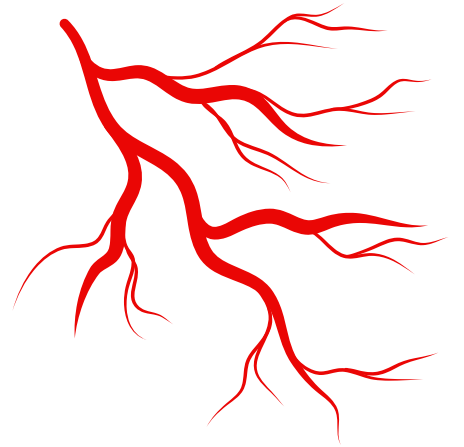




The **Blood** Project

Patient Information

Immune Thrombotic
Thrombocytopenia
Purpura (TTP)



A Pocket Resource for Patients

William C. Aird, MD



What does TTP Stand For?

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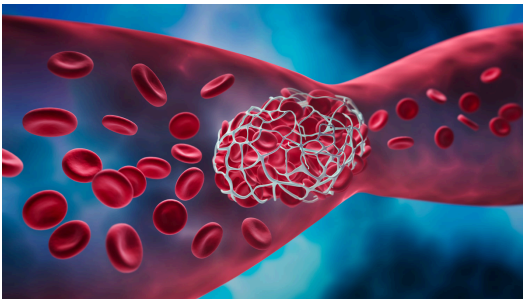
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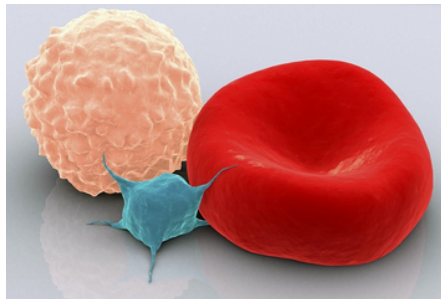
Thrombotic

Thrombocytopenia

Purpura



Thrombosis
(blood clot)



Thrombocytopenia
(low platelet count)

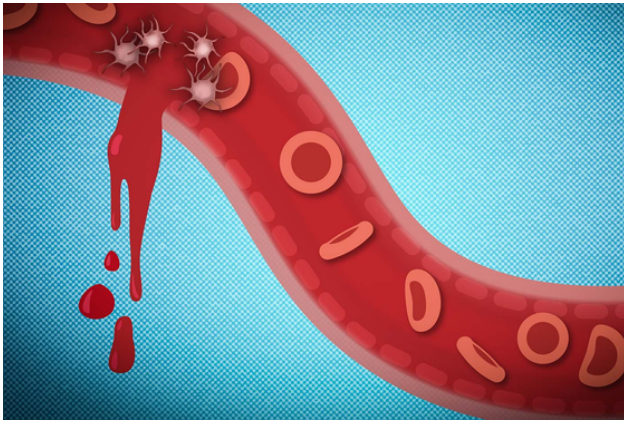


Purpura
(bleeding into skin)

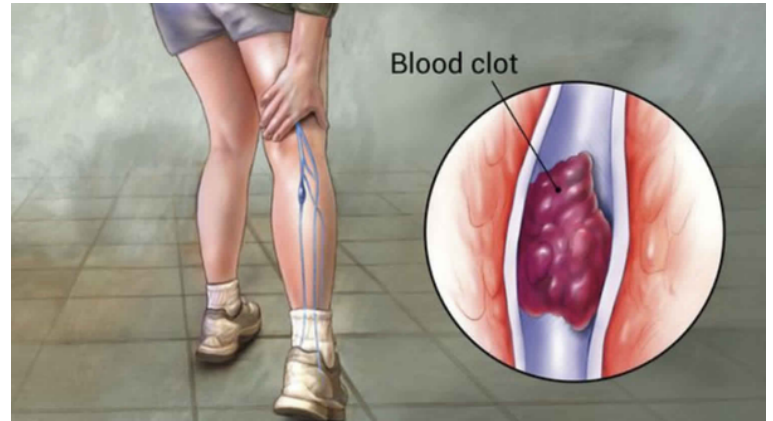


What is Thrombosis?

