

# Communities Living with Sickle Cell Disease/Trait: Nursing Roles & Collaboration

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# Overview & Objectives

- 1) Pathophysiology and management needs of individuals with SCD/T over the lifespan
- 2) Examples of Nursing Roles & Collaboration with families, providers, & communities:
  - Educating Clinicians & Community
  - Community trait screening
  - Appraising aspects of stigma that may impair access to & quality of care/services
- 3) Lessons Learned & Future Needs
- 4) Resources & References

# Sickle Cell Disease

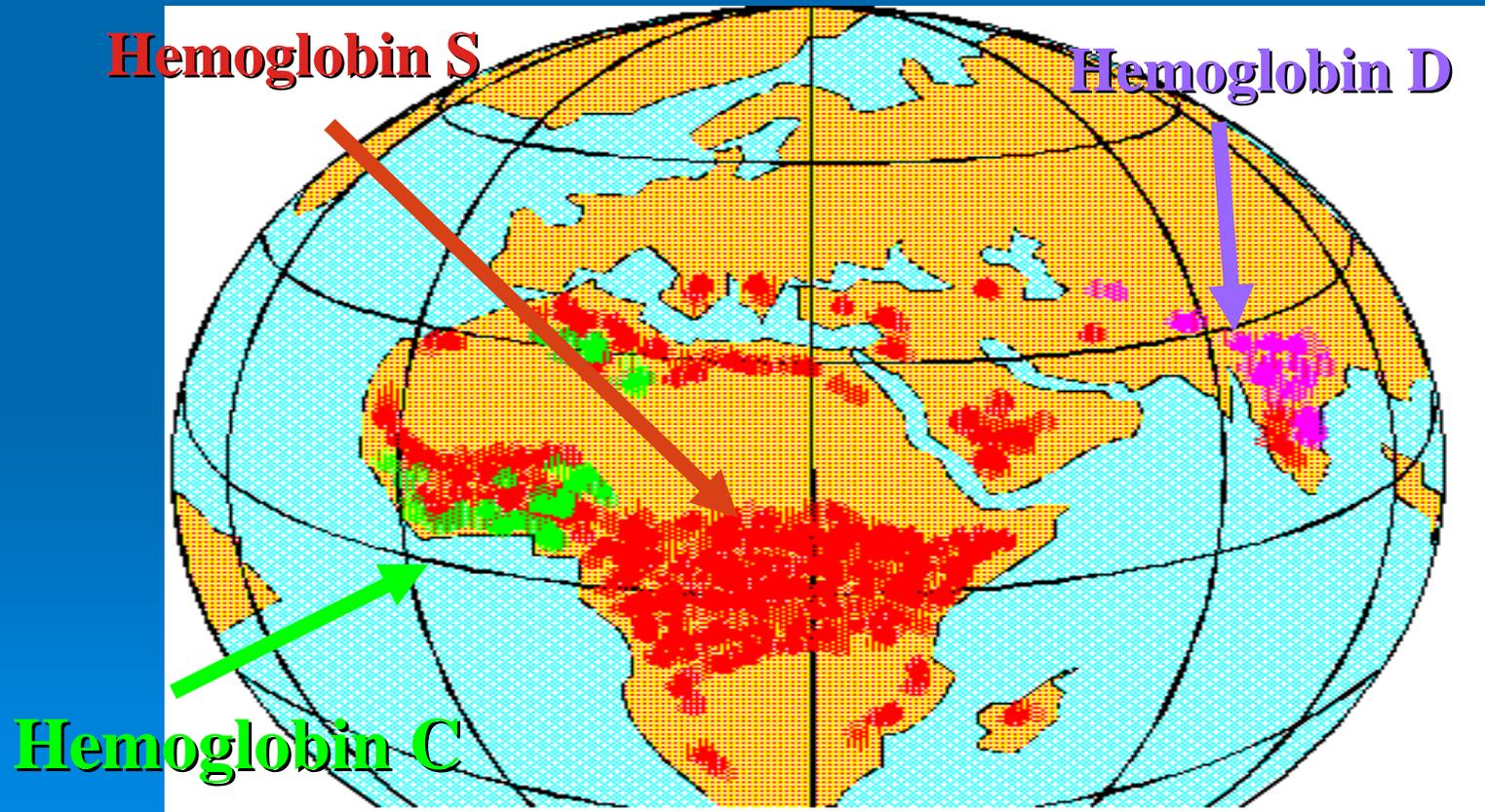
- Most common hereditary condition worldwide
- 2 altered hemoglobin genes
- Effects @ 1 in 475; 100,000 in the US
- Different genotypes (SS, SC, SD, etc) and phenotypes (mild, moderate, or severe symptoms & clinical outcomes effecting HRQOL)
- Chronic anemia, hemolysis
- Sickling & inflammation episodes lead to organ damage and disability

