

Sickle Cell Disease: A family Perspective



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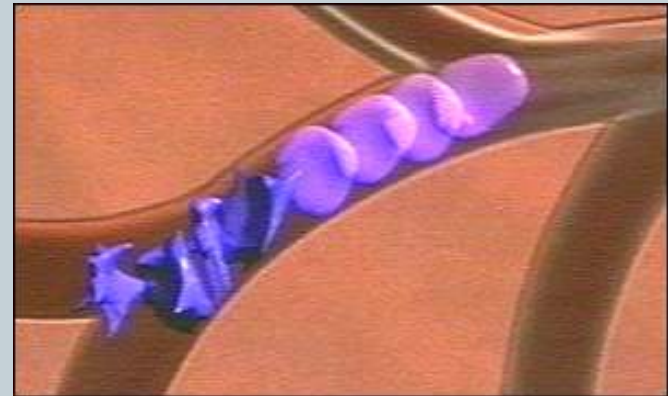
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Sickle Cell Disease



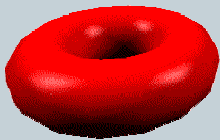
- Genetic disorder affecting the red blood cells (RBC)
- RBC changes to sickle shape-blocking blood vessels
- Results in lack of oxygen to tissues
 - tissue damage causes pain (mild, moderate, severe)
- Destruction of RBCs
- Chronic anemia/fatigue
- Organ damage
- Death



Normal vs. Sickle Hemoglobin

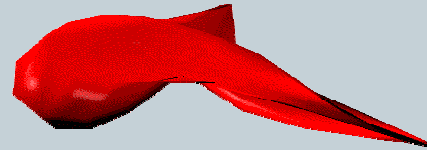
Normal

- disc-Shaped
- soft (like a bag of jelly)
- easily flow through small blood vessels
- lives for 120 days

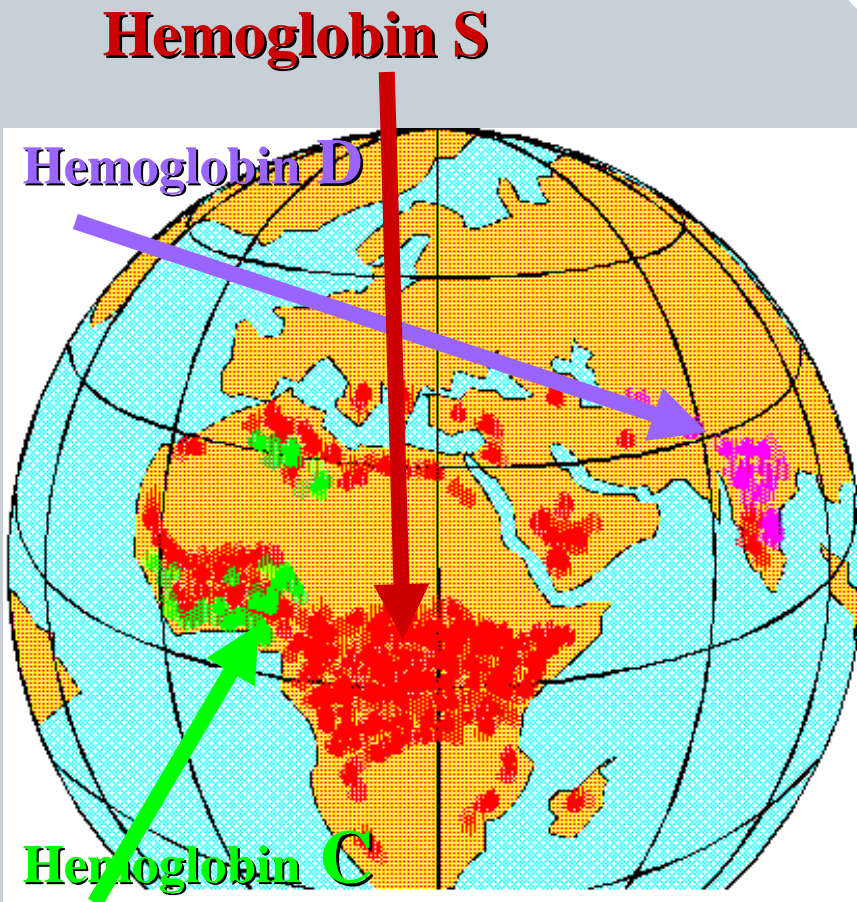


Sickle

- sickle-Shaped
- hard (like a piece of wood)
- often get stuck in small blood vessels
- lives for 20 days or less



