School-Based Care for Children with Sickle Cell

Cris Hogue, PharmD, BCPPS
Pediatric Pharmacy Clinical Coordinator
Baylor Scott and White McLane Children's Medical Center
Temple, TX

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Objectives

Review sickle cell disease (SCD) and its common complications

- Describe the basic pathophysiology of SCD
- Identify common complications seen in SCD
  - Pain crises, Anemia, Infections, Acute chest syndrome, Stroke/neurocognitive concerns
- Recognize signs and symptoms requiring urgent medical evaluation at school

Discuss prevention and treatment of complications related to SCD

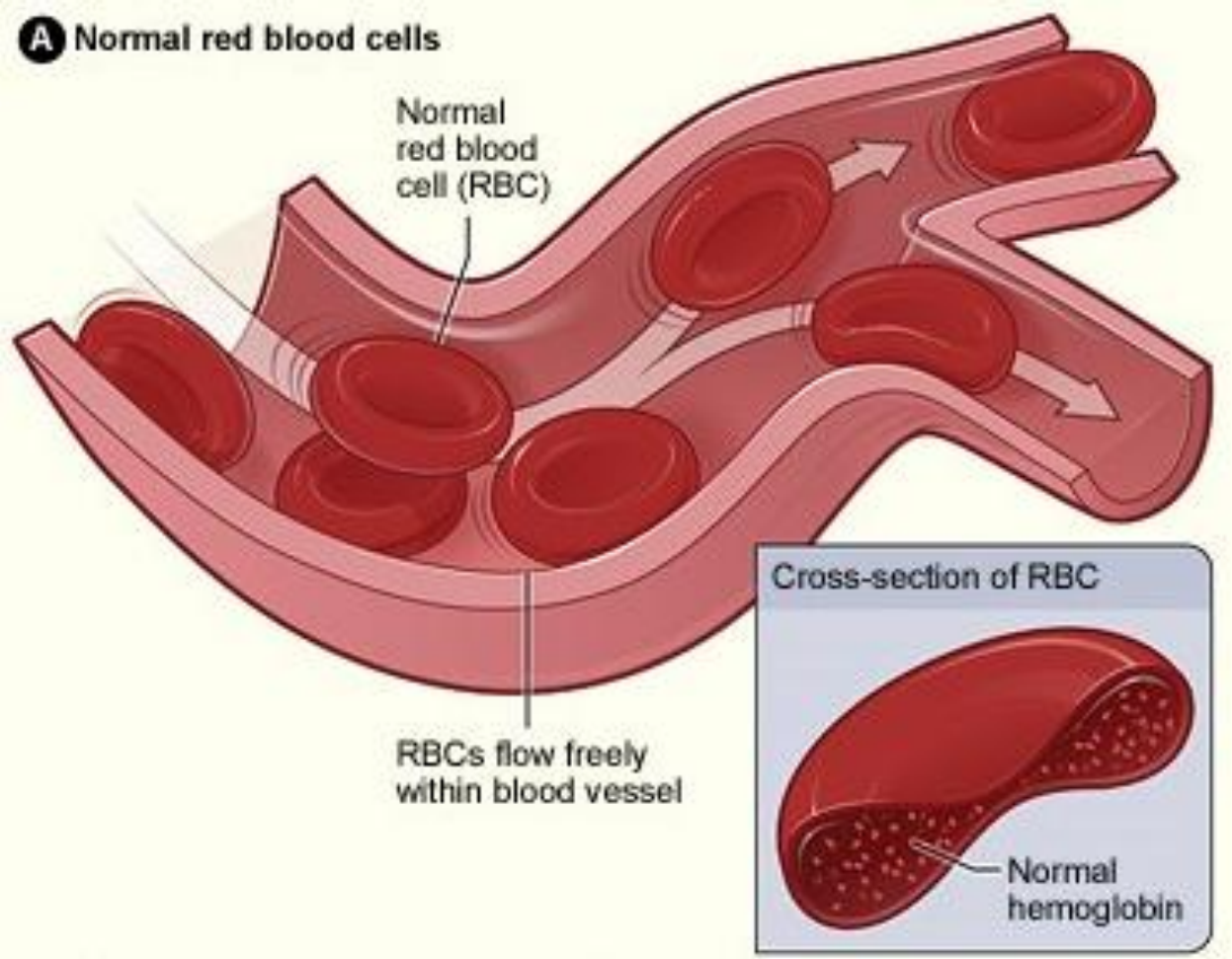
- Review prevention measures, including:
  - Hydration, Infection prevention/immunizations, Temperature regulation
- Review common medications prescribed in SCD and treatment approaches for pain management, fever/suspected infection, dehydration.

