California Sickle Cell Disease Longitudinal Data Collection Project

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The findings and conclusions in this presentation have not been formally disseminated by the Centers for Disease Control and Prevention, and should not be construed to represent any agency determination or policy.
Public Health Surveillance

- The ongoing, systematic collection, analysis, interpretation, and dissemination of data regarding a health-related event
Reasons to do surveillance in sickle cell disease (SCD)

Sickle cell disease summit: From clinical and research disparity to action

Kathryn Hassell, Betty Pace, Winfred Wang, Roshni Kulkarni, Naomi Luban, Cage S. Johnson, James Eckman, Peter Lane, and William G. Woods

The American Society of Pediatric Hematology/Oncology Sickle Cell Summit brought together a broad range of constituencies to identify a unified approach to healthcare and research disparities for sickle cell disease. Recommendations included the following: (1) speak with a unified voice representing all constituencies; (2) optimize access to care; (3) integrate health care provider and patient responsibilities at home for all individuals with the disease; (4) develop overall approaches to basic, translational, clinical, and health services research; (5) enhance the community role in advocacy, education, service, and fundraising. Taskforces were identified to effect implementation. Am. J. Hematol. 84:39–45, 2009. © 2008 Wiley-Liss, Inc.
Reasons to do surveillance in SCD

NIH State-of-the-Science Conference Statement on Hydroxyurea Treatment for Sickle Cell Disease

5. What Are the Future Research Needs?

We support the use of hydroxyurea for the treatment of sickle cell disease but recognize that additional research is required to provide information that will ensure the most appropriate application of this therapy.

A surveillance system is needed for patients with sickle cell disease who will be followed prospectively. This system should contain demographic, laboratory, clinical, treatment, and outcome information.
Reasons to do surveillance in SCD

Evidence gaps in the management of sickle cell disease: A summary of needed research

To begin addressing the many unmet research needs, development of coordinated databases are necessary. This can be facilitated by widespread use of electronic health records and enhanced interoperability across sites, as envisioned in meaningful use requirements. Current newborn hemoglobinopathy screening programs represent one resource that can be better leveraged. Expanded data systems should enable the development of clinical and health care resource utilization phenotypes of individuals with SCD.
History of CDC surveillance in SCD

- **Registry and Surveillance System for Hemoglobinopathies (RuSH)**
  - Collaboration with NIH/NHLBI
  - 2 year project; 7 states
  - Goal: Identify all individuals in each state with an SCD or thalassemia diagnosis using pre-existing data sources

- **Public Health Research, Epidemiology, and Surveillance for Hemoglobinopathies (PHRESH)**
  - 2 year project; 2 RuSH states + 1 new state
  - Goals: Evaluate and validate RuSH methods; Implement activities in a new state with reduced funding amount; Disseminate information
Longitudinal Data Collection System for SCD

OUR MISSION

The CDC Foundation helps the Centers for Disease Control and Prevention do more, faster by forging effective partnerships between CDC and others to fight threats to health and safety.
California and SCD data collection

- Newborn screening (NBS) as public health surveillance
- Clinical research/cohorts
- Administrative data research
- CDPH joins CDC efforts
  - CDPH Genetic Disease Screening Program, Dr. Lisa Feuchtbaum
  - Inclusion of 90% of expected cases
Key Findings in CA (RuSH/PHRESH)

- Pediatric vs. adult clinical populations
- Death certificates unreliable
- Multiple data sources needed for complete surveillance
- High health care utilization is intermittent
- Adults over 40 have different needs
- 51% of adults live in Los Angeles County
CA Rare Disease Surveillance

- Part of the CA Environmental Health Tracking Program, a partnership between CDPH and the Public Health Institute
- Staff flexibility and capacity
- Key personnel:
  - Paul English, Co-PI
  - Faith Raider, Health Educator
  - Jhaqueline Valle, Data Analyst
  - Justin Howell, IT and Data Linkage Oversight
  - Eric Roberts, Biostatistics
  - Dan Meltzer, GIS/Mapping
Data Collection and Methodology

Phase I

2015-2016:

- State IRB approval (submitted)
- Approval to use data already acquired
- Requests for new state data
- Standardization of data
- Approach clinical sites about case reports
Data Linkage and Case Profiles

2016-2017:
- Refine data linkage algorithms
- Link and de-duplicate state data
- IRB approval from clinical sites
- Contracts and data use agreements from clinical sites
- Data collection from clinical sites
- Integration of clinical data into system

