Sickle Cell Disease

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Sickle Cell Disease- What Is It?

- Sickle Cell Disease (SCD) or Sickle Cell Anemia (SCA) is a hereditary blood disorder, characterized by an abnormality in the hemoglobin.
- Valine substituted for glutamic acid in the 6th position of the β–globin chain.
- When deoxygenated, these abnormal (sickle) cells change shape and can get “stuck” in the blood vessels, causing damage and pain.

Complications of SCD

- Pain
- Infection
- Splenic Sequestration
- Acute Chest Syndrome
- Chronic Lung Disease
- Pulmonary Hypertension
- Congestive Heart Failure
- Leg Ulcers
- Stroke
- Gallstones
- Transient red cell aplasia
- Priapism
- Poor Growth
- Retinopathy/ Blindness
- Decreased brain function, mental delays, memory problems

Sickle Cell Disease- Treatments

- Routine screenings- We can prevent complications by monitoring patients closely.
- Penicillin, Folic Acid, Hydroxyurea and other drugs
- Research studies to improve care
- Excellent pain management
- Extensive family and patient education
- Symptomatic care for specific problems- need experts in order to do this well

Sickle Cell Disease

- Incidence of sickle gene among black Americans is about 8%
- About 2000 babies with SCD are identified yearly through the U.S. neonatal screening programs

California Statistics- RuSH Project Findings

- 6207 people identified with SCD in California between 2004-2008
- 43% younger than 18 years of age
- 21% 18-29 years
- 25% 30-50 years
- 11% 51 or older
- 1 of every 8000 live births in the CA general population
- 1 of 500 live births of Black or African Americans
- 1 out of 99,000 live births of Hispanic Americans

Sickle Cell Care Pediatrics vs. Adult Care

Pediatric Care (RCHSD)

- CDC lists 19 Centers for SC Care in CA
- Sickle Cell Team: 1MD, 1NP, 1RN, 1SW(~120 patients)
- Routine Appointments- available within 2 weeks.
- Urgent Appointments- available same day in our clinic with expert care for sickle cell patients

Adult Care (UCSD)

- CDC lists 3 Centers for SC Care in CA
- Sickle Cell Team: 1MD
- Routine Appointments- can be 3 months
- Urgent Appointments- nothing. Emergency Room is the only option for adult patients.

Sickle Cell Care Pediatrics vs. Adult Care

Pediatric Care (RCHSD)

- Comprehensive Sickle Cell Center for CCS
  - Yearly Comprehensive Exams and Visit
  - Other routine exams as needed based on severity of disease, treatments
- When hospitalized, children are cared for by physicians/ nurses who have expertise and education in Sickle Cell Care

Adult Care (UCSD)

- Seen in "benign hematology" when they make an appointment.
- When hospitalized, adults are cared for by general medical staff with little to no education about Sickle Cell Disease and appropriate care.

Sickle Cell Care Pediatrics vs. Adult Care

Pediatric Care (RCHSD)

- RN Case Manager- family can call with questions, for triage, advice, and/or education.
- Can refer for clinic visit, talk family through home treatment of pain crisis, or other appropriate treatment options.
- Can assist patient/family in navigating healthcare system.
- After hours- family can call on call hematologist with urgent questions.

Adult Care (UCSD)

- Patient can call hematology office, but may or may not get a call back.
- Patient can call the on call hematologist after hours, but that person is likely to have no advice for patient, more likely to send patient to ED.