Pathophysiology of Sickle Cell

Red Blood Cells (RBC)

- Function: Carry oxygen to tissues and return carbon dioxide to the lungs
- Old/damaged RBC: Filtered out by spleen and removed from circulation

Life Span of normal RBC: 120 days



Ref 1, 2, 3

Pathophysiology of SCD—Sickled RBC

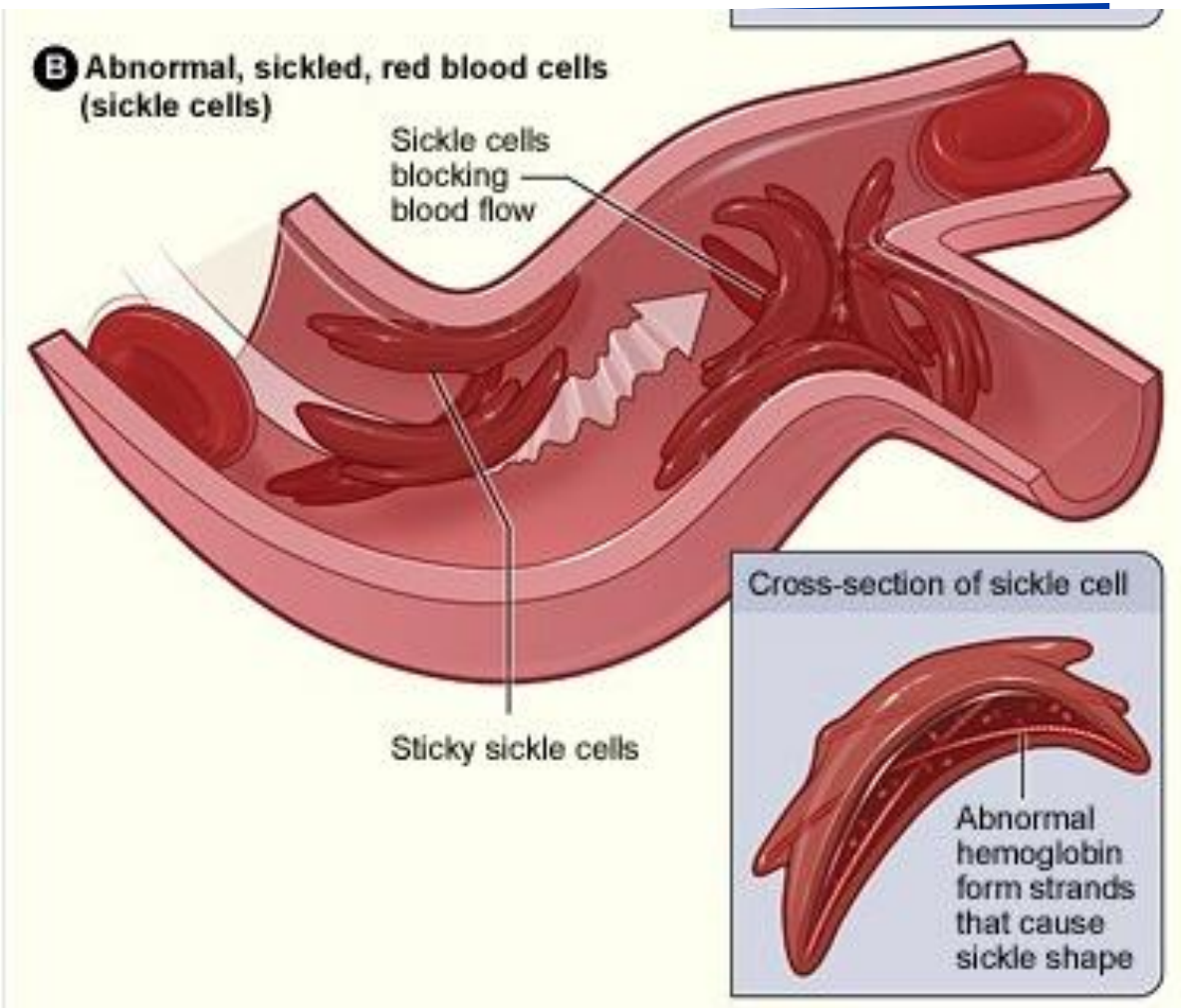
Abnormal, rigid sickle shape:

- Stick to each other and vessel walls
- Creates blockages—slows or stops blood flow

Life Span of Sickle RBC: 10-20 days

Genetics:

- Hemoglobin change
  - HgbA → HgbS
- Autosomal Recessive Disorder
- Part of newborn screen in US



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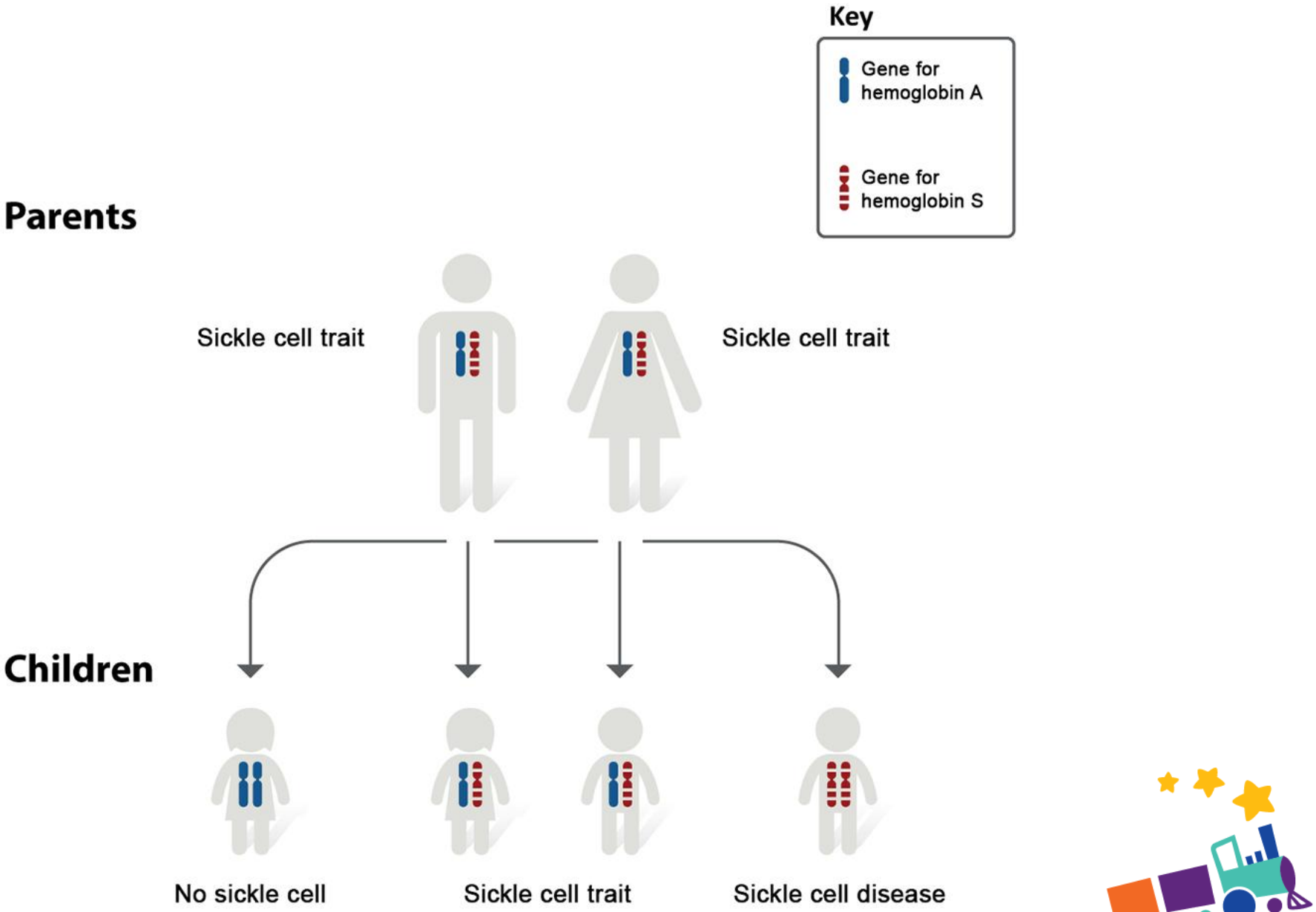

