Sickle Cell Disease Awareness and Primary Care Management

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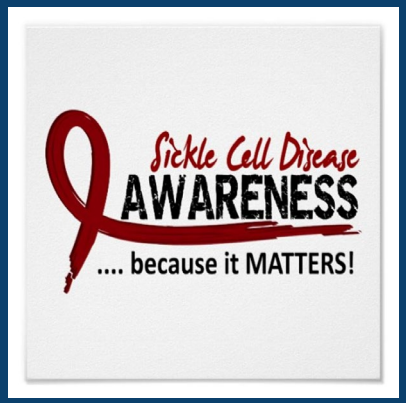
Our Lady of the Lake Adult Sickle Cell Clinic



Objectives & Goals

- Increase knowledge and understanding of Sickle Cell Disease (SCD)
- Primary care management of SCD
- OLOL Adult Sickle Clinic overview

Remember **Invisible ≠ Imaginary**

Statistics

United States

Sickle Cell Anemia is one of the most common inherited blood disorders in the United States.

- 90-100,000 individuals in the US are affected
- 1 in 365 African-American births have SCD
- 1 in 16,300 Hispanic- Americans births have SCD
- 1 in 13 African-Americans have Sickle Cell trait (AS)

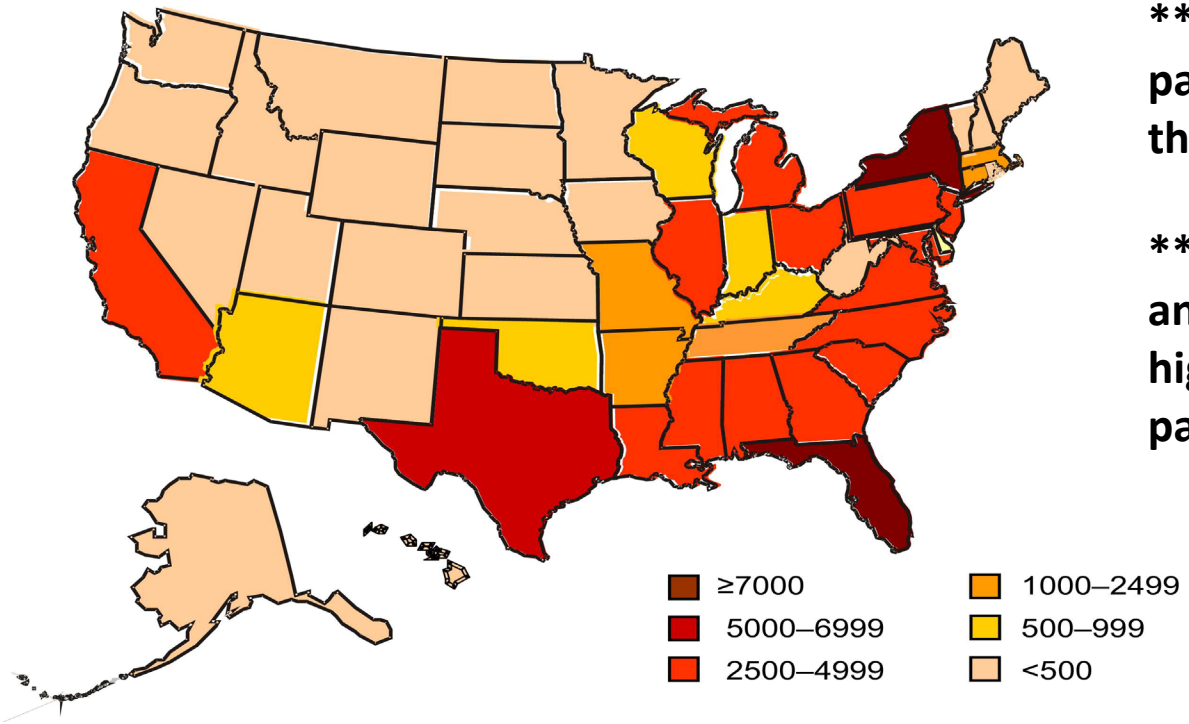
Louisiana

- Approximately 4,000 people living in Louisiana with Sickle Cell Disease

Baton Rouge- OLOL Sickle Cell Clinic/St. Jude Affiliate

- 260+ adult patients
- Approximately 400 pediatric patients (including Lafayette area)

Sickle cell disease rates in the US vary by state, but patients are geographically concentrated



