

TTP and HUS

A plain-English guide for you and your family

WHAT IS HAPPENING

Normally, a sticky protein called von Willebrand factor (vWF) floats in the blood in huge clumps. An enzyme named ADAMTS13 acts like scissors, cutting vWF down into safe, smaller pieces. Platelets can then do their clotting job without getting tangled. In TTP (thrombotic thrombocytopenic purple bleeding blotches (purpura)), ADAMTS13 stops working—either because the immune system attacks it or because a person is born without enough of it. Without those scissors, giant vWF tangles stay in the bloodstream and grab onto platelets, forming tiny clots (microthrombi) that clog small blood vessels all over the body. Red blood cells try to squeeze through these clogged, damaged vessels and get shredded, spilling their insides. Platelets are used up in the clots, so the count drops. The five classic TTP signs—fever, broken red cells, low platelets, brain problems, and kidney problems—don't always appear together, but the triad of low platelets, broken cells, and no other clear cause is the key. HUS (hemolytic uremic syndrome) looks similar but starts differently: Shiga toxin (usually from *E. coli* in contaminated food) or an overactive complement system (part of the immune system) directly damages the lining of blood vessels, especially in the kidneys. That injury also triggers clots, breaks red cells, and drops platelets. In HUS, bloody diarrhea often comes first, and kidney failure is the main danger. Both conditions are life-threatening. TTP requires emergency plasma exchange to replace the missing enzyme and wash out the antibodies. HUS is treated with supportive care—fluids, dialysis if needed, and sometimes a complement blocker (eculizumab) for the atypical form. Every finding traces back to one upstream break: no enzyme scissors in TTP, or vessel injury in HUS.

What you might feel

- Broken red blood cells (schistocytes) on the blood smear
- Very low platelet count (thrombocytopenia)
- Low haptoglobin and high LDH in the blood
- Dark or tea-colored urine
- Sudden confusion, trouble speaking, or weakness on one side of the body
- Sharp drop in urine output or swelling in the legs and face

- Fever (more common in TTP)
- Severe abdominal pain or bloody diarrhea (more common in typical HUS)

Call the doctor right away if...

- New confusion, slurred speech, or one-sided weakness (signs of stroke or brain bleeding)
- Urine output drops sharply or stops completely (acute kidney failure)
- Platelet count falls dangerously low (often below 20,000) with active bleeding (nosebleeds, bleeding gums, or bruises that spread quickly)
- Severe chest pain or trouble breathing (possible heart or lung clots)
- Seizures or sudden loss of consciousness

What the care team will do

- Monitor neurological status every 2 hours using Glasgow Coma Scale and check for new confusion, headache, vision changes, or seizure activity
- Measure strict intake and output every hour and notify provider immediately if urine output drops below 0.5 mL per kg per hour
- Hold all platelet transfusions unless approved by hematology and the patient has life-threatening bleeding
- Coordinate daily plasma exchange treatments through a large-bore central line and monitor vital signs every 15 minutes during the procedure
- Monitor hemoglobin, platelet count, LDH, and haptoglobin levels daily before morning rounds
- Assess for signs of bleeding including checking all IV sites, gums, stool, and urine for blood and applying pressure for 10 minutes to any puncture sites
- Avoid all unnecessary needle sticks, use electric razors instead of blades, and keep fall precautions in place at all times
- Monitor for signs of fluid overload including lung sounds, weight, and peripheral swelling (edema), especially if kidney function is declining

Medicines you may be given — and why

Plasma exchange (plasmapheresis)

In TTP, the body attacks ADAMTS13 (the enzyme that cuts up big sticky von Willebrand factor proteins). Without it, huge sticky clumps form in the blood and trap platelets, blocking tiny vessels. Plasma exchange pulls out the patient's plasma (the liquid part of blood containing the

attacking antibodies) and swaps in fresh donor plasma that has working ADAMTS13. This stops new clots from forming and lets the body recover.

Corticosteroids (prednisone or methylprednisolone)

Steroids quiet down the overactive immune cells that are making antibodies against ADAMTS13. They block the inflammatory signals (like NF-kappa-B inside immune cells) so fewer antibodies are produced and the attack on the enzyme slows.

Rituximab

Rituximab is a lab-made antibody that locks onto CD20 (a marker on the surface of B cells, the immune cells that make antibodies). Once rituximab sticks to CD20, it marks those B cells for destruction. Killing the B cells stops them from churning out the anti-ADAMTS13 antibodies, so the attack on the enzyme ends and TTP is less likely to come back.

Caplacizumab

Caplacizumab is a tiny engineered antibody piece that grabs onto von Willebrand factor (the sticky blood protein) and covers the spot where platelets normally attach. Even though the big sticky clumps are still floating around (because ADAMTS13 is missing), platelets can't stick to them anymore, so the dangerous clots in tiny vessels stop forming right away.

Supportive care for STEC-HUS (fluids, dialysis if needed)

In typical HUS (caused by Shiga toxin from E. coli O157 or other STEC bacteria), the toxin damages the cells lining blood vessels in the kidneys and gut. The best treatment is careful IV fluid to keep blood flowing through the injured kidneys and dialysis (a kidney machine) if the kidneys stop making urine or toxins build up. Antibiotics are avoided during the diarrhea phase because killing the bacteria can make them dump more toxin all at once and worsen HUS.

Eculizumab (for atypical HUS)

Atypical HUS happens when the complement system (a cascade of immune proteins that normally punches holes in bacteria) is stuck in the 'on' position and attacks the body's own blood-vessel linings, especially in the kidneys. Eculizumab is a lab-made antibody that blocks C5 (one of the key complement proteins). By stopping C5, it halts the formation of the membrane attack complex (the final hole-punching weapon), so vessel damage stops and the kidneys can heal.

Red blood cell transfusion (as needed for severe anemia)

Both TTP and HUS cause destroying red blood cells (hemolytic) anemia (red cells break apart as they slam through tangled clots in tiny vessels). If hemoglobin drops so low that the patient is dizzy, short of breath, or the heart is racing dangerously, packed red blood cells are transfused to carry oxygen while the underlying disease is treated.

Where this information comes from

- ASH (American Society of Hematology) 2020 TTP guideline
- KDIGO (Kidney Disease: Improving Global Outcomes) clinical practice guideline for acute kidney injury
- CDC information on hemolytic uremic syndrome
- NEJM review: Thrombotic Microangiopathies

Last reviewed: 2026-07-07