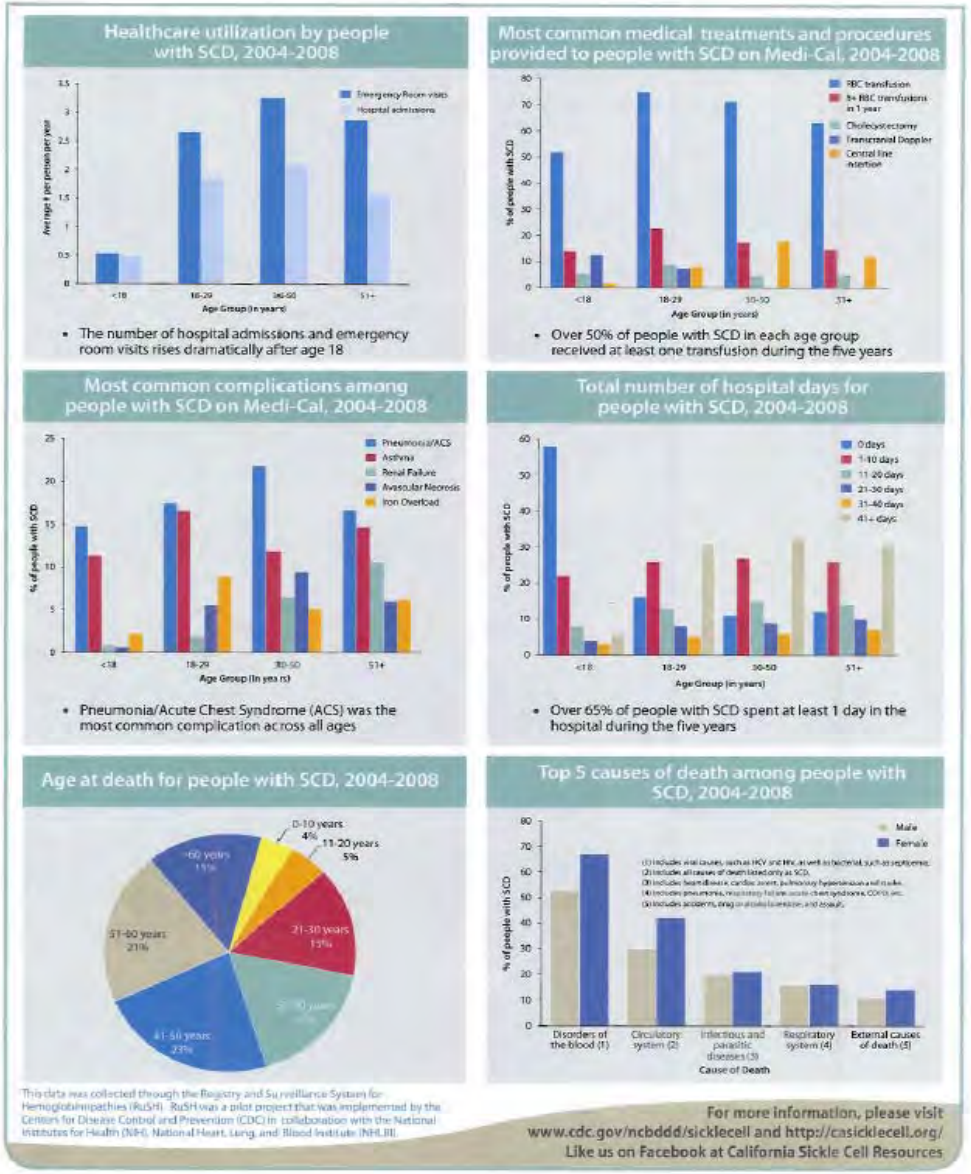
Sickle Cell Care Pediatrics vs. Adult Care

Pediatric Care (RCHSD)

- Emergency Room Care:
  - Patients come to RCHSD ED when possible, call the hematologist on call when on their way.
  - ED consults with hematology for assistance with patient care.
  - Patients are triaged with a high level for fever or pain crisis. (Triaged right under trauma patients)
  - Specific algorithm used for appropriate treatment
  - ED Staff are educated about Sickle Cell Pain and how to treat.

Adult Care (all hospitals)

- Emergency Room Care:
  - Adults go to any hospital as they don't usually have a "medical home".
  - Often must wait for long periods of time before being seen.
  - Often treated as "med seeking" or "drug addict"
  - Adults who do not look like they are "in pain" are often not taken seriously.
  - May be required to have a urine tox screen to prove that they are taking the narcotics (not selling them) prior to getting a prescription.



RuSH Project Findings:

"Hospital Admissions and Emergency Room Visits rise dramatically after age 18."

In Conclusion…

- The care for children (0-18/21) with Sickle Cell Disease has improved over the past years. We are now able to keep these children alive and well with the screenings, treatments, and support available.
- We need to look carefully at the lack of adult care for people with SCD… we are seeing increased morbidity, mortality, and higher costs associated with this population.