NORTHERN CALIFORNIA
NETWORK OF CARE
FOR SICKLE CELL DISEASE

Sickle Cell Disease: Myths and Realities

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Oakland | San Francisco

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

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

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

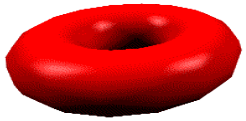
- 60% Medi-Cal
- Catchment area: culturally and sociodemographically diverse Northern California Region

What is Sickle Cell Disease?

A group of inherited blood disorders

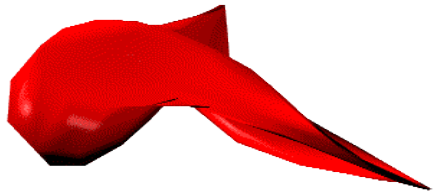
Normal Red Blood Cells

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

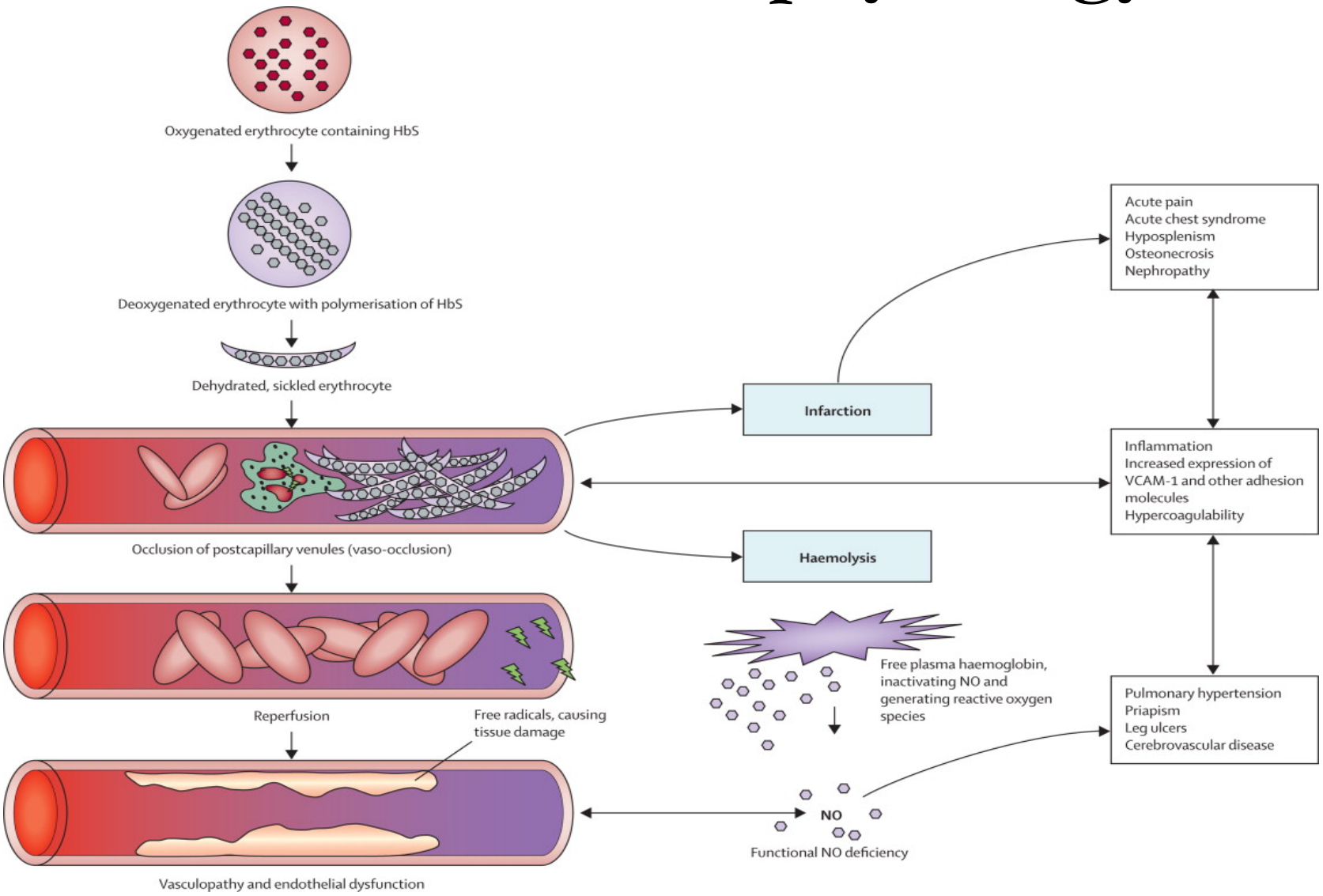


Sickle Cells

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



Sickle Cell Pathophysiology



Rees et al. *Lancet* 2010; 376: 2018-31

Myth

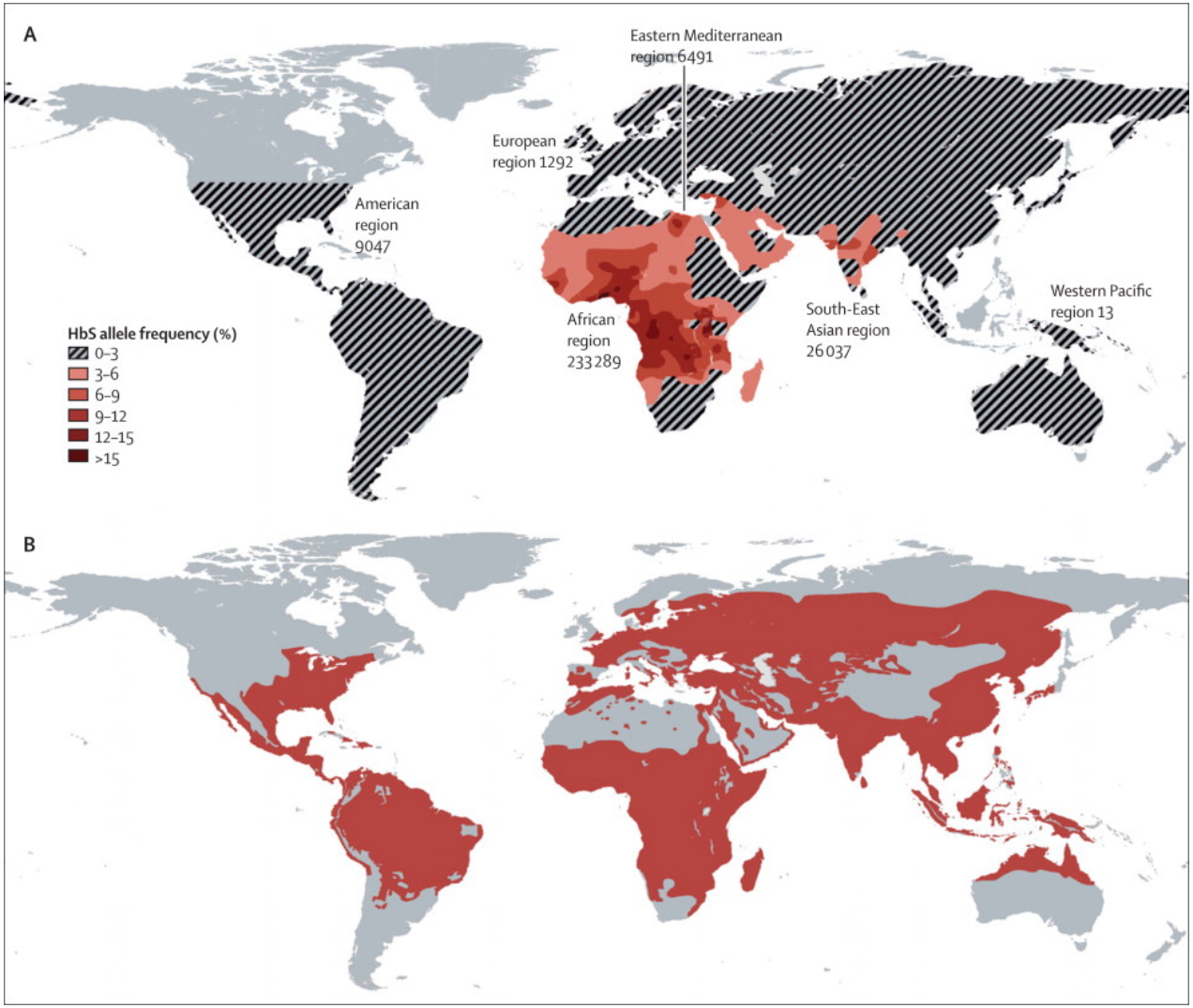
Sickle Cell Disease is a "Black" Disease