Phase II
Data Linkage and Case Profiles

2017-2020 and beyond:

- Continued integration of new state and clinical cases annually
- Analysis, dissemination, collaboration, education
Establishing Goals for Surveillance

Prioritizing use of the data

- Why collect data?
- To change care, outcomes and quality of life
- Collaborating with and supporting local and national efforts
- Bringing attention to healthcare disparities
- Answering questions
Engagement of Stakeholders

- June / July Meetings
- Information needed

- Needs of the community
- Feasibility
Engagement of Stakeholders

- American Society of Hematology
- Association of Public Health Laboratories
- Blood Centers of the Pacific/Blood Systems Research Institute
- California Health Care Foundation
- Center for Inherited Blood Disorders
- Children’s Hospital Los Angeles
- Children’s Hospital Orange County
- Health and Human Services/Office of Minority Health
- Health Resources and Services Administration/Maternal and Child Health Bureau
- Kaiser Permanente Southern California
- The KIS Foundation
- Loma Linda University Medical Center
- National Institutes of Health/National Heart, Lung, and Blood Institute
- National Medical Association
- Sickle Cell Community Advisory Council of Northern California
- Sickle Cell Disease Association of America
- Sickle Cell Disease Foundation of California
- UC Davis Hematology Clinic
- UC Irvine Medical Center
- UCSF Benioff Children’s Hospital Oakland
- Parents of children living with sickle cell disease
- Adults living with sickle cell disease
- Expert on hemophilia Universal Data Collection system
- Los Angeles City Council Sponsored Meeting on SCD
Geography of Patient Population

- Maps telling us where patients are located
  - How far away is quality care and other services?
  - Are patients in some areas denied access to care?
- Targeted outreach (partners)
- Environmental issues that impact health
- Reports or fact sheets to support decision making and policy change
Transition from Pediatric to Adult Care

- Where and when are patients seen?
- What happens to young adults in adult care?
- What conditions in childhood predict poor young adult outcomes?
  - High emergency department usage, mortality, complications
  - Peer reviewed publications
Hispanic/Latino (H/L) SCD Cases

- Proportion of H/L cases remains stable in NBS
- New immigrant cases (adult and children)
- Diagnosis challenges
- Are there health status/health outcome differences?
- Educational materials for providers, data to support outreach
The Aging SCD Population

- Who cares for older adults with SCD?
- What are the complications?

- Routine screenings/common illness of older adults
- Predictors of complications and/or death
- Peer reviewed publications, data to support outreach and policy change
High ED Utilization Patterns

- What does high ED utilization look like over time?
- What factors start and stop periods of high ED utilization?
- How does outpatient care and/or admission from the ED impact ED utilization?
- Educational materials for providers, data to support Medicaid policy, peer reviewed journal articles
Support of Local and National Programs

- Community-based organizations
- Policy makers
- Grant applicants
- Researchers
Other Areas (Limited Resources)

- Health care quality for people with SCD
- Costs of care
- Narcotic use/pain management
- Transfusions
- Quality of life/burden of disease
- Preventative treatments and screenings
- Rates of complications
- Fertility and pregnancy
- Sickle cell trait
## Comparison of SCD registries

<table>
<thead>
<tr>
<th>Registry</th>
<th>HHS Agency</th>
<th>Population</th>
<th>Data Source</th>
<th>Connection to other registries</th>
</tr>
</thead>
<tbody>
<tr>
<td>Get Connected</td>
<td>HRSA</td>
<td>Nation-wide; all ages</td>
<td>Patient-entered</td>
<td>Mechanism for disseminating findings</td>
</tr>
<tr>
<td>Sickle cell disease implementation consortium</td>
<td>NIH/NHLBI</td>
<td>Up to 7 geographic areas; 15-45 years old</td>
<td>Clinical data</td>
<td>High detail of data</td>
</tr>
<tr>
<td>Longitudinal data collection system</td>
<td>CDC/DBD</td>
<td>California; all ages</td>
<td>Pre-existing data (state-based, administrative)</td>
<td>Benchmark of all SCD patients</td>
</tr>
</tbody>
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Resources for More Information

- Quarterly webinars with guest speakers and project updates
- CDC Project Website:
  
  http://www.cdc.gov/ncbdd/hemoglobinopathies/scdc.html

- California Sickle Cell Website:
  
  www.casicklecell.org

- California Sickle Cell Resources Facebook Page
  
  https://www.facebook.com/SickleCellResources/
Stay in Touch

- Like us on Facebook: CA SCD Resources Facebook page

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