Hemolytic Uremic Syndrome (HUS)

HUS is a thrombotic microangiopathy characterized by over-activation of platelets that form thrombi and subsequently amass in microvascular beds leading to various sequelae including microangiopathic hemolytic anemia, organ dysfunction and thrombocytopenia. This condition has significant overlap with thrombotic thrombocytopenic purpura (TTP), also a thrombotic microangiopathy; however, neurologic involvement is typically not a feature in HUS as it is in TTP.



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Children

Children

HUS primarily affects children under 10 years old, often after an E. coli O157:H7 diarrheal illness, but it can also present in the elderly, particularly in atypical HUS caused by complement dysregulation. Pediatric cases usually have a better prognosis than adult cases.

Pathophysiology

Preceded by Acute Diarrheal Illness

Acute-angle Toilet

HUS is commonly preceded by an infectious gastroenteritis. Shiga toxin from E. coli O157:H7 enters the bloodstream through the inflamed intestinal mucosa, binds to Gb3 receptors on endothelial cells, and triggers endothelial damage and platelet activation. This is the mechanism behind typical, diarrhea-associated HUS, which is the most common form in children.

Endothelial Damage

Inner-layer Damaged

Shiga toxin directly injures endothelial cells, particularly in renal glomeruli. The endothelial damage exposes the subendothelial matrix, which triggers platelet aggregation and microthrombus formation, a hallmark of HUS. This is the pathophysiologic basis of microangiopathic hemolytic anemia and thrombocytopenia.

Classic Triad of Symptoms

Microangiopathic Hemolytic Anemia (MAHA)

Microscope-angel with Hemolysing-RBCs from Anemia-Anemone

Thrombi in small vessels shear passing red blood cells, producing schistocytes (helmet cells) on peripheral smear. This destruction of red blood cells leads to hemolytic anemia, elevated LDH, indirect hyperbilirubinemia, and low haptoglobin, which are important USMLE high-yield lab findings.

Thrombocytopenia

Trombone-side-toe-peanut

Platelet consumption within microvascular thrombi causes low platelet counts. Clinically, this may manifest as easy bruising, petechiae, or mild bleeding, although severe bleeding is uncommon.

Acute Kidney Injury

Acute-angle Kidney-injured

Microthrombi deposition in glomeruli causes renal ischemia, tubular injury, and acute kidney injury. Lab findings include elevated BUN and creatinine, hematuria, proteinuria, and electrolyte abnormalities such as hyperkalemia and metabolic acidosis. This is the reason HUS is a leading cause of acute kidney injury in children.

Labs

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Helmet cells

Helmet Cell

Schistocytes appear as helmet-shaped RBC fragments on peripheral smear. This is a high-yield USMLE sign of MAHA in thrombotic microangiopathies like HUS and TTP.

LDH

Ladies Disc Hockey

Lactate dehydrogenase (LDH) is elevated due to red blood cell destruction in MAHA. High LDH is a supportive lab marker for HUS and reflects cellular injury.

Treatment

Dialysis

Dial-machine

Dialysis is required in severe cases with BUN >80 mg/dL, symptomatic uremia, or refractory electrolyte disturbances, especially hyperkalemia or fluid overload. Pediatric patients may recover renal function after supportive care, but some may progress to chronic kidney disease.

Supportive

Supportive IV Bags

Treatment is primarily supportive, including intravenous hydration to maintain renal perfusion, electrolyte monitoring and correction, and RBC or platelet transfusions if indicated. Antibiotics are generally avoided in typical HUS because they may increase Shiga toxin release. In atypical HUS, complement inhibition with eculizumab is used.

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