

What is Thrombotic Thrombocytopenic Purpura (TTP)?

- TTP is a very rare blood disorder. Between 2 and 6 new cases per million people are diagnosed each year.
- It can affect people of all ages.
- It is a serious and life threatening condition.
- It is a life-long condition and patients may experience relapses after treatment for the initial event. Relapses can be managed and often avoided with careful follow up care.

Thrombotic thrombocytopenic purpura describes the three main features of TTP

T - Thrombotic refers to the formation of a blood clot.

T - Thrombocytopenic means the number of platelets (blood clotting cells) is reduced

P - Purpura refers to purple bruises caused by bleeding under the skin

- In health, the body aims to keep blood liquid when it is in the blood vessels and to clot when the vessel wall is damaged, to limit blood loss.
- When the body loses control of this process, abnormal clotting occurs which can lead to **strokes, heart attacks, deep vein thrombosis (DVT), kidney damage, pulmonary embolus (PE), etc.**
- The problem in TTP is a lack of an enzyme called **ADAMTS13**.

Common Sign and Symptoms of TTP

- **Headache**
- **Disturbed vision**
- **Jaundice (yellow tinge to eyes and skin)**
- **Stroke like symptoms**
- **Abdominal pain**
- **Anaemia**
- **Confusion**
- **Reduced platelet count**
- **Anxiety**
- **Dark urine**
- **Fever**
- **Chest pain**
- **Kidney problems**

It is important to remember that symptoms may differ from the list above and will be different for everyone. Symptoms at relapse may be different to those experienced when first diagnosed.

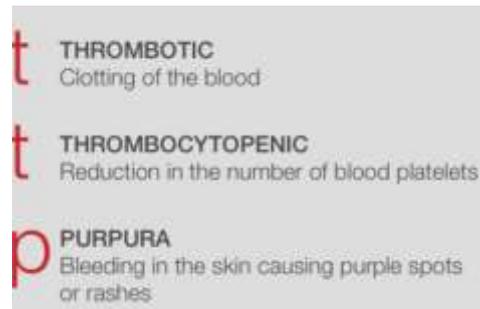
What happens during an episode of TTP?

- The body is missing ADAMTS13 so **blood clots can form anywhere** in the body. Where the blood clots form determines the type of symptoms that develop.
- The **platelet count is reduced** because the platelets are stuck in clots and therefore can't be measured.
- These blood clots make it difficult for the red blood cells to circulate around the body, breaking them up into small pieces. This can cause **anaemia** which can turn your skin yellow (**jaundice**), and lead to shortness of breath and fatigue (extreme tiredness).

Causes of TTP

There are two types of TTP

- In **Acquired TTP**, people develop an **antibody** that stops ADAMTS13 working. It is not known why this happens but sometimes it can be associated with other autoimmune conditions (where your immune system attacks healthy cells in your body by mistake), or it may be triggered by pregnancy or surgery.
- Very rarely, people can be born with TTP (called **Inherited or Congenital TTP**) and this means that there is a mistake in their genes that means that they don't make ADAMTS13. This is usually diagnosed in childhood.



Treatment of TTP

- When you are unwell with TTP, you need to be admitted to hospital.
- The main treatment is **plasma exchange therapy**, where the antibodies to ADAMTS13 are removed and you receive a transfusion of plasma as it contains ADAMTS13.
- **Steroids** and **Rituximab** (immunosuppressant agents) are given to stop your body producing the antibody against ADAMTS13.
- **Caplacizumab**, is a drug that stops the blood clots forming.
In congenital TTP, as there is no antibody, patients are usually just given a **plasma** transfusion.

Monitoring patients after an episode of TTP

- Patients who have had an episode of TTP need long-term monitoring by a specialist doctor
- Platelets, haemoglobin, LDH and ADAMTS13 levels are monitored after treatment has finished.

Useful link:

www.tpnetwork.org.uk

- (1) National Institute of Health. What Is Thrombotic Thrombocytopenic Purpura? [online]. 2014. Available at: [https://www.nhlbi.nih.gov/health/health-topics/topics/ ttp](https://www.nhlbi.nih.gov/health/health-topics/topics/ttp)
- (2) National Institute of Health. What Causes Thrombotic Thrombocytopenic Purpura? [online]. 2014. Available at: [https://www.nhlbi.nih.gov/health/health-topics/ topics/ttp/causes](https://www.nhlbi.nih.gov/health/health-topics/topics/ttp/causes)
- (3) Zheng et al. ISTH guidelines for treatment of thrombotic thrombocytopenic purpura. Journal of Thrombosis and Haemostasis (2020)

October 2020 CUH

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Thrombosis Ireland
Spot The Signs... Save A Life

Registered Charity No: 20154240

Thrombotic Thrombocytopenic Purpura (TTP)



Patient Information Leaflet



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