Immune Thrombocytopenic Purpura
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A HANDBOOK FOR FAMILIES

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WHAT IS ITP?

ITP stands for immune thrombocytopenic purpura. ITP occurs when the body’s immune system does not work correctly and results in thrombocytopenia, a decrease in the number of platelets. Platelets are cells in our blood that help form clots to prevent or stop bleeding. When the platelet count is low, there is an increased risk of bruising and bleeding. One may also develop tiny blood spots under the skin or inside the mouth (petechiae).

A normal platelet count is between 150,000 and 400,000. In children with ITP, the platelet count is less than 100,000. Bleeding after a minor injury may occur when the platelet count is less than 50,000. The most serious risk of bleeding occurs when the platelet count is less than 20,000. When the count is this low, bleeding may occur without any injury.

The cause of ITP is not always known, but it often occurs after a viral illness or infection. It can also be associated with some medications and autoimmune disorders such as lupus.

To fight infections, white blood cells make special proteins called antibodies. These antibodies attach to infection-causing agents such as bacteria and viruses that have invaded the body. Anything the antibody attaches to is seen as foreign by the body. The spleen is an organ that filters the antibody-coated viruses or bacteria out of the blood. In ITP, these same antibodies attach themselves to healthy platelets. The antibodies tell the body that these platelets are foreign and the spleen destroys them. As long as antibodies are attached to the platelets, the spleen will continue to remove as many platelets as possible from the body, resulting in a decreased number of platelets in the blood.

Platelets are made in the bone marrow by large cells called megakaryocytes. A protein made by the liver called thrombopoietin controls how many platelets are made by the megakaryocytes. In some cases of ITP, not enough thrombopoietin reaches the bone marrow. When this happens, the megakaryocytes do not make enough new platelets to replace those that are being destroyed.

Most cases of ITP are temporary, or acute. More than 80% of ITP cases in children go away in fewer than 6 months. This happens when the body stops making antibodies against the platelets. Depending on the number of platelets and the risk for serious bleeding, your provider may recommend medications to help manage ITP. If ITP lasts more than 6 months to a year, it is called chronic. Chronic ITP may also need to be managed with medications or just observed to make sure no serious bleeds occur.

WHO GETS ITP?

ITP can occur in children and adults. Newborns can develop ITP shortly after birth if the child’s mother had ITP during pregnancy. Children are more likely to have acute ITP, while teenagers are more likely to have chronic ITP. Boys and girls are affected equally. ITP is not contagious; it cannot be passed from one person to another. ITP is not inherited.
WHAT ARE SOME SYMPTOMS OF ITP?

- Bruising, especially if bruises cannot be explained by injury, are located in uncommon places (head, chest), or more bruises than expected exist.
- Petechiae (flat, pinpoint-sized red or purple dots) caused by minor bleeding appear under the skin.
- Nose bleeds and mouth bleeding from gums occur.
- Heavy menstrual bleeding (heavy periods) occurs.
- Blood appears in urine or stool.
- Confusion, seizures, weakness, or loss of consciousness occur. In rare cases, these symptoms may indicate intracranial (brain) hemorrhage or other internal (stomach, lungs) bleeding.

HOW IS ITP DIAGNOSED?

Most children are diagnosed with ITP when their platelet count drops very low and they develop symptoms. To diagnose ITP, your healthcare provider will draw a sample of blood from your child to check the number of platelets. A low platelet count (less than 100,000) but a normal number of other blood cells (red cells and white cells) will usually indicate ITP. Your healthcare provider may ask questions about medications that your child is taking and recent infections, vaccinations, or immune disorders that may be affecting your child. The healthcare provider will also examine your child to see if he or she has signs of a low platelet count (bruising, petechiae) or infection (enlarged lymph nodes, liver, or spleen).

The healthcare provider may also do blood tests to determine if antibodies are being made against the platelets or if there is a higher than normal percentage of immature (young) platelets in the blood. Low platelet counts also may be found in children with other conditions that affect the bone marrow. For this reason, the healthcare provider may ask to do a bone marrow aspirate and biopsy. This test will show if the platelets are still being made as expected and if there are no abnormal cells in the bone marrow that are causing your child’s symptoms. During a bone marrow aspirate and biopsy, the provider inserts a needle through the bone, usually the hip, and takes a sample of the liquid bone marrow from the spongy space in the bone (aspirate) and another sample from the hip bone called a core (biopsy). These samples will then be examined under a microscope in the lab by a specially trained doctor (pathologist or hematologist). Children are usually given sedation or general anesthesia during the procedure to prevent discomfort. Upon waking, your child may complain of pain at the bone marrow biopsy site. This pain is usually mild and resolves within a day or two. If your child is uncomfortable, a mild pain medication such as Tylenol® may be given, but not aspirin or ibuprofen (Motrin®/Advil®).
How is ITP Treated? What are the Side Effects of Treatment?

