Sickle Cell Disease: Myths and Realities

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Outline

- Who we are
- Sickle cell disease pathophysiology
- Sickle cell trait versus sickle cell disease
- Clinical complications
- Treatments and interventions
- Emergency department care
- Advocacy
- Resources
Who we are
Northern California Comprehensive Sickle Cell Center

- Multidisciplinary team provides comprehensive care
- Day hospital for transfusion services
- Inpatient services for pediatrics
- Outpatient services for adult and pediatrics
- Bone Marrow Transplant Unit
Our Program

Sickle cell population N = 732

- 52% female
- 84% African American; 3% Hispanic; 13% mixed or other
- 60% Hb SS; 26% Hb SC; 10% Hb Sbeta+ or 0
- Age breakdown
  - 33% 0 - 12 years
  - 30% 13 - 24 years
  - 37% 25+ years

- 60% Medi-Cal

Catchment area: culturally and sociodemographically diverse Northern California Region
What is Sickle Cell Disease?
A group of inherited blood disorders

Normal Red Blood Cells
- soft, flexible, disc-shaped
- easily flow through small blood vessels
- live for 120 days

Sickle Cells
- rigid, misshapen
- often get stuck in small blood vessels
- live for 20 days or less
Sickle Cell Pathophysiology

Oxygenated erythrocyte containing HbS

Deoxygenated erythrocyte with polymerisation of HbS

Dehydrated, sickled erythrocyte

Occlusion of postcapillary venules (vaso-occlusion)

Reperfusion

Free radicals, causing tissue damage

Vasculopathy and endothelial dysfunction

Infarction

Acute pain
Acute chest syndrome
Hypersplenism
Osteonecrosis
Nephropathy

Inflammation
Increased expression of VCAM-1 and other adhesion molecules
Hypercoagulability

Haemolysis

Free plasma haemoglobin, inactivating NO and generating reactive oxygen species

Pulmonary hypertension
Priapism
Leg ulcers
Cerebrovascular disease

Functional NO deficiency

Myth

Sickle Cell Disease is a “Black” Disease
Reality

SCD is one of the most common inherited conditions in the world with over 300,000 affected births annually.

Affects approximately 100,000 people in U.S.

- 1 in 12 Blacks
- 1 in 1400 to 1 in 36,000 Hispanics
- 1 in 58,140 Caucasians

Individuals of Middle Eastern, Asian, Indian, and Mediterranean descent are also commonly affected.
Hemoglobin Trait Advantage
Myths

You can catch sickle cell disease

“How long have you had sickle cell disease?”
Reality: SCD is Inherited

Probability for each pregnancy when two individuals have sickle cell trait

- 25% Hgb SS
- 50% Hgb AS
- 25% Hgb AA
Myths

Sickle cell trait can “turn into” sickle cell disease.

Sickle cell trait is a form of sickle cell disease
Reality: Sickle Cell Trait is NOT a Disease

Complications associated with SCT are UNCOMMON

Complications associated with EXTREME conditions

- Severe low oxygen, such as in a unpressurized aircraft
- Severe dehydration
- Excessive physical exercise under severe conditions

Mitchell J Natl Med Assoc 2007;99:300-05
Reality:
Sickle Cell Trait is NOT a Disease

U.S. Army demonstrated that universal precautions to reduce dehydration and allow for gradual conditioning, were effective in reducing heat/exercise related collapse for ALL recruits.

Hematuria (blood in urine) may occur – consult with your doctor.
Sickle Cell Disease: Clinical Complications

- Cerebral infarcts - stroke
- Pneumonia or Acute Chest Syndrome
- Splenomegaly/sequestration
- Increased infections
- Pain episodes
- Bone disease
- Kidney disease
- Cholelithiasis
- Delayed puberty, priapism, high risk pregnancy, skin ulcers, retinopathy
Pain

- Most common complication of SCD
- Most common reason to seek treatment
- BUT – majority of pain is managed at home
Myths

You can tell how much pain someone with sickle cell disease is in by looking at them. Pain is just something that people with SCD have to live with – it is not that serious.
Realities

The ONLY way to measure pain is by ASKING!