Formation of a blood clot (thrombus) inside a blood vessel, obstructing normal blood flow



Normal clotting
(plugs the hole in vessel wall)



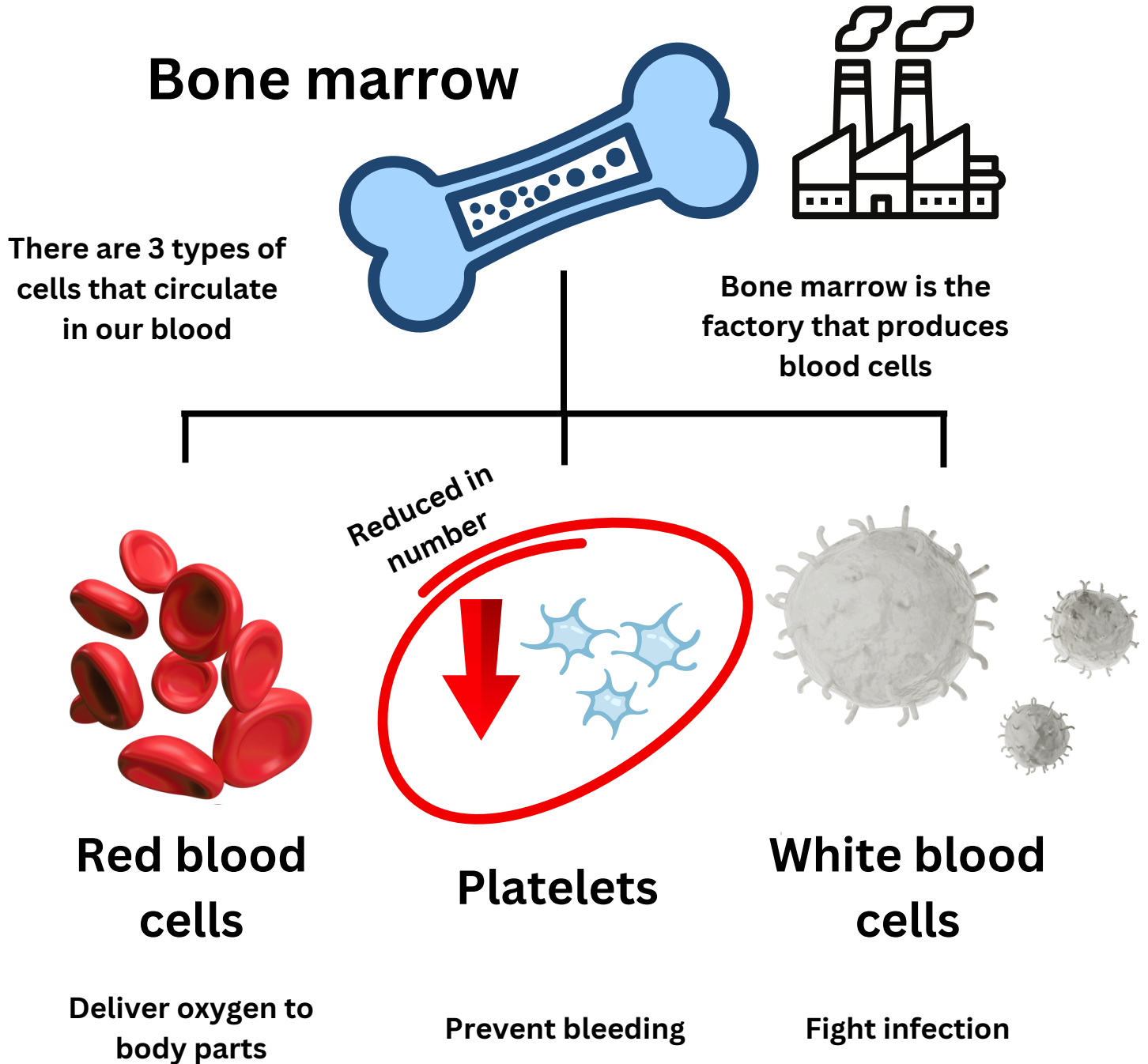
Thrombosis
(plugs the lumen of the blood vessel)