Most cases of childhood ITP get better without treatment. Simply monitoring your child’s platelet count, known as observation, is one type of treatment. With time, the body usually stops making antibodies against the platelets. Your child’s platelet count may be low for several weeks or months. As the antibody level decreases, the platelets are no longer destroyed and the platelet count increases.

While waiting for the white blood cells to stop making these antibodies, the body is aware of the low platelet count. As a result, the bone marrow (where platelets are made) will work harder to produce young, larger platelets that can do the job of the many older, smaller platelets. Your healthcare provider may order blood tests to monitor your child’s platelet count. As the platelet count improves, these tests will be needed less often. If the platelet count is low because of a medication, then the medication may be stopped or changed. If the low platelet count is caused by an infection or immune system disorder, then that infection or disorder will be treated, if possible.

Your child will only require treatment for ITP if he or she has a very low platelet count, is bleeding, or wants to participate in sports or other activities that require a higher platelet count. Treatments for ITP include medications and surgery. Your doctor or nurse practitioner will talk to you about the possible benefits and risks of different treatments. Together you can decide what treatment, if any, is best for your child.

Medications do not cure ITP. The most common medications used to treat ITP decrease the number of platelets being destroyed by the spleen. This will increase the platelet count to a safe range so that there is a lower risk of serious bleeding. The body may still be making antibodies against the platelets, and treatments may need to be repeated for several weeks or months until the body stops destroying its own platelets. These medications include intravenous immune globulin (IVIG), anti-D immune globulin, and steroids. A newer class of medications, including romiplostim and eltrombopag, work by increasing the number of platelets made in the bone marrow.

IVIG

IVIG is made from the plasma of human blood. Blood is made up of three different types of cells (red cells, white cells, and platelets) and plasma (the liquid part of the blood). Immunoglobulins, or antibodies, are proteins in the plasma that help the body fight infections. Healthy people can donate their blood or plasma. Immunoglobulins are removed from the plasma of thousands of people and pooled together to make one dose of IVIG. The IVIG is treated to make the product pure (free of other blood proteins) and safe (free of viruses that can cause infection).

It is not clear how IVIG works to treat ITP. In ITP, the spleen destroys antibody-coated platelets in the blood. The IVIG may bind to antibody receptors in the spleen. If the receptors in the spleen are blocked by IVIG, they cannot attach to the antibodies on the platelets.

IVIG does not cure ITP but will usually result in a rapid (24–48 hours) platelet increase in more than 75% of children with ITP. This increase in platelets may last for several weeks. If the body is still making antibodies against the platelets when the IVIG is gone, the platelet count may start to drop again. IVIG is a liquid that is given intravenously (into a vein). Medicines can be used to numb the skin on your child’s
hand or arm before the needle is placed into his or her vein. The IVIG infusion rate is started slowly and increased if there are no side effects. The infusion may take several hours. IVIG is usually given in the infusion unit or a hospital. A nurse will monitor your child for side effects. Most reactions to IVIG occur during the infusion, and some side effects may occur 1–3 days after the infusion. Your child may be given diphenhydramine (Benadryl®) or acetaminophen (Tylenol®) before the IVIG infusion to decrease the chance of side effects.

Common side effects of IVIG include
- headache
- fever or chills
- nausea or vomiting
- hives or rash
- fatigue (feeling tired)
- muscle or joint pain.

There is a small chance that your child may have a severe allergic reaction to IVIG. If this happens, the infusion is stopped and medications to treat the allergic reaction are given immediately. There is a very small risk that the IVIG may contain viruses that cause infection.

IVIG may prevent some live vaccines (childhood shots) from being effective. Live vaccines (measles-mumps-rubella [MMR], chickenpox) should not be given for several months after a child receives IVIG.

