

# HUS and TTP

## Haemolytic Uraemic Syndrome (HUS) & Thrombotic Thrombocytopenic Purpura (TTP)

HUS and TTP cause kidney problems and problems with the blood. They have different names but share a lot in common. The two disorders together are 'thrombotic microangiopathies', making TMA the third abbreviation.

[What are HUS and TTP?](#)

[What about E.coli and HUS?](#)

[What else can cause HUS and TTP?](#)

[What tests are needed?](#)

[What is the treatment?](#)

[How serious is it?](#)

[Where can I get further information?](#)

### What are HUS and TTP?

#### HUS:

- **H-aemolytic** part of the title is because the red blood cells are *haemolysed*, in which some of the blood cells are in fragments. The consequence of this is anemia (a low blood count) and fragments when looked at through a microscope.
- **U-remia** occurs when the kidneys are not working. Urea is a waste product which should be cleared when the kidneys are working properly. Kidney failure occurs because the lining of small blood vessels is damaged (endothelium) resulting in clotting in the very small blood vessels of the kidney causing kidney damage. This is a rare cause of kidney damage.
- **S-syndrome** is the name given to a group of symptoms. It does not reflect how severe it is or its cause.

#### TTP:

- **T-hrombotic** meaning clotting in the blood vessels.
- **T-hrombocytopenic** meaning a low level of platelets, tiny blood cells


which cause clotting.

- **P-urpura** meaning tiny bruises on the skin due to abnormal clotting.

Both of these diseases cause narrowing and blockage of small blood vessels because of tiny thromboses (clots). In these damaged vessels, red blood cells become broken up, and platelets are used up. This leads to anaemia, and low platelet counts, which can cause bleeding. The kidney may be affected in both disorders, but other organs may also be involved.

### **Are there different types of HUS and TTP?**

HUS mainly affects the kidney (as well as blood). A common cause in children is infection with a rare type of *E.coli* (see below). In adults, HUS is often not related to *E.coli* O157 and there is a list of several causes as described in the next section.

*E.coli* O157 is commonly found in cattle but may also be present in sheep, goats and other domestic animals 

**TTP** is quite similar but is more likely to affect the brain than HUS (although severe HUS can do this).

There may be other important differences between HUS and TTP, at least with some of the causes. HUS often seems to start with damage to tiny blood vessel cells. The first problem in many patients with TTP seems to be deficiency of a molecule in the blood called **von Willebrand protease**. Occasionally this is an inherited problem, but in other patients, an antibody in the blood seems to inactivate it.

### **What about E.coli and HUS?**

*E.coli* O157 is a rare subtype of the *E.coli* bacteria, a bug which lives in cattle and in other farm animals. It is a well publicised cause of gastroenteritis, with bloody diarrhoea. In Britain, it is more common in Scotland. Once infected with this bug there is a small risk that HUS may develop. The infection is more likely to occur in the summer months. It is much more common in children under 5 years and in very elderly people. Infection with this bug has occurred in outbreaks when several people become infected from one source. However most of the cases in

Scotland occur outside of the outbreaks and are called sporadic cases. Often the source of infection is never found. *E.coli* induced HUS follows the diarrhoea by 2-5 days. If you develop *E.coli* O157 gastroenteritis there is usually about a 5-10% chance you will get HUS.

### **What else can cause HUS and TTP?**

Reactions to some drugs can cause HUS. Quinine is a common one, but there are others, including some chemotherapy.

An illness very like HUS/TTP can develop in late pregnancy. It is usually short-lived but may be severe.

Inherited conditions sometimes cause diseases of this type. Patients with this type of HUS/TTP may get further attacks, and the condition may also affect transplanted kidneys if their own kidneys are destroyed. This type is called atypical HUS.

HUS/TTP sometimes occurs as part of other diseases, for instance after bone marrow transplantation, and very rarely in some kinds of cancer.

### **What tests are needed?**

Blood tests will show the presence of anaemia and fragments in the blood. The urine output will start to reduce and kidney blood tests will become abnormal. There is always a reduction in platelets (the tiny blood cells that cause clotting). Tests will be sent to check for *E.coli* O157. A kidney biopsy is often needed to show that HUS is causing the kidney problem, unless the cause is very clear (eg *E.coli* O157 infection). Tests for causes of HUS/TTP are improving.

### **What is the treatment?**

The illness varies from person to person, and depends on the probable cause. Sometimes the symptoms are mild and needs no further treatment, other than close observation. The main part of therapy is to treat symptoms as they arise and to prevent complications.

- People with this illness need to be treated in a specialised ward, usually in a renal unit.
- If the cause is known, it can sometimes be removed.
- If the blood count is very low, blood transfusions may be required.

- Platelets are usually given only if it is absolutely essential, as they may make the clots worse.
- Antibiotics do not prevent HUS caused by *E.coli* infection, and they may make it worse.
- High blood pressure is often present and needs to be treated.
- In some cases the kidneys may stop working and dialysis treatment may be needed.
- Plasma Exchange is used for patients with TTP and some patients with HUS.

### **Plasma exchange?**

This is a treatment which uses a machine to clear the blood of its own plasma and replace it with donor plasma. It may remove substances which are involved in making clotting worse and replace some deficient substances. It carries the same (low but real) risks of infection and reaction as blood transfusion, and large amounts of fresh plasma may be required.

**In HUS** plasma exchange has not been shown to make any difference to the outcome of the illness when it is caused by *E.coli*, and it is not recommended. It is recommended for most cases of 'atypical' HUS.

**In HUS/TTP occurring as a complication of pregnancy** it often seems to help.

**In TTP** occurring because of a lack of von Willebrand protease, plasma exchange can remove the antibody causing the deficiency (if there is one) and replenish the protease. It is the recommended treatment for TTP.

### **How serious is it?**

This is a serious condition, but as our understanding and treatment improves the prognosis also improves.

Survival of sufferers from HUS caused by *E.coli* O157 is now 90-95%. The kidney function may not fully recover in 1 in 10 of individuals with the illness. 40% of people may have resulting scarring of the kidneys which may affect kidney function in later life.

People who have HUS /TTP caused by other things are at greater risk of longterm kidney damage.

Severe disease may also affect blood vessels in other organs like the brain.

### **Where can I get further information?**

- There is a UK support organisation for patients, called [HUSH](#), which can be contacted at PO Box 159, Hayes, UB4 8XE.
- The [NKF](#) is the American National Kidney Foundation. Its information on HUS is relevant to HUS in childhood.
- [Lois Joy Galler](#) foundation is an organisation to provide information and promote research into HUS set up by Lois Joys parents in her memory.
- See our [links page](#) for other sources of information about kidney diseases.
- Rare renal pages on [STEC HUS](#) and [Atypical HUS](#).

### **Eculizumab**

This is a new treatment that seems to be effective in many patients with 'atypical HUS' caused by abnormal complement molecules. It is probably not effective in other types of HUS.

**Acknowledgements:** The author of this page was Gemma Browne. It was first published in March 2001, and extensively reviewed in March 20. The date it was last modified is shown in the footer.