# SCD in Connecticut

- CT Department of Public Health
  - Newborn screening program
  - Stakeholders Group coordinated by Hospital for Special Care
  - Grant funded initiatives & Public Awareness
- ~ @ 400 Children & 300 Adults with SCD
- ~ 10,000 individuals with Sickle Cell Trait
- Pediatric Sickle Cell Treatment Centers
  - Connecticut Children's Medical Center
  - Yale New Haven Hospital

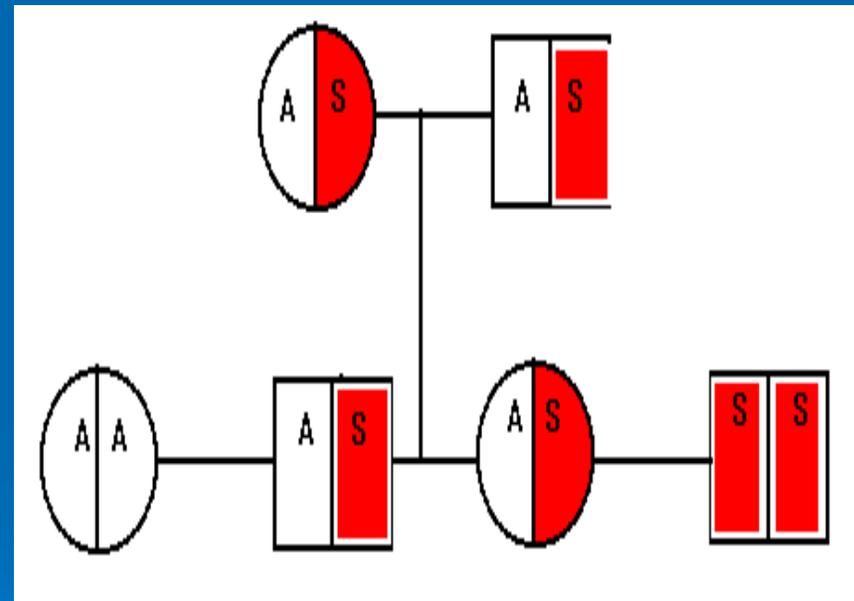
# Worldwide Distribution

- SCD is found in Africans, Turks, Greeks, Saudi Arabians, Egyptians, Iranians, Italians, Latin Americans and Asiatic Indians
- SCD is present in one out of four hundred African Americans in the United States. It is the **most common** genetic disease in this country



