guilt—thinking he or she has caused the ITP; about being mean or rude to the sister or brother who has ITP.

Try to keep these children informed about ITP as well, and help them find ways to express their

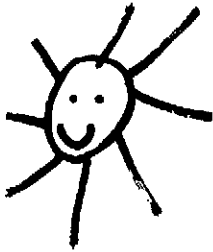


feelings. Do what you can to keep them feeling "special" too, so ITP is not seen as a withdrawal of your attention and love for them.

As with any family issue, talking things over and finding ways to express your emotions while on this "roller coaster" is key. Many health professionals are available to help you and your family sort these feelings out if you feel the need. Many hospitals and clinics offer social support services; this may be a good place to start.

Where can I learn more about ITP?

Learning as much as possible can help too. There are a number of sources for more information, a few of which are detailed below.



- Your child's hematologist and other health professionals
- The ITP Society of the Children's Blood Foundation, 333 E. 38th Street Room 830 New York, NY 10016, 1-800-ITP-7010
- Libraries in your city, town, or in nearby hospitals and medical universities
- The National Heart, Lung, and Blood Institute Information Center, P.O. Box 30105 Bethesda MD 20824-0105, 301/251-1222
- The National Organization for Rare Disorders P. O. Box 8923, New Fairfield CT 06812 203/746-6518
- Health Data Brokers and Computer Bulletin Boards (These networks and businesses provide information for a fee. Your local library may have more specifics.)





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Illustrations by Sara Radin

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