Sickle Cell Disease: Public Health Agenda and Social, Economic and Healthcare Implications

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National Center on Developmental Disabilities
Centers for Disease Control and Prevention

The findings and conclusions in this report are those of the authors and do not necessarily represent the official position of the Centers for Disease Control and Prevention.
Outline

- Burden of Disease
- Mortality as an indicator
- Challenges
- Efforts to address the challenges
BURDEN OF DISEASE AND ECONOMIC IMPACT
US Prevalence and Incidence

• *In the United States, SCD affects approx. 80,000-100,000 people* \(^1,^2\)
  – Precise prevalence is unknown
• Approximately 1,800 to 2,000 infants are born with SCD each year in the US\(^1,^2\)
• Among newborn American infants, SCD occurs in approximately
  – 1 in 400 Blacks
  – 1 in 1,400 to 1 in 36,000 Hispanics
  – 1 in 80,000 Whites
• Over 3 million Americans have sickle cell trait \(^3\)
  – 1 in 12 (or 8%) African Americans

Major Complications of SCD
National Heart, Lung, and Blood Institute (NIH)

- Infections
- Anemia
- Painful Crisis
- Stroke
- Asthma
- Acute chest syndrome
- Renal failure
- Pulmonary Hypertension
- Others
Burden

• About 20-30% of children with Hb SS experience stroke, and cognitive impairment is common.\(^1\)

• Accounts for 23% of pediatric stroke cases among African-Americans.\(^2\)

• Due to frequent pain crises, children with SCD miss an average of 18 days of school each year. \(^1\)

\(^1\)Swanson et al. 2011
\(^2\)C.Baker and A.Grant – in preparation
Burden – Health System

• The number of hospitalizations among adults with sickle cell disease (SCD) in 2004 was 83,149.

• The total hospital costs for hospitalizations principally for SCD were approximately $488 million.
  – Adults aged 18-44 accounted for 66% of hospitalizations.
  – 3 emergency department visits a year, typically due to painful episodes.

• Among those hospital stays principally for SCD, 66 percent were paid by Medicaid and 13 percent were paid by Medicare.

• Emergency department visits associated with SCD average 200,000 per year, or roughly 2 per person per year.

• Frequent rehospitalizations.

1 Healthcare Cost and Utilization Project (HCUP), ARHQ – 2004
Cost

- The total healthcare cost associated with sickle cell disease estimated $2 billion annually.\(^1\)
  - Medicaid patients total
    - Health care costs: $1,400 per month ($1,700*)
    - Annual costs:
      - Children ages 0-9: $10,700 (12,900*)
      - At ages 30-39: $34,300 (41,000*)
  - Mean medical expenditures (2005) for children (<20 years).\(^2\)
    - $11,000 for those enrolled in Medicaid ($13,300*)
    - $14,800 for those with private insurance ($17,700*)

* In 2010 dollars

Prevention of Complications

- Newborns screening
- Penicillin Prophylaxis
- Pneumococcal vaccination and other vaccinations
- TCD screening
- Therapeutic transfusion
- Iron chelation
- Hydroxyurea
MORTALITY
**Mortality Rate of Black Children with SCD**

**Table 1—Deaths of Black Children with Sickle Cell Disease by Age Group and Year of Birth: United States**

<table>
<thead>
<tr>
<th>Age Group, y</th>
<th>Cohort Group</th>
<th>Years of Birth</th>
<th>Deaths per Year of Birth, Mean No.</th>
<th>Deaths per 1000 Persons with Sickle Cell Disease in Birth Cohort, Rate (95% CI)</th>
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<tbody>
<tr>
<td>1–4</td>
<td>Early</td>
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<td>55</td>
<td>37 (32, 43)</td>
</tr>
<tr>
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<td>Early</td>
<td>1963–1965</td>
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*Note.* Deaths occurred in 1968 through 1992. Deaths due to trauma, congenital anomalies, or perinatal conditions were excluded. Sickle β-thalassemia is not included in the definition of the **International Classification of Diseases** codes for sickle cell disease used in this study. CI = confidence interval.
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Mortality Rate of Black Children with SCD

## Survival of children with sickle cell disease

**Quinn et al Blood 2004;103:4023**

<table>
<thead>
<tr>
<th>Cohort</th>
<th>Dates of enrollment</th>
<th>Total</th>
<th>SS</th>
<th>Patient-years of observation</th>
<th>Age at entry</th>
<th>Uniform PCN prophylaxis</th>
<th>Ongoing accrual</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jamaica</td>
<td>1973–1981</td>
<td>563</td>
<td>315</td>
<td>NR NR</td>
<td>Birth (NBS)</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>CSSCD</td>
<td>1978–1988</td>
<td>694</td>
<td>427</td>
<td>2908 1781</td>
<td>&lt; 6 mo</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Dallas</td>
<td>1983–2002</td>
<td>711</td>
<td>431</td>
<td>5648 3571</td>
<td>Birth (NBS)</td>
<td>Yes</td>
<td>Yes</td>
</tr>
</tbody>
</table>

### Diagram

![Graph showing survival rates](image)
Trends in SCD Mortality rate, 1986-2005

Year per 100,000 blacks by age:
- 15-19 years
- 20-24 years
- 25-34 years
- 35-44 years
- 45-54 years
SCD-related mortality rate in adolescents versus early adults, 1985-1989 versus 2002-2005
Figure 1: Sickle Cell Disease: Distribution of Age at Death

Age at Death, SCD – 2006 US

Median age at death SCD (African-Americans, 2006) = 41

Median age at death (all African-Americans, 2006) = 73 males, 75 females
Life expectancy for Americans with SCD