- Our **heart** pumps blood around the body
- This pumping function generates **blood pressure**
- Blood is normally contained within **blood vessels**
- Blood vessels can leak from normal wear and tear
- The **clotting system** evolved to patch defects in the blood vessel wall
- It consists of **platelets** which rush to the site of injury, and the formation of a glue called **fibrin** (think of the platelets as the bricks and fibrin as the mortar)
- In some diseases, the clotting system is overactive and leads to spontaneous brick-and-mortar formation inside otherwise healthy blood vessels
- Instead of patching holes, these “clots” or “**thrombi**” can obstruct the lumen (the tube) of the blood vessel and impede blood flow to body parts

What is **Thrombocytopenia**?

Thrombocyte = platelet Penia = low

Low platelet count (low numbers in blood)



What is purpura?

Bleeding in the skin



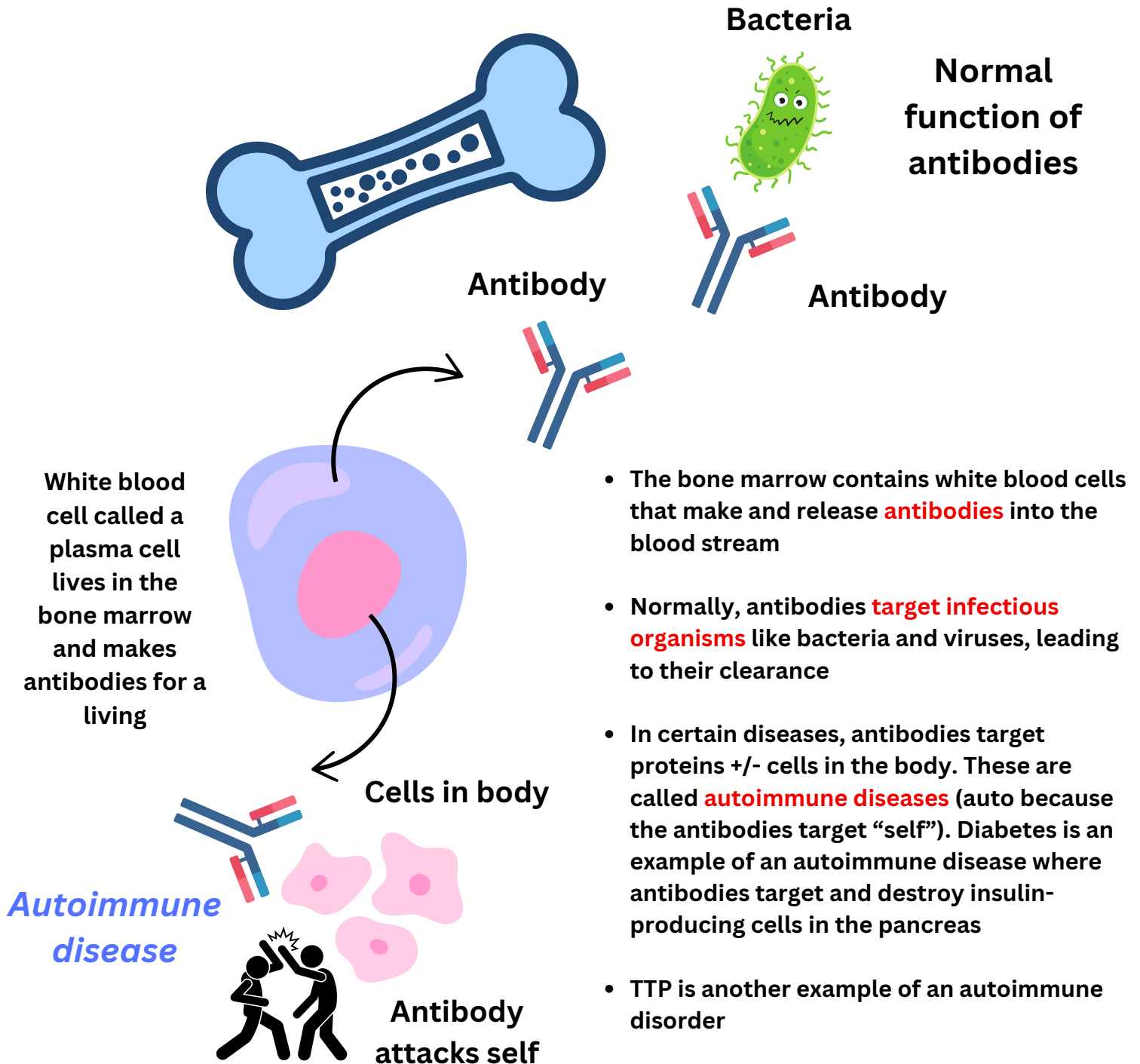
Purpura causes red, purple or brown blood spots on your skin. It happens when small blood vessels leak blood under the skin's surface.