**Anti-D Immune Globulin**

WinRho®, an anti-D immune globulin, is an immunoglobulin (antibody) that binds to the “Rh” protein on red blood cells. Like IVIG, anti-D immune globulin is removed from the donated plasma of a specific group of donors and pooled together to make anti-D immune globulin, also called Rho(D) or WinRho®. WinRho® is treated to make the product pure (free of other blood proteins) and safe (free of viruses that can cause infection).

Unlike IVIG, WinRho® is only effective in children who have a positive blood type (A+, B+, O+, or AB+) and in children who have not had their spleen removed. It is not clear how it works to treat ITP. WinRho® coats a number of the red blood cells in the blood with an antibody. These antibody-coated red blood cells are destroyed by the spleen. Because the spleen is “busy” destroying the antibody-coated red blood cells, fewer antibody-coated platelets are destroyed. WinRho® does not cure ITP, but increases the platelet count in more than 75% of children. If the body is still making antibodies against the platelets when the WinRho® is gone, the platelet count may drop again.

WinRho® is a liquid that is given intravenously (into a vein). Medicines can be used to numb the skin on your child’s hand or arm before the needle is placed into the vein. WinRho® is given over a few minutes. If your child experiences side effects, future doses will be given at a slower infusion over 30–60 minutes. WinRho® is usually given in an infusion unit or a hospital. A nurse will monitor your child for side effects. Most reactions occur during the infusion. To decrease these symptoms, your child may be given diphenhydramine (Benadryl®) or acetaminophen (Tylenol®) before WinRho® is administered. Your child may also receive intravenous (IV) fluids before or after the WinRho®. Some side effects may occur 1–3 days after the infusion.
Common side effects of WinRho® include:

- small drop in red blood cell count (mild anemia)
- headache
- fever or chills
- nausea or vomiting.

There is a small chance that your child may have a severe allergic reaction to WinRho®. If this happens, the infusion is stopped and medications to treat the allergic reaction are given immediately. There is a very small risk that the WinRho® may contain viruses that cause infection.

Some children may develop severe anemia (a large drop in red blood cell count) a few days after receiving WinRho®. Contact your healthcare provider immediately if your child has any of the following symptoms:

- back pain
- dark or cola-colored urine
- fever or shaking chills
- pale complexion, fatigue (feels tired), or dizziness
- problems breathing or shortness of breath
- decrease in amount of urine
- sudden weight gain or puffy appearance.

WinRho® may prevent some live vaccines (for example, MMR, chickenpox) from being effective; they should not be given for several months after a child receives WinRho®.

**Steroids (Glucocorticoid Steroid Hormones)**

Cortisone is a chemical that is made by the adrenal glands, which are small organs above the kidneys. Cortisone is a type of steroid, a natural substance made by the body. Steroids are important for many body functions. Prednisone and dexamethasone are synthetic (man-made) medicines that are similar to cortisone. These types of steroids are different than anabolic steroids that are used to build muscles.

It is not clear how steroids work to treat ITP, but they may decrease the number of antibodies that are made against the platelets and may slow the rate that the antibody-coated platelets are removed by the spleen. Steroids may also make the blood vessels more stable, lowering the risk of bleeding.

Steroids do not cure ITP, but increase the platelet count in more than 75% of children with ITP. If the body is still making antibodies against the platelets when the steroid dose is decreased or stopped, the platelet count may drop again.

Steroids can be given intravenously (into a vein) or by mouth. If intravenous (IV) steroids are used, medicines can be used to briefly numb the skin on your child’s hand or arm before the needle is placed in the vein. The steroid is given over a few minutes. IV steroids are usually given in an infusion unit or a hospital.

Typically, the steroids are given by mouth as a pill or a liquid. Your child’s healthcare provider will prescribe how much medicine your child should take. Steroids may be given in different ways: very high doses over a few days, high doses for several days followed by a special schedule to stop the medication (a taper, where the amount of medicine is slowly decreased), or low doses of medicine for several weeks or months. Make sure that you understand how to give the medicine and what to do if your child refuses the medicine or vomits a dose.
Common side effects of steroids include

- heartburn (reflux)
- increase in appetite or weight gain
- mood changes
- puffiness in the face or behind the neck
- problems with sleep
- acne.

Most of the common side effects of steroids will get better once the medication is stopped. Children who take steroids for weeks or months may have other side effects, including increase in blood pressure, increased sugar in the blood or urine, risk of infection, thinning of the skin, problems with growth, or calcium loss from the bones. Because of these risks, steroids are usually only given for a few weeks or months at a time.