National Sickle Cell Act

Sickle Cell Anemia
Current Challenges

• Two-three decades shorter life expectancy (Platt et al. 1994)
• Significant pain and other complications
• Transition Issues
• Education
• Employment
• Insurance
• Psycho-social concerns
Current Challenges

• Lack of data sources to monitor burden and impact
• Lack of access to quality care for adults
• Lack of uptake of effective treatments
• Lack of community awareness
• Lack of alternatives for treatments
Background: Mortality Rate of Black Children with SCD

Public Health Needs

- Specialty care especially for adults
- Coordination of health service other services
- Accurate estimates of health indicators
  - Prevalence, Mortality, Healthcare Utilization and Others
- Knowledge of risk factors and complications over the lifespan
- Community awareness
- Increased utilization of effective interventions like hydroxyurea
- Updated and uniform clinical care guidelines
- Healthcare quality indicators
Going Forward – Addressing the Challenges and Closing the Gap
Healthcare Types

- Tertiary Care
- Acute Care
- Preventive and Primary Care

Cost vs. Impact
Models for Care for Adults

Sickle Cell Day Hospital — Sickle Cell Treatment Services

A Leader in Sickle Cell Treatment

The Sickle Cell Day Hospital at Memorial Regional Hospital is a leader in sickle cell pain treatment for patients age 16 and older. One of only a few Sickle Cell Day Hospitals in the country, we offer a comprehensive approach to the management of acute and chronic sickle cell syndromes. Our dedicated staff of expert physicians, nurses and support personnel works closely with the patient's primary care physician or with a Sickle Cell Day Hospital primary care provider to deliver appropriate sickle cell treatment. In addition, the staff works in partnership with the patient and family members to manage and relieve pain as quickly and effectively as possible.

SCD, a group of inherited red blood cell disorders. The disease most commonly affects the African-American population, occurring in every one in 400 births.
Clinical Practice Guidelines

Sickle Cell Disease Guidelines
Overview

The National Heart, Lung, and Blood Institute (NHLBI) has launched an initiative to develop evidence-based clinical practice guidelines to manage Sickle Cell Disease (SCD) across the lifespan. We have appointed an expert panel to identify the clinical questions and to develop recommendations based on a systematic review of the relevant literature. The panel began work in February 2009 and expects to finish in Late Spring 2012.

Our goal is to develop guidelines that address key aspects of care in clinical settings, focus on primary care, and identify evidence-based best practices. The areas for these guidelines are health maintenance, acute and chronic care, hydroxyurea usage, and transfusion therapy.

http://www.nhlbi.nih.gov/guidelines/scd/overview.htm
Developing New Therapies

- More therapies
- Over 1 billion in R&D investment in 2012 alone
10 New Health People Objectives

Blood Disorders and Blood Safety

Hemoglobinopathies

BDDBS-1  (Developmental) increase the proportion of persons with hemoglobinopathies who receive recommended vaccinations

BDDBS-2  (Developmental) increase the proportion of persons with a diagnosis of hemoglobinopathies and their families who are referred for evaluation and treatment

BDDBS-3  (Developmental) increase the proportion of persons with hemoglobinopathies who receive care in a patient/family-centered medical home

BDDBS-4  (Developmental) increase the proportion of persons with a diagnosis of hemoglobinopathies who receive early and continuous screening for complications
42% of African-Americans
53% of Asian-Americans
Summary

- Progress has been made in improving survival and delivery of preventive care to children with SCD.
- Adults with SCD are a growing and diverse population facing many healthcare challenges.
- Collaborative widespread initiatives are needed.
- Innovative healthcare delivery models are being developed to address the needs of adults with SCD.
- More investment in developing and disseminating effective therapies is needed.
- More and better data needed to identify issues and monitor the effect of interventions at the population level.
Thank You

Twitter: @DrGrantCDC
Email: Agrant@cdc.gov
http://www.cdc.gov/ncbddd/sicklecell/

For more information please contact Centers for Disease Control and Prevention
1600 Clifton Road NE, Atlanta, GA 30333
Telephone, 1-800-CDC-INFO (232-4636)/TTY: 1-888-232-6348
E-mail: cdcinfo@cdc.gov  Web: www.cdc.gov

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EXTRA SLIDES
Number of States by Year Universal Screening for Sickle Cell Disease Started
## Prophylactic Penicillin Trial

<table>
<thead>
<tr>
<th>Group/Number</th>
<th>Penicillin</th>
<th>Placebo</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>105</td>
<td>110</td>
</tr>
<tr>
<td>Serious Infections</td>
<td>2</td>
<td>13</td>
</tr>
<tr>
<td>Infectious Deaths</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td><strong>p= 0.0025</strong></td>
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Number of States by Year Universal Screening for Sickle Cell Disease Started

NIH Recommends NBS for SCD 1987

NEWBORN SCREENING FOR SICKLE CELL DISEASE AND OTHER HEMOGLOBINOPATHIES
National Institutes of Health Consensus Development Conference Statement
Volume 6  Number 9  April 6-8, 1987
Number of States by Year Universal Screening for Sickle Cell Disease Started

In 2001: 1 state did not offer screening, 4 states offered screening by request
In 2002: 1 state did not offer screening, 3 states offered screening by request
In 2003: 2 states offered screening by request; 2004-5: 1 state offered screening by request
Effective Interventions

- Penicillin Prophylaxis
- Vaccination
- Parental Education
- Comprehensive Healthcare