**** 91% of the 97,930 patient with SCD in the US live in 23 states**

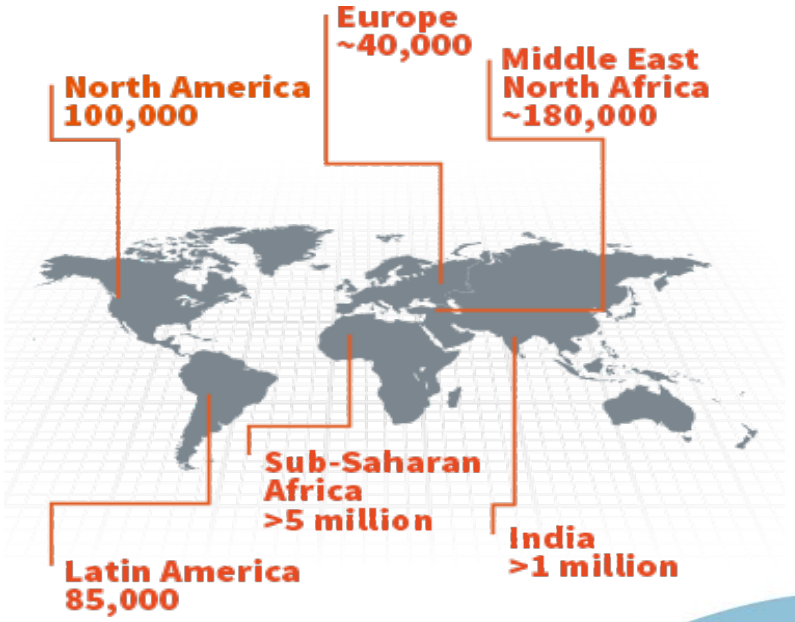
****Florida, New York and Texas have the highest prevalence of patients**

*1. Hassell K.L. Population estimates of sickle cell disease in the U.S. American journal of preventive medicine.*2010:38(4 Suppl:S512-521)

Global Burden of SCD

SCD disproportionately impacts Black and Hispanic Americans, however…..

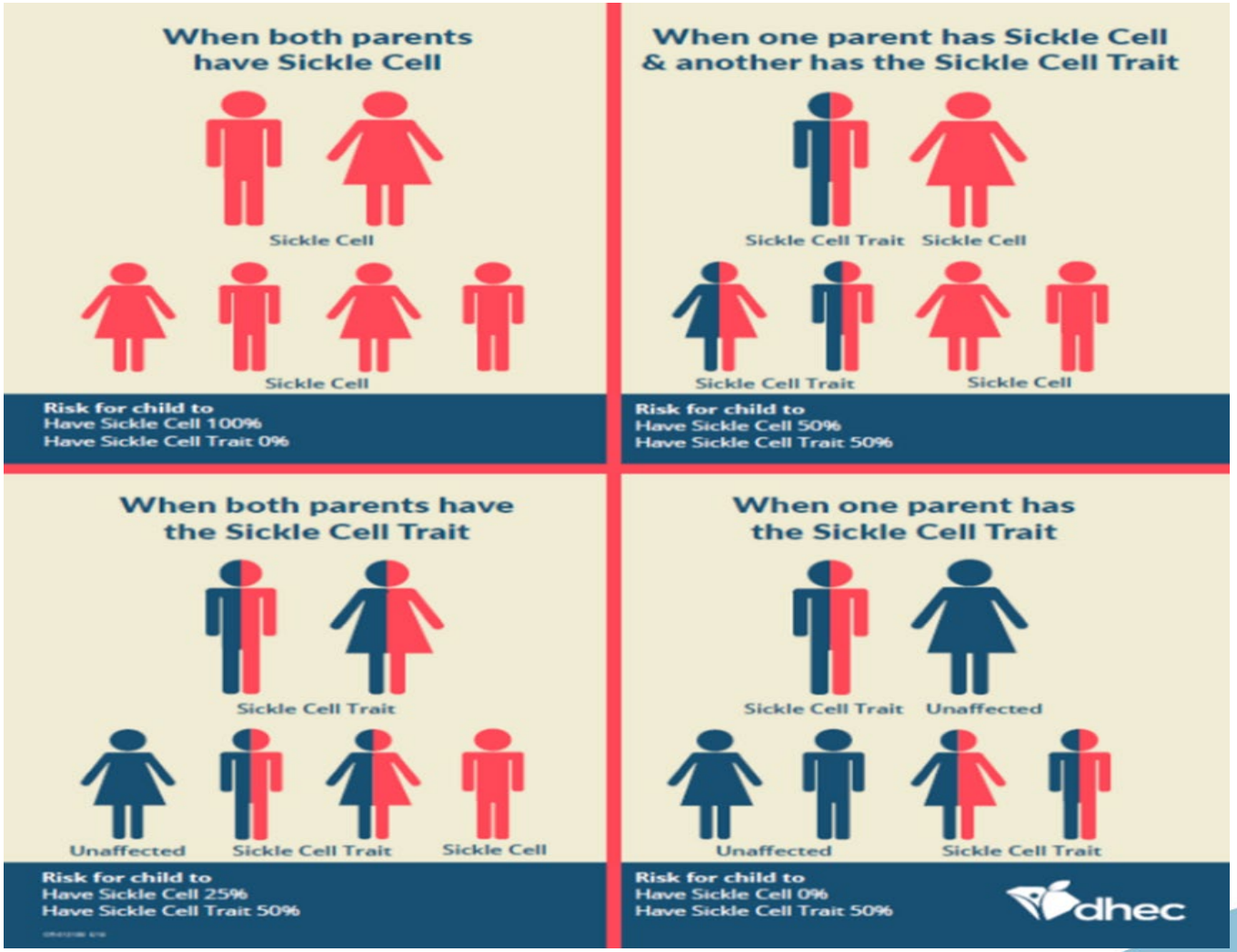
Sickle cell disease is a **global disease**. People with SCD also come from Southern European, Middle Eastern, or Asian Indian backgrounds