Contact your healthcare provider immediately if your child

- is exposed to chickenpox and has not yet had this infection or received the vaccine
- is drinking more or has an increase in the amount of urine
- has black, tar-like stools.

**Thrombopoietic Agents**

Thrombopoietic agents are man-made proteins that increase the number of platelets made in the bone marrow. They are typically only used for patients who have chronic ITP or who have ITP that has not responded to medications that decrease platelet destruction such as immune globulin (IVIG, WinRho®) or steroids. Romiplostim and eltrombopag (Promacta®) are two thrombopoietic medications that are used to treat ITP.

Romiplostim (Nplate®) is given by subcutaneous (under the skin) injection once a week. The amount given is based on the platelet count. Romiplostim must be given in a doctor’s office. It is important not to miss an injection. The platelet count may drop very low if the medication is not given on a regular basis. Common side effects of romiplostim include

- headache
- dizziness
- trouble sleeping (insomnia)
- abdominal pain
- joint and/or muscle pain
- rebound thrombocytopenia
- antibody formation.

Less common but serious side effects of romiplostim include blood clots and changes inside of the bone marrow. Your healthcare provider will discuss how to watch for these side effects and if any special tests are needed to monitor for these side effects. There is an increased risk of developing bone marrow changes in people who use romiplostim. Your doctor may check your bone marrow to monitor for this.

Eltrombopag is given by mouth daily. The dose may be adjusted based on platelet count. Liver function tests will also be monitored while taking this medication. Persons of East Asian descent require a lower starting dose and monitoring for dose adjustment. Eltrombopag should be taken 4 hours before or after antacids and
multivitamins as they may interact with these medications.

Common side effects of eltrombopag include

- nausea, vomiting
- diarrhea
- muscle aches, back pain
- abnormal liver function tests
- flu-like symptoms (fever, headache, sore throat, cough, body aches)
- skin rash, itching
- runny or stuffy nose.

Less common but serious side effects of eltrombopag include blood clots, flu-like symptoms, and liver problems. Your healthcare provider will discuss how to watch for these side effects.

**Transfusions**

Most children with ITP make enough platelets, but they also make antibodies against their platelets. The spleen quickly destroys platelets that are coated with an antibody. These antibodies can also bind to platelets that are transfused. When transfused platelets are coated with the antibody, the spleen quickly destroys them. For this reason, platelet transfusions are only used if a child has serious bleeding.

**Other Medical Treatments**

Some children with ITP do not respond to IVIG, anti-D immune globulin, or steroids. If your child has a low platelet count or bleeding despite these treatments, your doctor or nurse practitioner may discuss other treatments. Alternate treatments for ITP include chemotherapy (vincristine, cyclophosphamide, mercaptopurine), drugs that suppress the immune system (mycophenolate, cyclosporine), or a medication (rituximab) that decreases the number of B-lymphocytes, the white blood cells that make antibodies.

**Complementary Therapies**

Some families want to try complementary or natural therapies to treat their child’s ITP. Complementary therapies may include the use of supplements, herbs, vitamins, homeopathy, or a change in diet. These treatments have not been studied in children with ITP and we do not know if they are helpful. We do know that some treatments may be harmful because they decrease the platelet count or affect how well the platelets work to stop bleeding. Please talk to your child’s healthcare provider if you want to try complementary or natural therapies.

**Medicines to Avoid**

Children with ITP should not take medicines that decrease their platelet count or inhibit the platelets’ work to stop bleeding. These medications include aspirin and ibuprofen (Advil® or Motrin®). It sometimes is not immediately clear if a product contains aspirin or ibuprofen. You should always check the medication label and call your healthcare provider if you have any concerns.
Some herbs and supplements may also be harmful. Be sure to tell your child's healthcare provider all the medicines that your child is taking, including medications that do not require a prescription.

**Surgery**

The spleen is an organ on the upper left side of the abdomen under the ribs. The spleen cleans the blood and removes old blood cells. The spleen also helps to fight infections. In ITP, white blood cells in the spleen called B-lymphocytes produce antibodies against platelets. Platelets that are coated with antibodies are removed from the blood by the spleen. Removing the spleen often helps to increase the platelet count.