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

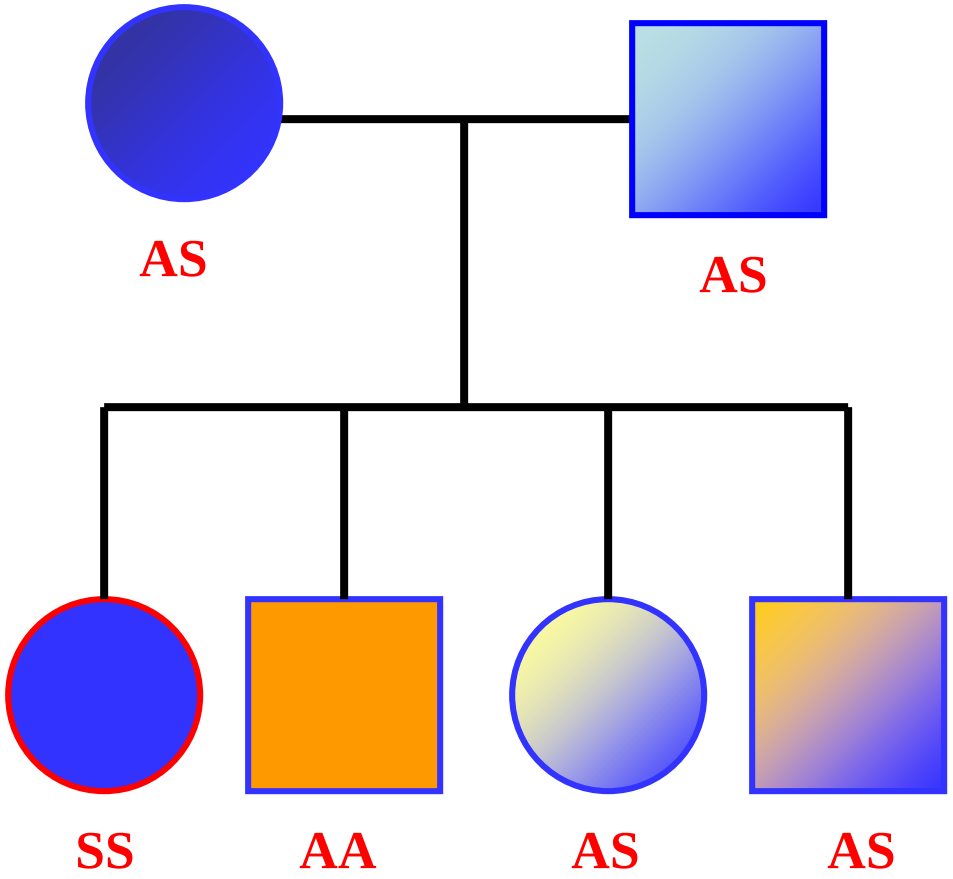


Myths

You can catch sickle cell disease

"How long have you had sickle cell disease?"