Types of Sickle Cell Anemia Disorders

Sickle Cell Trait = Carrier not Disease (1 in 13 African Americans)

Inheritance of Sickle Cell Disease



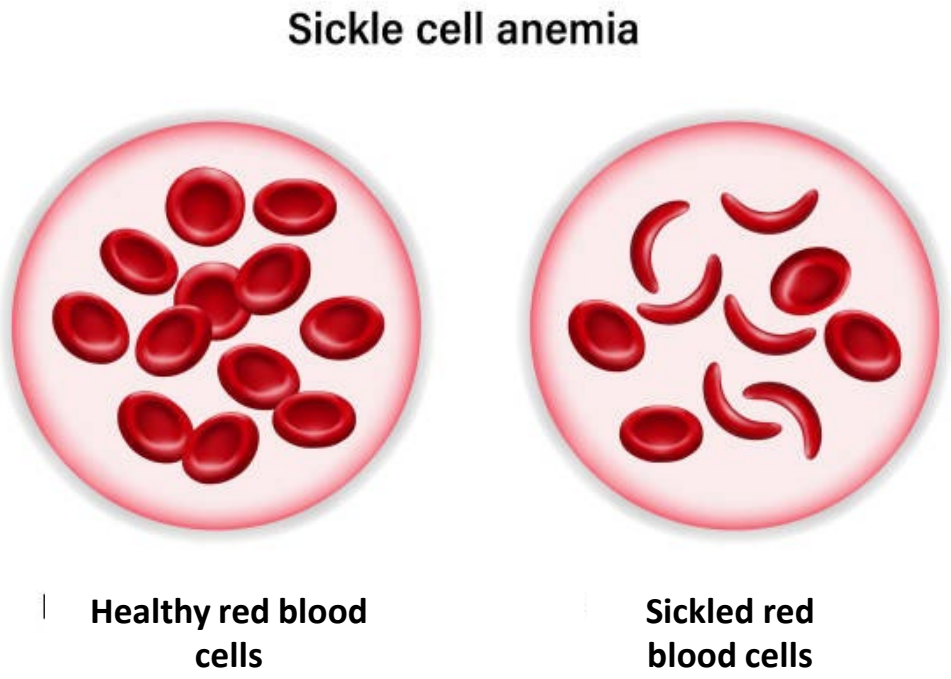
What is Sickle Cell Disease?

- SCD is an inherited blood disorder caused by a mutation on the 6th position on the B-globin change. There is a single amino acid substitution of valine for glutamic acid.

- Hemoglobin (Hb) is a special protein located within red blood cells (RBCs) that help carry oxygen throughout body.

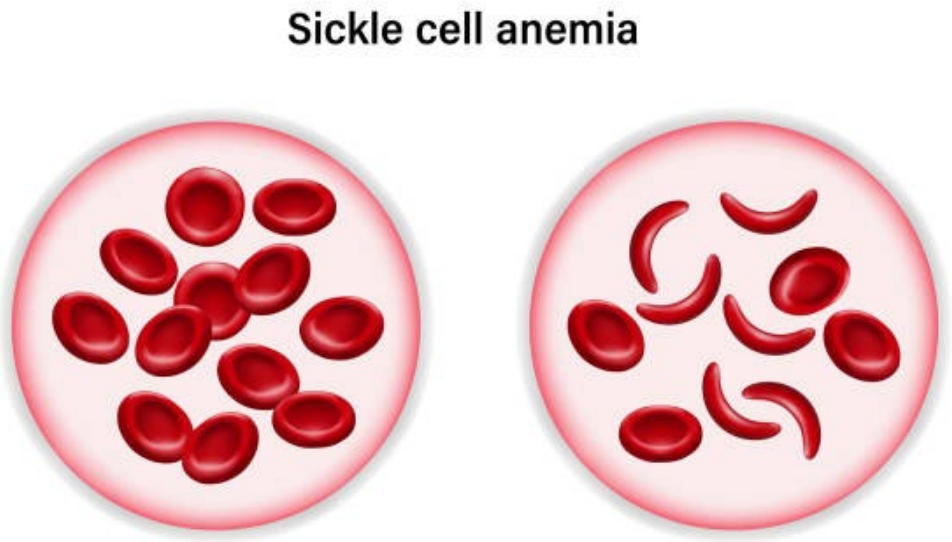
- SCD results in a mutated form of Hb called Hb S.

