Sickle Cell Disease: A family Perspective

VICTORIA, LOLA AND SOLA ODESINA
CITIZENS FOR QUALITY SICKLE CELL CARE, INC
NEW BRITAIN, CT
WWW.CQSCC.ORG

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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
<table>
<thead>
<tr>
<th>Normal</th>
<th>Sickle</th>
</tr>
</thead>
<tbody>
<tr>
<td>disc-Shaped</td>
<td>sickle-Shaped</td>
</tr>
<tr>
<td>soft (like a bag of jelly)</td>
<td>hard (like a piece of wood)</td>
</tr>
<tr>
<td>easily flow through small blood vessels</td>
<td>often get stuck in small blood vessels</td>
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<tr>
<td>lives for 120 days</td>
<td>lives for 20 days or less</td>
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</tbody>
</table>
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.
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 -opthalmologist, 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.