Reality: SCD is Inherited



Probability for <u>each</u> 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

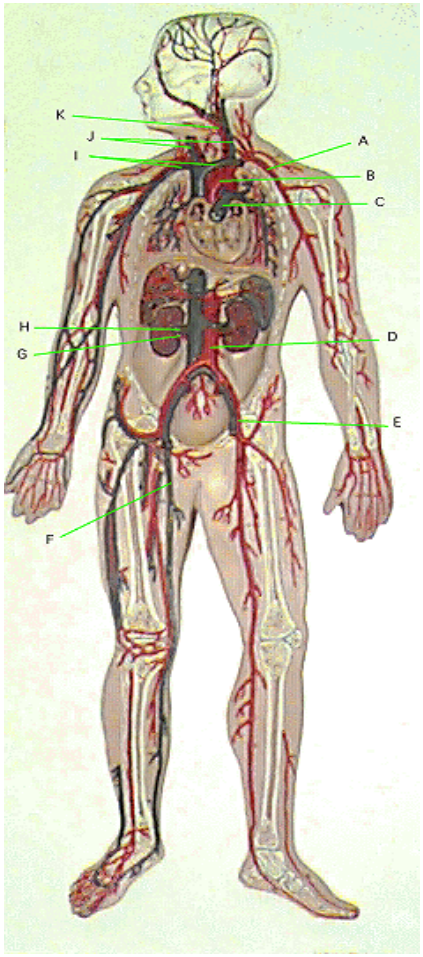
Reality:
Sickle Cell Trait is NOT a Disease

- 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

Mitchell *J Natl Med Assoc* 2007;99:300-05

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

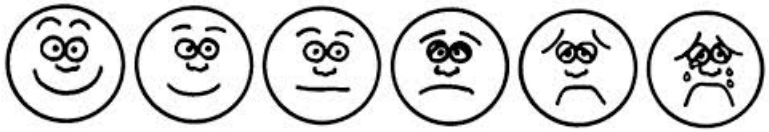
Smith, Scherer *ASH Education Program* 2010; 409-15

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

Ballas *ASH Education Program* 2007; 97-105
Taylor et al *Pain Manag Nurs* 2013; 14: 287-301

Myths

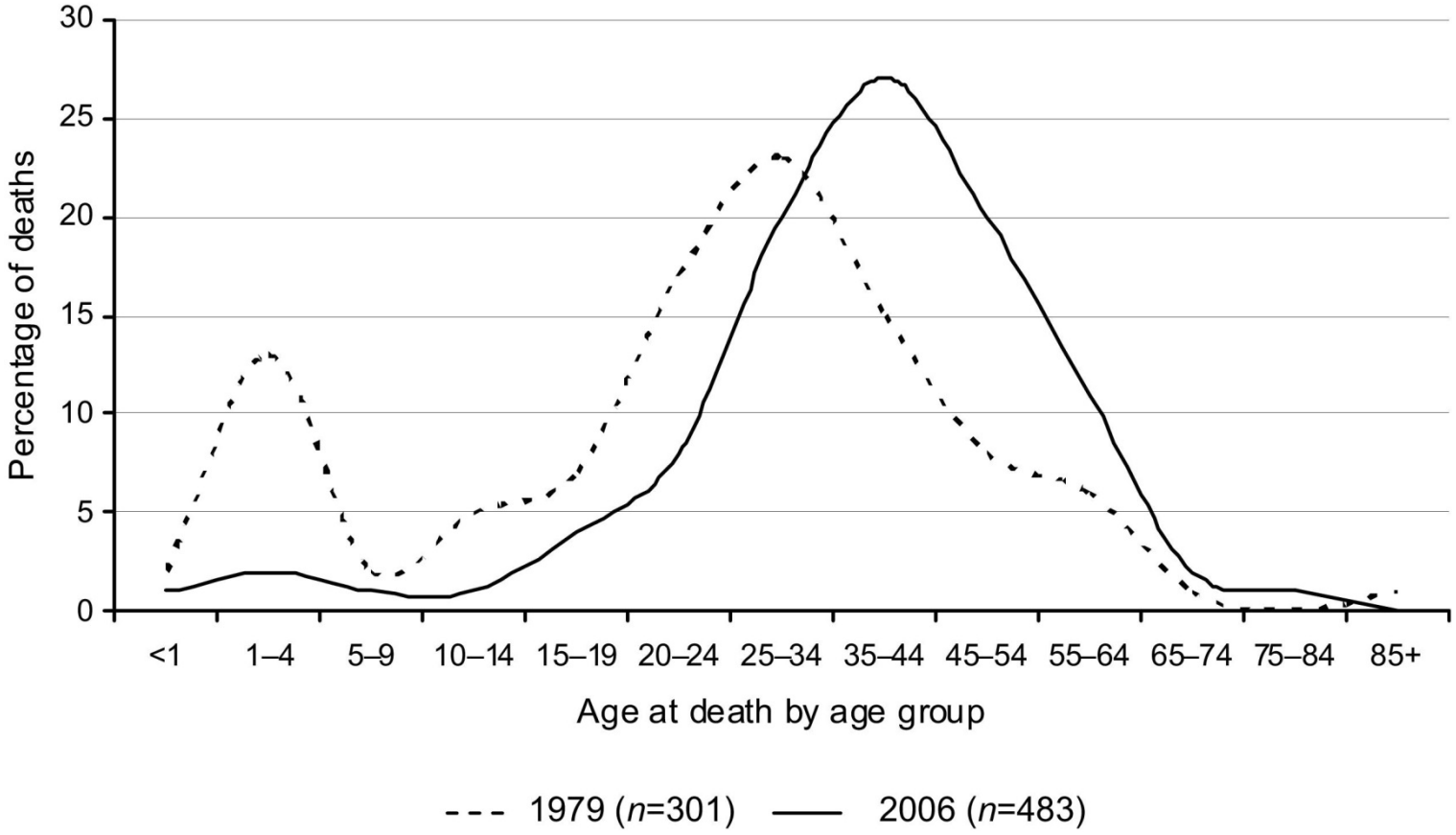
People with sickle cell disease cannot live normal, productive lives.

