

Coping with ITP

FREQUENTLY ASKED QUESTIONS



PLATELET DISORDER
SUPPORT ASSOCIATION

for People with ITP



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When a patient has a chronic disease like ITP, finding means of coping is very important. This booklet provides information about ITP and offers suggestions for how to cope with the disease and with side effects of its treatments. It also offers ways to cope with the psychological aspects of having a chronic illness. Coping means “dealing successfully with something difficult” and “struggling to overcome problems or difficulties.” Coping means managing, succeeding, and carrying on. The information in this booklet will help you successfully cope with ITP.

Q What is ITP?

A Auto immune diseases are disorders in which the body mounts an immune attack toward one or more seemingly normal organ systems. Immune thrombocytopenia (ITP) is an autoimmune disease in which platelets are the target. They are marked as foreign by the immune system and eliminated in the spleen and sometimes, the liver. In addition to increased platelet destruction, most people with ITP also have impaired platelet production.

The specific cause of ITP is often unknown. Some cases occur after a viral or bacterial infection, immunizations, exposure to a toxin, or in association with another illness such as lupus or HIV. It is important to recall what was happening in your life before you began having symptoms of low platelets. This information may help your physician in diagnosing and treating your low platelet count.

Q What are platelets?

A Platelets are relatively small, disc-shaped cells in our blood. They are required to maintain the integrity of our blood vessel walls and for blood to clot. Without a sufficient number of platelets people with ITP are subject to spontaneous bleeding and bruising, or they may bleed a long time if they get a cut.

Q What is a normal platelet count?

A According to the Merck Manual for Health Care Professionals (2011 Ed.) normal platelet counts range from 140,000 to 440,000 per microliter of blood. Mosby's Manual of Diagnostic and Laboratory Tests (4th Ed. 2010) states that normal platelet counts range from 150,000 to 400,000 per microliter of blood. Therefore, reference ranges vary slightly depending upon the specific laboratory performing the test. The lower bound (140,000 or 150,000) is important because counts less than that indicate thrombocytopenia. However, a platelet count less than 100,000 might be considered ITP according to the International Consensus Report on the Investigation and Management of Primary Immune Thrombocytopenia (*Blood*, Jan. 14, 2010). People with ITP who have platelet counts under 10,000 have a severe case. For many, a count of 30,000 or more is sufficient to prevent bleeding. Individual responses to low platelet counts differ. Determining a safe platelet count is a decision to be made in consultation with an experienced treating physician.

"I hope I can experience a long-term recovery, but one never knows with this illness, so I take one week at a time."

— SONIA

Q What are the symptoms of ITP?

A The symptoms vary greatly from person to person. Most people with ITP experience spontaneous bruising. Some find they have petechiae (*pe-TEEK-ee-ay*), tiny red dots on the skin caused by broken blood vessels or leaks in a capillary wall. If your platelet count is very low you may have other bleeding symptoms including blood blisters on the insides of your cheeks, nosebleeds, or blood in your urine or stool. Women may have prolonged menstrual bleeding. In general, the more bleeding symptoms you have, the lower your platelet count.

Q Can ITP be treated?

A Treatment improves the platelet count in most patients, which decreases problems with bleeding. The difficulty for many patients is finding the treatment that works for them without unwanted side effects. This is not easy. Some patients report that changing their diet or lifestyle helps them feel better. While the disease can go into remission for a long time, perhaps for the remainder of the person's life, ITP can also recur. There is currently no way to predict the course of the disease.



Q Are there differences in remission?

A In children, who usually have the acute, short-lived form of ITP, most often associated with an infectious agent, there is a high rate of spontaneous remission. In adults, who usually have the chronic form of ITP, some patients may go into remission for periods of time.

Coping With the Physical Symptoms of ITP

Q How can I minimize the bleeding symptoms related to having ITP?

A Know and understand the symptoms of ITP and its flare ups, including bruising, petechiae, bleeding gums, blood in urine or stool, or excessive bleeding from a cut. Take precautions to avoid injury, bumps, cuts, and bruises. Be extra careful when using sharp tools such as knives, razors, and scissors.

Keep copies of lab reports and all blood work. Maintain a log of the medications used, dosages, your platelet count, and how they made you feel. Pay attention to your lifestyle. Notice if there is any connection between your low count and the food you eat, places you visit, possible toxins in your environment, etc.

Include plenty of dark, leafy greens loaded with Vitamin K (a vitamin needed for your blood to clot) in your diet. Avoid alcohol, which damages the bone marrow and harms the liver where your body makes clotting factors and the protein that signals platelet production of thrombopoietin. Know the foods and substances that can interfere with the clotting process or reduce the number of platelets. PDSA maintains a list on the Web site. Visit www.pdsa.org and click on 'About ITP' and then 'Warnings.' Avoid medications such as aspirin and NSAIDS that may reduce platelet function and increase bleeding.

Q What symptoms should prompt me to call my health care provider?

A Call your doctor or other healthcare provider *immediately* if you hit your head or have a serious accident. Watch for a large number of bruises and petechiae, or other signs of severe bleeding, which can indicate a very low platelet count. Signs of bleeding in the brain include: a headache that won't go away, dizziness, vomiting, unusual sleepiness, confusion, slurring of speech, eyes not moving together, weakness on one side of the body,



stiff neck or back, seizures, and inability to see or hear. Notify your doctor about nosebleeds, bleeding gums, or blood in urine, stool or vomit. Even when ITP patients are in remission they should watch for these symptoms so that they can contact their healthcare provider immediately if any of these occur.

Q How can I stop worrying about my platelet count and symptoms?

A Many people with ITP find they obsess about their platelet count, bruises or petechiae. When the platelet count goes up it is easy to become elated and when the platelet count goes down, many people become depressed. Staring at bruises and looking for red dots can become habitual.