- In deoxygenated conditions, Hb S polymerizes causing the RBCs to take on a sickled shape.



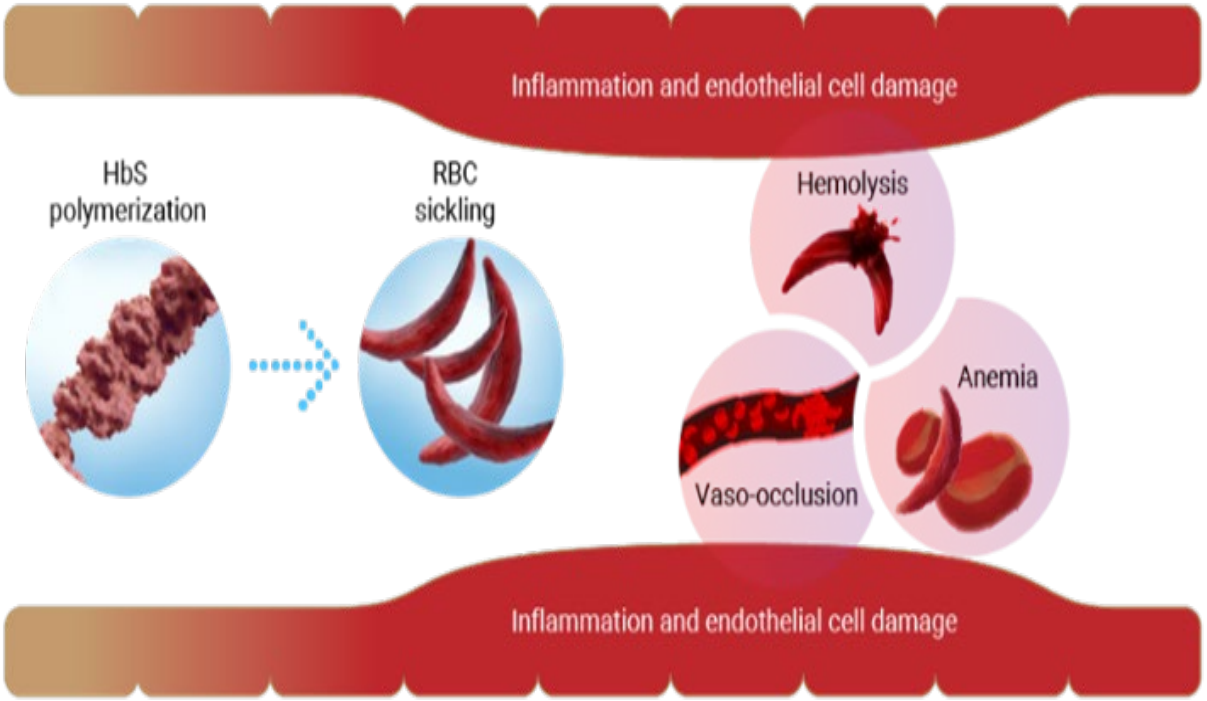
Anemia and Hemolysis

- Blockages and decreased oxygen delivery to tissue/organs
- Hemolysis- Breakdown of RBCs
- Anemia



Pathophysiology of Sickle Cell Disease

Anemia	Hemolysis	Hemolysis-mediated endothelial dysfunction	Inflammation	Adhesion-Mediated vaso-occlusion

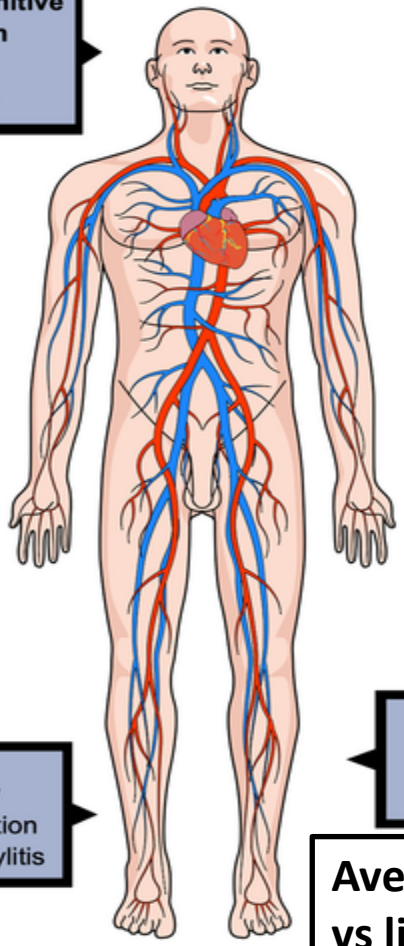


SCD complications (eg, organ damage)

PAIN: The Hallmark of SCD

- Primary reason people seek care
- Unpredictable in nature
- Secondary to vaso-occlusion
- Triggers vary
- Most will manage at home (important to assess)