People with sickle cell disease die young.

Reality:

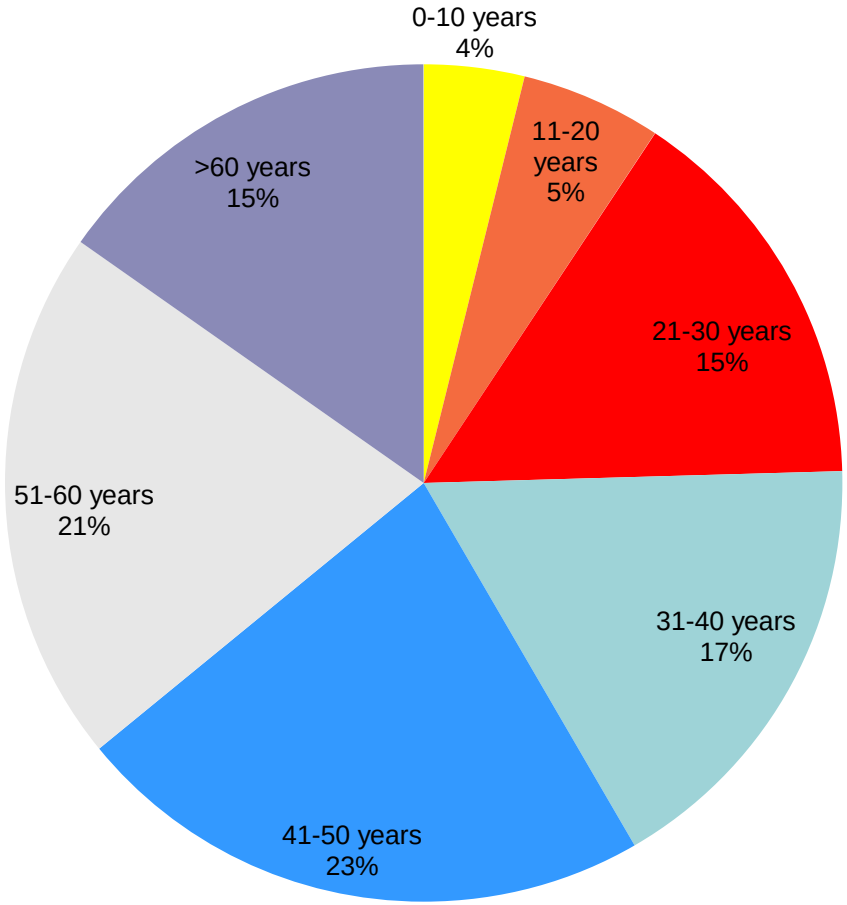
Sickle cell disease is serious but with comprehensive and preventive care, and strong support systems, people with SCD are living longer and more productive lives

