The Impact of Sickle Cell Disease and Trait in Adolescence and Young Adulthood

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Faculty

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Objectives

- **Review some of the major morbidity-causing SCD complications and unique presentations and therapeutic responses in adolescents and young adults (Painful VOC, Chronic Pain, Rx of Chronic Pain, SCI)**
- **Review evolution of one major cause of SCD mortality, Renal Disease**

Objectives

- **Review aspects of SCD that affect the reproductive health of patients**
- **Discuss barriers and benefits to pediatric to adult medicine care transition**
- **Discuss medically significant features of sickle cell trait**

Who Should be Considered for HSCT?

- **Anyone offered hydroxyurea**
- **CVA patients**
- **Recurrent ACS patients**
- **Patients requiring chronic tranfusion for pain management**
- **Chronic pain patients who want to hear about this alternative**

Benefits of Hematopoietic Stem Cell Transplantation (HSCT)

- **Cure (~85-87% in large US and French studies with related, allogeneic HSCT)**
- **In chronically transfused patients with CVA no further transfusion**

Benefits of Hematopoietic Stem Cell Transplantation (HSCT)

- Exact cure rates with non-related HSCT not certain due to limited reported experience to date
- Non-myeloblative transplants are also now being tried but less experience that non-related to date re long term cure

Risks of Hematopoietic Stem Cell Transplantation (HSCT)

- Short Term
  - Infection
  - Bleeding
  - Graft versus host disease (GVHD)
  - Donor HSC rejection
  - ~ 8% mortality

Risks of Hematopoietic Stem Cell Transplantation (HSCT)

- Long Term
  - GVHD
  - Short stature
  - Sterility
  - Leukemia or late cancer

Reproductive Health

Priapism, Pregnancy, Azoospermia

Priapism

- Priapism is the spontaneous occurrence of unwanted, painful penile erection of prolonged duration
- Most frequent cause in man is sickle cell disease (SCD)
- 30-45% of SCD males disease will have an episode of priapism
- Adolescent SCD males must learn to distinguish normal from pathologic erection

Priapism

- SCD males are predisposed because they are chronically nitric oxide deficient
- Repeated episodes including so called “stuttering priapism” lead to:
  - Recurrent ischemic priapism,
  - Erectile dysfunction,
  - Penile fibrosis,
  - Chronic pain
  - Psychological and social dysfunction

Priapism

- Treatment paradoxically involves use of sildenafil (Viagra) and similar drugs orally, chronically

Pregnancy in Sickle Cell Diseases

- Morbidity and mortality (2.1% per Sargeant; 72.4 deaths / 100,000 vs nonsickle women 12.7 deaths / 100,000 with US current rate 8 / 100,000) of pregnancy is increased in SCD
- Later onset menarche
- Increased rate of spontaneous abortions, 36% (10% controls)

Pregnancy in Sickle Cell Diseases

- Increase in retained placenta for delivery > 24 weeks' gestation
- Fewer live born infants
- Greater proportion of prematurity
- Lower mean gestational age at delivery
- Lower birth weights (41.7% <2500 g)

Pregnancy in Sickle Cell Diseases

- Rate of stillbirths (7.1% in SCD, 0.7% normal)
- All findings affected by the number of sickle related events (eg: pain crisis, acute chest, urinary tract infection) during pregnancy

Non-Reproductive and Reproductive Side Effects of Hydroxyurea in Sickle Cell Diseases

- Non-Reproductive
  - Lowers platelet, white cell and red cell counts (dose dependent)
  - Eczema-like rash
  - Mild nausea
  - Minimal to mild hair loss

Non-Reproductive and Reproductive Side Effects of Hydroxyurea in Sickle Cell Diseases

- Skin and more commonly nail darkening
- Unlikely but unknown risk of caner or blood disorder

Non-Reproductive and Reproductive Side Effects of Hydroxyurea in Sickle Cell Diseases

- **Reproductive**
  - **Recommend birth control to avoid fetal malformation during pregnancy**
  - **Men have reversible azoospermia (decreased sperm count) with prolonged use**

What is Health Care Transition?

