

ttpnetwork

THROMBOTIC
THROMBOCYTOPENIC
PURPURA
(TTP)

*A GUIDE TO
UNDERSTANDING TTP
FOR PATIENTS, CARERS,
RELATIVES AND FRIENDS*

We hope you have found the information in this leaflet helpful. If you have any questions or feedback please email us at:-

jo@ttpnetwork.org.uk

The medical information within this document has been verified as accurate by:-

We have written this booklet to help patients, carers, families and friends understand the rare blood condition known as TTP.

The following pages explain the disease and the many feelings and emotions a patient might experience when diagnosed with TTP.

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Transfusion Liaison Nurse Manager, National Blood Service

Anja Tusold,
*Research Nurse, Clinical Haematology,
John Radcliffe Hospital, Oxford*

The survival of a TTP patient is reliant on blood donations. Even though once you have received blood you will be ineligible to donate blood yourself, you can still help others by encouraging your friends and family to do something amazing -

GIVE BLOOD - 0845 7 711 711
<http://www.blood.co.uk>

Acknowledgements:-

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Post - Trauma Stress, Frank Parkinson (Sheldon Press)



TTP PATIENT AND CARERS GUIDE

It might help to make contact with others who understand what you as a patient, family or friend have been through because they have been there themselves. There is a UK based patient support group called TTPNetwork.

This is run via the internet with a website. There is also a message board where patients, ex-patients, relatives and friends can leave messages for one another. They ask questions and share experiences. Some people just visit the website and the message board, but never leave a message. This is fine. We would rather you came and got some answers by reading other messages or found some support (even if you don't leave a message) than sit feeling lonely and scared.

The website address is :-

www.ttpnetwork.org.uk

You will find that the internet has many sites containing information - be cautious and always check with your consultant before taking advice from the internet. It's important to remember that anyone can put information onto the World Wide Web and its accuracy cannot always be ensured.

Thrombotic Thrombocytopenic Purpura (TTP) is known, in the majority of patients, as an autoimmune condition. It is a rare blood disorder.

Below we describe what each word of the name means:-

THROMBOTIC = *Clotting of the blood;*

THROMBOCYTOPENIC = *Reduction in the number of platelets in the blood;*

PURPURA = *Bleeding in the skin causing purple spots / rash;*

People with TTP have a fault in the enzyme that breaks down the von willebrand factor, which, with platelets, normally prevents bleeding. This enzyme stops working properly and the platelets become sticky and misshapen.

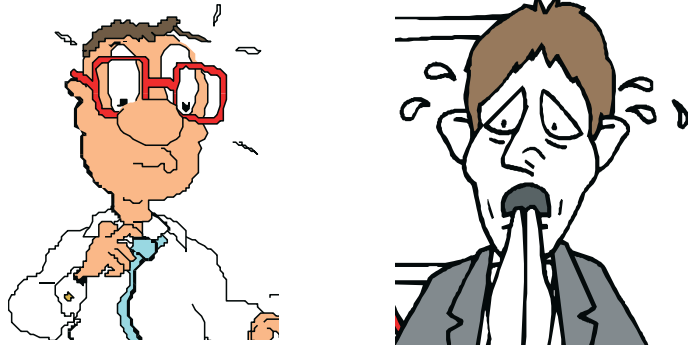
This causes them to clump together and form small clots in the body, typically in the blood vessels supplying the brain and kidneys. This is dangerous and could lead to strokes or failure of the major organs.

Whilst the platelets are clotting in this way they are not available for use at a site of injury - where you would expect them to be working. The normal function of platelets is to work with clotting factors in plasma to form a plug that stops, or prevents bleeding.

Because they are misshapen they become an obstacle to the red cells that are trying to travel around the body. This causes the red cells to break up. You may hear this referred to as fragmentation

SYMPTOMS OF TTP

A patient becomes anaemic and would feel tired, they might have headaches. They may be feeling anxious, confused or even paranoid about things around them. They might have a fever or stomach upset.



These are all typical symptoms of TTP.

