HEMOLYTIC-UREMICSYNDROME

- ETIOLOGY.
- PATHOGENESIS.
- CLINICAL MANIFESTATION.
- INVESTIGATION.
- D.DX.
- TREATMENT.
- PROGNOSIS.

- HUS is a common cause of community-acquired ARF in young children.

- **<u>It is characterized by the triad of:</u>**

- ***MICROANGIOPATHIC HEMOLYTIC ANEMIA***
- ***THROMBOCYTOPENIA,***
- ***RENAL INSUFFICIENCY.***

Etiology:

HUS can be classified according to etiology as follows:-

- *Infection-induced* (most common); it include: Verotoxin-producing E coli (most common 057:H7) , Shiga toxin-producing Shigella dysentereriae type 1 (common), Streptococcus pneumoniae (rare), and HIV (rare).

- *Genetic (Atypical)* HUS include: Familial AR & AD of undefined etiology, recurrent, undefined etiology without diarrhea prodrome.

- *Diseases associated* with microvascular injury include: SLE, Following BM transplantation, Malignant hypertension.

- *Medication-induced* include: some immunosuppressant & cytotoxic medications.

Pathogenesis:

- **Microvascular injury with endothelial cell damage** is characteristic of all forms of HUS, capillary and arteriolar endothelial injury in the kidney particularly in glomeruli, leads to localized thrombosis causing a direct decrease in GFR.
- Progressive platelet aggregation in the areas of microvascular injury results in consumptive thrombocytopenia.
- Microangiopathic hemolytic anemia results from mechanical damage to red blood cells as they pass through the damaged and thrombotic microvasculature

Clinical manifestation:

HUS is most common in preschool and school-aged children.

- In HUS caused by E. coli, onset of HUS occurs a few days (as few as 3 days up to 3 wk.)

after onset of gastroenteritis with fever, vomiting, abdominal pain, and diarrhea which is often bloody, but not necessarily, especially early in the illness.

Following the prodromal illness, a sudden onset of pallor, irritability, weakness, lethargy and Oliguria .

- Patients can develop petechiae, but significant or severe bleeding is rare despite very low platelet counts.
- Patients with pneumococcus-associated HUS usually are ill with pneumonia and empyema when they develop HUS.

- E coli is usually transmitted by undercooked meat,unpasteurized milk , contaminated apple cidar or bathing in contaminated swimming pool.

Investigation:

1. **CBP shows :**

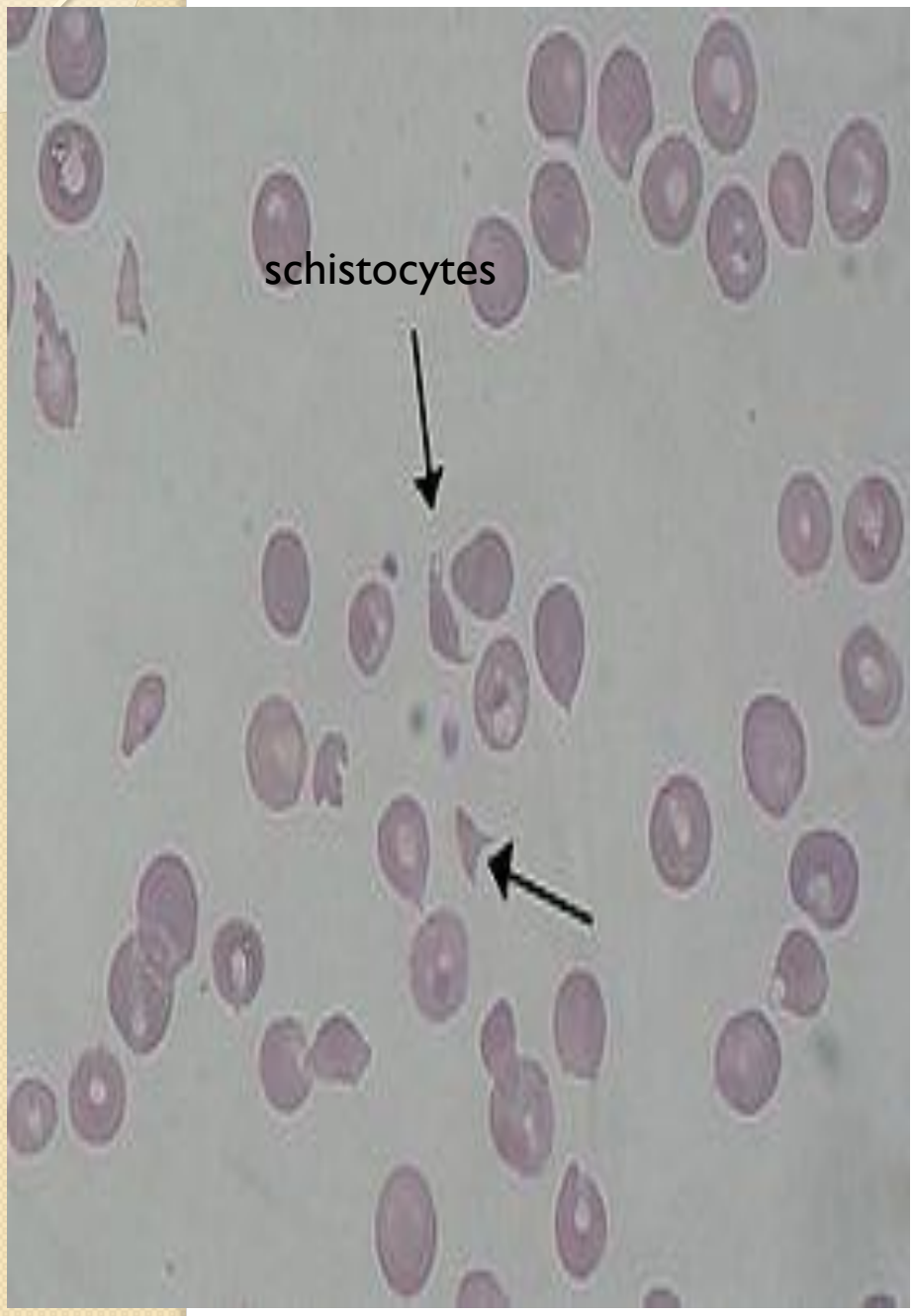
- **Hb** is in the range of **5-9gm**/dl.
- **Thrombocytopenia** is an invariable finding in the acute phase
- Leukocytosis is present