Key
Gene for hemoglobin A
Gene for hemoglobin S
Parents
Sickle cell trait
Sickle cell trait
Children
No sickle cell
Sickle cell trait
Sickle cell disease

Ref 1

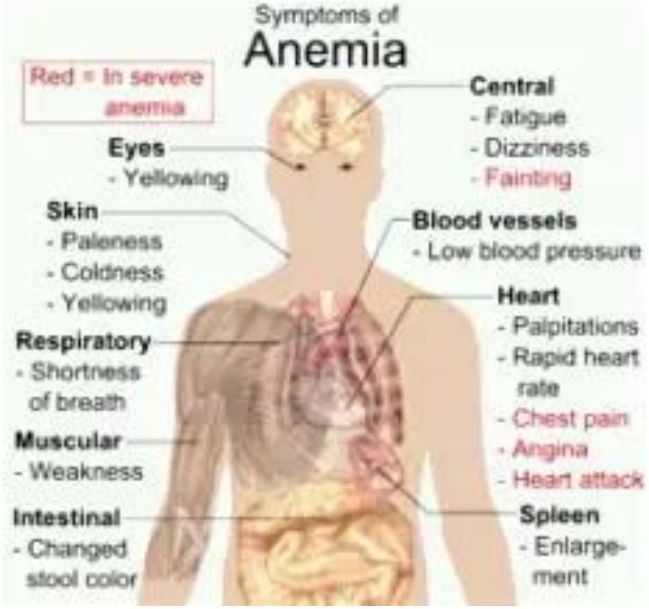
What are the Complications of Sickle Cell?

Related to the abnormal shape of the cells "sticking" vs. smoothly flowing through the blood vessels and decreased life span of RBC

- Anemia
- Infection
- Acute Chest Syndrome
- Stroke/Neurocognitive concerns
- Pain

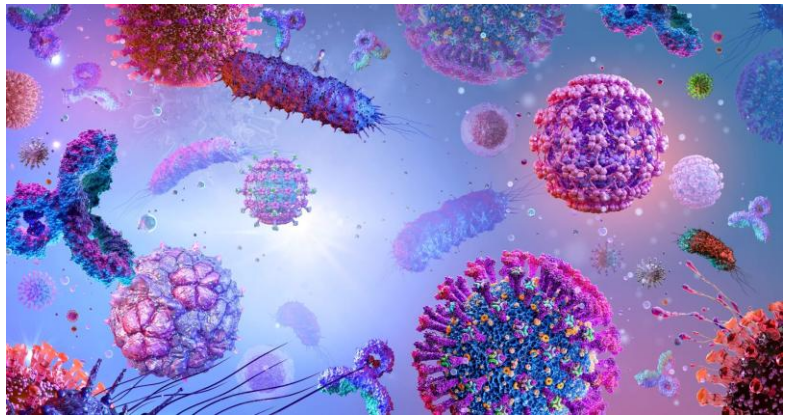
Anemia

- Sickle RBC become fragile
  - Go to spleen to be removed
    - eventually spleen becomes scarred/fibrotic then dysfunctional
  - Sickle RBC live 10-20 days vs 120 days in normal RBC
- Chronic transfusions to treat anemia
- Signs & Symptoms
  - Fatigue
  - Dizziness/Lightheadedness/Weakness
  - Pale Skin (or nailbeds)
  - Tachycardia
  - Jaundice
    - Due to increased bilirubin from RBC destruction