Complications of Sickle Cell Disease



Psychosocial
-Anxiety
-Depression

Neurocognitive Dysfuntion
-Stroke
-Meningitis

Retinal
-Post-hyphaemia glaucoma
-Retinal infarction
-Retinal Detachment

Pulmonary
-Hypoxia
-Acute Chest Syndrome
-Pulmonary Hypertension

Reproductive
-Priapism
-Delayed Puberty
-Infertility
-Erectile Dysfunction

Children
-Cognitive Delay
-Growth Retartation
-Hand-foot Dactylitis

Immune
-Immunocompromised
-Prone to Infections

Hepatobiliary
-Hepatic Sequestration
-Hepatomegaly
-Cholecystitis
-Gallstones

Cardiovascular
-Vascular Occlusive Crises
-Anaemia
-Vasculopathy
-Diastolic Heart Failure
-Deep Venous Thrombosis
-Pulmonary Embolism

Spleen
-Splenic Infarction
-Splenic Sequestration
-Functional Splenia/ Hypoplasia

Renal
-Renal Medullary Carcinoma
-Hematuria
-Renal Failure

Bone
-Bone Marrow Infarction
-Osteomyelitis

Average Life expectancy: 52.6 years vs life expectancy in US 73(M) 79(F)

VOC Management

VOCs are managed in various settings

Patients surveyed report that **~4 of 5 VOCs are managed at home without a health care visit.** [1]

VOCs are the **leading cause of ER visits.** [2]

VOCs are the **leading cause of hospitalizations.** [3]

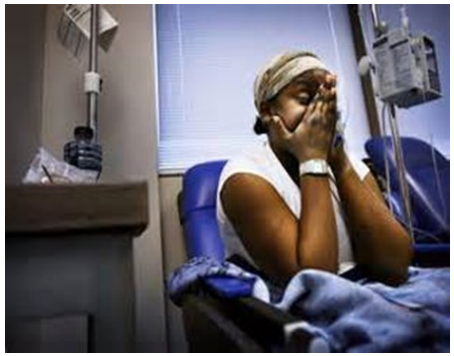
Regardless of where a VOC is managed, the etiology is the same. VOCS start with vaso-occlusion [4]

1Data form 182 adult patients with sickle cell anemia followed prospectively over 5 years at the sickle cell center of the Cardeza Foundation for Hematologic Research of Jefferson Medical College and Thomas Jefferson University Hospital, Philadelphia, from December 1998 to January 2002

2National Estimate of SCD-related ER visits. Data from the Nationwide Emergence Department Sample (NEDS) of 50,418 ER visits for patient with SCD in 2006.

3 According to PISCES (Pain is Sickle Cell Epidemiology Study),in which 232 adults with sickle cell disease completed daily pain diary longs (for up to 6 months), home management of pain episodes (as self-reported) constituted about 13% of the total days. The use of health care facilities, constituted less than 4% of the total days

VOC management



Outpatient

- IV fluids
- IV/SQ (Dilaudid, Morphine)

Individuals are assessed and medication administered within 1 hour of their arrival. Pain is frequently reassessed q 30-60 minutes for consideration of additional doses of pain medication to optimize their pain control

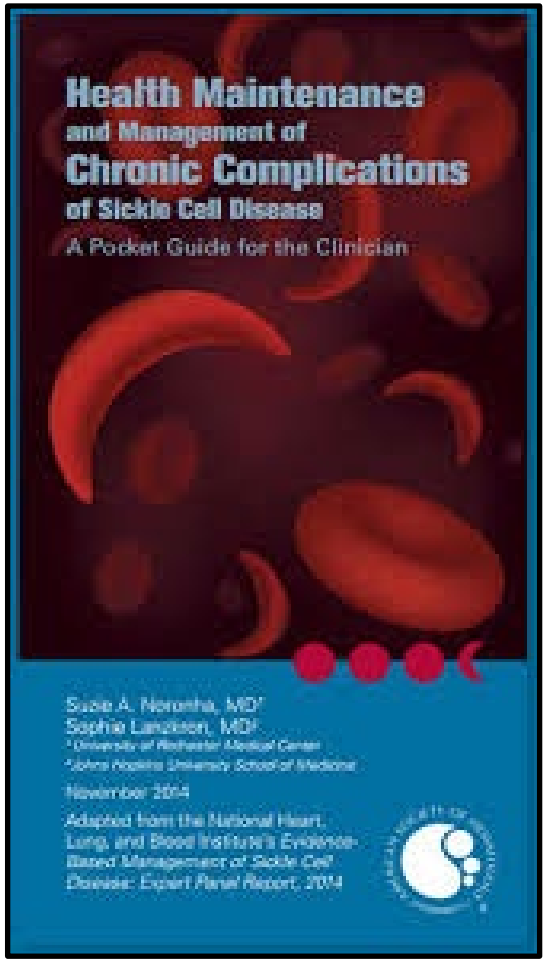
- Toradol if not contraindicated
- Oral short acting opioids (i.e. Norco, Percocet etc)

