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Answering TTP Foundation  
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## EXECUTIVE DIRECTOR'S TTP STORY

I regularly suffer from visual migraines, so when I couldn't see through a dark hole in the center of my vision on September 2, 2008, I was not overly concerned. But over the next 10 days, the course of my life changed. I went from being an indestructible 28 year old newlywed, to a patient faced with a life threatening autoimmune blood disorder creating blood clots throughout my body. I was lucky that the irreversible damage was limited to my eyes.

My first 3 week stay in hospital for treatment was confusing and filled with tremendous anxiety. I had never heard of TTP before, and besides the description from the doctor, my only sources of information were the internet (filled with many scary stories) and complex papers from medical journals.

I relapsed on November 13, 2009, on June 17, 2010, and the latest relapse was diagnosed on November 25, 2011. Each time I have endured the plasma exchange treatments, and both myself and my supporters have felt the side effects of the dreaded prednisone. Moreover, I have completed two rounds of Rituximab, a drug approved for Lymphoma (cancer) and more mainstream autoimmune diseases, in hopes of reducing the chance of further relapse by "knocking out" some of my immune system.

I was not well informed and proactive in participating in a clinical trial prior to my second relapse and this meant that I was excluded from clinical trials for Rituximab. Not only does this mean that I've had to apply for compassionate coverage from the drug company twice, but my results will not help expedite the potential availability of this drug to all TTP patients.

I remain baffled as to how I ended up with this very rare disorder and I am concerned for the future that I used to take for granted. But I am also hopeful, because together we will raise awareness and money for research and patient care! Where there is research, there is hope for a cure!

Join the Answering TTP Community today to connect with other patients, stay informed and participate.

Take care,

*Sydney Kodatsky*

Executive Director, Answering TTP Foundation  
Sydney@AnsweringTTP.org

## PROGNOSIS

Without treatment 95% of patients succumb to the disease, however; with treatment 80 – 90% of Idiopathic TTP patients achieve remission. Of these, about 30% will relapse and early detection of such a flare of the disease is critical to minimize the risk of death or irreversible injury to vital organs.

## SUPPORT

A TTP diagnosis is scary and complex. Many patients have never heard of this 3 letter acronym before, nor do they have any idea of its ramifications. Moreover, patients are told over and over that we just don't know:

- why it happens
- what may trigger a relapse from remission
- why some patients relapse and others do not
- what the long term prognosis is
- how to ease treatment
- how to cure TTP



**Patient Connect** is an initiative designed to connect newly diagnosed TTP patients with more seasoned patients to provide peer support. Register at [www.AnsweringTTP.org](http://www.AnsweringTTP.org) or call us (416) 792 4656 | toll free 1-888 506 5458.

## Answering TTP Together

The purpose of Answering TTP Foundation is to help find answers to these questions by connecting patients and supporters. Together we can support each other, raise awareness and raise funds towards support programs, treatment and research. Join the Answering TTP community today. Register at [www.AnsweringTTP.org](http://www.AnsweringTTP.org) or complete the mail in tear off form attached. Each person touched by TTP brings further insight to the community.

Answering TTP is committed to connecting patients through support group meetings and other events. All community members will receive our electronic quarterly newsletter. Our website [www.AnsweringTTP.org](http://www.AnsweringTTP.org) contains additional information and links to help patients and supporters with this complicated disease.

*Note: The content herein is intended for informational purposes only, and is not meant to substitute consultation from a recognized health professional. Special thanks to the Canadian Apheresis Group for reviewing the content herein, and to Octapharma Canada Inc. for contributing publishing funds.*

# Thrombotic Thrombocytopenic Purpura

## INFORMATION FOR PATIENTS & SUPPORTERS



[www.AnsweringTTP.org](http://www.AnsweringTTP.org)  
[Contact@AnsweringTTP.org](mailto:Contact@AnsweringTTP.org)  
(416) 792- 4656 | Toll free. 1-888-506-5458  
Charitable Registration # 84600 4802 RR0001

## WHAT IS TTP?

Thrombotic Thrombocytopenic Purpura is a rare blood disorder that is considered a true medical emergency. TTP is diagnosed at a rate of 3-4 in 1 million people per year. Potentially fatal complications can result from internal blood clotting, with damage to critical organs such as the brain, heart and kidneys.

The cause of TTP continues to evade us. What is known is that blood becomes "sticky" and forms clots in blood vessels throughout the body. These clots are made up of platelets, one of the elements in blood. Vital blood flow to the body's organs is restricted, placing the organs at risk for damage due to a lack of oxygen and nutrients from the blood.