Purpura and other types of bleeding occur in TTP because the platelet count is often very low and platelets normally function to prevent bleeding

What Causes TTP?

TTP is an autoimmune disease

Bone marrow



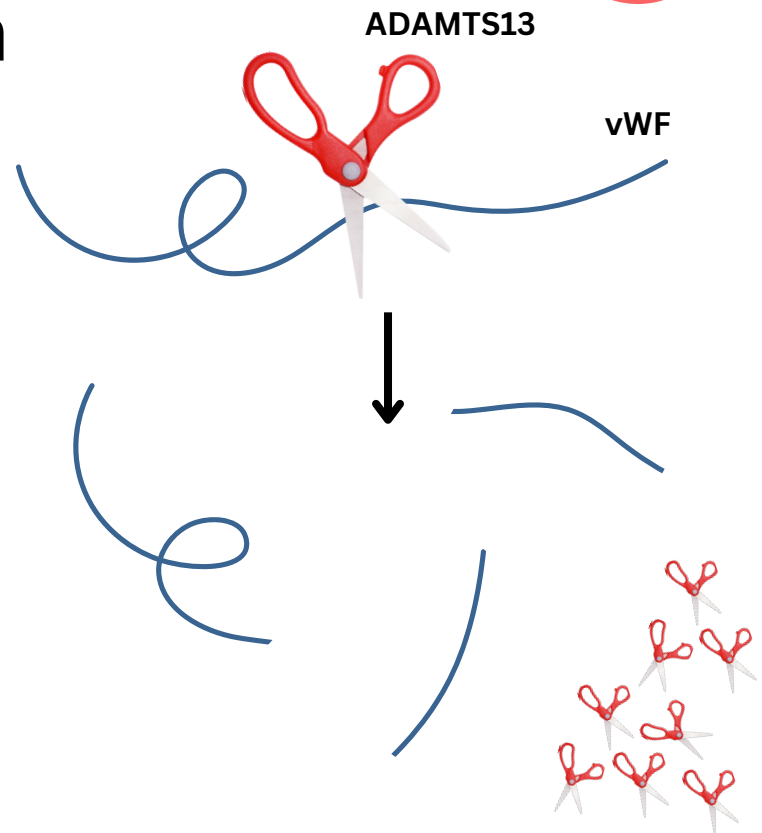
What Causes TTP?

