

# Sickle Cell Anemia

### Signs & Symptoms

- Chronic hemolytic anemia
  - frequent infections, fatigue, delayed physical growth
- Signs of Vaso-occlusive Crisis: pain, fever, painful edematous hands and feet, arthralgia, leg ulcers, cerebrovascular accident

### Pathophysiology

- Abnormal adhesion, entanglement, & emmeshing of rigid sickle-shaped RBCs → inflammatory process → intermittent blocking or microcirculation → vaso-occlusion → absence of blood flow to adjacent tissues → local hypoxia → tissue ischemia and infarction

### Diagnostics/Labs

- Prenatal
  - Chorionic villus sampling from prenatal tissue
  - amniocentesis
- After birth
  - newborn screening
  - Sickledex (sickle turbidity test)

### Treatment/Medication

- Blood transfusions
- Hydroxyurea, L-glutamine, crizanlizumab, voxelator, pain meds (acetaminophen, ibuprofen)
- Crisis: hydration, analgesics, blood transfusion, antibiotics, hydroxyurea

### Nursing Interventions

- Manage pain (meds, warm compresses)
- Oral or IV hydration
- Prevention of infection
  - standard precautions
  - handwashing
- Monitor I/Os
- Emotional support

### Patient Teaching

- Prevent hypoxia by:
  - avoid strenuous activity/exercise
  - avoid high altitudes
  - avoid anyone who is sick
- seek care with any signs of infection
- prophylactic penicillin if prescribed
- keep child hydrated, & do not withhold fluids at night

### Other

### Priority Nursing Diagnosis

- hypoxia and dehydration
- pain