Moreover, since platelets are being used to form numerous unnecessary blood clots, their availability to perform their normal function, which is to seal injury sites to prevent excess bleeding, is compromised. Therefore, life threatening bleeding may occur.

Research has shown that in some cases the ADAMTS 13 enzyme is deficient. This finding can be used to explain blood clotting; however, while ADAMTS 13 enzyme deficiency is found in hereditary TTP cases, this is not always true of adult acquired cases of adult TTP. So we know that there is more to the recipe for TTP. Much more research is required!

## TYPES

### Hereditary TTP

Less than 10% of TTP cases are due to an inherited deficiency or abnormality of the ADAMTS 13 enzyme.

### Idiopathic or Acquired TTP

45% of TTP cases are of the idiopathic form, meaning there is no defined cause. Some cases have been linked to a decreased level of the ADAMTS 13 enzyme as a result of antibodies to the enzyme.

### Secondary TTP

45% of TTP cases are of the secondary form which is diagnosed when a predisposing factor is present including: autoimmune diseases; cancer; bone marrow transplantation; pregnancy; use of certain medications (quinine, platelet aggregation inhibitors, and immunosuppressants); HIV infection; pancreatitis; and hepatitis. Usually ADAMTS 13 activity is normal in secondary TTP.

## SYMPTOMS

- fatigue
- fever
- bleeding (from nose, gums)
- diarrhea
- chest pain
- kidney failure (dark urine, jaundice)
- neurologic symptoms (confusion, headaches, visual changes)
- thrombocytopenia (bruising, purpura, petechiae)

### 'EARLY DETECTION SAVES LIVES'

## KNOWN TRIGGERS

- pregnancy
- cancer
- infections and live vaccines
- underlying autoimmune conditions such as Lupus
- medical procedures, surgery and blood and marrow stem cell transplant
- medicines such as quinine, chemotherapy, ticlopidine, clopidogrel, cyclosporine A, hormone replacement therapy and estrogens

## DIAGNOSIS

A medical history indicating any of the listed triggers, and a physical exam for symptoms, in combination with a complete blood count (CBC), lactate dehydrogenase level (LDH) and blood smear are used to determine a diagnosis of TTP. An ADAMTS 13 enzyme level test may be used, not to rule out, but, to help confirm the diagnosis. Importantly, diagnosis and immediate treatment should not await the results of an ADAMTS 13 assay.

## TREATMENT

### Hereditary TTP

Monthly prophylactic plasma is administered to

patients to replenish and maintain adequate levels of functioning ADAMTS 13, the enzyme which the patient is unable to produce themselves.

### Idiopathic & Secondary TTP

In some patients the steroid, prednisone, has been used to slow the immune system and therefore the progression of this autoimmune disorder. The side effects of prednisone can be challenging and can include but not be limited to:

- increased appetite
- indigestion
- anxiety
- facial flushing
- sweating
- mood swings
- vision changes
- acne
- moon face
- easy bruising
- tiredness
- unusual hair growth

In all cases of idiopathic or secondary adult TTP, plasma exchange is the basic treatment of choice. Plasma exchange involves the use of automated machinery which permits the removal of the patient's plasma and replacement with donor plasma during a 3 to 4 hour treatment. Plasma exchange both removes antibodies and replenishes normal plasma proteins. To treat TTP, a series of daily or every other day plasma exchanges is used.



Plasmapheresis Blood Treatment  
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Other medications, such as Rituximab, and/or removal of the spleen are used when patients fail to achieve remission from first line therapy. Links to current TTP clinical trials and journal articles can be found on our website [www.AnsweringTTP.org](http://www.AnsweringTTP.org).

 **Answering T.T.P.**  
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## Join the Answering TTP Community

### ANSWERING TTP TOGETHER

Fill in the tear off form and mail or visit [www.AnsweringTTP.org](http://www.AnsweringTTP.org) to sign up electronically.

Name: \_\_\_\_\_

Address: \_\_\_\_\_

Tel/Cell: \_\_\_\_\_

e-mail: \_\_\_\_\_

\*Answering TTP community members are required to provide an email address to receive our quarterly newsletter and updates regarding our organization, upcoming support group sessions and fundraisers. If you do not have internet access please ensure your telephone number is accurate so that we can call you to arrange alternate communication.

I am interested in:

- participating in local support group sessions
- helping to organize support group sessions
- participating in local fundraisers
- organizing local fundraisers
- making a cash donation
- sharing my experience with TTP
- organizing local blood drives
- information about Peer Connect

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