A TAKEDA SUMMARY

Thrombotic thrombocytopenic purpura: recognise and refer

TTP is a rare but life-threatening medical emergency. Without prompt diagnosis and treatment, it can result in **irreversible organ damage or death.**[1]

In the UK, TTP affects approximately **6 people per million every year**.[2]

Women are notably affected, as TTP affects approximately **1 in 200,000 pregnancies**.[3]

Children are also impacted, with one study reporting that **38%** of cases are diagnosed in individuals **under 12 years old.**[4]

TTP requires urgent referral and immediate treatment.[1]

Without intervention, mortality rates are as high as 90%.[1] Even with treatment, **survivors remain at risk of long-term complications**, including strokes, heart attacks and organ damage.[1,2]

If you suspect TTP, urgently refer your patient to a specialised centre for diagnosis and treatment.[1]

Click or scan the QR code to locate your nearest specialised centre:[5]

This QR code will take you to an external website that is not owned, or controlled by Takeda Ltd.

WHAT IS TTP?

TTP results from severely reduced activity of the enzyme **ADAMTS13**.[1] Without sufficient ADAMTS13, platelet-rich blood clots form in small vessels, **leading to organ ischaemia and dysfunction**.[1]

TTP can be:

IMMUNE-MEDIATED

Acquired autoantibodies that inhibit ADAMTS13 activity;

>80% of all cases[1,2]

CONGENITAL

Inherited mutations in the ADAMTS13 gene;

2-10% of all cases[1,2]

	IMMUNE-MEDIATED	CONGENITAL
Age at diagnosis	Typically adults aged 40–60 years;[6] ~5–10% of cases occur in children[7]	38% in childhood; 62% in adulthood; 69% of adult cases occur during pregnancy[1,4]
Incidence	~5–6 cases per million people per year[2]	~1 case per million people per year[1]

TTP can mimic other conditions such as autoimmune haemolysis, pregnancy-associated HELLP syndrome and infections. In suspected cases, **urgent assessment of ADAMTS13 activity is essential**.[1]

SIGNS AND SYMPTOMS TO LOOK OUT FOR

International consensus defines TTP as **microangiopathic haemolytic anaemia, with moderate to severe thrombocytopenia** with associated organ dysfunction.[1]

For a complete list of signs and symptoms, consult the 2023 British Society of Haematology guidelines (QR code below).

Neurological symptoms are the most common, affecting ~70% of patients

including headaches, seizures, paraesthesia and speech changes.[1,2]

Cardiac involvement occurs in approximately 40% of cases

Patients may also present with gastrointestinal and renal symptoms.[1,2]

REFERRAL PATH FOR ACUTE TTP EPISODES

Urgent investigations

If TTP is suspected, order the following tests: FBC, reticulocyte count, blood film, LDH, coagulation screen, B12/folate, liver and renal function tests, troponin. **Do not wait for results to refer.**[1]

Discussion with specialist centre

Contact your nearest specialised centre urgently to discuss clinical findings and agree on diagnosis.

Plan for immediate transfer.[1]

Urgent transfer for plasma exchange

Immediate "blue light" transfer to specialised centre. Consider the safety of transfer and possible need for intubation where unstable or airway threatened (involve the anaesthetic team early).[1]

In acute TTP, the 2023 **BSH guidelines state that plasma exchange should be commenced urgently to replete ADAMTS13**. Plasma infusions are only indicated if there is an unavoidable delay in commencing plasma exchange.[1]

Resources: click or scan the QR codes to access

These QR codes will take you to an external website that is not owned, or controlled by Takeda Ltd.

BSH 2023 Guidelines
National guidance for the diagnosis and management of TTP in the UK[1]

UK TTP Registry
A nationwide database collecting clinical data and samples from newly diagnosed TTP patients to support research and improve care[8]

ABBREVIATIONS: ADAMTS13, a disintegrin and metalloproteinase with thrombospondin type 1 motifs, member 13; FBC, full blood count; HELLP, haemolysis, elevated liver enzymes, and low platelet count; LDH, lactate dehydrogenase; TTP, thrombotic thrombocytopenic purpura.

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