Age at death for individuals with SCA in 1979 and 2006



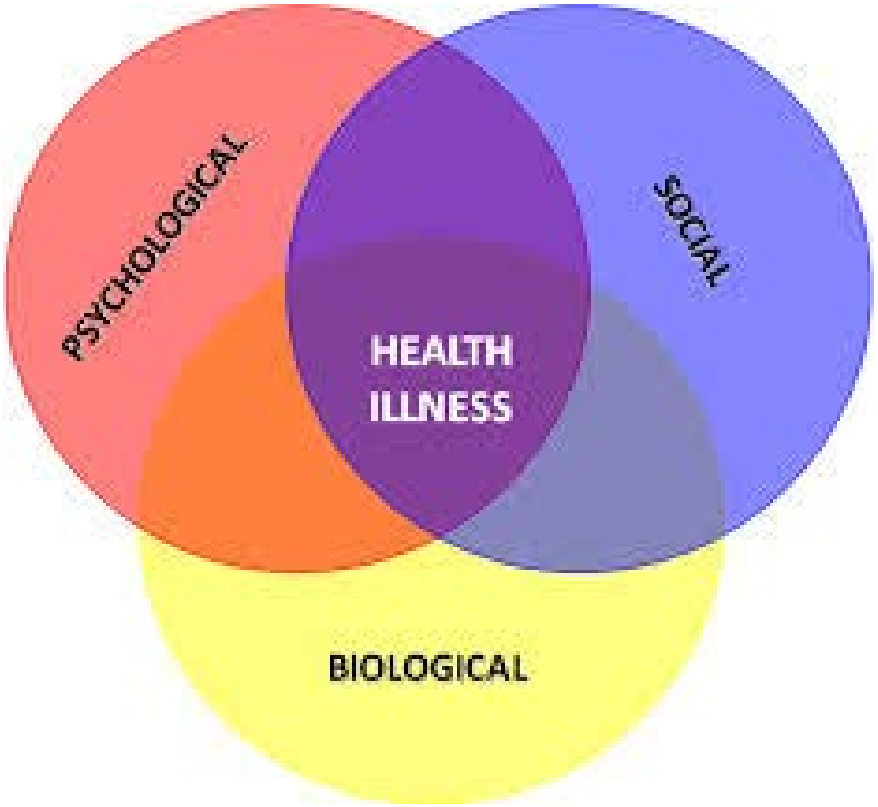
Hassell *Am J Prev Med* 2010; 38: S512-21

