

Autoimmune Hemolytic Anemia

Autoimmune destroying red blood cells (hemolytic) anemia is a condition where the body's immune system attacks and destroys its own red blood cells. This causes anemia (low red blood cell count) because the bone marrow cannot make new red cells fast enough to replace the ones being destroyed.

THE ONE-LINE TRUTH

The immune system mistakes the patient's own red blood cells for foreign invaders and destroys them faster than the bone marrow can replace them.

The chain — how it unfolds

1. Autoantibody formation and red cell coating — why: Immune system produces antibodies against red blood cell surface antigens, coating the cells and marking them for destruction within hours to days instead of the normal 120-day lifespan
2. Accelerated extravascular red blood cell destruction (hemolysis) in spleen — why: Splenic macrophages rapidly remove antibody-coated red cells, causing unconjugated hyperbilirubinemia (elevated indirect bilirubin from hemoglobin breakdown) and depleting red cell mass faster than bone marrow can replace it
3. Progressive anemia despite compensatory erythropoiesis — why: Hemoglobin drops because red cell destruction outpaces production; reticulocyte count rises as bone marrow works overtime, but patient becomes increasingly anemic with fatigue, pale skin (pallor), and trouble breathing (dyspnea) (shortness of breath)
4. Tissue low oxygen (hypoxia) and organ dysfunction — why: Low hemoglobin means inadequate oxygen delivery to heart, brain, and kidneys; patient develops a fast heart rate (tachycardia) (fast heart rate), confusion, angina (chest pain), and decreased urine output as organs suffer
5. Cardiovascular strain and high-output state — why: Heart pumps harder and faster to compensate for low oxygen-carrying capacity, increasing myocardial oxygen demand and risking heart failure, especially in patients with pre-existing cardiac disease
6. Severe intravascular red blood cell destruction (hemolysis) with complement activation — why: In severe cases or cold agglutinin disease, complement proteins punch holes in red cells inside blood vessels, releasing free hemoglobin into plasma, which is toxic to kidneys and causes hemoglobinuria (dark urine from hemoglobin)

7. Acute kidney injury from hemoglobin precipitation — why: Free hemoglobin filtered by kidneys precipitates in renal tubules, causing acute tubular tissue death (necrosis), rising creatinine, and potentially requiring dialysis if not corrected rapidly
8. Life-threatening anemic crisis and multi-organ failure — why: Severe untreated red blood cell destruction (hemolysis) drops hemoglobin to critically low levels (sometimes below 4-5 g/dL), causing cardiovascular collapse, lactic acidosis (acid from oxygen-starved tissues) (buildup of lactic acid from oxygen-starved tissues), altered mental status, and death without urgent transfusion and immunosuppressive therapy

Spot it — what you see, and why

- Fatigue and weakness: Low hemoglobin (oxygen-carrying protein in red blood cells) means less oxygen reaches muscles and brain because antibodies destroy red blood cells faster than the bone marrow can make new ones
- Pale skin and mucous membranes: Decreased number of circulating red blood cells reduces the pink color normally seen in skin, gums, and inside the eyelids
- Yellowing of the skin and eyes (Jaundice) (yellow skin and eyes): When red blood cells break apart, hemoglobin releases bilirubin (a yellow pigment) faster than the liver can process it, causing yellow discoloration
- Dark urine: Hemoglobin and bilirubin spill into urine when red blood cells are destroyed in large numbers, turning urine tea-colored or cola-colored
- Shortness of breath with exertion: Low red blood cell count means less oxygen delivery to tissues, so the body tries to compensate by breathing faster during activity
- Chest pain or rapid heartbeat: The heart pumps faster and harder to deliver enough oxygen when hemoglobin is low, increasing cardiac workload and oxygen demand

Red flags — act now

- Hemoglobin below 7 g/dL with chest pain, confusion, or severe shortness of breath at rest requiring immediate transfusion
- New swollen lymph nodes (lymphadenopathy) or an enlarged spleen (splenomegaly) suggesting underlying lymphoma or chronic lymphocytic leukemia driving the red blood cell destruction (hemolysis)
- Drug-induced destroying red blood cells (hemolytic) anemia with continued exposure to the causative medication
- Cold agglutinin disease with distal starved blood flow (ischemia) (blue fingers or toes) from red blood cell agglutination blocking small vessels

Treat it — in this order

1. Corticosteroids (prednisone or methylprednisolone) → Suppresses the immune system's production of antibodies against red blood cells and reduces removal of antibody-coated cells by the spleen, raising hemoglobin in 70 to 85 percent of warm autoimmune destroying red blood cells (hemolytic) anemia patients within 1 to 3 weeks
2. Packed red blood cell transfusion → Immediately increases oxygen-carrying capacity and prevents organ damage from severe low oxygen (hypoxia) when hemoglobin drops below 7 g/dL or patient has chest pain, confusion, or severe shortness of breath
3. Rituximab (anti-CD20 monoclonal antibody) → Destroys B-lymphocytes (cells that make antibodies) and achieves remission in 80 to 90 percent of patients with warm autoimmune destroying red blood cells (hemolytic) anemia who fail steroids or relapse after steroid taper
4. Splenectomy (surgical removal of the spleen) → Eliminates the major site where antibody-coated red blood cells are trapped and destroyed, producing long-term remission in 60 to 70 percent of warm autoimmune destroying red blood cells (hemolytic) anemia patients who do not respond to steroids and rituximab
5. Cold avoidance measures and warming strategies → Prevents activation of cold-reactive antibodies in cold agglutinin disease by keeping body and extremities warm, reducing red blood cell destruction (hemolysis) episodes triggered by temperature below 30 degrees Celsius

Your top priorities at the bedside

1. Monitor for signs of acute red blood cell destruction (hemolysis) and severe anemia — why: Autoimmune destroying red blood cells (hemolytic) anemia (AIHA, a condition where the immune system destroys red blood cells) can cause rapid drops in hemoglobin leading to low oxygen (hypoxia) (low oxygen), heart failure, and shock requiring immediate intervention.
2. Assess respiratory status and oxygen saturation continuously — why: Severe anemia reduces oxygen-carrying capacity of the blood, so patients may develop trouble breathing (dyspnea) (shortness of breath), fast breathing (tachypnea) (rapid breathing), and low oxygen in the blood (hypoxemia) (low blood oxygen) requiring supplemental oxygen.
3. Monitor vital signs for hemodynamic instability — why: Acute red blood cell destruction (hemolysis) causes a fast heart rate (tachycardia) (fast heart rate), low blood pressure (hypotension) (low blood pressure), and signs of shock as the body tries to compensate for decreased oxygen delivery to tissues.