It is important to remember that the automated platelet counters are not always very accurate when reporting the number of platelets when the count is low and bruises are not fatal. If you find yourself thinking about your platelet count and ITP for most of the day, consider substituting thoughts about something fun and fulfilling you are planning in the future or just stop the ruminating and count the ceiling tiles.

Talk with someone who has a positive attitude, as well as try to maintain a positive attitude yourself. Having that positive attitude, along with a sense of humor can be helpful in managing the stress of having ITP. You may benefit by joining one of the local support groups sponsored by PDSA, where you can talk with others who have been touched by ITP either as a patient or a family member or caregiver.

Q In addition to a low platelet count, I'm often tired. Is fatigue a normal part of the disease? Will anything improve this?

A Fatigue is a common experience for people with ITP. It may be caused by the disease or it could be a response to your medications or other treatments. Some patients report that changing to a healthy diet (especially adding dark, leafy greens) increases their energy level and reduces fatigue. Be sure to get adequate rest but be careful with using "sleep aids" because of possible side effects. Include as many activities in your life as you can that feed your soul and make you happy. Try to avoid draining, exhausting situations that will only sap your energy. Get plenty of exercise (check with your doctor for types of exercise appropriate for you). Breathe clean air and drink pure water.



Q Should ITP patients wear medical alert jewelry?

A It is recommended that you wear some type of identification of your low platelet situation. Wearing a medical alert bracelet or necklace can be used to indicate you have ITP, a bleeding disorder, have had splenectomy, and/or are on steroids. This would help emergency medical staff quickly and correctly attend to your needs in an emergency situation. In addition, you should carry an identification card in your wallet with a list of your medications and dosages and a brief note on your illness. For more details visit: <https://www.medicalalert.com> or call 1-800-432-5378. PDSA provides free wallet medical cards. While traveling, it is especially important to wear some type of medical alert jewelry.

"Another way to make it a little easier is to always be honest with yourself and loved ones...making sure that everyone who is important knows what's going on."

— MIKE

Q As a parent, how can I protect my busy, active child with ITP?

A Encourage your child to live as active a life as possible. Talk to your child's doctor about those activities that may be appropriate for your child. Explain to older siblings the need to avoid rough play with the child with ITP and enlist their help in looking out for their sibling. When deciding if an activity is okay for your child, consider allowing anything within reason where the child's feet never leave the ground. This usually excludes bikes, motorcycles, skates, and swings. Avoid sharp-edged toys, popsicle sticks, and drinking straws.

Use rubber no-slip mats in the tub or shower. Pad the sides of cribs for small children and for older children who may fall out of bed, put their mattress on the floor or add a side rail. Don't allow running through the house or on stairs. Clear obstacles such as tables and chairs that you frequently have to go around. For small children, pad the corners of furniture and counters where they might run into them.

In addition, parents should build and maintain a strong relationship with their child's health provider.

Q What else can parents do to help a child cope with physical symptoms of ITP?

A Examine your child's skin for bruises or petechiae during regular bath time. Check your child's stool or have them report to you any blood in their stool or urine. Blood in their urine may be red or brownish (tea or cola colored). Blood in the stool may look red or black like tar. Be sure your child is eating plenty



of fruit, fruit juice, water, and fiber foods to avoid constipation.

Watch for bleeding in the mouth or nose, especially in the dry air season. Encourage only gentle nose blowing. Use a saline nose spray to moisten nasal passage during dry weather to help prevent nosebleeds. In addition, regular application of a Vaseline®-based compound to the child's nasal mucosa helps prevent bleeding.

Using a room or house humidifier can also help. To minimize gum bleeding, have them use the softest toothbrush possible, using plenty of toothpaste, and avoid using dental floss until their platelet count is up. Use lip balm to keep their lips from drying and cracking. Report to your doctor or hospital immediately any blows or injuries to the child's head, belly or back.

Avoid giving medications to your child until you have discussed them with your doctor or nurse practitioner. Check to be certain that any medicines recommended by your health provider do not reduce platelets. Your child's platelet count can be affected by many over-the-counter pain, cold, and cough medicines. To minimize bleeding, aspirin or medicines containing salicylates (such as Pepto Bismol®) should not be used. Also avoid ibuprofen/other NSAIDS. Usually acetaminophen products (Tylenol®, Tempra®, etc.) are safe to give your child for a fever or pain while their platelet count is low.

Continue to learn all you can about ITP and its treatment and share more information with your child as they are able to understand it. Try to stay upbeat and avoid transmitting unnecessary anxiety and concern to your child. Explain to them that most children recover completely from ITP.

Q What are the signs of serious injury or bleeding in children?

A In children, signs of serious injury or internal head bleeding include: dizziness, headache that doesn't go away or gets worse, vomiting, confusion, unusual sleepiness, slurred speech, weakness, numbness or tingling in hands or feet, stiff neck, seizures, eyes not moving together, or inability see. Call your doctor or hospital immediately if your child falls resulting in hitting their head or for any event that causes your child to lose consciousness.

"Educate yourself and take responsibility, both for what you can control and for knowing what you cannot. Mostly, remember that you've got ITP, but ITP doesn't have to have you."

— LIZABETH



Coping with the Side Effects of Treatments for ITP

Q What are the main treatments available for ITP?
What are the goals of treatment?