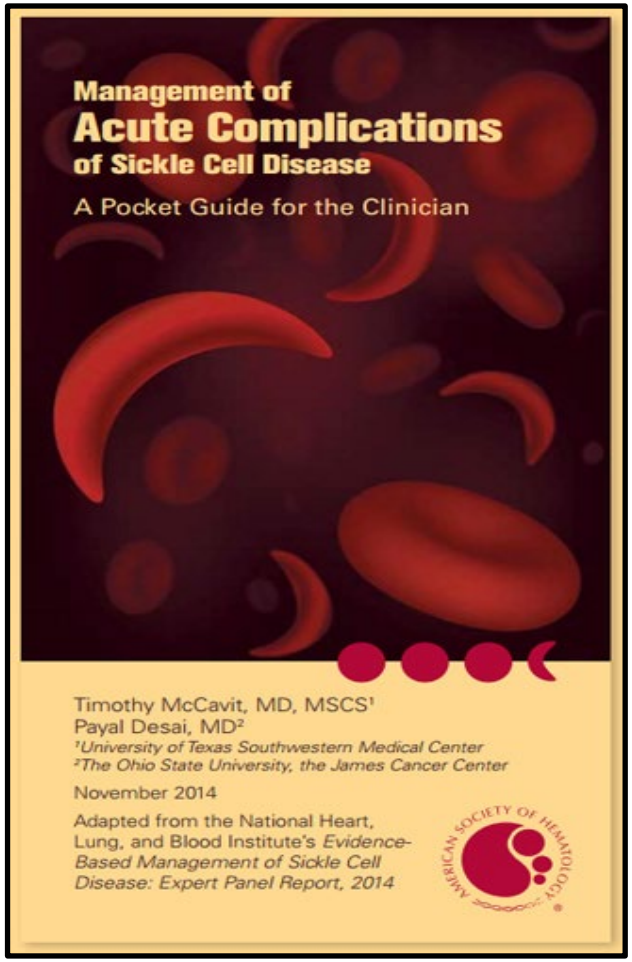
Inpatient

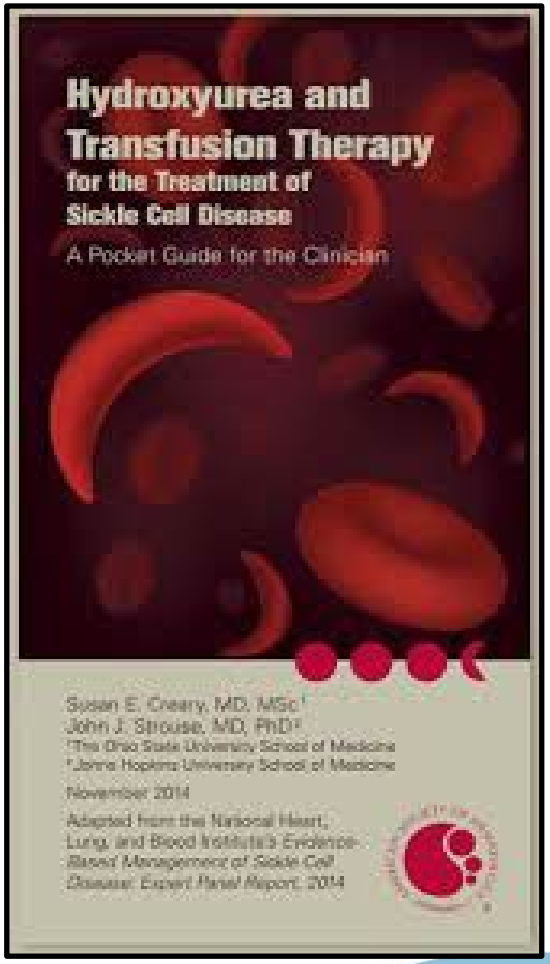
- PCA use with basal dosing
- PCA use with demand dosing only
- PCA use with addition of oral long -acting opioid