Surgery to remove the spleen is called a splenectomy. About 75% of children with ITP are cured with splenectomy; however, this is a major surgery with a risk of serious bleeding during the procedure. After the spleen is removed, your child will have an increased risk of infection for the rest of his or her life. Your healthcare provider may recommend that your child take antibiotics by mouth once a day to prevent blood infections. Fever in a child who has had a splenectomy is a medical emergency. If your child has had his or her spleen removed and has a fever, he or she should be seen by your healthcare provider right away. Because of these risks, splenectomy is only used to treat children who have had a low platelet count for more than 1 year or who have serious bleeding that does not respond to medications.

**HOW CAN I WORK WITH THE HEALTHCARE TEAM?**

You know your child better than anyone else, and your input is important. Other members of your child’s healthcare team include doctors, nurses, and social workers.

Always communicate openly with the other members of your child’s healthcare team. Ask questions when there is anything you are not sure about. It may help to write down your questions as you think of them. Important questions you may want to ask include

- Is the ITP chronic or acute and what does that mean?
- What treatment choices are available?
- What treatment do you recommend, and why?
- What are the risks or side effects during the treatment?
- What are the risks and side effects after the treatment?
- How will ITP impact my child's daily life and future?
- What are the chances for recurrence?
Use the lines below to write down any other questions you might want answered.

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**CAN MY CHILD ATTEND SCHOOL AND PARTICIPATE IN ACTIVITIES DURING THERAPY?**

Children with ITP may attend school. When your child’s platelet count is low, your healthcare provider may limit the types of physical activities in which your child can participate. For a child who wants to play sports, this can be very frustrating. Explain to your child why this activity is not allowed at this time, and choose other activities that are okay even with a low platelet count.

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**ARE MY FEELINGS NORMAL?**

Hearing that your child has a medical problem is often shocking and overwhelming. ITP, its cause, and the uncertainty of how it will respond to treatment can be confusing. It is important to know this is normal and expected.

Many families feel somehow responsible for their child’s disease. Feelings of guilt that they could not protect their child from illness or about the amount of time it took to diagnose the child are also common. This disease is not caused by anything that you did or did not do.
HOW CAN I HELP MY CHILD?

Children often think that something they did caused their ITP. Reinforce that this is not the case. It is important to keep your relationship honest and to maintain closeness. Your child will need to share his or her feelings with someone that he or she trusts. Sometimes, they may choose to speak with someone other than their parents because they are afraid they might upset them. Don’t be afraid to ask your child about his or her feelings—it may be what your child is waiting for. Also, don’t be afraid to share information about what is happening and why it is happening with your child. The things children imagine on their own are often more frightening than what is actually happening.

Talk to your child about treatment using direct terms and explanations your child will understand. As a parent, it may be difficult to watch your child go through ITP therapy. The frequent blood tests and possible limits on their activities because of low platelet counts can be upsetting to you and your child. Children tolerate these things better if they understand why it is necessary and are allowed to help make decisions about their care when appropriate.

IS MY CHILD’S DIET IMPORTANT DURING THERAPY?

If your child is on steroid medications such as prednisone or Decadron (dexamethasone) to treat the ITP, his or her appetite will likely be increased. While on these medicines, your child frequently may be hungry and crave unusual foods or want one particular food. Try to keep healthy snacks on hand. Steroids may cause your child to gain weight and develop a round face and puffy appearance, but these side effects are temporary.

Some medications, including vitamins and herbs, can decrease the platelet count or interfere with the platelets’ ability to work to stop bleeding. Some medications may interact with your child’s treatment. Be sure to tell your child’s healthcare provider all the medicines that your child is taking, including those that do not require a prescription.

A dietician trained in the energy needs of children may be part of your child’s healthcare team. Your child’s healthcare team will monitor your child’s nutritional status, height, and weight during and after treatment to try to prevent problems. When there is a concern about nutritional problems, your child’s healthcare team will intervene.
RESOURCES

Children’s Cancer and Blood Foundation
333 E. 38th Street
Suite 830
New York, NY 10016
212.297.4336
http://www.childrenscbf.org/content/itp#.VrEVq53nbcs

ITP Foundation
30 Old Kings Highway, South Suite 275
Darien, CT 06820
203.655.6954
www.itpfoundation.org

ITP Support Association
Kimbolton Road
Bolnhurst
Bedfordshire
MK44 2EW
United Kingdom
www.itpsupport.org.uk

Platelet Disorder Support Association (PDSA)
133 Rollins Avenue, Suite 5
Rockville, MD 20852
877.528.3538
www.pdsa.org

BIBLIOGRAPHY