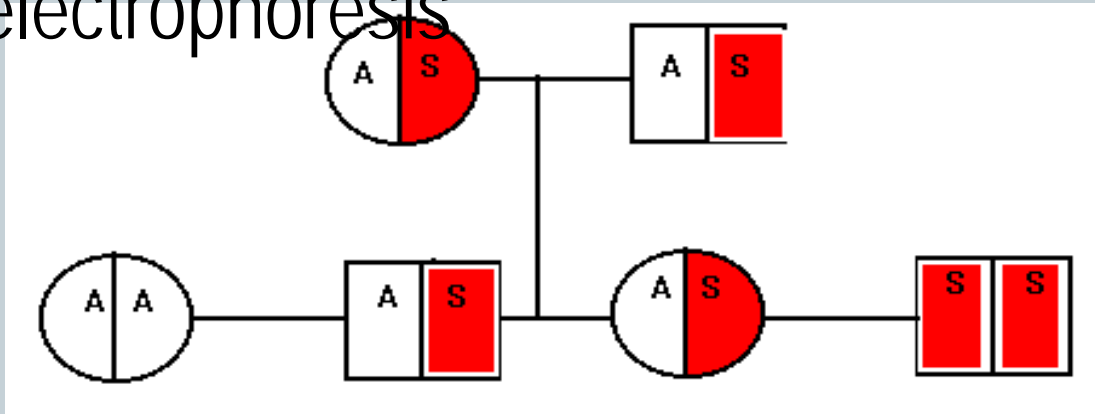
Worldwide Distribution



- Sickle Cell Disease (SCD) is found in Africans, Turks, Greeks, Saudi Arabians, Egyptians, Iranians, Italians, Latin Americans and Asiatic Indians.
- Present in 1:400 African Americans in the United States. 1:1100 Hispanics.
- It is the most common genetic disease in this country.

Pedigree and Testing

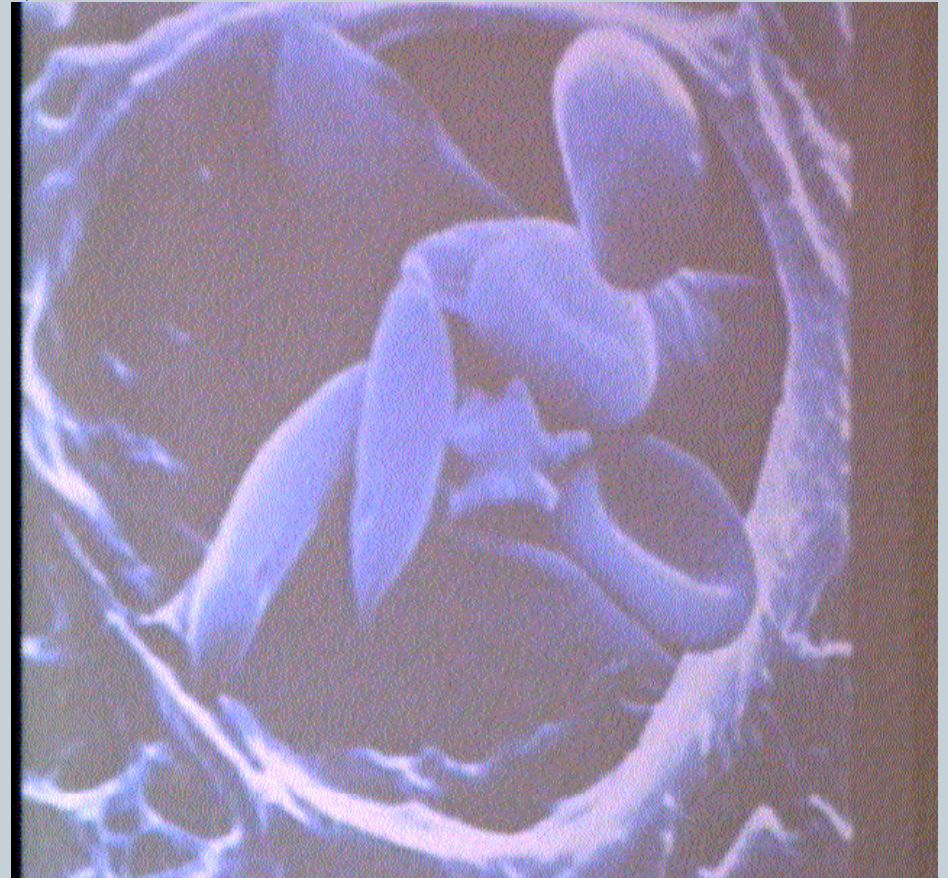
- Testing method by hemoglobin electrophoresis



Both parents have sickle trait, 1 in 4 children will have Sickle Cell Disease, one half will have trait and 1 in 4 will have normal hemoglobin (with each pregnancy)

Sickle cell trait

- Inheritance of one sickle hemoglobin gene and one normal hemoglobin gene
- Red blood cells sickles under severe low oxygenation
- Blood in the urine
- Risk of major health problems/death
 - extreme physical activity
 - severe pressure changes
 - deep sea diving



