An Overview of Sickle Cell Disease and Current Services in the Commonwealth

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Presentation Objectives

- Describe sickle cell disease (SCD) and its complications
- Describe Virginia's resources and efforts to improve the quality of life for individuals and families affected by SCD

What is Sickle Cell Disease (SCD)?

- A collective term used to describe a group of inherited blood disorders that affect the shape and function of the red blood cell (RBC)
- RBCs form crescent or sickle shape and slow or block blood flow
- Constant breakdown of damaged RBCs
- Results in pain and damage to multiple organs

Complications from Sickle Cell Disease

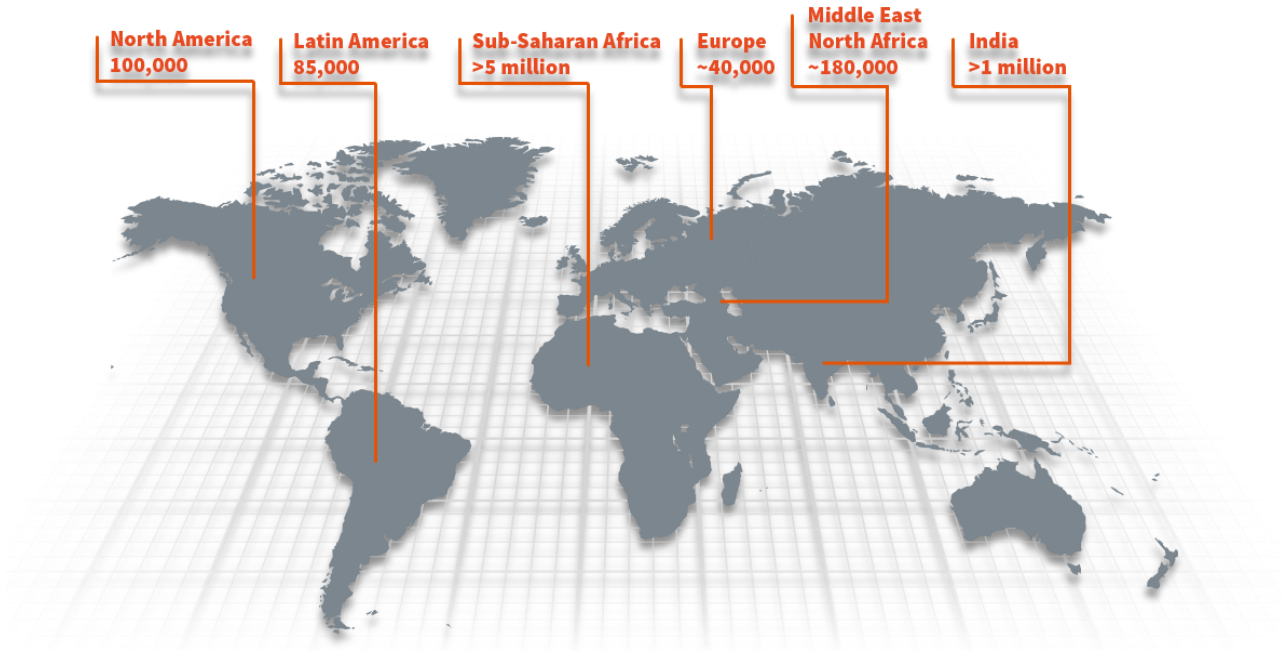
- Acute chest syndrome
- Anemia
- Stroke
- Splenetic sequestration
- Bacterial infections
- Leg ulcers
- Detached retina