Sickle Cell Disease Management

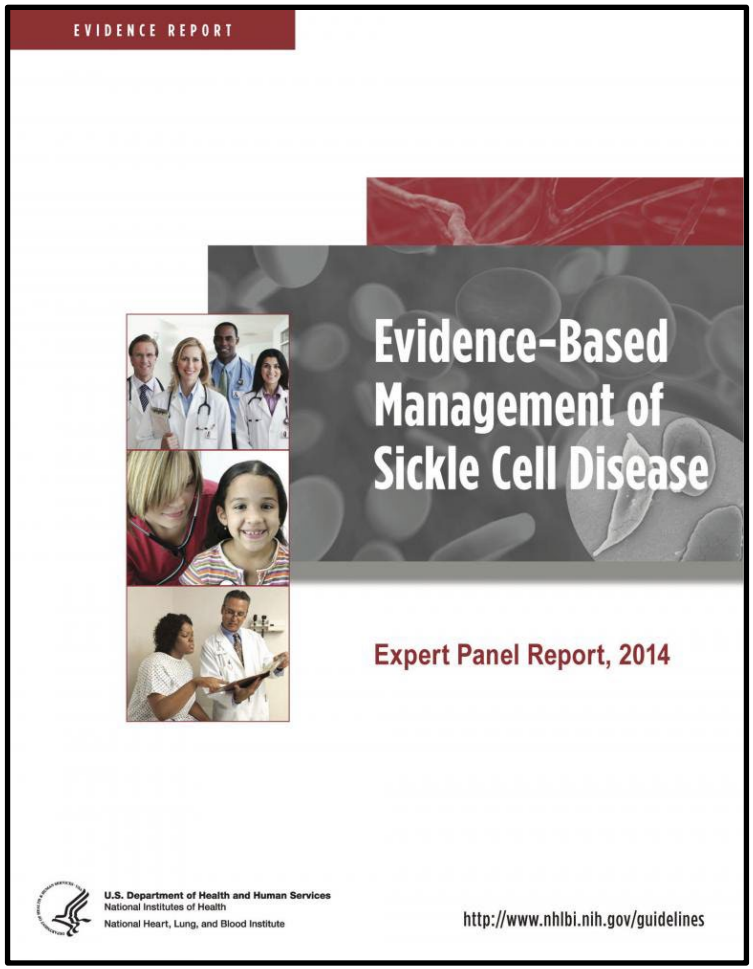
American Society of Hematology Guidelines



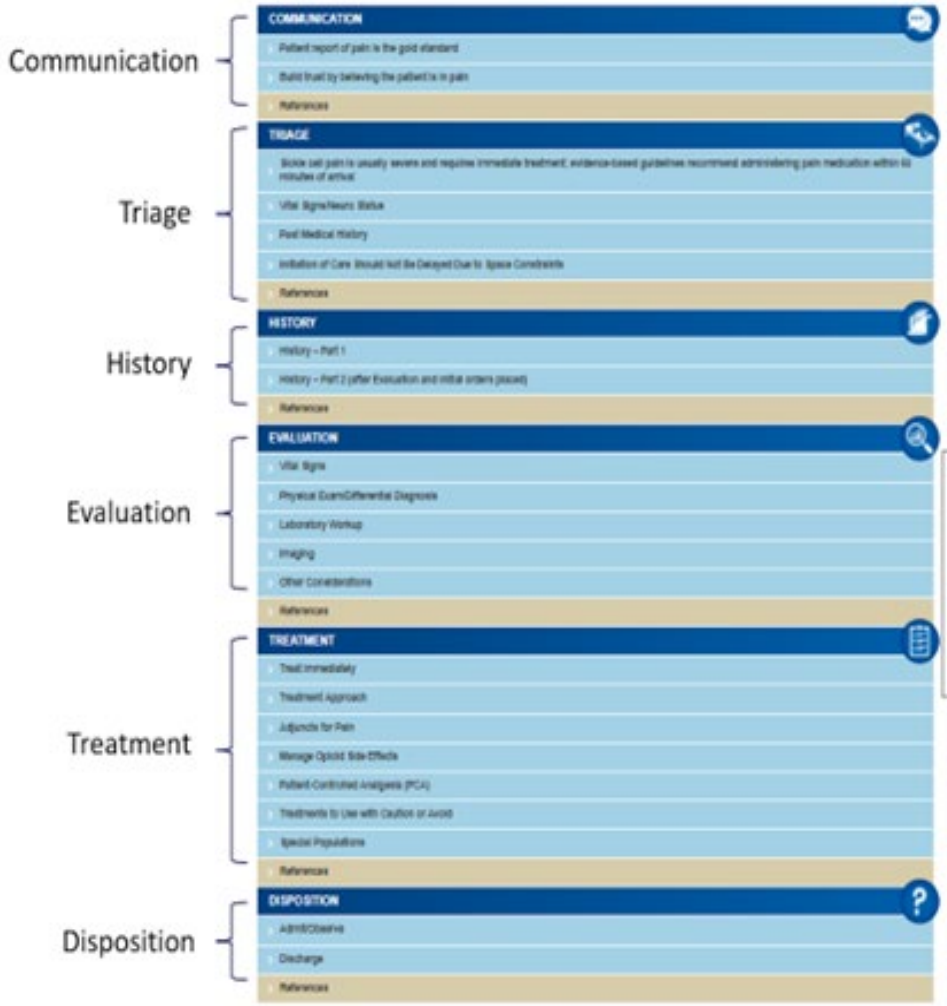




National Heart Lung Blood Institute Guidelines (NHLBI)



American College of Emergency Physician Guidelines (ACEP)



FREE!

https://www.acep.org/sickle-cell/

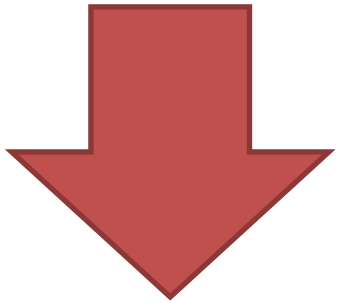
American College of Emergency Physicians

Primary Care Management of SCD

Importance of Primary Care Provider

Unfortunately, many adults with SCD may not have access to a hematologist for care.