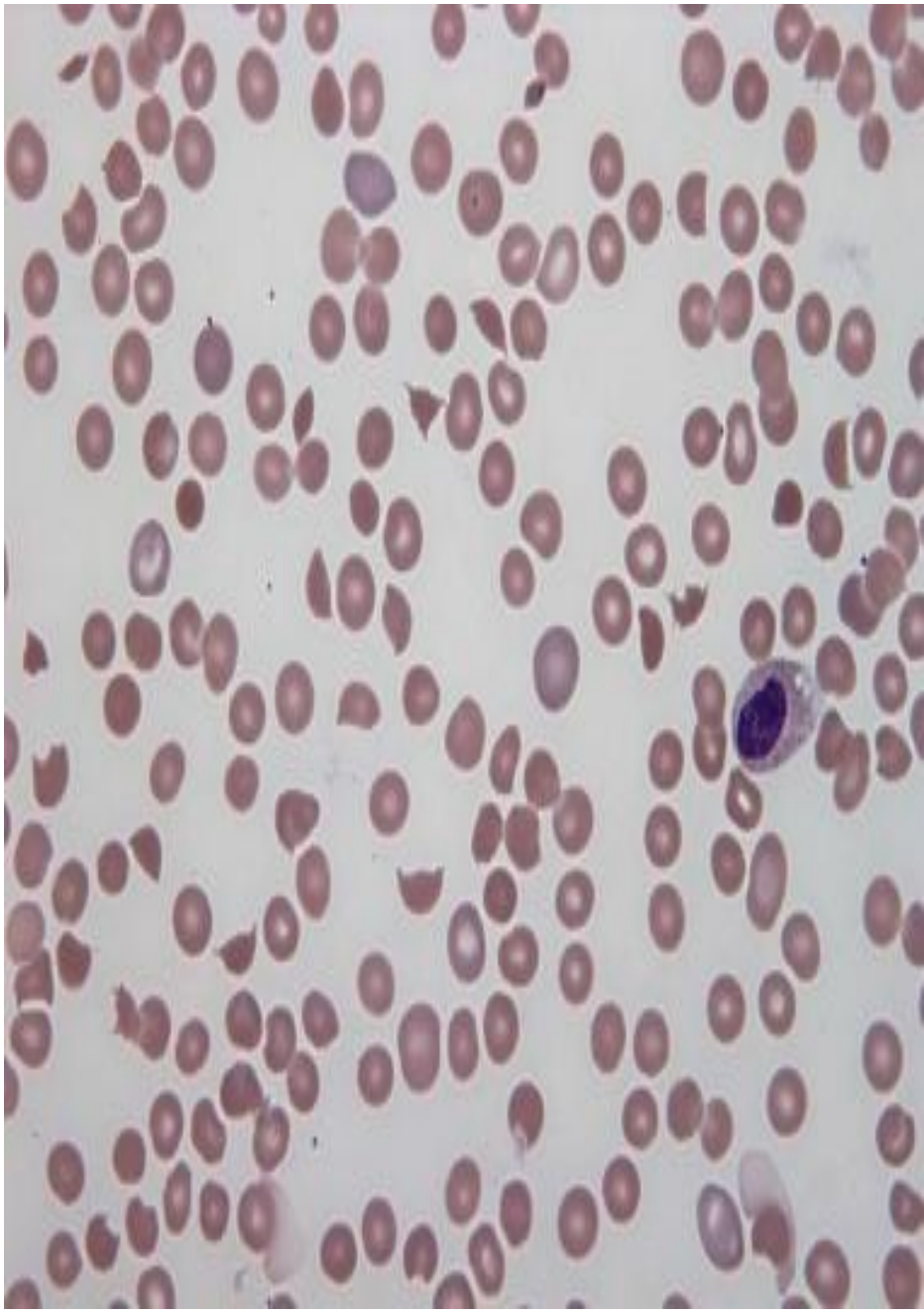
i. **Blood film** : **microangiopathic hemolytic anemia with schistocytes, burr cells, helmet cells and fragmented RBCs**.

ii. **Coombs test is negative**.

iii. **PT & PTT** are usually **normal**.

iv. **RFT**: Renal insufficiency can vary from mild elevations in BUN to ARF.


schistocytes



D.Dx.

- THROMBOTIC THROMBOCYTOPENIC PURPURA (TTP)
- SLE.
- MALIGNANT HYPERTENSION.
- BILATERAL RENAL VEIN THROMBOSIS.

Treatment:

- Careful management of fluid and electrolytes e.g. correction of volume deficit, control of hypertension, and early institution of dialysis if the patient becomes anuric .

  plasmapheresis or FFP has been recommended.

- Red cell transfusions are usually required because hemolysis can be brisk and recurrent until the active phase of the disease has resolved.

- Platelets should not be administered, regardless of platelet count because they are almost immediately consumed by the active coagulation and can theoretically worsen the clinical course.
- Antibiotic therapy is not recommended as it result in increased toxin release, potentially exacerbating the disease.

PROGNOSIS:

- The mortality rate for diarrhea-associated HUS after careful supportive care has declined to <5%. Most recover renal function completely, but of surviving patients, 5% remain dependent on dialysis,.
- The prognosis for HUS that not associated with diarrhea is more severe,with mortality reported ≈20%. The familial forms of HUS have a poor prognosis

Thank you