Pain is whatever the person says it is

Patients with SCD are routinely undertreated for pain

Under- or over-treatment of pain can lead to longer pain episodes and longer hospital stays

Acute pain should be promptly treated

High rates of pain associated with early mortality

RN.com Acute and chronic pain. 2010
Benjamin et al Blood 2000;95:1130-37
Platt et al NEJM 1991; 325: 11-6
Correlates of Frequent Pain

- Negative self-concept
- Increased anxiety and depression
- Negative body image
- Poor school performance
- Social isolation, poor peer relationships
- Decreased participation in normal activities of daily living
Myths

People with sickle cell disease cannot live normal, productive lives.

People with sickle cell disease die young.
Reality:
Sickle cell disease is serious but with comprehensive and preventive care, and strong support systems, people with SCD are living longer and more productive lives
Age at death for individuals with SCA in 1979 and 2006

--- 1979 (n=301)  --- 2006 (n=483)
Age at Death for People with SCD in California, 2004-2008

- 0-10 years: 4%
- 11-20 years: 5%
- 21-30 years: 15%
- 31-40 years: 17%
- 41-50 years: 23%
- 51-60 years: 21%
- >60 years: 15%

www.cdc.gov/ncbddd/hemoglobinopathies/phresh
Biopsychosocial Model: Systems

- Person
- Family
- Community/
  Health Care System
- Society/Culture
Psychological Adjustment

People with SCD have shown increased vulnerability to

- Poor academic performance
- Lowered adaptive functioning
- Internalizing rather than acting out behavior problems
- Depression
- Low self esteem
- Difficulties in social and vocational functioning
**Treatments**

**Goals**

- Relieve pain
- Prevent infection, organ damage, stroke
- Control complications

- Pain medicines and fluids
- Hydroxyurea
- Transfusions
- Daily antibiotics (infancy to age 5), routine vaccinations
- Screenings – eye damage, stroke risk, body iron, bone density, vitamin D levels, mental health symptoms…
Pain Treatment Planning

Identify appropriate interventions based on comprehensive assessment

PHARMACOLOGICAL
- Acetaminophen or NSAIDS
- Opioids
- Adjuvants

BEHAVIORAL
- Relaxation
- Deep Breathing
- Behavior Modification
- Biofeedback
- Exercise

PSYCHOLOGICAL
- Cognitive Therapies
- Hypnotherapy
- Imagery
- Distraction
- Social Support

PHYSICAL
- Hydration
- Heat
- Massage
- Hydrotherapy
- Ultrasound
- Acupuncture/ Acupressure
- TENS
- Physical therapy

Identify patient/family educational needs

Formulate treatment plan

Benjamin et al. 1999, American Pain Society
Successful Transition

Pediatric medical home:

- Evaluates transition readiness goals and increases interactions until they are reached
- Identifies available and qualified adult care providers, along with family
- Ensures appropriate communication of any and all medical needs to receiving provider

Adult medical home optimizes patient’s self-management skills by providing support through care coordination and consultation
New Treatments

- Blood and Marrow stem cell transplant
- Gene therapy
- New medicines
  - Decitabine
  - Adenosine A2a receptor agonists
  - 5-HMF
  - Statins
How to have a Successful Emergency Room Visit
Keep Calm and Bring a Wingman or Wingwoman
Have Backpack Will Travel

- Government ID / Picture
- Dr.’s Information
- Cell Phone & Charger
- Insurance Card
- Medication List
- Note Pad & Pen
- Reading Glasses
- Glamour bag
Notebook & Pen

- Sign In Time
- Triage Time
- Where Is My Room
- Dr. Visit Time
- Medication Time
- Test & Review Time
- Admit or Not
Discharge

- Instructions
- FollowUp
Survey Says

Who pays attention to the Survey

Should I sign my name
Myth

I thought sickle cell disease was cured. I never hear anything about it, so I thought it must not be around anymore.
### Realities: Support for SCD

