Answering T.T.P.

Thrombotic Thrombocytopenic Purpura Foundation

INFORMATION FOR PATIENTS & SUPPORTERS

SYDNEY'S STORY

I was diagnosed with TTP in 2008 after my mom took life-saving action by seeking out a 3rd opinion. I was extremely lucky to not have experienced a heart attack or stroke due to the delay in diagnosis.

After almost 9 years, 7 relapses, plasma exchange using over 700 blood donations, spleen removal surgery, chemotherapy, and years of immune suppression medication, I am still learning to live with TTP and its effects. I believe that my non-targeted TTP treatments resulted in my subsequent battle with metastatic cancer at only 35 years old.

Despite these years of challenges I have remained hopeful. I founded Answering TTP Foundation because I have big ideas for our small and geographically dispersed patient group to ease the burden of living with TTP. Our recognized Canadian charity with international reach is founded on 2 beliefs:

#1. A worldwide patient and supporter effort to fund the most effective TTP research will propel TTP research forward. TTP treatment will be improved and we will find a cure. *I will watch my daughter grow up.*

#2. Peer support helps patients to help themselves. I knew nobody affected by TTP when I was diagnosed. Connecting with other patients brought me great piece of mind. *I want to ensure that even the most remote patients have that opportunity.*

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

Take care,
Sydney Kodatsky
Chair, Answering TTP Foundation

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 and heart.

Due to a deficiency in the ADAMTS13 enzyme, 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.

Video animation at
AnsweringTTP.org

TYPES OF TTP

Hereditary TTP (hTTP)

1% of TTP cases are due to an inherited deficiency or abnormality of the ADAMTS 13 enzyme.

Immune-mediated TTP (iTTP)

99% of TTP cases have no defined cause. In all cases, there is a decreased level of the ADAMTS 13 enzyme as a result of antibodies attacking the enzyme.

SYMPTOMS

- fatigue
- fever
- bleeding (from nose, gums)
- diarrhea
- chest pain
- abdominal pain
- neurologic symptoms (confusion, headaches, visual changes)
- thrombocytopenia (bruising, purpura, petechiae)

DIAGNOSIS

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

TREATMENT

Hereditary TTP (hTTP)

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

Immune-mediated TTP (iTTP)

Most patients receive steroids, e.g. prednisone, to slow the immune system and therefore the progression of this autoimmune disorder. The side effects of steroids can be challenging.

In all cases of iTTP, 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 iTTP, a series of daily or every other day plasma exchanges is used. Rituximab is increasingly used to achieve and maintain remission.

Other medications and/or removal of the spleen are used when patients fail to achieve remission from first line therapy.

PROGNOSIS

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

Pregnancy may be a trigger for women with both hTTP and iTTP. Women considering pregnancy should discuss their individual case with their TTP specialist. Research is showing an ADAMTS 13 assay to be helpful in evaluating the risk of relapse during pregnancy, and in suggesting a risk mitigating prophylaxis treatment.

A growing percentage of patients are recognized with anxiety, depression and neurocognitive deficits after recovering from an episode of TTP.

RESEARCH

Links to current iTTP and hTTP research and journal articles can be found on our website:
AnsweringTTP.org

Answering TTP Foundation

A TTP diagnosis is scary and complex. Many patients have never heard of this 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

The purpose of Answering TTP Foundation is to help find answers to TTP by funding TTP research, connecting patients and supporters, and providing education and support.

SUPPORT

TTP affects a small and geographically dispersed population. Visit **AnsweringTTP.org** to join the international Answering TTP community today for supportive newsletters, and to learn about peer, virtual, and face-to-face support initiatives. Each person touched by TTP brings further insight and strength to the community.

International TTP Day

Raising funds for Research

Yearly Event for Anyone, Anywhere.

Join the global effort to raise funds for international TTP research in your community.

www.AnsweringTTP.org
Contact@AnsweringTTP.org
Toll free. 1-888-506-5458

Charitable Registration # 84600 4802 RR0001

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 Answering TTP Foundation's Medical Advisory Board of experts for reviewing the content herein.