Sickle Cell Disease
Access to Care

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Sickle Cell Disease: A True Story

- Patient is a 28 yo female with sickle cell disease, lives in Loris, SC
- She has Medicaid, on disability, no children
- She last saw a primary care doctor >1 year ago
- She is unsure (or if) she has seen a sickle cell specialist
- Goes to the emergency room 3x/week due extreme pain
- She is not receiving ANY disease modifying therapy
- She gets intermittent pain medication, no one monitoring prescriptions
- Transfused intermittently “whenever I get admitted”
- Medically abused
U.S. Sickle Cell Disease Population

• Calculation based on birth prevalence and census data, correcting for early mortality:

  • Total: ~100,000 individuals
    • 60% adults (at least)
    • 90% Black (African-American), 10% Hispanic
      • Areas with African immigrant populations

  • Genotype distribution:
    • At birth: HbSS 60%; HbSC 30%; HbSβthalassemia 10%
    • In adulthood: HbSS 50% age at 30, 25% at age 60

SCD and Mortality in the U.S.

- Childhood survival 96-98% for all genotypes
- In 2014, most deaths (66%) occur at ages 25-54 yrs
- More recent surveillance data from Georgia and California show mean age at death 43 yrs for women, 41 yrs for men

Geographic Distribution: Black/African American

Source: U.S. Census Bureau, 2010 Census Redistricting Data (Public Law 94-171) Summary File, Table P1.
Geographic Distribution: Hispanic Ethnicity

Source: U.S. Census Bureau. 2010 Census Summary File 1
Distribution of U.S. SCD Population

Adult Sickle Cell Providers by State
April 2012

5-Canada
1-India
1-Tobago (Caribbean)
1-Brazil
1-Puerto Rico

5-MA
8-MA
3-RI
5-CT
5-NJ
11-MD
1-DE
8-DC

www.scapn.net; www.hematology.org
Emergency Care for Sickle Cell Disease

• In 2013, 315,000+ ED visits were to
  • Metropolitan hospitals:  94%
  • Teaching hospitals: 65%
• Area of country
  • South    56%
  • Midwest  20%
  • Northeast 17%

• 61% resulted in discharge rather than admission
• 74% were in 18- to 44-year-olds

http://hcupnet.ahrq.gov/HCUPnet.jsp, Nationwide Emergency Department Sample, report run 21 May 2016
Hospital Care for Sickle Cell Disease

- Hospital inpatient care: 120,000+ discharges in 2013 were from
  - Larger facility >75-100 beds 88%
  - Teaching hospital 71%
  - Urban area 95%
  - Area of the country
    - South 51%
    - Northeast 21%
    - Midwest 19%
    - West 8%

- Mean length of stay 5 days at a cost of $8235, with 0.24% in-hospital mortality

http://hcupnet.ahrq.gov/HCUPnet.jsp
National Inpatient Sample Report, run 21 May 2016
SCD patients in South Carolina and Acute Care