# Hemoglobinopathies a brief overview: Genetics

- Inherited ~ single gene
- Substitution of amino acid
- Pedigree

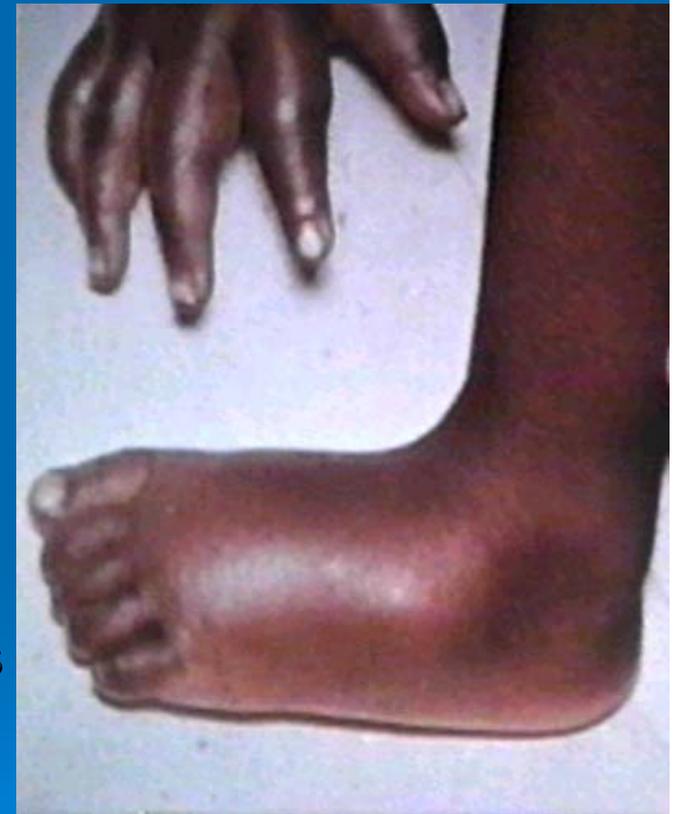


# Sickle Cell Disease

- An inherited, autosomal recessive, disease of red blood cells and endothelium; Single point mutation – valine for glutamic acid.
- Genetic screening ~ many hemoglobinopathy variants “SS” *is more sever*
- Affects the oxygen carrying protein, hemoglobin, in red blood cells (old term: sickle cell anemia). Sickle-shaped red cells interrupt blood flow by blocking small blood vessels (vaso-occlusion event)
- Tissue (at any organ site) that has no blood flow is damaged and causes pain. High risk for disability and organ disease over the life-span

# Disease Complications

- Sickle cells become trapped and destroyed in the spleen causing Splenic Sequestration
- Anemia - hemolysis
- Pain episodes
- Hand foot syndrome-Dactylitis, < 2yrs
- Gall Stones
- Strokes (silent) or aneurysms
- Chronic renal disease/failure
- Pneumonia or Acute Chest Syndrome
- Increased Infections
- Bone/joint infarctions/avascular necrosis
- Priapism, > 24hrs dysfunction
- Retinopathy, hearing loss
- Iron Overload from Transfusion Therapy



# Dependence ~ Tolerance ~ Addiction

- Pain Crisis (VOEs): per year / per individual
  - 90% have 0 – 3/yr
  - 5% have 3 – 12/yr
  - 5% > 13/yr (consume 50% resources)
- **Physical Dependence**
  - Anyone after 7 days continuous opiate use
- **Tolerance**
  - Anyone on continuous opiate- increased dosing is needed
- **Addiction:** Life revolves around drug = @ 5%
- **Pseudo-addiction cycle :** Under treatment → return → under treatment → return = **ED & Hospital recidivism!**

# Managing SCD

- National Newborn Screening - since 1990 in CT
- Prophylactic Penicillin, start age 2 months
- Vaccination Prevnar – pneumococcal
- Pain Management – acute & chronic; life-span
- Prevention of infections
- Chronic blood transfusions
- Antibiotics/Surgery
- Bone marrow transplantation
- Hydroxyurea
- Nitric Oxide
- Increased Fluid Intake
- Healthy lifestyle -> HRQOL!