A The main treatments for ITP are medications (drugs) or splenectomy. The main drug treatments include corticosteroids (ex., prednisone), intravenous gamma globulin (IVIg), and anti-Rho(D) immune globulin (ex., WinRho SDF®). Some additional drugs used to treat ITP may include azathioprine (Imuran®), cyclophosphamide (Cytoxan®), cyclosporine (Sandimmune®), danazol (Danocrine®), mycophenolate mofetil (ex. Cellcept®), rituximab (ex., Rituxan®), vinca alkaloids (Vincristine®), and the thrombopoietin mimetics [ex. eltrombopag (Promacta®), romiplostim (Nplate®)]. In some cases, surgery to remove the spleen (splenectomy) is recommended. Treatment for ITP is most often coordinated by a hematologist (doctor who specializes in blood disorders). The goals of ITP treatment are to ensure a safe platelet count, prevent bleeding complications, and minimize treatment side effects.

Q How can I find out about the latest developments in ITP treatment?

A Join the Platelet Disorder Support Association at www.pdsa.org.

Q What are the side effects of ITP treatments?

A All of these treatments have their own benefits and risks. Side effects have been reported for each of the drugs and treatments for ITP. These side effects will vary from one person to another. You may experience all, some, or none of the side effects. Here is a description of the different treatments for ITP and their side effects.

First-Line (Initial) Therapies

Prednisone

Q What is prednisone?

A Prednisone, in a class of medications called corticosteroids, is similar to cortisone, a natural substance produced in the body's adrenal glands. Cortisone helps the body use carbohydrates, proteins, and fats and helps the body deal with stress.



Q What are the possible side effects of prednisone?

A Prednisone is generally given for only a few weeks at a time because it can have serious side effects with long-term use. Prednisone affects every system in your body. Even when prednisone is given for a short time, though, you may feel irritable and experience stomach upsets, sleep disturbances, headaches, increased appetite, weight gain, puffy cheeks, frequent urination, sugar in the urine, loss of bone density, loss of muscle mass (including the heart muscle), and skin problems. Once the medicine is stopped, most side effects will begin to disappear. As prednisone is tapered, patients may experience extreme fatigue. This occurs because their adrenal glands haven't resumed their function of producing adrenal hormones, which normally gives a person energy and vitality.

Q Are there dietary changes that can help minimize prednisone weight gain?

A Taking corticosteroids may cause increased appetite, weight gain, and changes in electrolytes, even when you watch your diet carefully. Seek nutritional advice and counseling. Talk with your doctor about a vitamin/mineral supplement to assure adequate replacement of lost electrolytes. Follow these dietary suggestions to lessen prednisone weight gain. **Avoid salt** (sodium) to help decrease fluid retention. Don't add salt to food. Find unsalted versions of foods you eat. Avoid salty snacks, such as chips and pretzels. Eat more fresh, unprocessed foods. **Avoid fat.** Steroids can increase levels of blood fats. A lowfat diet may help. Use good fats, such as cold pressed olive oil, for cooking and avoid fried foods and foods with trans fats. **Avoid sugar.** Refined white sugar can leach B vitamins, calcium, phosphorus, iron, etc. from our teeth, bones, and tissues. Try alternative sweeteners such as rice syrup or barley malt for baked goods and cereal. **Eat complex carbohydrates**, which contain more fiber and are less sweet, including whole grain breads and cereals, beans, brown rice, whole grain pasta, and root vegetables. **To combat increased hunger — snack more frequently** on low-calorie fruit, vegetables, low fat dairy, and low salt rice cakes.

Q What else helps in coping with prednisone?

A Try to avoid stressful situations as much as possible. To cope with side effects of prednisone try using stress reduction tapes, relaxation tapes, yoga, and meditation.



Also, avoid coffee and other products with caffeine. While taking steroids you may experience personality changes. Consider getting counseling if the stress of prednisone becomes too difficult. Exercise to minimize muscle loss. See your doctor to monitor your health for signs of osteoporosis (bone thinning), high blood pressure, and diabetes.

IVIg

Q What is IVIg?

A IVIg (intravenous gamma globulin), which comes in slightly different formulations made by a number of manufacturers, is a liquid concentrate of antibodies purified from the plasma (the liquid portion of the blood that doesn't contain blood cells) of healthy blood donors. IVIg treatment usually results in a rapid (24 to 48 hours) increase in the platelet count; however, any improvement is generally short-lived, usually a month or less. IVIg is delivered by intravenous infusion directly into an arm vein for several hours a day over a period of 1 to 5 days.

Q What are the possible side effects of IVIg?

A Patients treated with IVIg may experience side effects that include headache, fever, local skin reaction or pain at the infusion site, nausea and vomiting, leg cramps, rash, and flu like symptoms. Rare but more severe side effects can include aseptic meningitis, abnormal blood clots or kidney failure. Patients on IVIg treatment should contact their doctors immediately if they experience these more serious side effects: decreased urination, sudden weight gain, swelling of legs or ankles, chest tightness, or shortness of breath. IVIg is made from a protein extracted from human plasma and carries a small risk of transmittable disease. Because of difficulty in manufacturing and extensive screening that it must undergo, the cost of IVIg is quite high, causing a limitation on treatment for some patients.

Q What can help in coping with side effects of IVIg?

A Talk with your doctor about pre-medications to reduce the side-effects of the infusion. If side effects occur during the infusion of IVIg, the rate of infusion can be slowed down, which helps. Most of these side effects will disappear after the infusion is completed.

"I would have to say positive people and positive influences have helped me deal with my ITP. I have a great family and amazing friends. My outlook has totally changed... I believe our thoughts affect our health... I only want to attract positive and healthy things."

— NATALE



Anti-Rho(D)

Q What is Anti-Rho(D)?

A Anti-Rho(D) (such as WinRho®) is also a liquid concentrate of antibodies derived from healthy human plasma. However, this medicine is targeted against the Rh factor* on red blood cells. It is thought that anti-Rho(D) binds to red blood cells to such an extent that the spleen is fully occupied eliminating red blood cells and does not have much opportunity to remove the antibody-coated platelets.