<table>
<thead>
<tr>
<th>Age, y</th>
<th>Patients, No.</th>
<th>Encounters per Patient, No. (95% CI)</th>
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</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>ED</td>
</tr>
<tr>
<td>Total</td>
<td>2313</td>
<td>2.90 (2.63-3.17)</td>
</tr>
<tr>
<td>0-9</td>
<td>473</td>
<td>1.08 (0.97-1.18)</td>
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<tr>
<td>10-17</td>
<td>272</td>
<td>1.31 (1.12-1.50)</td>
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<tr>
<td>18-30</td>
<td>713</td>
<td>4.92 (4.24-5.60)</td>
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<tr>
<td>31-45</td>
<td>478</td>
<td>3.77 (3.05-4.84)</td>
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<tr>
<td>46-64</td>
<td>290</td>
<td>1.75 (1.33-2.17)</td>
</tr>
<tr>
<td>≥65</td>
<td>87</td>
<td>0.30 (0.19-0.41)</td>
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<tr>
<td>Region</td>
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<tr>
<td>Lowcountry</td>
<td>808</td>
<td>3.83 (3.23-4.44)</td>
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<tr>
<td>Midlands</td>
<td>613</td>
<td>2.44 (2.08-2.80)</td>
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<tr>
<td>Pee Dee</td>
<td>541</td>
<td>2.74 (2.20-3.28)</td>
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<tr>
<td>Upstate</td>
<td>351</td>
<td>1.80 (1.39-2.21)</td>
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<td>Expected payer</td>
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<tr>
<td>Medicaid</td>
<td>1057</td>
<td>2.84 (2.46-3.22)</td>
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<tr>
<td>Medicare</td>
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<td>4.57 (3.77-5.36)</td>
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<tr>
<td>Private</td>
<td>486</td>
<td>1.62 (1.30-1.94)</td>
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<tr>
<td>Self-pay/uninsured</td>
<td>211</td>
<td>1.74 (1.44-2.05)</td>
</tr>
</tbody>
</table>
READMISSION for SCD in South Carolina
What are the barriers for care in sickle cell disease?

- Large patient population in certain areas with minimal doctors
  - Rural communities
  - Poverty
- More pediatric doctors but more adults living with SCD
- Lack of Disease-Specific Education
  - Getting worse with less physicians getting necessary training by specialists
- Pain
- STIGMA
Partnerships to improve care for SCD

• **Health Resources and Services Administration**
  • Sickle Cell Disease Treatment Demonstration Program: Established Sickle Cell networks within regions

• **CDC**
  • Sickle Cell Longitudinal Data Collection (California)
    • Develop a system for long-term monitoring of health/care
  • Reducing Transfusion Complications
  • Best Practices for Hemoglobinopathy Screening

• **American Society of Hematology**
  • The ASH Sickle Cell Disease (SCD) Initiative addresses the state of SCD treatment in the US and globally by uniting people who care about SCD to improve care, early diagnosis, treatment, and research.
  • This is the first time that ASH is undertaking an initiative in support of a single disease.
Partnerships to improve care for SCD

- **NIH**
  - Continual increase in funding for sickle cell disease (estimated $78 million in FY2017)
  - Draft Strategic Plan Research Priorities, naming sickle cell explicitly:

- **Increasing clinical research**
  - Partnership with Industry
  - Orphan Drug Act
  - 187 open studies for sickle cell disease, 139 recruiting in U.S.
  - 28 with industry partnership and 32 by federal agencies

- **SCDAA**
  - "To advocate for and enhance our membership's ability to improve the quality of health, life and services for individuals, families and communities affected by sickle cell disease and related conditions, while promoting the search for a cure for all people in the world with sickle cell disease."
One Solution to Improve the Situation

- Sickle Cell South Carolina (SC)$^2$
- Hub and Spoke care delivery model
- The SC$^2$ program is designed to increase access to care for all persons with SCD in South Carolina
- SC$^2$ includes both specialty and primary care
- This approach will both harness the resources of the state to approach SCD and will also use a technologic based approach to increase education of providers.
(SC)^2

- Beaufort
- Georgetown
- Columbia
- MUSC
- Florence
- Upstate
Sickle Cell Disease in the U.S.: A Perspective

• A rare disease that still limits life expectancy through chronic complications despite childhood survival and availability of therapies

• At the **individual** level, it is about where you live:
  • NBS, healthcare and insurance is basically state-based and even more local than that
  • **Insurance (may be) more available but ≠ access to necessary care**
  • Best care often equilibrates to best advocates

• Shifting programmatic emphasis on building systems (hub-spoke), enhanced coordination of care
23 year old female, SCD, lives in Florence, SC
Overall, she is healthy, has rare pain, working in a clothing store
Hospitalized due to “severe pain” and was given limited pain medications and sedatives
Her mother called me at 10pm to beg for transfer to MUSC
She was transferred at 3am and in the ICU by 5am on a ventilator by 3pm, all organs failing
After two weeks-she is out of the hospital, doing well, looking for a new job