Antibody against a scissor-like protein

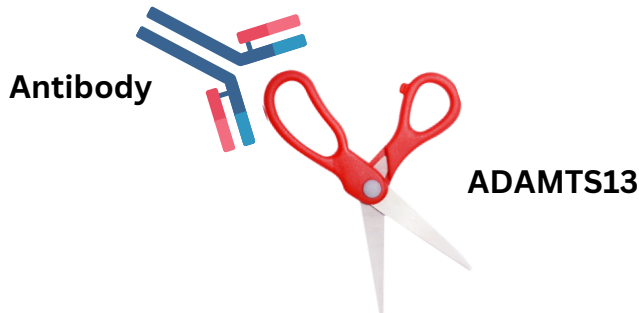


Healthy condition

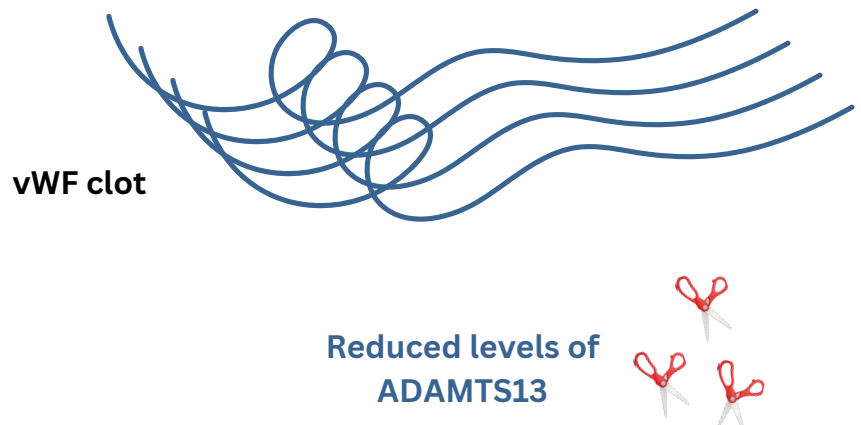
Normally, a scissor-like protein called **ADAMTS13** cleaves another protein in the blood called **von Willebrand factor (vWF)**. vWF promotes clumping of platelets (the bricks in the brick-and-mortar analogy) in blood vessels. So, ADAMTS13 is a protective mechanism to prevent excessive platelet clots/plugs