# Sickle Cell Trait

- Sickle cell trait is present in 1/10 African Americans and % varies in others. About half of the hemoglobin in the red cell is sickle and the RBCs will exhibit sickling when under severe conditions of low oxygenation
- Resistance to Malaria
- Hematuria or blood in the urine is the most common problem
- Those with trait should be advised of risks of extreme physical activity, severe pressure changes, deep sea diving, dehydration, and possibilities of hematuria --> sever (rhabdomyolysis) & life threatening conditions

# Disparities in Health & Community Services:

Unequal treatment, access, or opportunities due to:  
Historical & Environmental Contexts

- Social Economic Status (SES-ism)
- Race
- Ethnic or language (cultural barriers)
- Gender (sexism) \* Age (ageism)
- Infectious/contagious/“contaminated”; “*it’s in the blood*” (L. Pauling)
- Chronic illness
- Mental health condition
- Disability

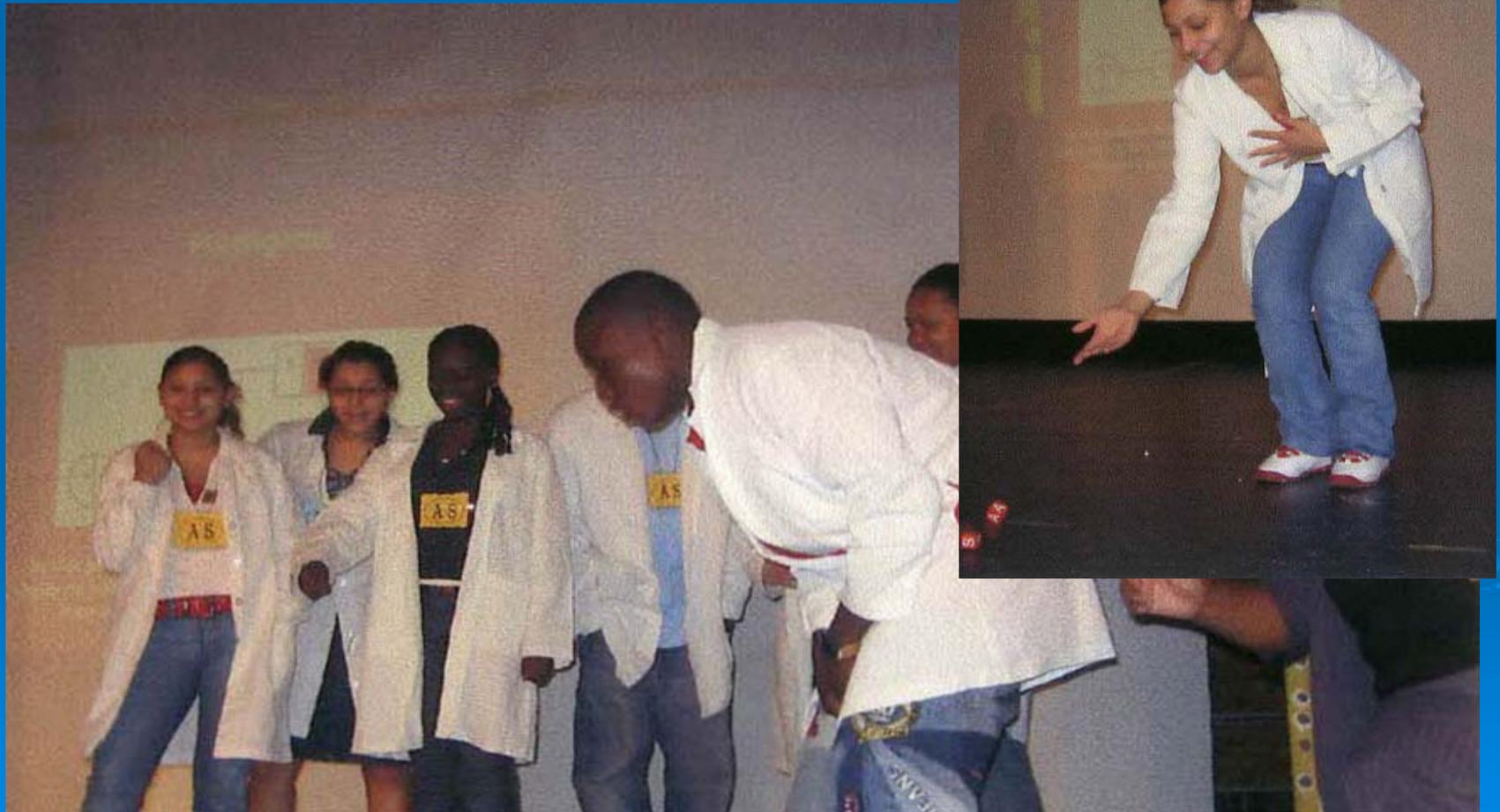
# Hemoglobinopathy Community Testing: Troubled History Current Practice