Access to care



Morbidity/Mortality

Screenings

Ischemic retinopathy

Refer patients with SCD to an ophthalmologist for an annual dilated retinal exam ≥ 10 years old annually

Renal disease

Begin annual screenings for microalbuminuria and proteinuria with spot urine testing ≥ 10 years of age; Refer to nephrology for proteinuria (>300 mg/24hours)[1]

Screen for Iron overload

Patients with transfusion history >10-15/lifetime ; suspect iron overload. Check ferritin levels[1]

Vascular Disease and Stroke Risk

Screen annually with transcranial doppler (TCD) beginning at age 2 and continuing until at least 16 years of age. Refer children with conditional (170 to199 cm/second)or elevated (>200 cm/second) to a subspecialist with expertise in long term transfusion therapy aimed at preventing stroke. In children or adults who have had a stroke, initiate monthly simple or exchange transfusions. If no transfusion program available, initiate hydroxyurea therapy. [1]

**Do not perform screening with neuroimaging (TCD, MRI, CT)on asymptomatic adults with SCD

**Do not perform screening with CT or MRI on asymptomatic children with SCD.

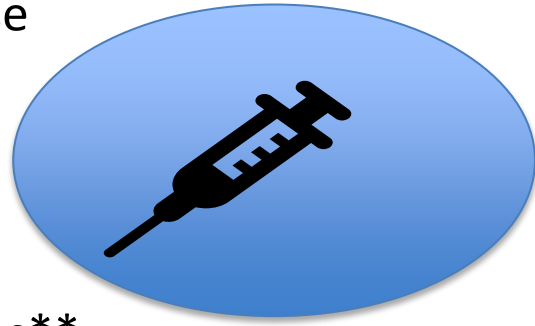
1 Health maintenance and management of chronic complications of sickle cell disease. Adapted from the National Heart, Lung and Blood Institute's *Evidence Based Management of Sickle Cell Disease; Expert Panel Report, 2014*

Preventive strategies and immunizations

Prophylactic Penicillin

** Children with SCD have an increased susceptibility to bacterial infections, especially to those caused by *Streptococcus Pneuomoniae*

- Administer prophylactic penicillin BID until 5 years of age, longer if they have had splenectomy or invasive pneumococcal disease



Immunizations

** Please ensure that patients are up to date on all vaccines**

- Meningococcal vaccine
- *Haemophilus influenzae* Type b vaccine (if not previously received Hib vaccine)
- Pneumococcal vaccine
- Influenza vaccine

Current Therapies for SCD

Disease management

Hydroxyurea
- First approved therapy (1998 for adults)
- Indication: 2-3 pain episodes yearly
- Modifies the course of SCD ; Increases Fetal Hg
- Decreases the frequency of pain crisis, ACS and need for blood transfusions in SCD patients
- Not universally accepted (oral chemotherapy)

L-Glutamine (Endari)
- Anti-inflammatory
- Reduces oxidative stress in red blood cells
- Naturally occurring amino acids made by body
- Oral powder packs BID
- Decreases frequency of pain crisis and acute chest syndrome in SCD patients

Crizanlizumab (Adavkeo)
- Decreases frequency of pain crisis in SCD patients
- Once monthly IV infusion monoclonal antibody

Blood Transfusions
- Improve stroke risk
- Decrease recurrent acute chest syndrome in several studies
- Multiple unwanted complications

Palliative management
- Pain management/opioids
- Anti-depressants, anti-convulsants, muscle relaxers

Curative therapies

Bone Marrow (STEM CELL) Transplant

Gene Therapy

OLOL Adult Sickle Cell Clinic

- OLOL Adult Sickle Cell Clinic est. 2014 - 260+patients
  Servicing patients from: Natchez, MS, Alexandria, Bunkie, Lafayette, New Iberia, Opelousas, Walker, Bogalusa, Sorrento, Gonzales, Houma etc.
- Staffed by 2 nurse practitioners, LCSW, physician
- OLOL St. Jude Affiliate-Children's Hospital Transition program
- Outpatient infusion (fluids, IV pain medication, blood products, Crizanlizumab)
- Screenings/Vaccinations
- Disease Modifying Therapies
- Mental Health counseling
  (Collaborative Care Model)
- Clinical Trial Participation

Saving Lives Every Day !!

Acknowledgments

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