Ref 1, 2

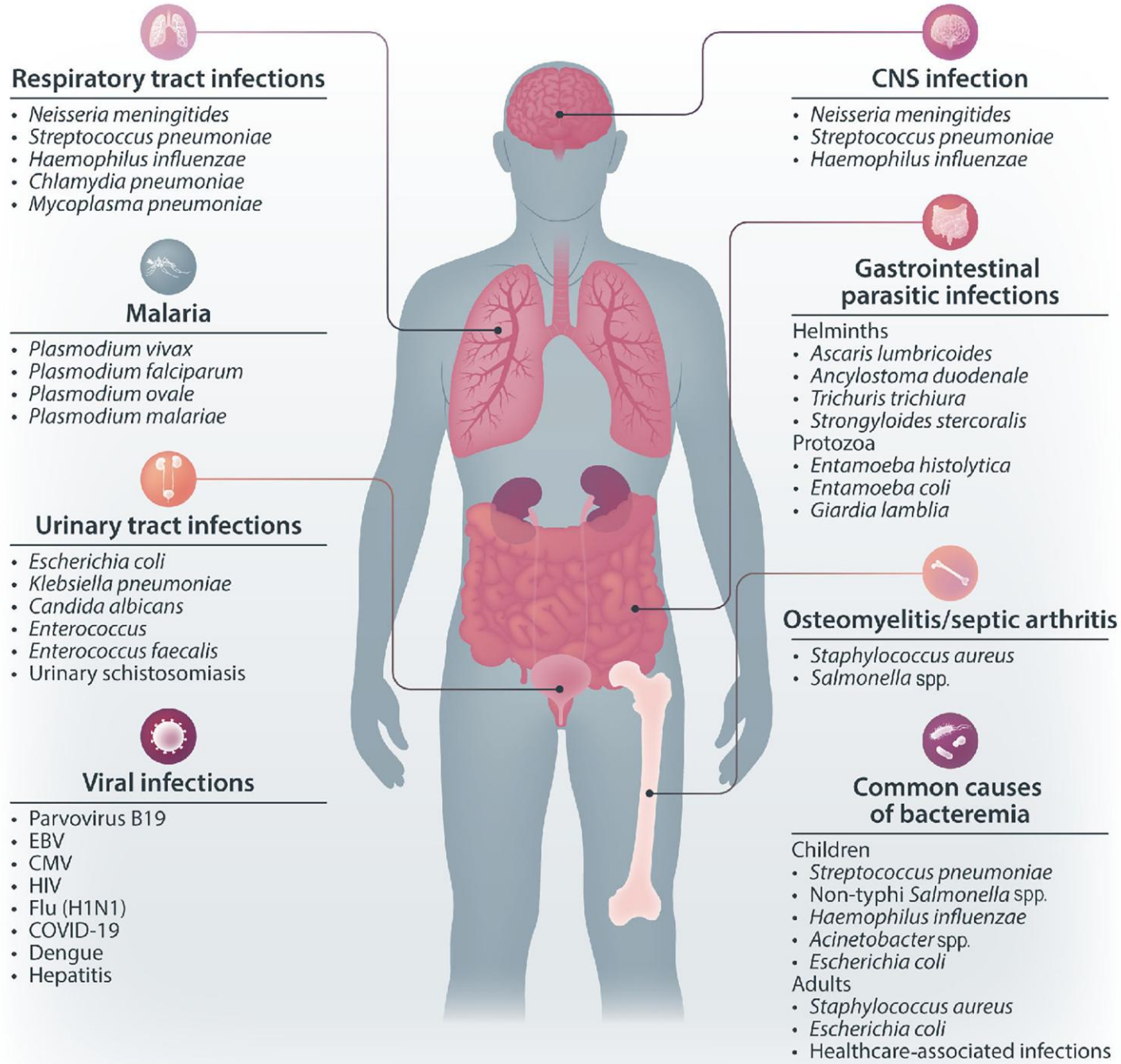
Infection



Why high risk for infections?

