

# Sickle Cell



#### The University of Zambia

School of Public Health o Family Medicine MMed Program

#### Education

- Prophylactic mediation
  - Folic acid
  - Anti-malaria
  - Penicillin
  - Hydroxyurea
- Vaccinations
  - Pneumonia
  - Meningitis
- Teach spleen palpation
- Precautions against
  - Dehydration
  - Overexertion
  - Exposure to cold
- Delayed puberty
- When to seek care
  - Enlarging spleen
  - Temperature elevation
  - Pallor of skin, lips or nail beds
  - Respiratory symptoms
  - Signs of pain or inability to move extremities

## **Blood transfusions**

Simple	Exchange	Chronic Hypertransfusion
Surgery	ACS	Prevention of stroke
Splenic sequestration	Recurrent stroke	Prevention of repeat stroke
Hepatic sequestration	Multiorgan failure	Recurrent VOC
Sepsis		Recurrent ACS
Acute anemia		Pregnant with previous OB complication
		Delayed growth & development

#### Acute chest syndrome



- Mimics severe pneumonia, usually due to infection
- FBC, retic count, group and match, blood cx, total bili, LDH, CXR (60% have infiltrate with normal PE), CRP/ESR, U&Es, LFTs, pulse ox
- Hydration IVF @ max of 2/3 maintenance if needed.
  Overhydration can lead to pulmonary edema
- Continuous monitoring of pulse, RR, O2 sats (>94%), BP
- Blood transfusion: simple pRBC at 10-15mL/kg if severe anemia (<7 or drops > 2 from baseline), significant hypoxia, worsening respiratory status
- Antibiotics for pna: cefotaxime 200mg/kg q8 IV + Erythromycin 10mg/kg q4 for PO 10-14 days
- Bronchodilators if wheeze/reactive airway disease

## **Painful Crisis**



- IVF with 5% dextrose at 1.5x maintenance
- Assess cause, reassurance, warm packs, reposition, massage, distraction
- IV antibiotics if febrile
- Adequate analgesia started within 15 minutes of triage
- Mild: paracetamol 15mg/kg QID or adult 1g/ qid
- Moderate: mild + ibuprofen 5mg/kg tds OR diclofenac
  1mg/kg tds
- **Severe:** moderate + oral morphine 0.2-0.3mg/kg q4 as needed, adult 5-10mg q4 hours
  - Night dose 1.5-2 times higher

## Splenic sequestration



- Tender enlarged spleen up to 3yo, peak at 1 year
- Pallor, tachycardia, signs of hypovolemic shock
  - FBC, retic count, urgent transfusion of 20mL/kg of whole blood
  - IVF with NS if blood is not available while transferring to higher care
- Splenectomy when stable