Like IVIg, the response is usually rapid but temporary. Anti-D is given by intravenous infusion usually lasting less than an hour. Anti-D will generally not work if you are Rh-negative or have had a splenectomy (removal of the spleen).

Q What are the possible side effects of anti-Rho(D)?

A Temporary side effects from anti-Rho(D), also called anti-D, (seen in about 2 percent of patients in clinical trials) can include fever, headache, chills, nausea and vomiting, and anemia. Because anti-D is prepared from human plasma, it carries a very small risk of developing a viral infection. Use of any anti-D drug causes hemolysis (destruction of red blood cells). However, an extremely rare but serious side effect with anti-D is development of intravascular hemolysis. Symptoms of intravascular hemolysis may include back pain, dark or discolored urine, shaking chills, decreased urine production, swelling, or shortness of breath. These can occur as soon as four hours following the intravenous infusion. Patients should contact their doctor immediately if they experience any of these symptoms.

Q What can help in coping with side effects of Anti-Rho(D)?

A Talk to your doctor about pre-medication to reduce the infusion reaction. If side effects occur during the infusion of anti-Rho(D), the rate of infusion can be slowed down, which helps. Most of these side effects will disappear after the infusion is completed.

* Most people have Rh-positive blood. This means they produce the Rh factor, an inherited protein found on the surface of red blood cells. A small percentage of people lack the Rh factor. They are considered Rh-negative.

Second-Line Therapies

Q If “front-line” therapies are not effective what are the “second-line” medical treatments that the doctor is most likely to use to raise platelet counts?

A If corticosteroids, IVIg, and anti-D are not effective in raising platelet counts to a safe level or the side effects pose problems, a number of “second-line” treatments may be tried.

The two primary types of second-line treatment are either medical (drugs) or surgical (spleen removal). Second-line treatments include: the drugs danazol, dapsone, monoclonal antibodies (e.g, rituximab), and several immunosuppressant drugs; surgery to remove the spleen (splenectomy); and the thrombopoietin mimetics or platelet growth factors.

Danazol is a steroid-sparing agent. It is a synthetic androgen (male sex hormone) also known as 17-alphaethinyl testosterone. Danazol was marketed as Danocrine® in the United States. In the early 1970s it was approved by the U.S. FDA as the first drug to treat endometriosis. Danazol has also been used off-label for other disorders, including immune thrombocytopenia (ITP).

Danazol may decrease removal of platelets by the immune system. About 50% of patients who are treated will respond to danazol although responses may be slow (average response time is 2.7 months with 85% of responses occurring within 4 months). The reported response rate has been about 30%. Its main side effects are its masculinizing effects in sensitive women. This is due to the fact that danazol disrupts the action of the pituitary gland by suppressing the output of some hormones, which causes the reduction of estrogen, which may increase androgens, halt menses, promote growth of facial hair, and cause acne. Baseline and periodic liver function tests should be performed in all patients.

Dapsone (diamino-diphenyl sulfone) is an antibacterial drug commonly used in combination with rifampicin and clofazimine as multidrug therapy (MDT) for the treatment of *Mycobacterium leprae* infections (leprosy). As an antibacterial, dapsone inhibits bacterial synthesis of dihydrofolic acid. Dapsone has also been used to treat chronic ITP. Several studies reported a response rate of 40% to 60%, with average response time of 3–5 months. Responses last for several months but relapse usually occurs unless therapy is continued.

Dapsone may be considered as a substitute in patients whose platelet counts can be maintained with



small doses of prednisone. It may be added to the prednisone and after a few weeks the prednisone can be slowly tapered. Side effects include destruction of red blood cells (hemolysis) at higher doses and sometimes anemia. Hypersensitivity reactions occur in some patients and involve a rash and possibly fever, jaundice, and eosinophilia (increased white cell count).

Monoclonal Antibodies

Q What are monoclonal antibodies and how do they work? What are the side effects?

A **Monoclonal antibodies** are antibodies produced artificially from a cell clone and consist of a single type of immunoglobulin. **Rituximab** (trade name: Rituxan®) is one type of monoclonal antibody that was approved by the FDA to treat lymphoma and rheumatoid arthritis. There has been some success in its use to treat ITP. Clinical studies are in progress.

Mechanism of action: rituximab destroys both normal and malignant B lymphocytes. It is used to treat diseases characterized by having too many B cells, dysfunctional B cells, or overactive B cells (as in ITP). Thus, rituximab reduces the number of B cells in the body. B cells are a type of white blood cell that, when activated, multiply and produce antibodies.

Since rituximab reduces the number of B cells, it also reduces the number of cells that produce antibodies, including the antibodies that attack platelets, thereby enabling the platelet count to rise. Most recent reports from studies in the U.S. on the effects of Rituxan® showed that after being given to patients for four weeks, 32% of patients had an increase in platelets lasting up to a year. Side effects of Rituxan® were reported in 87% of the patients. Ten percent of patients reported very serious adverse effects that included fever, chills, weakness, nausea and headaches. Some hypersensitive patients may get serum sickness. Some rare cases of progressive multifocal leukoencephalopathy due to a specific virus have been reported to the FDA.

Immunosuppressant Drugs

Q What are immunosuppressant drugs and how do they work?

A **Immunosuppressants** are a class of drugs that are capable of inhibiting the body's immune system. Many of the agents in this category are also cytotoxic (cell poisons) and are used in the treatment of cancer.

In ITP, the immune system is hyperactive and produces auto-antibodies at a rapid rate of growth. Immunosuppressive chemotherapy medicines have their greatest effect against rapidly dividing cells and, therefore, can be beneficial in the treatment of ITP by suppressing the cells involved in the hyperactive immune response.

