HEMOLYTIC ANEMIA

Hemolytic anemia is a group of disorders where red blood cells are destroyed. Hemolytic anemia can be classified into inherited or immune-mediated causes. The etiology of hemolytic anemia is relevant to management.

CLINICAL PRESENTATION

SIGNS AND SYMPTOMS

- Pallor
- Fatigue and or lethargy
- Lack of activity
- Changes in school performance
- Dark urine
- Scleral icterus
- Jaundice
- Fever
- Abdominal or back pain
- Dizziness and confusion

PHYSICAL EXAM

- Vital signs
- Level of activity/consciousness
- May appear fatigued
- Check for pallor

HEENT
- Lymph node exam

Cardiac examination
- Tachycardia
- Early systolic flow murmur

Abdo examination
- Splenomegaly
- Hepatomegaly

ETIOLOGY

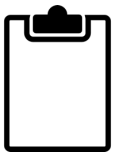
Intrinsic causes of hemolytic anemia:
- ❑ Hemoglobinopathies: sickle cell disease, thalassemia and unstable hemoglobin
- ❑ Red cell membrane defects: hereditary spherocytes and elliptocytosis
- ❑ Enzyme deficiencies: G6PD deficiency and pyruvate kinase deficiency

Extrinsic causes of hemolytic anemia:
- ❑ Immune hemolysis:
  - Hemolytic disease of the newborn
  - Warm/cold autoimmune hemolytic anemia
  - Acute or delayed transfusion reaction
- ❑ Drug-induced hemolysis
- ❑ Microangiopathic hemolytic anemia (MAHA)
  - Can have infectious triggers

Examples of MAHA:
- Disseminated intravascular coagulation (DIC), including sepsis with DIC
- Hemolytic uremic syndrome (HUS)
- Thrombotic thrombocytopenic purpura (TTP)

INVESTIGATIONS

If you suspect hemolysis:
- ❑ Hemoglobin
- ❑ Reticulocyte count
  - ❑ High: hemolysis/blood loss
  - ❑ Low: decreased bone marrow response
- ❑ Bilirubin
- ❑ LDH
- ❑ Haptoglobin
- ❑ Urinalysis
- ❑ Peripheral smear
- ❑ Type and screen
- ❑ Renal function: urea, creatinine



To determine cause of hemolysis:
- ❑ DAT
- ❑ CBCd for other cell lines
- ❑ PTT/INR
- ❑ Fibrinogen
- ❑ G6PD def screening
- ❑ Hemoglobinopathy investigation for thalassemia and sickle cell disease
- ❑ EMA flow cytometry +/- osmotic fragility
- ❑ Pyruvate kinase enzyme activity
- ❑ Genetic testing as indicated

Hemolysis Labs

Test	Result
Bilirubin	↑
LDH	↑
Reticulocytes	↑
Haptoglobin*	↓

*Serum haptoglobin is not reliable in infants under 4 months of age.

Hemolytic anemia should be treated as an **emergency** until you know the cause and rate of hemolysis due to the risk of **severe anemia** and **renal failure**.

MANAGEMENT

Acute management of all hemolysis:
- ❑ Stop cause if found (ie. medication)
- ❑ Hydrate at 1.5x maintenance
- ❑ Watch urine output
- ❑ Serial monitoring of hemoglobin
- ❑ Consider pRBC transfusion depending on cause and rate of hemolysis

Management can depend on specific etiology:
- ❑ Warm AIHI: corticosteroids
- ❑ Cold AIHI: supportive care if mild, consider rituximab or exchange transfusion if severe
- ❑ Transfusion reaction: STOP transfusion, supportive care
- ❑ Treat cause of hemolysis (e.g. sepsis)

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