- SCD Treatments:
  - Hydroxyurea (used to prevent complications of SCD)
  - Chronic transfusions (for anemia)
- Functional loss of spleen at early age [Asplenia]
  - Increased risk of certain bacterial infections
    - *Streptococcus pneumoniae, Haemophilus influenzae, Neisseria meningitidis, Salmonella sp.*
- Infection can trigger other complications
- Signs & Symptoms
  - Fever [Temperature over $101^0F/38^0C$]
  - Fatigue/Lethargy
  - Cough/Respiratory distress

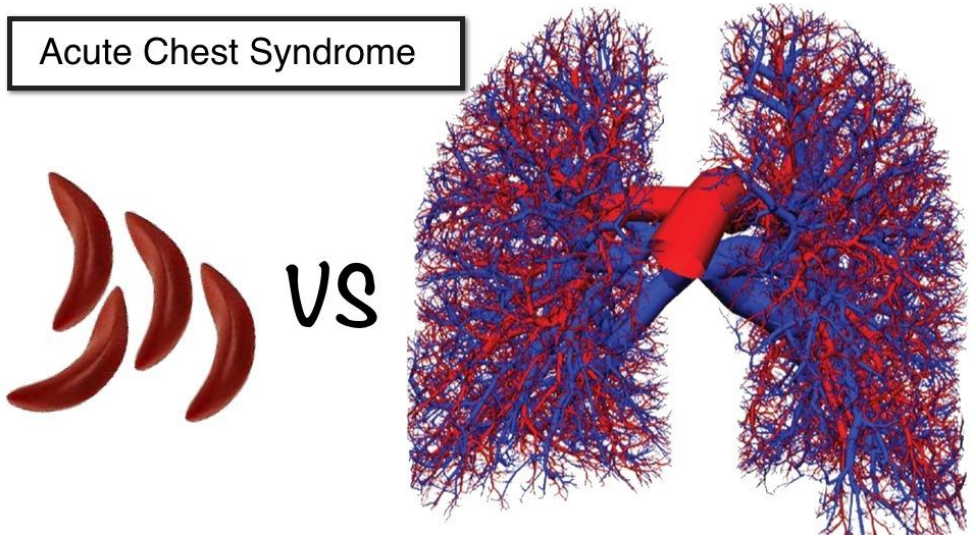
Ref 7


Respiratory tract infections
• Neisseria meningitides
• Streptococcus pneumoniae
• Haemophilus influenzae
• Chlamydia pneumoniae
• Mycoplasma pneumoniae
Malaria
• Plasmodium vivax
• Plasmodium falciparum
• Plasmodium ovale
• Plasmodium malariae
Urinary tract infections
• Escherichia coli
• Klebsiella pneumoniae
• Candida albicans
• Enterococcus
• Enterococcus faecalis
• Urinary schistosomiasis
Viral infections
• Parvovirus B19
• EBV
• CMV
• HIV
• Flu (H1N1)
• COVID-19
• Dengue
• Hepatitis
CNS infection
• Neisseria meningitides
• Streptococcus pneumoniae
• Haemophilus influenzae
Gastrointestinal parasitic infections
Helminths
• Ascaris lumbricoides
• Ancylostoma duodenale
• Trichuris trichiura
• Strongyloides stercoralis
Protozoa
• Entamoeba histolytica
• Entamoeba coli
• Giardia lamblia
Osteomyelitis/septic arthritis
• Staphylococcus aureus
• Salmonella spp.
Common causes of bacteremia
Children
• Streptococcus pneumoniae
• Non-typhi Salmonella spp.
• Haemophilus influenzae
• Acinetobacter spp.
• Escherichia coli
Adults
• Staphylococcus aureus
• Escherichia coli
• Healthcare-associated infections

