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WHAT IS TTP?
Thrombotic Thrombocytopenic Purpura (TTP) is a rare autoimmune 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 congenital 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

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

II. 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.

III. 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 and pancreatitis
- 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

IV. 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.

V. 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.

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 can be found on our website www.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 have they any idea as to 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 our organization, which is called Answering TTP, 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. So far we have raised over $22,000 for TTP research in 2010.
PATIENT 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 and I did not suffer damage to other vital organs.

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 source of information was the internet which was filled with many scary stories and complex papers on the subject.

After 9 months in remission, I was diagnosed with a relapse on November 13, 2009 and again on June 17, 2010. Again, I had the apheresis treatments and both myself and my supporters have felt the side effects of the dreaded prednisone. Moreover, I have completed a round 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 did this mean that I had to apply for compassionate coverage from the drug company, but my results will not help expedite the availability of this drug to all TTP patients.

I remain baffled as to how I got this very rare disorder in the first place 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
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Answering TTP Together
Join the Answering TTP community today by visiting www.AnsweringTTP.org and filling in the electronic submission form. All community members will receive our electronic quarterly newsletter.
Answering TTP is committed to connecting patients through support group meetings and other events. Moreover, our website www.AnsweringTTP.org contains additional information and links to help patients and supporters with this complicated disease.

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