

Microangiopathic Hemolytic Anemia (MAHA)

Microangiopathic destroying red blood cells (hemolytic) anemia happens when red blood cells get torn apart while traveling through damaged or blocked tiny blood vessels. This releases hemoglobin into the bloodstream and causes anemia, yellowing of the skin and eyes (jaundice), and organ damage. Common causes include TTP, HUS, DIC, and severe high blood pressure.

THE ONE-LINE TRUTH

MAHA is red blood cell destruction (hemolysis) caused by red blood cells being physically sheared apart as they squeeze through abnormal clots or damaged tiny blood vessels.

The chain — how it unfolds

1. Endothelial injury or intravascular clot formation begins in microvessels — why: Triggering cause (Shiga toxin, ADAMTS13 deficiency, DIC, malignant high blood pressure (hypertension), or cancer) damages vessel walls or activates clotting.
2. Fibrin strands and platelet microthrombi form mesh-like networks in capillaries — why: Damaged endothelium exposes tissue factor and von Willebrand factor, triggering localized clotting and platelet aggregation.
3. Red blood cells fragment into schistocytes as they pass through narrowed vessels — why: Physical shearing forces exceed membrane strength, creating helmet cells, triangular fragments, and cell rupture.
4. Intravascular red blood cell destruction (hemolysis) releases free hemoglobin, LDH, and depletes haptoglobin — why: Broken red cells spill their contents into plasma, overwhelming the body's ability to bind and clear hemoglobin.
5. Anemia worsens as red cell destruction exceeds bone marrow production — why: Ongoing red blood cell destruction (hemolysis) consumes red cells faster than reticulocytes can replace them, dropping hemoglobin to 6 to 8 g/dL or lower.
6. Tissue low oxygen (hypoxia) develops as oxygen delivery falls — why: Fewer functional red cells combined with blocked microvessels reduces oxygen reaching brain, kidneys, heart, and other organs.

7. Organ starved blood flow (ischemia) and tissue death from blocked blood flow (infarction) occur from combined anemia and microvascular blockage (occlusion) — why: Brain (stroke, confusion), kidneys (acute kidney injury), heart (ischemia, arrhythmias), and gut (ischemia) sustain damage from clots and low oxygen.
8. Multi-organ failure and death if untreated — why: Without treatment (plasmapheresis for TTP, supportive care for HUS, treating DIC cause, controlling high blood pressure (hypertension)), progressive starved blood flow (ischemia) leads to renal failure, neurologic injury, cardiac arrest, or fatal bleeding.

Spot it — what you see, and why

- Dark or tea-colored urine: Hemoglobin (oxygen-carrying protein from broken red blood cells) passes through kidneys into urine when cells rupture in small vessels
- Pale skin and mucous membranes: Anemia (low red blood cell count) reduces hemoglobin available to give skin its pink color
- Fatigue and weakness: Fewer red blood cells deliver less oxygen to muscles and organs, forcing body to work harder for basic tasks
- Shortness of breath with mild activity: Reduced oxygen delivery from anemia forces faster breathing to compensate for decreased oxygen-carrying capacity
- Yellowing of skin or eyes (jaundice): Bilirubin (yellow breakdown product of hemoglobin) accumulates when red blood cells are destroyed faster than liver can process the waste
- Pinpoint bleeding spots (Petechiae) or purple bleeding blotches (purpura) (small red or purple spots on skin): Low platelets (blood-clotting cells) from consumption in microthrombi (tiny clots) allow small bleeds under skin

Red flags — act now

- Platelet count below 30,000/microL with confusion or neurologic changes suggests TTP requiring immediate plasma exchange
- Hemoglobin below 7 g/dL with chest pain or severe shortness of breath needs urgent transfusion
- Creatinine rising despite fluids with decreased urine output indicates acute kidney injury needing nephrology consultation
- New pinpoint bleeding spots (petechiae) or purple bleeding blotches (purpura) with any mucosal bleeding requires platelet count check and hematology evaluation within hours

Treat it — in this order

1. Plasma exchange (plasmapheresis) → Removes ADAMTS13 autoantibodies and replenishes functional enzyme in TTP; reduces mortality from 90% to 10-20% when started immediately
2. Packed red blood cell transfusion → Improves oxygen delivery when hemoglobin drops below 7 g/dL or patient has chest pain, severe shortness of breath, or altered mental status from anemia
3. Supportive care with IV fluids and electrolyte monitoring → Prevents dehydration-induced kidney damage in HUS; corrects electrolyte imbalances (hyperkalemia, metabolic acidosis (acid building up in the blood)) from kidney injury; maintains adequate blood flow (perfusion)
4. Delivery of fetus and placenta in pregnancy-related MAHA → Removes the trigger for HELLP syndrome and severe preeclampsia; usually resolves red blood cell destruction (hemolysis) and a low platelet count (thrombocytopenia) within 48-72 hours postpartum
5. Blood pressure control to target <140/90 in malignant high blood pressure (hypertension) → Reduces shear stress on vessel walls and stops endothelial injury that triggers microthrombi; allows vessel healing over days to weeks

Your top priorities at the bedside

1. Monitor for signs of organ damage from microthrombi (tiny blood clots) — why: MAHA causes small clots in tiny blood vessels that block blood flow to the brain, kidneys, and heart. Early detection of stroke symptoms, decreased urine output, or chest pain allows rapid intervention.
2. Assess for bleeding tendencies despite low platelet count — why: The destruction of platelets (blood-clotting cells) in small vessels causes a low platelet count (thrombocytopenia) (low platelet count). Patients may bruise easily, have pinpoint bleeding spots (petechiae) (tiny red spots), or bleed from gums or nose.
3. Evaluate neurologic status every 2-4 hours — why: Brain microthrombi cause fluctuating confusion, headache, vision changes, or seizures in thrombotic thrombocytopenic purple bleeding blotches (purpura) (TTP) and other MAHA causes. Subtle changes require urgent provider notification.