Ref 7

Acute Chest Syndrome – Medical Emergency

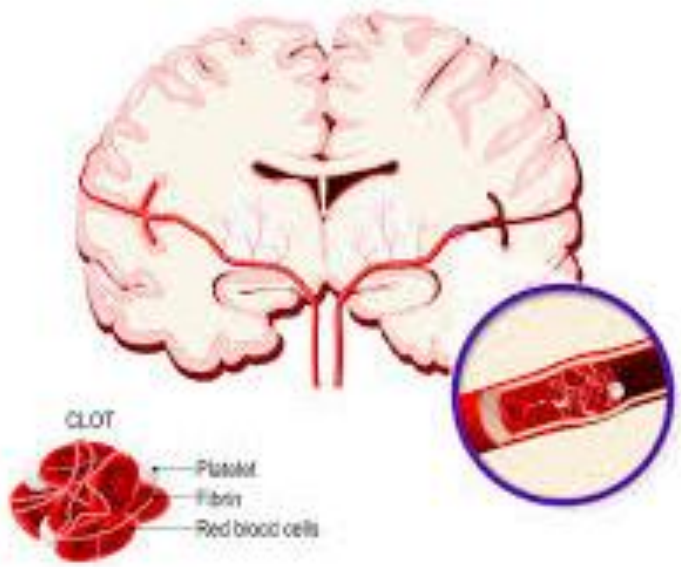
- A leading cause of hospital admission/death in SCD
- RBC sickle in small vessels of the lungs
  - Injury to vessels, respiratory distress, hypoxia
  - May be triggered by infections, asthma, pain crisis
- Signs & Symptoms
  - Fever
  - Pain: Chest, back and/or stomach
  - Low oxygen levels
  - Shortness of breath/Wheezing
    Tachypnea/Cough



Ref 1,2,7,8

Stroke – Medical Emergency

- RBC sickle and damage blood vessels in brain
  - Decreased blood flow/oxygen to brain
    - Annual screening (Transcranial Doppler)
- Signs & Symptoms
  - Weakness/Numbness (usually one sided)
  - Trouble speaking/Confusion
  - Headache
  - Visual Changes
  - Droopy face/mouth
  - Seizure



Ref 4, 9

Pain Crisis [Vaso-occlusive Episode]

- Pain Location
  - Bone/joints most common but can be anywhere
- Acute Treatment
  - Non-pharmacological:
    - Hydration
    - Heating pads—**never** cold
    - Soothing techniques: music, meditation, yoga/physical therapy
  - Medications
    - Non-narcotic pain medications: NSAID, Acetaminophen
    - Opiates

Ref 6, 10

Common Medications in SCD – Focus on Pain Management

- Acute Pain Management:
  - Over the counter:
    - Acetaminophen 15 mg/kg (max 1,000 mg/dose)
    - Nsaids [Ibuprofen/Naproxen]
      - Ibuprofen 10 mg/kg (max OTC: 400 mg/dose but 800 mg – with provider prescription)
      - Naproxen 5 mg/kg (max OTC: 440 mg /dose but 750 mg – with provider prescription)
  - Prescriptions:
    - Opiates [Morphine, Oxycodone, Hydromorphone]
- Maintenance Medications
  - Hydroxyurea – decreases HgbS to lessen complications of SCD
  - Gabapentin – nerve pain + adjunct with other pain medications
  - Duloxetine – antidepressant + adjunct for pain
  - Opiates – chronic use not preferred but may be needed

Ref 6, 10

Opiate Use in Schools

- Need an Individual Health Plan
  - Hematologist directed
  - Stepwise approach to care
- Follow your district's guideline for administering a prescribed opiate in the school

Prevention of SCD Complication

- Dehydration – Hydration
  - More volume in blood vessels = less chance to sickle cell to “stick”
- Temperature regulation
  - Overheating = dehydration
  - Too cold = constriction of blood vessels
- Infection prevention/immunizations
  - Precautions for viruses
  - Penicillin prophylaxis [Up to Age 5 years]
  - Vaccines *[Streptococcus pneumoniae, Haemophilus influenzae, Neisseria meningitidis]*

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2026 SCHOOL NURSE conference

Champions of Student Health

Resources:
Sickle Cell Association of America

Home - SCDAA

American Society of Hematology (ASH)

Clinical Practice Guidelines on Sickle Cell Disease - Hematology.org

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