Immunosuppressant chemotherapy drugs have been used for patients with chronic ITP. **Vincristine** (vinca alkaloids), **Cytoxan®** (cyclophosphamide), and **Imuran®** (azathioprine) are those most frequently prescribed. Each drug has a slightly different profile of side effects. These effects may include hair loss, decreased immunity, gastrointestinal symptoms, bleeding from the bladder, damage to the central and peripheral nervous systems, bone marrow suppression, liver toxicity and risk of developing leukemia.

Splenectomy

Q If ITP patients do not respond to “second-line” drug therapies what else can be done?

A Another second-line treatment for ITP is surgery to remove the spleen (splenectomy) in patients whose platelet count remains below 30,000 and who have serious bleeding. The decision as to whether to undergo a splenectomy is a complex issue that each patient must consider carefully depending on their personal medical situation and medical history. The issues are too complex to fully cover in this brochure. Please consult with your doctors and consider a second opinion.

On the positive side, the medical community has considerable experience with splenectomy. It has been used on thousands of patients over the decades in countries throughout the world. It has a high cure rate and benefit-to-risk ratio. Patients and their doctors may wish to defer splenectomy until drug therapies have failed or side effects become intolerable.

Q What is splenectomy?

A Splenectomy is the surgical removal of the spleen, an organ that filters and stores blood, destroys old red blood cells, and produces antibodies to fight infections.

Removing the spleen may raise the platelet count by removing the site of platelet destruction. It does not work for all cases of ITP. Surgery is performed in a hospital with the patient under general anesthesia

(asleep). Many patients may be able to have laparoscopic surgery, which uses small incisions and has faster recovery time.

Q How can a splenectomy stop platelet destruction?

A Surgically removing the spleen should significantly reduce platelet destruction because the spleen acts as the primary site of platelet removal and of antiplatelet autoantibody production.

Q What are the side effects of having splenectomy?

A Possible short-term complications of splenectomy immediately following surgery may include infection, incisional bleeding, deep vein blood clots, pneumonia, incisional hernia, pancreas inflammation, and pulmonary embolism (blood clot that travels to the lungs). While general anesthesia prevents pain during the surgery, there may be incision pain for several days after surgery. The spleen filters your blood and produces antibodies to help fight infections. There may be a slightly increased risk of certain bacterial infections (Pneumococcus, N. meningitides, and H. influenzae) and prior to splenectomy your doctor will recommend vaccination for preventing those infections.

Q How can I cope with side effects of a splenectomy?

A For incisional pain after surgery, your doctor can prescribe pain medication. You should avoid vigorous activity such as heavy lifting and driving as directed by your doctor.

Over the long term, patients who have had a splenectomy should seek medical attention for fever and major infections, as these may require antibiotics to prevent a more serious infection. Always tell any doctor treating you that you do not have a spleen. When traveling outside the U.S., take special precautions against malaria, babesiosis, and other infections that may cause a threat. Talk to your health provider about immunizations you may need.

Platelet Growth Factors

Q What about the platelet growth factors?

A **Platelet Growth Factors** are the newest approved form of treatment for patients with chronic ITP. Nplate™/romiplostim completed all clinical testing and was approved in August 2008 by the Food and Drug Administration (FDA) for use in the U.S. for chronic

ITP in splenectomized and non-splenectomized adult patients. Another platelet growth factor, PROMACTA®/eltrombopag, completed all clinical testing and received FDA approval in November 2008 for use in the U.S. for chronic ITP. In 2015 Promacta was FDA approved for treatment of chronic ITP in children, ages 1-17. Both of these new treatments have been found effective for raising platelets in ITP. Nplate™ is given by weekly subcutaneous injections. PROMACTA® is a pill taken once a day.

Mechanism of action: Most previous treatments for ITP have focused on stopping destruction of platelets in the spleen and elsewhere. The platelet growth factors or thrombopoietin (TPO) receptor agonists are a new class of treatments that stimulate platelet production. TPO, a protein made in the liver, naturally stimulates platelet production in the bone marrow. TPO receptor agonists bind to the same receptor as the TPO produced in the body and stimulate the bone marrow to produce more platelets.

While ITP is often considered a disease characterized by platelet destruction, recent research has shown that many people with ITP also have inappropriately low platelet production. The bone marrow stimulation prompted by the platelet growth factors (e.g., Nplate™ or PROMACTA®) creates a sufficient number of platelets to overcome the platelet production defect in most people who receive these treatments. The result is a safe level of platelets with minimal side effects.

Nplate™/romiplostim — The most commonly reported side effect of romiplostim was headaches (35% in those taking romiplostim, 32% in those taking the placebo). Other reported side effects included dizziness, insomnia, pain in arms or legs, abdominal pain, shoulder pain, indigestion, and paresthesia (feeling of pins and needles in hands and feet). Potential adverse events included bone marrow reticulin deposition and worsening thrombocytopenia after discontinuation of romiplostim therapy. Other risks include blood clots and potential for increased blasts in patients with myelodysplasia.

More information on Nplate™/romiplostim is available at PDSA's Web site. See www.pdsa.org and click on 'Treatments'.

PROMACTA®/eltrombopag — The most common side effect reported in short-term studies was headaches, in 8% and 11% of patients receiving eltrombopag and placebo, respectively. Other common side effects occurring in at least 5% of eltrombopag patients included nausea, common cold, diarrhea, and vomiting. Potential adverse events of eltrombopag

include development of reticulatin in bone marrow, changes in liver function tests, and a drop in platelet count after stopping the drug. Other risks include blood clots from excessive platelet formation and potential for increased blasts in patients with myelodysplasia. PROMACTA® may cause hepatotoxicity (drug-induced liver damage). Patients receiving therapy with PROMACTA® must have regular monitoring of serum liver tests.

