Sickle Cell Disease in the US: An Overview of Public Health Burden and Opportunities to Improve Health Outcomes

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The findings and conclusions in this presentation are those of the author and do not necessarily represent the views of the Centers for Disease Control and Prevention.
What is Sickle Cell Disease (SCD)?

*National Heart, Lung, and Blood Institute (NIH)*

- Hemoglobin gene makes abnormal hemoglobin.
- *In a state of low oxygen, the abnormal hemoglobin deforms red blood cells and they stick together when it gives up its oxygen to the tissues. This causes blockage in the small vessels.*
Major Complications of SCD

- Anemia
- Splenic sequestration
- Infection
- Painful Crisis
- Stroke
- Acute chest syndrome
- Organ damage
- Pulmonary Hypertension
- Others
Common Variants of SCD

- **Homozygous sickle cell disease (HbSS)**, commonly known as sickle cell anemia
- **Sickle Cell Trait (HbAS)** – Not the disease. Usually can live normal life, but can pass on the S gene.
- **Hemoglobin SC disease (HbSC)**
- **Sickle beta (β) -thalassemias**: (HbSβ0) and (HbSβ+)
Who Does Sickle Cell Affect?

**Malaria**
- Areas with endemic falciparum malaria

**Sickle Cell Disease**
- Percent of population that has the sickle-cell allele (Hemoglobin S)
  - 14+
  - 12-14
  - 10-12
  - 8-10
  - 6-8
  - 4-6
  - 2-4
  - 0-2

**Ancestry**
- Sub-Saharan Africa
- India
- Saudi Arabia
- Mediterranean
Sickle Cell Disease in the United States

- 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 365 Blacks\(^4\)
  - 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

4. NNSIS- National Newborn Screening Information System
Sickle Cell Disease: History

- 1910 - Herrick – first description in western lit
- 1949 - Neel & Beet – inherited disorder
- 1951 - Pauling and Itano – molecular disease
- 1956 - Ingram & Hunt protein identified
- 1972 - National Sickle Cell Anemia Control Act
1972 National Sickle Cell Anemia Control Act

Goals

- Reduce the morbidity and mortality of SCD
- Increase awareness by educating the public
- Develop new modes of therapy.

The Act created funds for

- Mandatory carrier screening and counseling programs,
- Information and educational activities for health care professionals and the general public,
- Research training in diagnoses, treatment, and management of sickle cell disease.

Sickle Cell Disease in the United States

- Significant child mortality in 1970's and before
- Currently all 50 states and the DC conduct universal screening of sickle cell disease (1975-2006) (1987 NIH Recommendation)
- Over 90% of children with SS live past 18
SICKLE CELL DISEASE, HEALTH DISPARITIES AND HEALTH EQUITY
What is a Health Disparity/Inequality?

- "Health disparities are differences in the incidence, prevalence, mortality, and burden of diseases and other adverse health conditions that exist among specific population groups in the United States."

- National Institutes of Health

<table>
<thead>
<tr>
<th>Measure</th>
<th>Comparison</th>
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<tbody>
<tr>
<td>Prevalence</td>
<td>≠</td>
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<tr>
<td>Burden/Morbidity</td>
<td>≠</td>
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<tr>
<td>Mortality or Survival</td>
<td>≠</td>
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<tr>
<td>Access to Care</td>
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Opportunities to Improve Outcomes: Stroke

- About 20-30% of children with Hb SS experience stroke, and cognitive impairment is common.
- Accounts for 23% of pediatric stroke cases among African-Americans.
- Transcranial Doppler Screening (TCD) is an ultrasonography tool used to identify children with SCD who are at high risk for stroke.
- High risk can begin chronic red-cell transfusion which has been shown to reduce the risk of a first stroke by 90%.
- Selected studies suggest about 1/3 of children with SCD may not be receiving TCD screening.
Opportunities to Improve Outcomes: Hydroxyurea

- Only one FDA-approved drug for SCD – Hydroxyurea
- Decreases the morbidity and mortality
  - HU reduces total health care costs (is cost-saving)
    - Children who received HU treatment had nearly a third (30%) fewer hospital stays between ages 1 and 3 years.
    - Total costs were reduced by 21% for children who received HU treatment compared to those who did not receive this type of treatment.
    - Could reduce cost of SCD to Medicaid programs and other public health insurers
- Only 14-17% of SCD patients are prescribed HU and that among those prescribed HU only about 1/3 are adherent to therapy.
Growing need for carrier education