<table>
<thead>
<tr>
<th>Variable</th>
<th>SCD</th>
<th>CF</th>
</tr>
</thead>
<tbody>
<tr>
<td>US Prevalence</td>
<td>80,000</td>
<td>30,000</td>
</tr>
<tr>
<td>Federal support, millions</td>
<td>$90</td>
<td>$128</td>
</tr>
<tr>
<td>Total NIH and private support, millions</td>
<td>$90.4</td>
<td>$280.2</td>
</tr>
<tr>
<td>Total support per affected person</td>
<td>$1130</td>
<td>$9340</td>
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</tbody>
</table>

- Millions of people live with SCD around the world
- In the U.S., disparities remain in survival, quality of life and quality of care
- There is a pressing need for advocacy

Smith et al *Pediatrics* 2006;117: 1763-70
Call to Action

Ask questions and insist upon answers about sickle cell care and treatment

- Your hospital administrators and insurance companies
- The California Public Health Department and your local legislators
- Your representatives and senators
- Private philanthropic groups
People with Sickle Cell Disease Need Your Support

- By educating yourself, you reduce their isolation
- Your activism can lead to reducing other health disparities
- Our community is strengthened when we take individual and collective responsibility for our health
Local Resources

Comprehensive Sickle Cell Center – Children’s Hospital & Research Center Oakland

- Elliott Vichinsky, MD Director (510) 428-3651
- Marsha Treadwell, PhD Project Director, Network of Care for SCD (510) 428-3356

Sickle Cell Community Advisory Council

- Wanda Williams, Chair (510) 888-4568
Celebrating Sickle Cell Awareness Month

Art Contest
Everyone ages 5 and up can participate! Select Artwork will be displayed at the John ‘Larry’ Valley - 104 Heroes Blood Drive on Saturday 20, 2014

Resources
Check out our library of articles, publications and research on Sickle Cell Disease. More >>

Programs
The Northern California Network of Care for Sickle Cell Disease, at Children’s Hospital & Research Center Oakland, launched in 2009 as one of the Health Resources and Services Administration’s (HRSA) programs for sickle cell disease (SCD)....More >>

The California PHRESH Project is a two-year project funded by the Centers for Disease Control and Prevention. PHRESH stands for the Public Health, Research, Epidemiology & Surveillance in Hemoglobinopathies. More >>

Sickle Cell Trait
Sickle cell trait can only be inherited- In other words, something a parent passes to a child like eye or hair color.

Sickle cell trait is not contagious- you cannot "catch" it. More >>

Gallery

What is Sickle Cell Disease?
A genetic disorder of hemoglobin, a protein in red blood cells that carry oxygen.

The conditions cause a change in hemoglobin A that create hemoglobin S.

“Sickle cell anemia” refers to the condition where both hemoglobin are S (hemoglobin SS).

Common symptoms include: Pain: acute chest syndrome...More >>

Standards of Care Guidelines

Watch video and take survey

Sickle Cell Champion

www.casicklecell.org

California Sickle Cell Resources

@SickleCellCalif
Fighting a Global Disease

Many people in this country think Sickle Cell Anemia is a blood disorder that only affects African Americans and it is a disease that is disappearing. But, these are just two of the misconceptions surrounding a world-wide disease. On this show, my guests and I discuss both the facts and the misconceptions surrounding the blood disorder and we also discuss why having the facts about the disease can be critical for its treatment and for those who have it.

In this photo, Dr. Theopila Jackson, show host Henrietta J. Burroughs, Fred McFadden and Wanda Williams are shown on the set of the Talking with Henrietta television show after their discussion about a blood disease that affects millions around the world. Dr. Kim-Anh Nguyen also participated on this show.

This edition of Talking with Henrietta can be viewed on Channel 27, on the Midpeninsula and on the Internet from Sunday, September 16, 2012 through Saturday, September 29, 2012. The show can be seen on Sundays@5 p.m., Tuesdays@6 p.m., Wednesdays 4 a.m. and 11 a.m., Thursdays@6 p.m. and Fridays@6 a.m. and 12 p.m. and Saturday at 5 p.m. This show streams on the web at the above days and times at www.midpenmedia.org. Click the schedule for additional information about the show. Click here to see more Talking with Henrietta video excerpts.
SICKLE CELL SUPPORT GROUP

CHORI Library
5700 Martin Luther King Jr. Way
Oakland, CA 94609
Once Monthly on Saturdays
Time: 11:00 a.m. – noon
Thank You!