If you need additional information about PROMACTA®, visit the PDSA Web site. See www.pdsa.org and click on 'Treatments'.

Other Factors Affecting ITP

Q What is the role of the bacteria *H. pylori* in ITP?

A Some studies from Italy and Japan have shown if the bacteria *Helicobacter pylori* (*H. pylori*) was present in the stomach and was treated and eradicated with antibiotics, many (37% Japan; 68% Italy) patients increased their platelet counts and recovered from ITP. However, studies performed in Spain, France, England and the U.S. showed less benefit. Indeed, in the United States, tests for *H. pylori* are not routinely performed when ITP is being diagnosed in the absence of gastrointestinal symptoms but the testing may be useful in some cases of chronic ITP.

Q What is the role of thyroid disease in ITP?

A A small percentage of patients with ITP have or will develop immune thyroid disorders (hyper- or hypothyroidism). Platelet survival is reduced in patients with hyperthyroidism (which may also impair response to ITP-directed therapy) and platelet production may be impaired in those with hypothyroidism, with platelet values returning to normal as the thyroid condition is corrected.

Alternative Treatments

Q What about alternative medicine treatments?

A Some patients report success with herbs, supplements, energy work, diet changes, and other alternative treatments. While there are many reported cases of their success, there are currently no formal studies. Many alternative treatments attempt to correct

underlying problems rather than treat the symptoms of the disease. These treatments tend to take longer to become effective and have fewer unwanted side effects. As with more traditional treatments, alternative treatments do not have the same results in all who try them. Be sure to tell your doctor if you decide to use alternative or complementary therapies.

Coping with the Emotional Aspects of Having ITP

Q Is there a standard solution for coping with ITP?

A There is no “one size fits all” solution. Patients receive their information about medications, treatments, nutrition, etc. from many sources. Situations and coping strategies vary based on a patient’s genetics, lifestyle, influences, location, allergies, treatments, and available care.

Q How does having ITP change the patterns and relationships of life?

A We are used to the everyday routines of life and we struggle with new patterns that arise when we are dealing with a major life stressor like a chronic illness. As you cope with ITP, there may be changes in relationships and patterns in your family that can be upsetting for those involved. Successful coping means everyone must recognize and adjust to the new reality.

Q How do I cope with feelings of helplessness and dependency?

A Patients with ITP may react to their own feelings of helplessness by not relying on others for fear of overwhelming them or losing their support or love. Remember that people *can* fit into your life when you have a chronic illness. Tell them what you need or would like. When you are experiencing fatigue, request additional help.

Q What are the roles for family and caregivers in supporting the ITP patient?

A Those who are supporting a patient with ITP need to be *clear* about their role as parent, friend, spouse, sister, or child. They don’t need to be the nurse or doctor. When they fulfill their role, they support the ITP patient.



Q How do I cope with the unpredictability of the disease?

A Unpredictability of the disease is one of the worst aspects of having ITP. There is a “roller coaster” effect in dealing with the chronic form of this disease. Understand that some of your plans may not work out. Realize that feeling distress at facing ITP is “normal.” It may help to join a support group to meet others who are coping with ITP.

Q How can ITP patients maintain some control over their treatments?

A People with chronic illness need some element of control, to know in some way they are managing what is happening in their lives. Even though patients listen to their doctor and specialists for advice, it's each patient's own choice whether to follow that advice. Sometimes adding complementary or alternative approaches to health such as energy therapy, yoga, or stress reduction techniques gives the patient additional coping strategies to supplement their medical care.

Q What helps children cope with the emotional aspects of ITP?

A Children need to be respected and included in care giving decisions. An “indirect” approach method that helps some children cope with ITP is called bibliotherapy, where a child reads a book or watches a movie that shows another child coping with the same or similar illness.

Feelings Experienced by ITP Patients

ITP patients may experience many feelings during the course of their illness, including anxiety, anger, and depression. These feelings may be caused by the illness itself, from the treatments given, and/or from the process of coping with a chronic illness.

Q How can ITP patients cope with feelings of anxiety?

A Anxiety occurs when a patient feels something bad is about to happen and they can't control it. Adding new coping skills and knowledge, such as learning to recognize your own signs of stress, keeping a diary of stressful situations that make you feel anxious, and recognizing negative thoughts and making the effort to substitute more helpful thoughts can help you cope with anxiety. Talking about your feelings with a trusted friend, family member or medical professional

is usually helpful. Having some control of medical decision-making can help relieve your anxiety. Talking with your doctor and understanding your treatment options can help you in making those decisions.

Q How can ITP patients cope with anger?

A Patients feel anger when they think their rights have been denied or when they feel they have received something bad they didn't deserve. Much of this may be a carryover from childhood when we learned that when we did something "bad" we would be punished. So now we think, when things are going great, we must be doing something good, and when things are bad, we must have done something "bad". Patients need to realize that they didn't "cause" their ITP because they were "bad" and it won't go away if they are just "good" enough. Sometimes patients feel anger when they don't think their doctor is listening to them or offering them the best options for care. ITP patients must learn ways to understand and deal with their anger. They need to find a physician who will work together and collaborate with them.

Q What causes depression for ITP patients?

A Many people with ITP report being depressed. Several factors may cause this. One factor may be serotonin, a neurotransmitter carried by the platelets and delivered to the brain and other parts of the body. Serotonin helps regulate mood, so anything that interferes with serotonin processing could lead to depression. In ITP patients, the platelet count is low, resulting in less serotonin for the body. A second factor is that when a patient is dealing with a difficult, possibly chronic illness it can lead to feelings of isolation, fear, and anger. You feel that your own body has "turned against you." A third factor is the treatments for ITP. Many drugs used to treat ITP may have depression as a possible side effect.