- **Health care transition is the process of changing from a pediatric to an adult model of health care**
- **The goal of transition is to optimize health and assist youth in reaching their full potential**
- **To achieve this goal requires an organized transition process to support youth in acquiring independent health care skills, preparing for an adult model of care, and transferring to new providers without disruption in care**

National Health Care Transition Center (www.gottransition.org)

Six Core Elements of Health Care Transition

1. **Office health care transition policy**
2. **Staff and provider knowledge and skills**
3. **Identification of transitioning youth**
4. **Transition preparation of transitioning youth**
5. **Transition planning with transitioning youth**
6. **Transfer of care (when appropriate)**

Medical Home Health Care Transition Index for Youth Up to Age 18 (National Health Care Transition Center, www.gottransition.org)

Detailed Steps in Transition of Adolescent Sickle Cell Patients to Adult Care (Responsible Party; Planned)

1. **P H/O Announces Intent to Transition at First Visit of Baby STEP© (2min DVD as Intro to First Session, June 2012)**
2. **P H/O Re-States Intent To Transition at First Visit for Next STEP©**
3. **At Age 11-12 P H/O notifies patient and family by Transition Letter and verbally with signed Transition Letter the intention to transition at age 18 and out of school**
4. **At Age 11-12, P H/O teaches Next STEP© (Ready May 2012)**

Detailed Steps in Transition of Adolescent Sickle Cell Patients to Adult Care (Responsible Party; Planned)

5. **At Age 14, or as soon as Next STEP© complete, P H/O staff initiates Personal Medical History Education and Transition Readiness Checklist and update both annually**
   a. Med Hx Synopsis prepped prior to clinic visit in Word in My Passport Format
   b. Save Word Doc as "First Name ,Last Name, DOB, Date " on Shared Folder
   c. At Clinic Visit (any site, any time) enter Med Hx in Passport, review with patient
   d. Give patient Med Hx Trifold, offer opportunity to email to personal account, phone or Home Computer
6. **At Age 15 patients seen without parent, parent joining subsequently**

Detailed Steps in Transition of Adolescent Sickle Cell Patients to Adult Care (Responsible Party; Planned)

7. **Age 14-18 years patient completes and continuously renews and documents completion of Adolescent STEP© Education Modules**
   a. Medical History and Medications; b. Preparing for Future Education; c. Preparing for the Work Force; d. *Psychological Assessment (* avail June 2012); e. *Risk Avoidance; *Sex; g. Psychosocial Planning/Support; h. Self Help Skills (Avail Later) i. Support networks
8. **At Age 16 P H/O presents Adult Care Provider List and initiate discussions on choosing Adult Care Providers and patient chooses/establishes Adult Provider (in prep)**

Detailed Steps in Transition of Adolescent Sickle Cell Patients to Adult Care (Responsible Party; Planned)

9. At Age 17-18 patient attends at least one Adolescent Transition Clinic to assure all steps up to this time are completed.
10. At Age 18yo Peds H/O discusses individual case with adult hematologist or sickle cell specialists who will receive the patient during Adult Transfer Conference Calls
11. At Age 18yo patient establishes appointment with Adult Care Provider, P H/O Staff Prepares Adult Transfer Packet and when out of school transfer patient to adult PCP and adult hematologists or Sickle Cell Specialists.
12. Patient transfers to Adult Care

Barriers to SCD Adolescent Transition to Adult Care

- Number of SCD knowlegeable Health Professionals
  - Hematology subspecialists
  - Primary Care Physicians (PCP)
  - Improved with FQHCs and CHCs
- Funding for Health Care
  - Medicaid loss at age 18

Barriers to SCD Adolescent Transition to Adult Care

  - High Medicaid (65%) Population
- Limited access to State of Art Care
  - ~50% population s in rural counties Policy/Health Economic Barriers
- Intrinsic Patient Barriers
  - Limited knowledge of health care systems and funding

Barriers to SCD Adolescent Transition to Adult Care

  - Limited self help skills (banking, driving, social support etc)
  - Inconsistent technological connectedness
- Policy Barriers like National Focus on Abuse of Prescription Narcotics and its impact on population of patients in chronic pain