- Problems with testing included equipment sensitivity (false negatives)
- Discriminatory practices by employers, the military and insurers
- Misinformation
- Stigmatization of the condition; both Disease & Trait Status
- Impaired Access

# CT Hemoglobinopathy Educator & Counseling Course: (HRSA 05-027) (DPH 209-0911)

- Objectives:
  - Aligning with Newborn Screening Follow-Up
  - Expansion needed for community screening
  - CBO, Clinician and Health Educator Training
- Adapted Cincinnati Intermediate level
- Utilizes CT Clinical Faculty & CT DPH
- SC CBOs as admin. Support & faculty
- Outreach statewide through stakeholders
- National & International Participants
- 4 Levels of Certification

# “Making a Family”: High school students demonstrate genetic inheritance with SCD dice



# Hemoglobinopathy Curriculum:

- Overview of genetic patterns of inheritance
- Clinical management of hemoglobinopathies
- Affects on consumers & families across the life-span
- Identification of national guidelines, “best practice” standards of care, & research
- Cultural competencies, legal issues, & counseling skills.

## 4 Levels of Certification: Based on roles and educational preparation

- Hemoglobinopathy Peer Educator
- Hemoglobinopathy Community Educator
- Hemoglobinopathy Professional Educator
- Hemoglobinopathy Counselor



CITIZENS FOR QUALITY



SICKLE CELL CARE, INC.

**NOW WE HAVE A VOICE!**

***Mission Statement***

*To ensure available and accessible quality and comprehensive medical care and support services for children and adults in Connecticut with Sickle Cell Disease and related disorders.*

**[www.cqsc.org](http://www.cqsc.org)**

# Sickle Cell Trait: Why do community testing?

- Hemoglobin A ~> altered AS, AC, AE, etc.
- 1 in 10 African Americans + others worldwide
- Diagnosis by blood test: electrophoresis
- Under severe conditions may experience symptoms:
  - Blood in the urine
  - Enlarged spleen
  - Pain episodes or chronic
  - Sudden death?

...taking it to communities





# Stigma in Adults Living with Sickle Cell Disease and their Family/Caregivers Experience in the US & Nigeria (Leger, Odesina, Wagner & Knowlden; in process)

A pilot study was conducted to test for face validity and preliminary psychometrics of two self-reported scales for stigma (surveys for adults living with SCD & family members/caregivers).

Approved by UCHC, IRB # 06-210.

Component: Factor Analysis (N = 42)	1	2	3	4
1 Societal impact regarding the disease and isolation	.887	-.328	.146	.291
2 Personal feelings of shame, rejection, guilt, etc.	.249	.839	-.332	.352
3 Treatment when in pain & concerns for the future	.163	.433	.789	-.404
4 Sense of burden and needing assistance	-.354	-.032	.496	.792

# KEEP IN MIND!

- Genetics information is complex and may not be well understood by communities and families; results may raise questions about family histories
- Communities have unique attributes which can make providing awareness and health services both challenging & rewarding
- Community-Based Participatory Research is both a method & a guiding principle for practice