Depression can also occur when we experience a "loss" such as the loss of the healthy person we once were. We can experience loss of our job, our family and children's activities, and community involvement. Patients with a chronic illness like ITP may experience loss of friends because the patient is not the same as they once were in their friend's world. The friend doesn't know how to cope with these changes and isn't sure how to adjust to loss of some aspect of the person the patient once had been.

Q What can help ITP patients cope with depression?

A ITP patients can cope with their depression by understanding that this can occur naturally as part of having a chronic illness like ITP. In addition, they must realize that treatments for ITP, such as prednisone, can contribute to the depression. It helps to know that when the treatment ends, the depression will begin to improve.

Ask your doctor about the psychological effects of medication that you take to treat your ITP. Some ways to cope with depression include self-relaxation through tapes, meditation, prayer, and energy therapy. Coping with depression might include taking an anti-anxiety medication or getting psychological counseling for a while. Ask for understanding from your family and friends as you cope with having ITP and side effects of the treatments. They need to know that your depression, mood swings, and irritability are not caused by anything they have done. Try to find activities that bring you joy and help you focus on the positive.

Eating a balanced and nutritious diet, eliminating sugar, and getting enough rest can also help in coping with depression.

ITP patients and those with platelet disorders should be careful about using SSRIs (selective serotonin reuptake inhibitors), such as fluoxetine (Prozac®), paroxetine (Paxil®), sertraline (Zoloft®). These medications treat a variety of anxiety or mood disorders and depression. Studies found that SSRIs block serotonin uptake into platelets, thus decreasing, reducing, or inhibiting platelet aggregation, adhesion, and agglutination (clumping of red blood cells). These are important steps in blood clotting. Using SSRIs could lead to bleeding problems for the ITP patient.

Q Why should I join a support group?

A Many patients or parents of children with ITP benefit greatly from meeting, talking or chatting online with others touched by ITP. People who have been through, or are going through, a similar circumstance can do more than sympathize with you — they can relate to what you are going through and keep you from feeling alone. Support groups are a place for people to give and receive emotional and practical support as well as to exchange information. People with ITP, as well as their friends and families find support groups to be a valuable resource.

“At first I was
afraid, I was
petrified... but I
tried to educate
myself, to stay
positive, and as
time went by, I
felt less afraid and
knew that I had
to do what a lot
of others have to
do — live one
day at a time.”

— SALLY



Q How can family members and caregivers support the ITP patient?

A To overcome feelings of anxiety, anger, and depression ITP patients must offer their friends, family, and colleagues ways to reconnect and to continue their relationships. Care and support received from others is so important in helping the patient have a positive attitude.

Patients are often shocked to be diagnosed with ITP. Most probably have not heard of the disease and have no idea what to expect. Their initial reactions may include confusion, fear, and stress. It is hard to grasp all the new terms and understand the treatment options in a crisis situation.

You need to ask your family, friends, and other caregivers to be extra patient. Tell them that you have much on your mind. You are learning as fast as possible and dealing with side effects of very potent drugs. Explain to them that when your platelet counts are low, you may feel pretty awful, tired, moody, and sometimes sad. Side effects of your medications may make you feel bad as well. Even though you may look just fine on the outside, your body is waging an incredible battle inside. This is exhausting work.

Q As an ITP patient, what do I tell my child, siblings, parents, friends, teachers, and other adults about the disease and what's expected?

A Describe for them what ITP is. Tell them you are trying to understand more about the disease yourself and will share more as you learn it. Let them know there are a number of treatments that can make your platelet count rise and your bleeding symptoms decrease. Tell them you appreciate their care and support during this difficult time. You can encourage them to visit the PDSA Web site or contact PDSA for more information.

Q What can I do to support my caregiver(s) and reduce their stress?

A A caregiver provides assistance to another person who needs their help. In providing this care, caregivers often suffer from stress themselves, which can lead to mental and physical problems, including fatigue, anxiety, and depression.

You can encourage your caregiver. Give them credit and recognize their accomplishments in helping you as you cope with ITP. Remind them to take care of their own health, including keeping their own doctors'



appointments, taking their medications, eating right, exercising, and getting enough rest. Encourage them to take a break each day, whenever they need it. Have them maintain their own social network and spend time with their spouse, children, and friends. Urge them to enjoy a treat every now and then, such as good food, or going out to dinner. Have them join a PDSA support group or see a professional counselor to provide an outlet for their concerns, frustrations, and fears. Ask them to learn as much as they can about your illness.

What Else Can I Do to Take Care of Myself?

Q How can ITP patients have more effective interactions with their doctors?

A Follow up with your care providers on a regular basis. Learn to be a collaborator with your hematologist and primary care doctor and learn ways to interact effectively with them. If you take medicine for another condition, find out if it will affect your platelet count. Tell your doctor if you are trying any complementary or alternative treatments. Partner in your care.

Some important skills to have when talking to your doctor include writing down important questions/issues before meeting with the doctor, asking the doctor to explain something you don't understand, and not leaving the important questions until the end of your visit when you may run out of time. Bring a list of your current medications and dosages. Before your visit check with your doctor's office to be sure they have important lab results/doctor's reports.

If you feel unsure of your ability to interact effectively with your doctor, bring a family member or friend with you. Take notes or ask the doctor if you can record the session for later review.

Q What else should ITP patients do to take care of themselves?

A Decide how you want to approach the disease and your life. Sometimes a chronic disease is a 'wake-up call.' Ask yourself if there is anything in your life that is 'bleeding you'. Determine whether it is something you want to change and then make the life changes necessary to correct the situation. As much as you can, surround yourself with positive life situations and good feelings. Take them in and enjoy the moment.