Treatment/Medical Management

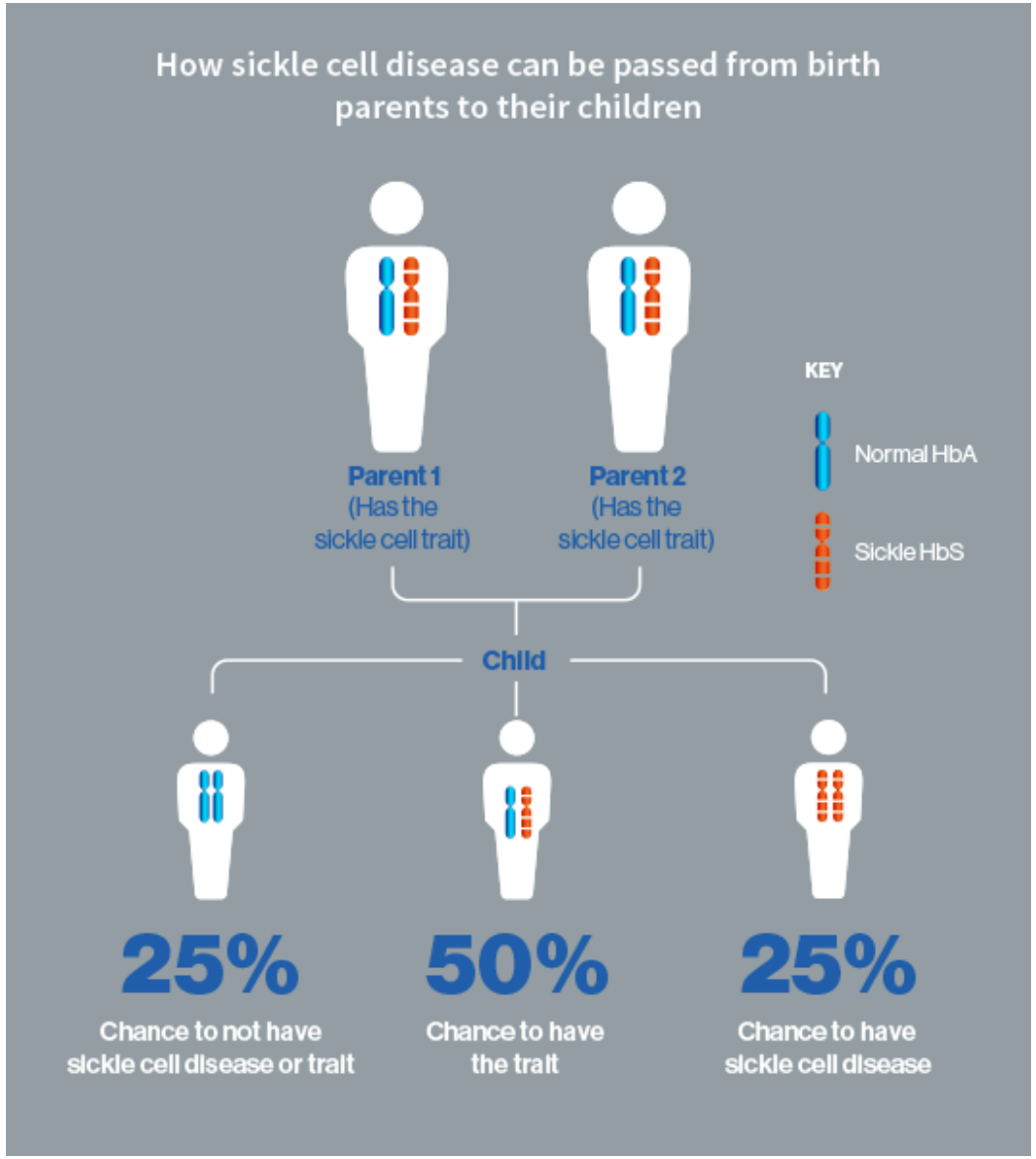
- Blood transfusions
- Antibiotics
- Comprehensive Care
- Medications that reduce symptoms
  - Hydroxyurea
  - Oxbryta
  - Endari
  - Adakveo
  - Pain medication
- Bone marrow transplant

Data and Statistics

- SCD affects approximately 6,400,000 people worldwide
- SCD affects approximately 100,000 Americans
- Globally, about 300,000 babies are born every year with SCD
- Approximately 300,000,000 people have sickle cell trait



SCD Inheritance Pattern



Sickle Cell Disease in Virginia

- ~75 newborns identified with disease yearly
- ~1000 individuals (0-21 yo) receiving care in the 5 pediatric comprehensive sickle cell clinics
- Current recipient of the Sickle Cell Data Collection Program Grant funded by CDC

Sickle Cell Services in the Commonwealth

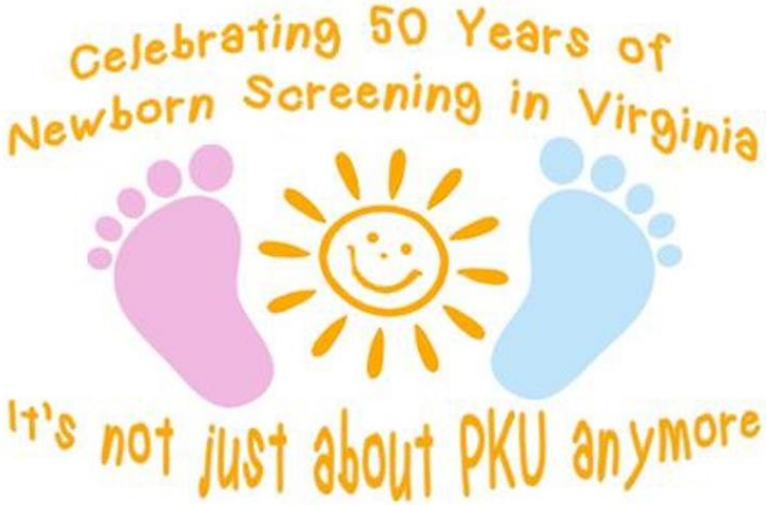
- The Virginia Sickle Cell Awareness Program
- The Virginia Newborn Screening Program
- Pediatric Comprehensive Sickle Cell Clinics
- Adult Comprehensive Sickle Cell Clinics