# Living with Sickle Cell Disease:

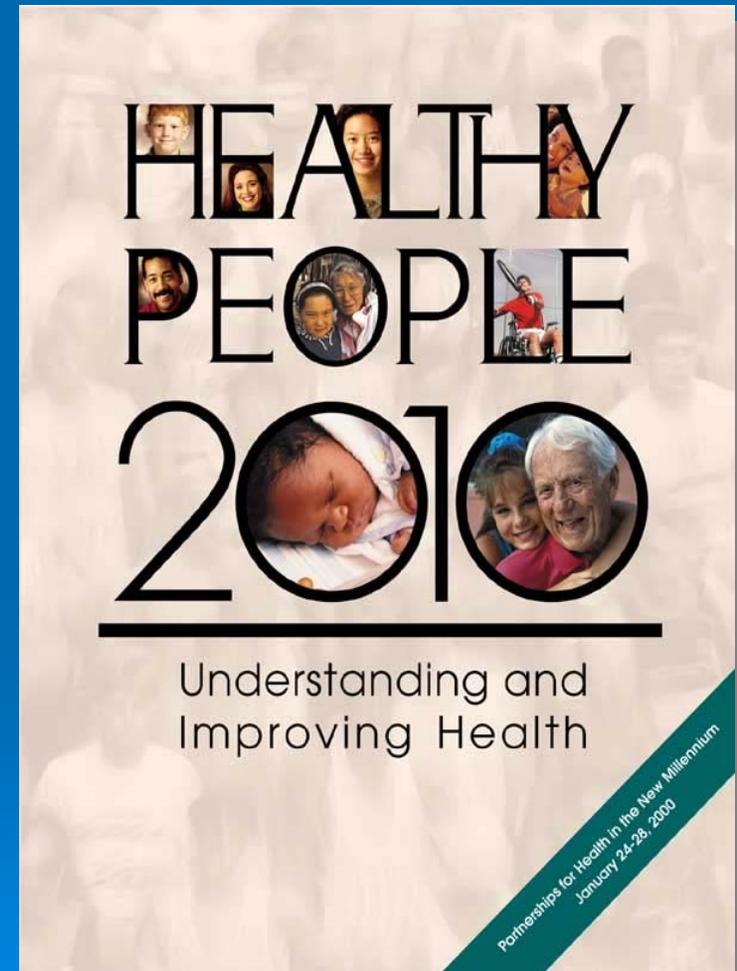
## “Old School” vs. New “Improved” Model

Hematology Specialty Clinic & Crisis Management via ED	Chronic Illness Model (Wagner): Collaborative and Continuous across environments
Pediatric Care	Life-span Approach
Regional Tertiary Medical Centers	Community Health Centers "Medical Homes"
Paternalistic Approach	CBPR with Community- Based Organizations (CBOs)
Prevention of VOE & Decrease Pain	Preventing organ damage & disabilities



# Major Areas for Intervention

- Improved Access to Care in Racial & Ethnic Disparities
- *Improved Quality of Care for Acute on Chronic Conditions*
- Continuity Across Environments of Care
- Pain Management (Acute & Chronic)
- Prevent Secondary Conditions!
- Functional Status (Ed., Voc., Psycho-social, Rehab)
- Schools & Athletics
- Health-Related **Quality of Life**
- Equity in Allocations of Resources and Research



# Resources

## ➤ 211 Info Line

- The Genetic Alliance [www.geneticalliance.org](http://www.geneticalliance.org)
- Citizens for Quality Sickle Cell Care (CQSCC)  
100 Arch St. New Britain, CT 06050 [www.cqsc.org](http://www.cqsc.org)  
860-223-7222 [citizensforquality@sbcglobal.net](mailto:citizensforquality@sbcglobal.net)
- Sickle Cell Disease Association of America of Southern Connecticut  
189 State St. Bridgeport, CT 06604  
203-366-8710 [scdaasouthernct@sbcglobal.net](mailto:scdaasouthernct@sbcglobal.net)
- Sickle Cell Disease Association of America, Inc.  
231 East Baltimore Street, STE 800  
Baltimore, MD 21202 [scdaa@sicklecelldisease.org](mailto:scdaa@sicklecelldisease.org)
- Grady Health System; Sickle Cell Information Center  
[www.SCInfo.org](http://www.SCInfo.org)

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