- Despite universal newborn screening, there is still a great unmet need for education and counseling of carriers
  - Over 3 million Americans have sickle cell trait but many may be unaware of their status or its implications
    - Born before universal NBS or in places with no screening and counseling programs
  - RuSH: NY we found that the majority (55%) of infants with sickle cell disease in that state were born to foreign-born mothers

- Parents of children with sickle cell trait less likely to receive genetic counseling than parents of cystic fibrosis carriers

Challenges with Healthcare Access and Quality

- Many adults do not have a medical home or primary care provider
- There are very few specialists (hematologists) for adults
- Frequent emergency department visits, hospitalizations, and rehospitalizations
  - ~200,000 per year: Emergency department visits associated with SCD
  - Adults account for 66% of hospitalizations each year
  - Adult healthcare costs are triple that of children
  - Adults account for the majority (60%) of people affected by SCD (RuSH)
- Among those hospital stays principally for SCD, 66 percent were paid by Medicaid and 13 percent were paid by Medicare
Frequent Emergency Department and Hospital Visits

California

Healthcare utilization by people with SCD, 2004-2008

- The number of hospital admissions and emergency room visits rises dramatically after age 18

Georgia

Average number of hospital encounters per individual with SCD, by age group, 2004-2008
Challenges with Healthcare Access and Quality

- Longer wait time in emergency rooms than other patients
- Disparities in pain management and stigmatization as “drug seeking”

# SCD: Inequity in Investments

<table>
<thead>
<tr>
<th>Variable</th>
<th>SCD</th>
<th>Cystic Fibrosis</th>
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<tbody>
<tr>
<td>US prevalence&lt;sup&gt;a&lt;/sup&gt;</td>
<td>80 000</td>
<td>30 000</td>
</tr>
<tr>
<td>Federal support</td>
<td></td>
<td></td>
</tr>
<tr>
<td>NIH fiscal-year 2004 funding, in millions of dollars&lt;sup&gt;b&lt;/sup&gt;</td>
<td>90</td>
<td>128</td>
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<tr>
<td>NIH funding per person with disease, $</td>
<td>1125</td>
<td>4267</td>
</tr>
<tr>
<td>No. of federal grants</td>
<td></td>
<td></td>
</tr>
<tr>
<td>No. of grants funded in 1968&lt;sup&gt;c&lt;/sup&gt;</td>
<td>22</td>
<td>65</td>
</tr>
<tr>
<td>No. of grants funded in 1972, after Sickle Cell Anemia Control Act&lt;sup&gt;d&lt;/sup&gt;</td>
<td>215</td>
<td>80</td>
</tr>
<tr>
<td>No. of grants funded in 2004</td>
<td>331</td>
<td>459</td>
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<tr>
<td>Private philanthropic support, $</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cystic Fibrosis Foundation 2003 annual revenue&lt;sup&gt;e&lt;/sup&gt;</td>
<td>152 231 000</td>
<td></td>
</tr>
<tr>
<td>Sickle Cell Disease Association of America 2003 annual revenue&lt;sup&gt;f&lt;/sup&gt;</td>
<td>498 577</td>
<td></td>
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<tr>
<td>Revenue per person affected with disease</td>
<td>6</td>
<td>5074</td>
</tr>
<tr>
<td>Total NIH and private support, in millions, $</td>
<td>90.4</td>
<td>280.2</td>
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<tr>
<td>Total support per person affected with disease, $</td>
<td>1130</td>
<td>9340</td>
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*Smith et.al. Pediatrics Volume 117, Number 5, May 2006*
Public Health Needs

• High quality and coordinated care especially for adults
• Updated and uniform clinical care guidelines
• Accurate estimates of health indicators
• Increased utilization of effective interventions like hydroxyurea
• Education and community awareness
Critical Role of Partnerships

- Federal Agencies
- State Health Departments
- Specialized Clinical Centers
- National and Local Community Based Organizations
  - *Sickle Cell Disease Association of America*
- Association of Public Health Laboratories (APHL)
- American Society of Hematology
- Private Sector and Industry
Closing The Gap

Life Expectancy

National Sickle Cell Act

All Americans

Sickle Cell Anemia

Years

0
10
20
30
40
50
60
70
80

Thank You

CDC

http://www.cdc.gov/ncbddd/sicklecell/index.html

Twitter:@DrGrantCDC

For more information please contact Centers for Disease Control and Prevention

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