A physical sign that the platelets are not working properly would be bruising or a rash on the body that looks like pin pricks. When this rash appears the platelets are already very low in number and a patient is very unwell.

If the disease progresses without treatment it is more likely the patient will collapse or become seriously ill.

The following pages describe some of the feelings and processes involved when a patient has been diagnosed with TTP.

LIFE AFTER TTP

When a patient is first diagnosed or after they are discharged from hospital they might continue to feel emotional.

When we are presented with a sudden shock or trauma such as being seriously ill, it's normal to put all our energy in to getting better. Once the initial crisis is over and we return to our normal life, no longer having medical staff fussing over us - things might feel odd. It's common to feel anxious, or to have flashbacks or trouble sleeping. These are completely normal reactions to an abnormal situation.

Post Traumatic Stress is a term often used to describe such reactions.

“ Post traumatic stress is the development of certain symptoms or reactions following an abnormal event. The event is abnormal in that it is life threatening or extremely disturbing, and can be anything from a minor accident to a major disaster. This includes other incidents such as a divorce, riots, war, bereavement or any event which causes trauma and shock. This trauma is the disturbance of our normal life beliefs, and turns our world upside down causing confusion, disbelief, feelings of vulnerability, a loss of meaning and purpose in life, and changes in self-image or self-esteem.”

Frank Parkinson - Post-Trauma Stress, (Sheldon Press)

The advice is: ‘ DON'T SUFFER IN SILENCE! ‘. Talk to the doctor or nurse. It may help to seek Counselling. The benefit of talking to a counsellor is that he or she will not know the patient or family / friends. Worries and concerns can be shared without fear of being judged. It may take more than one session before any benefit is felt. Allow some time and don't be afraid to cry. The counsellor will have seen this before and will not be uncomfortable.

The counsellor will not give advice or make judgements, but by listening and talking with the client (patient or family / friend) they can provide the time and space needed to come to terms with what has happened.

PROGNOSIS / SIDE EFFECTS

As recently as 15 years ago 80% of patients did not survive TTP. This statistic is now reversed. At least 80% of people now survive, but the key to this figure is in early diagnosis and treatment.

Many people only have TTP once, and go on to lead perfectly healthy lives. Others may relapse. There seems to be no pattern to either length of remission, age, sex or race of patient. Many patients that have relapsed several times still lead full and rewarding lives.

Sometimes people find they have side effects from their TTP experience. Those who were more severely ill might have more debilitating problems due to a stroke, or organ failure. Others sometimes find they have side effects such as joint pains, fatigue and vision or memory loss of varying degrees. These symptoms might appear worse when they are tired or under stress. Some of the side effects may lessen over time.

There is no 'one size fits all' for the experience of TTP as everyone seems to have a different story to tell. Doctors are becoming more experienced in treating the condition. Early diagnosis and perseverance with the treatment regime is essential.

The leading experts in the UK are Professor Sam Machin, Consultant Haematologist and Marie Scully, Consultant Haematologist and the team at London's University College London Hospitals. They have been conducting studies and have identified the enzyme to blame. Work continues.

Once a patient has been discharged from hospital and are trying to return to normal life, or are discovering a new type of 'normal' - they should pay attention to their body. If feeling tired, then take a rest. If feeling unwell, contact the consultant and ask for a blood test.

If increased bruising and / or a pin prick rash is noticed, even if they don't feel particularly unwell - contact the consultant and ask for a blood test.

It is better to have a wasted journey than to find out the patient has relapsed and not received treatment as soon as possible.



FEELINGS

When a patient is told they have TTP they may feel many different emotions. These might include :-

***SHOCK! ~ CONFUSION! ~ ANGER!
SADNESS! ~ FEAR! ~ RESENTMENT!***

All of these feelings are perfectly normal reactions to a unique situation.

It might have taken a long time for Doctors to diagnose TTP. This is quite common as the disease is estimated to affect only 1 to 4 people in every million. Most doctors will not have seen this condition before.

The patient may be feeling relieved that finally they have an answer to why they have been feeling so ill.