TTP



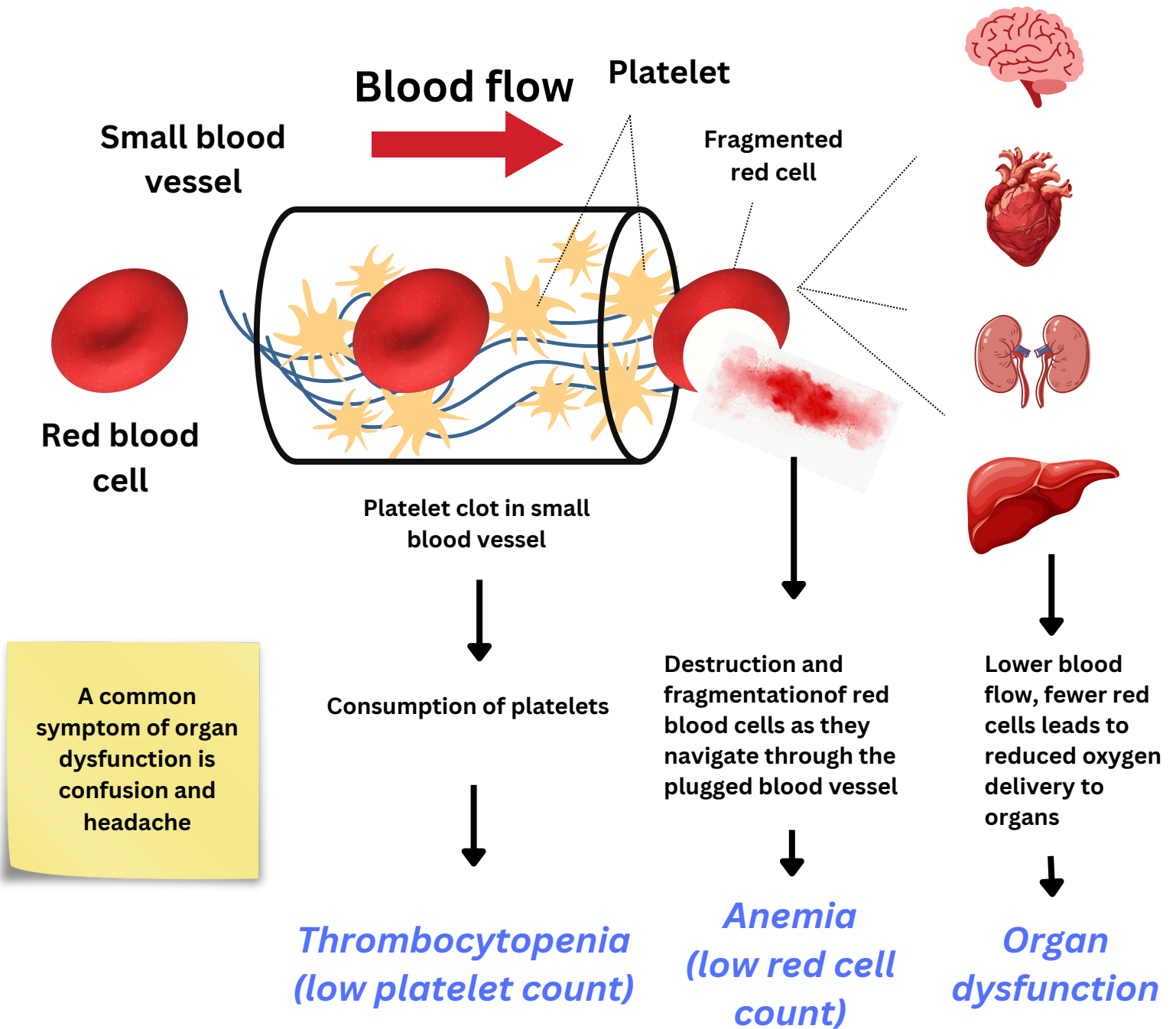
In TTP, antibodies bind to ADAMTS13, resulting in low levels of ADAMTS13 in the bloodstream. As a result, vWF cleavage is reduced with the result that platelets form clots in small blood vessels throughout the body



What Causes TTP?

Consequence of low ADAMTS13 levels

Thrombocytopenia, anemia, organ changes



Doctors will order frequent blood draws to follow the platelet count, the hemoglobin (a marker of red cell count) and markers of red cell destruction. They will also test for signs of organ dysfunction

How is TTP Treated?

