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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. It also affects the elderly.

Pathophysiology

Preceded by Acute Diarrheal Illness

Acute-angle Toilet

HUS is associated with infectious gastroenteritis caused by E. coli strain O157:H7 and leading to bloody diarrhea. The Shiga-like toxin that is formed then enters the circulation through inflamed gastrointestinal mucosa.

Endothelial Damage

Inner-layer Damaged

Once in the circulation, the Shiga-like toxin interrupts endothelial cell function causing damage and leading to platelet activation and aggregation.

Classic Triad of Symptoms

Microangiopathic Hemolytic Anemia (MAHA)

Microscope-angel with Hemolysing-RBCs from Anemia-Anemone

The thrombi formed in microcirculation shear blood cells as they travel through thus hemolyzing them. This results in the characteristic helmet cells. The word "microangiopathic" refers to small blood vessels.

Thrombocytopenia

Trombone-side-toe-peanut

Over-activation and aggregation of platelets leads to thrombocytopenia.

Renal Failure

Dead Kidney

Thrombi deposit in glomeruli of kidneys and can lead to infarcts and necrosis and possible renal failure.

Labs

Helmet cells

Helmet Cell

Sheared blood cells resemble helmets. Helmet cells are also known as schistocytes.

LDH

Ladies Disc Hockey

Elevated lactate dehydrogenase is characteristic in HUS as it is a chemical released from damaged cells.

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Treatment

Dialysis

Dial-machine

Dialysis is indicated in patients with BUN>80 mg/dL, symptomatic uremia, and electrolyte overload.

Supportive

Supportive IV Bags

Treatment for HUS is typically supportive consisting of IV hydration, monitoring of blood counts, monitoring of electrolyte counts and replenishing of electrolytes if needed. RBC and platelet transfusions are indicated if the hemoglobin drops to below 6 g/dL or if significant bleeding occurs, respectively.