They may wonder ***"WHY ME?"*** It's not clear why TTP occurs in some people yet not in others.

As TTP is described as an autoimmune condition, its most common to have no obvious associated cause.

Sometimes there are suspected triggers such as:-

- QUININE
- COMBINED ORAL CONTRACEPTIVE PILL
- VIRUS / INFECTION
- PREGNANCY
- HIV

There may be other triggers too.



We know that you cannot 'catch' TTP. It is not contagious. Your body will probably have been susceptible to it, but there will have been a trigger as previously mentioned



TREATMENT

The recognised treatment for TTP is to start a procedure called PLASMA EXCHANGE (*Plasmapheresis*). This involves removing the damaged platelets which are suspended in the plasma in blood, and replacing them with donor plasma containing healthy platelets.

This process does not hurt but it might feel slightly uncomfortable. The patient may feel particularly tired after the procedure - this is normal. The patient will have tiny tubes inserted into veins to allow the exchange procedure to take place. This is done in one of two ways :-

- Either via 2 needles, which are inserted into the arms at the start of the process and removed at the end. (It's possible to ask for a local anaesthetic to ease the discomfort).

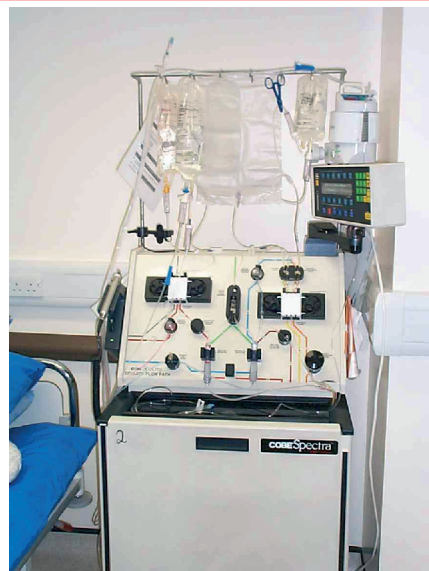
- Or via a plastic tube that is attached into a larger vein - known as a Central Line or a Hickman Line. This is a little bit like having a semi-permanent tap attached to the body. It will usually be fitted to the vein in the neck, groin or chest. It will be left in place for several days or even weeks. It is inserted under local anaesthetic.

The Central/Hickman Line might not look attractive, but patients may find it is more comfortable as it allows increased movement in the arms so they can read or write during treatment

The patient will be connected to a cell separator machine that carries out the exchange procedure.

As the blood comes out of the body into the tubes, it is spun in a centrifuge to separate the plasma and red cells.

The Plasma is discarded. The patients own red cells and the donor plasma are then returned to the body via the tubes



The patient may prefer not to see this. They can ask the nurse to cover the tubes.

Depending on the patients height and weight the exchange procedure will take several hours.

If the patient has any odd sensations whilst the blood is exchanged, they should inform the nurse immediately. Sometimes the process will reduce the Calcium levels in the body and this may be why the patient experiences sensations such as tingling. These nutrients can be replaced simply by taking a milky drink or a tablet.

Occasionally a patient will have an allergic reaction to donor plasma. Nurses will be looking out for signs of this and if it happens they will note the batch number to prevent it happening again.

Patients may need to have a plasma exchange once or twice a day for many days before their blood counts start to look normal again. It is recommended that regardless of how 'normal' their blood counts are - they should not have less than 7 days of plasma exchange.

Nurses talk of 'replacing a full volume', this simply means that the exchange process will flush the body's full volume of blood.

Steroids may be given at the same time as the exchange. This is to suppress the body's immune system and helps it accept the donor plasma.

Other drugs are sometimes used to help too. These might be chemotherapy drugs, but they are not used in the same way or at the same dose as they are with Cancer treatments, so patients are unlikely to lose their hair. TTP is *NOT* a Cancer.

More recently a drug called Rituximab is being used. This appears to help the enzyme work correctly. It can be used during an episode of TTP or when a patient is in remission. When in remission it might offer a longer remission period. It is *NOT* a cure.