Age at Death for People with SCD in California, 2004-2008



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

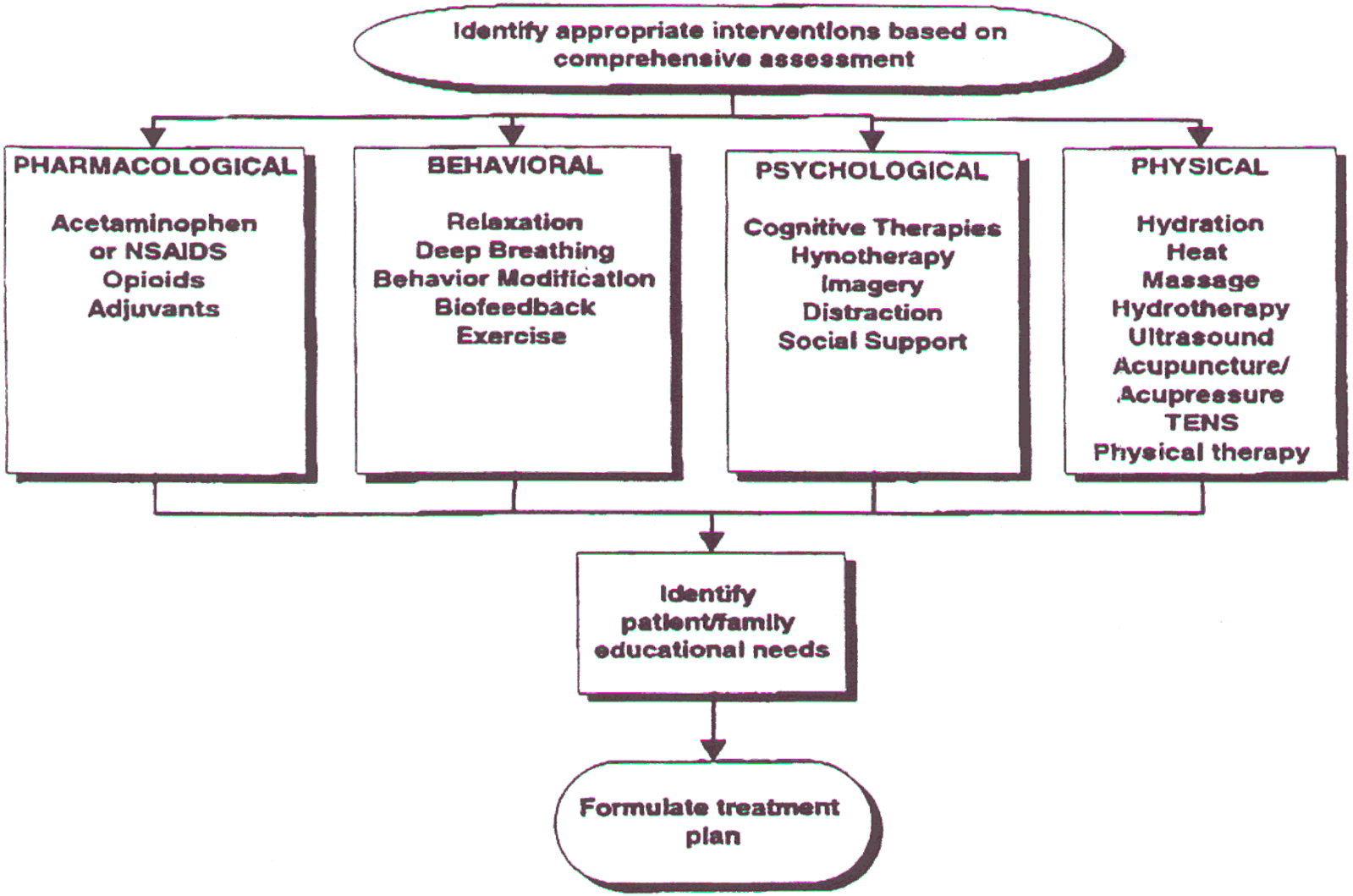
Treatments

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



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

Pediatrics 2011;128:182-200

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		
Variable	**SCD**	**CF**
US Prevalence	80,000	30,000
Federal support, millions	$90	$128
Total NIH and private support, millions	$90.4	$280.2
Total support per affected person	$1130	$9340

- 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

www.casicklecell.org

California Sickle Cell Resources

@SickleCellCalif

CHILDREN'S HOSPITAL & RESEARCH CENTER OAKLAND

NORTHERN CALIFORNIA NETWORK OF CARE FOR SICKLE CELL DISEASE

Sickle Cell Community Advisory Council of Northern California

S.C.C.A.C.

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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. Theopia 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@8 p.m., Wednesdays 4 a.m. and 11 a.m., Thursdays@8 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.

Mission Statement:

To provide information and assistance to families of sickle cell disease, to educate the public, and to assist health institutions and practitioners in the delivery of better care to sickle cell anemia patients.

Our Goals:

- Give people affected by sickle cell disease a say in deciding how their care will be given.
- Increase awareness of and support for those affected by sickle cell disease.
- Educate and increase access to available resources in the region and state for people with sickle cell disease.

Events:

August 2, 2014 11:00 am
Support group

October 11, 2014 11:00 am
Support Group

November 8, 2014 11:00 am
Support Group

www.sccac.net

CHILDREN'S HOSPITAL & RESEARCH CENTER OAKLAND

NORTHERN CALIFORNIA
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FOR SICKLE CELL DISEASE

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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

S.C.C.A.C.
Sickle Cell Community Advisory Council

Thank You!


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