Three-Pronged Attack

1

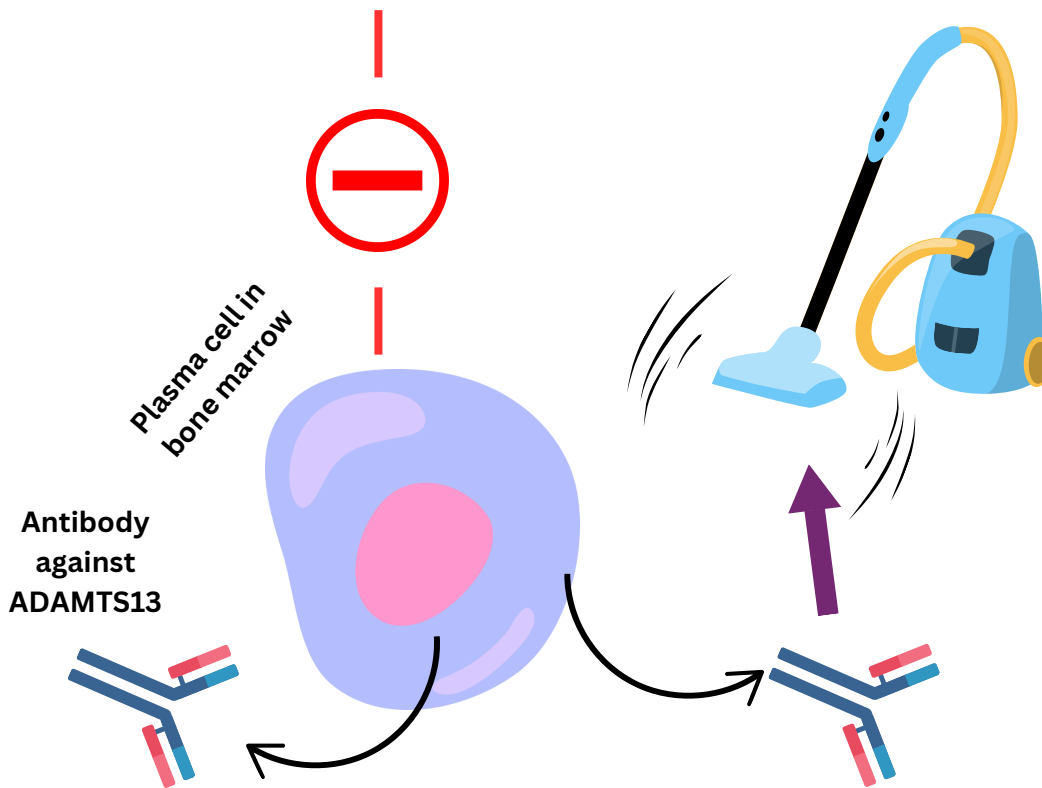
Suppress antibody production

(Corticosteroids)

2

Remove antibodies from the body

(Plasmapheresis)



Mainstay of treatment. All patients with a high suspicion of TTP should receive timely, daily plasmapheresis whereby plasma (the liquid part of blood) is removed and replaced with normal donor plasma; this not only removes antibodies against ADAMTS13 but it also provides ADAMTS13 from the donor plasma

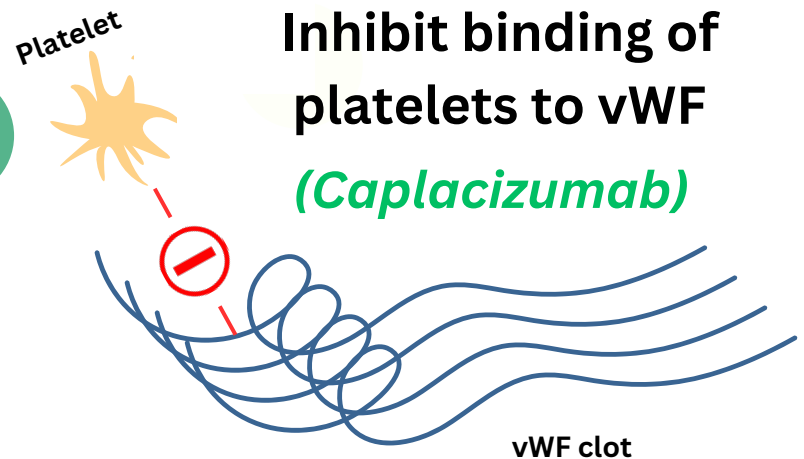
All patients with a new episode of TTP should receive high doses of daily corticosteroids (for example prednisone)

... many patients will also receive another immunosuppressant called *rituximab*, which is given as 4 weekly injections

3

Inhibit binding of platelets to vWF

(Cuplacizumab)



What to Expect

- You will likely present to an emergency department not feeling well, perhaps with some neurological or bleeding symptoms
 - You will have lots of blood tests, an EKG and X rays in the emergency room
 - Once TTP is suspected, you will be admitted to the hospital, possibly to an intensive care unit so that you can be carefully monitored
 - If your hospital does not have ability to perform plasmapheresis, you will likely be transferred to a hospital that does
 - You will have a special intravenous line placed in preparation for the plasmapheresis, which will typically be initiated within hours of admission and then performed once daily until the platelet count normalizes. This can take several days to achieve
 - You will be started on corticosteroids on the day of admission, either oral or by IV. These will be administered once daily
 - You may be started on caplacizumab, which is given as a subcutaneous injection under the skin once daily
 - You may also receive rituximab, which is given as a weekly injection for a total of 4 weeks
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