Virginia Sickle Cell Awareness Program (VASCAP)

- Title V funded statewide program for the education and voluntary screening of individuals for sickle cell disease, trait, and other related hemoglobinopathies.
- Screening Services
    - Family Planning
    - Maternity clinics
    - Newborn family studies (parent and siblings)
- Health education and promotion
- Contract management

Virginia Newborn Screening Program

- In July of 1989, Virginia began screening all newborns for SCD
- VASCAP collaborates with the Virginia Newborn Screening Program to provide:
  - Identification of newborns with SCD to ensure early entry to care



Pediatric and Adult Comprehensive Sickle Cell Clinic Network

- The goal is to enhance access to care.
- Clinics provide culturally competent comprehensive medical and support services that are:
  - Collaborative
  - Family-centered
  - And community-based

State Funded Pediatric Comprehensive Sickle Cell Center Catchment Areas:

State Funded Pediatric Sickle Cell Clinics

Northern Virginia

INOVA Fairfax - Falls Church

West

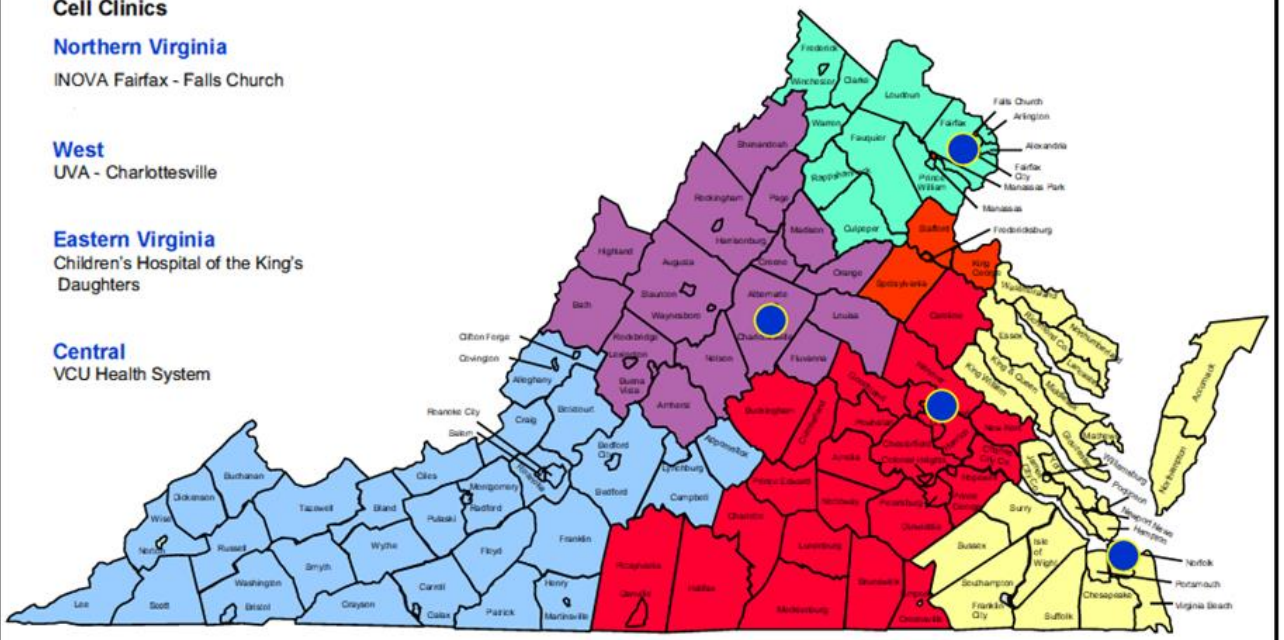
UVA - Charlottesville

Eastern Virginia

Children's Hospital of the King's Daughters

Central

VCU Health System



Challenges of Sickle Cell Disease

- Inconsistent care in emergency departments
- Healthcare providers lack familiarity with guidelines for treating the disease
- Stigma and medical bias associated with SCD
- Minimal or no health insurance coverage
- Poor access to appropriate health care
- Transition to the adult healthcare setting

Next Steps

- Apply for forecasted RFP for continuation of Sickle Cell Data Collection Program
- Finding a champion in the Hampton Roads area for adult sickle cell care

Thank you!

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