Complications

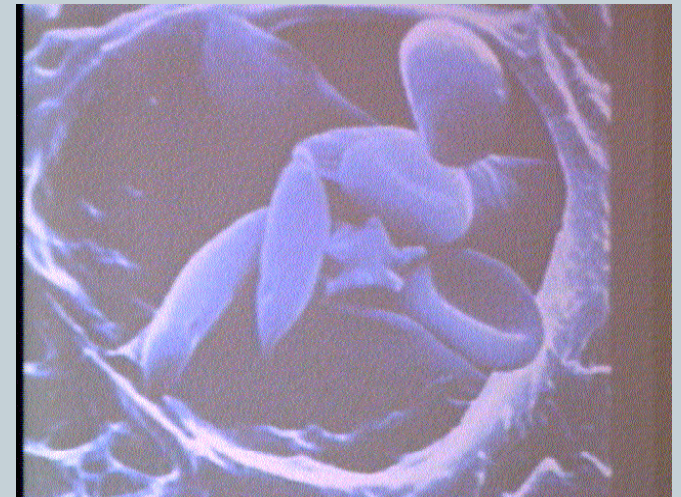
- Hand Foot Syndrome
 - Dactylitis
- Kidney problems
- Bone Pain
- Eye Problems
- Anemia - Jaundice
- Priapism
- Leg Ulcers



Management



- Preventive health maintenance
- Antibiotics/Fluids/Blood transfusion
- Pain medications (Codeine, Motrin, Morphine)
- Complimentary and
- Alternative medicine (CAM)
 - – herbs/vitamins, PT, heat
 - -yoga, faith, distraction



Management



- Family/community support/Respite
- Hydrea, ("no cure" - BMT, SCT)
- Specialists – Hematologists, surgeons, High risk OB
-ophthalmologist, dentists, Psychiatrists/psychologists

Flare Ups

- Extremes of temperature
- Stress and Infection
- Over exertion

Family Perspective



- “I attended pediatric/children's' hosp till age 21”
- “Nurses would mention my age and how old I was”
- “I wasn't sure where to go → wasn't aware of any adult SC care providers”
- Insurance – hard to get on your own b/c of “pre-existing condition” -why is that? It doesn't seem fair
- Difficulty with dental care-hard to get coverage

Family perspective



- “Family support important– full time job for families”
- “I get upset when they have to fight for what they need”
- “Medical providers need to work together – hematologists and primary care providers”
- “Lucky to have supportive employers about health condition, but always afraid of losing job”

Family Perspective



- “Our advocacy agency helped with insurance issues”
- “Patients and families need support to coordinate all aspects of care”
- “It would help to have some sort of program in place that guides people through transition”
- Orderly movement of a patient's medical home from one institution and set of providers to another, initiated at birth and ends at death.

Transition



- Occurs and exists between institution personnel - medical case manager
- Youth to assume health care responsibilities
- Care givers plan to relinquish roles
- Transition is about
 - WHO
 - TO WHERE
 - WHEN
 - HOW

Multidisciplinary Team

- PA/NP/MD as Consistent Providers
- RN/LPN as Care Managers, Teachers
- SW / CNS-Psych as Support Team
- Multimedia Vocational Rehabilitation
- Techs/Clerical - Patient Advocates
- Consultants - Eye, Nutrition, Audiology, Child Psychiatry, Physical Therapy
- School staff



Parent provider partnership



- Chronic care model for patient centered care
 - Medical case management/multidisciplinary team
- School –PPT for IEP and IHP
 - preschool evaluation and at regular intervals
 - tutorial support
 - health care needs (regular and urgent care)
- Family support- respite and guidance for resource referrals.
- Annual update, case review

Transition



- Needs assessment
 - request for individualized plan
 - consider severity of illness and capabilities
- Patients and care givers knowledge of disease, available social support and resources
- Providers knowledge of disease, process, needs, support and resources
- Anticipatory guidance/acquisition of life-skills
 - maximize potential (school, work, relationships)

Transition



- Disease prevention and general health maintenance
 - reduces complications, disability and rate of untimely death
 - Address non disease specific health issues (primary care provider)
- Start early and involve child/youth when appropriate
- Educate yourself and ask questions about health condition

Suggestions



- Find out about needs, available resources and contact information
- Clarify the transition schedule and responsible party
- Join a support group or seek out a Advocacy Organizations for support
 - attend meetings/social events/network to diffuse fears
- Confront loss of responsibility and control over medical care

Suggestions



- Address concerns about ill equipped provider
- Medical bag with all important documents
 - medical passports with medication, immunizations, treatment records and emergency contacts
- Advocate for, support and monitor transition programs
 - Positive outcomes (not guaranteed even with established program)
 - Seek out adult providers who focus on chronic illness
 - financial reimbursement deters profit and institutional commitment

Suggestions



- Seek assistance from Social service providers
 - care coordination
 - entitlements
- Schedule a case conference with care team (care plan)
- Visit adult team with pediatric staff and monitor outcomes
- Empower the young to self advocate
- Address "no shows" at the adult facility

Knowledge is Power!



**IF YOU DON'T KNOW, ASK
IF YOU ARE NOT SURE OR DO NOT
LIKE THE ANSWER, ASK SOMEONE
ELSE.**