Listen to your body. If your body tells you to rest, do that. If you are thirsty, drink. If you crave sitting in the sun, find some (note: be sure to discuss with your health provider to be sure it is safe for you to be exposed to sun). Your body wants to heal and has a wisdom of its own to help that happen. Try to avoid allergens as much as possible and try to decrease your exposure to chemicals, such as those found in many cleaning products, paints, and garden pesticides. See the PDSA Web site for a list of substances that may lower platelet counts.

Learn as much as you can about the disease. Meet others with ITP through discussion groups, local meetings, and the Name Exchange Program. Learn as much as you can about treatments and the disease. Truly, it is up to you to learn and help heal yourself.

Staying Healthy to Cope With ITP

Q What specific dietary changes should an ITP patient make?

A To stay healthy as you cope with having ITP, follow as many of the dietary suggestions as possible.

- Eat a wide variety of food.
- Eat whole foods, such as whole grain cereals and brown rice.
- Avoid foods sprayed or treated with fertilizers and pesticides.
- Reduce sugar, including fructose, corn syrup, honey, and other sweeteners.
- Limit dairy, reducing or eliminating milk, cheese, ice cream, and yogurt from your diet, based on your reaction to these foods and your dietary needs.
- Use healthy fats, such as cold-pressed olive for cooking and baking.
- Eat green leafy vegetables and sea vegetables often, such as kale and collards. They contain calcium, minerals, and Vitamin K to help with clotting.
- Rely on lean, white fish, whole grains and beans, and nuts for protein.
- Drink alcohol in moderation.
- Avoid foods that can thin the blood, such as blueberries, red/purple grapes, garlic, onion, ginseng, ginger, and tomatoes.

*"It is so important
not to cancel
one's life plans
when you have
ITP, but to
rearrange them
instead."*

— SUE



- Avoid food and drinks containing quinine as these can lower platelets. See <http://www.pdsa.org/about-ityp/warnings.html> for more information.
- Avoid allergic foods. If allergic, get tested. Delayed food allergies can produce vague and difficult-to-diagnose symptoms.
- Chew your food well to aid digestion.
- Drink plenty of pure water at room temperature or above. Ice water can slow and hinder the digestive process.

"The body cannot heal itself without proper nutrition and sleep, but in today's society, we are on a go-go-go schedule, and people forget to sit back and relax."

— SUMMER

Q Will regular exercise help ITP patients cope with their illness and depression?

A Exercise is another good way to cope with ITP. Exercise and stretching will give you a measure of power over your body by helping you gain strength, lessen bone and muscle loss, and regulate blood sugar levels. Ask your doctor what restrictions, if any, you may have on physical activity.

More Information

Q Where can I learn more information about coping with ITP?

A The Platelet Disorder Support Association (PDSA) has more information on all of the topics in this pamphlet. There are hundreds of pages of information on the PDSA Web site, www.pdsa.org. The organization publishes a monthly e-news, a quarterly newsletter, and makes available other publications and articles. PDSA holds an annual conference, regional meetings and has set up local support groups around the US and in Canada. PDSA members have access to the Name Exchange Program to communicate with other ITP patients. PDSA continues to expand its programs to offer more services and reach more people.

Acknowledgments

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Depending on your circumstances, one of our other guidebooks may also be helpful:

ITP in Adults – Frequently Asked Questions

ITP in Teens – Frequently Asked Questions

PTI en Adolescentes -- Preguntas Frecuentes

ITP in Children – Frequently Asked Questions

When a Child Has ITP – A Resource Guide for Parents

Understanding ITP – A Story for Kids about Immune Thrombocytopenia

*ITP and the Female Lifecycle – Bleeding Issues in the Stages
of a Woman's Life*

*La PTI y el ciclo de vida femenino -- Problemas Hemorrágicos
en las distintas etapas de la vida de una mujer*

PTI en la adultez – Preguntas frecuentes

PTI infantil – Preguntas frecuentes

PTI chez l'enfant – Questions Fréquemment Posées

PTI chez l'adulte – Questions Fréquemment Posées

The Role and Function of Platelets in ITP – Frequently Asked Questions

*Health Insurance & Assistance Programs for ITP Patients –
Frequently Asked Questions*

Living with ITP – Frequently Asked Questions

Cómo Sobrellevar la PTI – Preguntas Frecuentes

Vivre avec le PTI: Questions Fréquemment Posées

For more information about ITP, additional copies of this booklet, or to become a member of PDSA, please contact us:

Platelet Disorder Support Association

8751 Brecksville Road, Suite 150
Cleveland, OH 44141

tel 1-87-PLATELET (1-877-528-3538)

fax 844-270-1277

pdsa@pdsa.org

www.pdsa.org

The Platelet Disorder Support Association is dedicated to enhancing the lives of people with ITP and other platelet disorders through education, advocacy, research and support.

Membership benefits include a newsletter, discounts to the ITP Annual Conference, optional participation in the Name Exchange Program, and the good feeling of helping others.

PDSA is a 501(c)3 organization. All contributions are tax deductible.

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The information in this guide is for educational purposes only. For your unique medical condition, please consult a qualified medical doctor and/or health care provider.



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-

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Please send me fundraising information.

☐ I would like to receive an *ITP Emergency ID* card (1st one is free)

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Please help us update our records by completing this section of the form:

I am: ☐ an ITP patient ☐ parent of an ITP child ☐ family member
☐ friend/other ☐ health professional ☐ industry professional

For additional information about ITP and PDSA visit our Web site:

www.pdsa.org or send email to pdsa@pdsa.org

SEND THIS FORM TO: Platelet Disorder Support Association
8751 Brecksville Road, Suite 150
Cleveland, OH 44141

Call the PDSA office if you need assistance or to use a credit card:
877-528-3538 (toll-free) or (440) 746-9003

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for People with ITP

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