Transition of Care

Sickle Cell Disease

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GOAL

- To enhance the Health Home Care Manager’s knowledge of issues that youth and young adults with sickle cell disease may encounter.
Objectives

1) Care managers will understand the common medical problems/co-morbidities associated with sickle cell disease that could affect health and wellness in youth and young adults with sickle cell disease

2) Care managers will understand potential challenges that this group may face when transitioning from self-care and independent living.

3) Care managers will understand how social determinants of health may affect them

4) Care managers will understand why transition planning is especially critical to decrease morbidity and mortality in youth and young adults with sickle cell disease
Hemoglobin

- Hemoglobin is a protein in the red blood cell
- It carries oxygen from the lungs to the rest of the body
- Normal Hemoglobin - Hemoglobin A (Adult) made up of Heme + Two Alpha chains and two Beta Chains
- Sickle cell - Instead of Adult hemoglobin the patient has sickle hemoglobin
Hemoglobin

- 6th position of the Beta chain, substitution of an amino acid (valine for glutamic acid)

- Causes polymerization of Hb within the red cell when it is exposed to low oxygen or acidosis
Sickle Hemoglobin

- Polymerization makes the cell “Sickle shaped”
- This sickle shape contributes to a decreased RBC lifespan (hemolysis) and occlusion of small vessels.
- This causes all the complications
Clinical Complications

- **Anemia** - because the red cells break down easily and have a reduced life span.
- **Vaso Occlusion** - Blockage of blood vessels and inflammation within the blood vessels causing complications
Electron microscope

Normal red cell  Sickle red cell
A Normal red blood cells

Normal red blood cell (RBC)

RBCs flow freely within blood vessel

B Abnormal, sickled, red blood cells (sickle cells)

Sickle cells blocking blood flow

Sticky sickle cells

Cross-section of RBC

Normal hemoglobin

Cross-section of sickle cell

Abnormal hemoglobin form strands that cause sickle shape
Genetic Inheritance

Here, both parents carry S trait.
Types of Sickle cell disease

- Hemoglobin SS
- Hemoglobin SC disease
- Hemoglobin S-BetaThalassemia
Epidemiology

- Sickle Cell Disease occurs in endemic malarial areas.
  - 1/3 of all indigenous inhabitants of Sub-Saharan Africa carry the gene.
- Over 70-100,000 people in the US have Sickle cell disease\(^1\).
  - 1 in 400 African American babies
  - Sickle Cell Trait (Hb AS in 7.5%)


Before the 1970’s

• 1 out of 3 children with sickle cell disease died before their fifth birthday

• 50% of individuals with sickle cell disease died before their 20th birthday

Death was because of

- Sepsis or Infection- sudden onset of fever, infection which spread rapidly

- Splenic Sequestration crises- Sudden enlargement of the spleen causing a sudden drop in blood pressure
Clinical- Acute Problems

• **PAIN** is the HALLMARK of this disease.
• Sudden onset, excruciating pain which can last for a few days and responds usually only to Narcotic pain medications
• Recurrent vaso-occlusive crises swollen hands/feet, musculo-skeletal or abdomen
PAIN

- Because of Vaso-occlusive crises- blood supply is reduced to parts of the body like the bones in the arms, spine and chest, other organs

- Unfortunately for the patient- there is no “objective” measure of pain- you have to believe and trust the patient
Clinical Features

- **Infection** - Sepsis, Meningitis
- **Acute Abdominal Crisis**
  - Abdominal pain and distention
- **Stroke**
  - Occurs in 11% of patients, Peak at 6-9yrs
  - Subclinical Stroke in 30% of patients
  - Patient may have no symptoms
Clinical Features

- **Splenic Sequestration Crisis**
  - Rapid accumulation of blood in the spleen
  - Seen in patients < 3 years of age
  - Abdominal distention, enlarged spleen and shock.
  - High mortality
  - Also seen in adults

- **Aplastic Crisis**
  - Temporary stoppage of red cell production often caused by parvovirus B19.
  - Have pallor, fatigue and rapid heart rate
Clinical Features

- **Acute Chest Syndrome (ACS)**
  - Classified by new pulmonary infiltrates associated with cough, breathlessness and chest pain.
  - May be severe, attributed to approximately 25% of deaths in patients with SCD.
  - Often the cause of death in young adults
  - Requires hospitalization and transfusions and Oxygen
Chronic Problems

- **Anemia**, Jaundice (yellow discoloration)
- Spleen: Enlarged, non-functioning
- Cardiac: Enlarged heart and murmurs
- Kidney: Cannot concentrate urine, bed-wetting, leaking protein in urine
- Gall bladder: Gallstones
- Delayed growth and sexual maturation
- Opioid Tolerance
Chronic Problems

- **Lungs**: Restrictive lung disease, Pulmonary hypertension
- **Joints**: Avascular necrosis of hips needing replacement, shoulders
- **Eyes**: Retina issues - laser treatment
- **Leg ulcers**
- **Priapism**
- **Iron overload**
- **Complicated pregnancies** Miscarriages, Intra uterine growth retardation, Crises
Treatment

• Pain Management
  • Patients need Narcotics for pain and other pain medications, Medications like Morphine, Percocet, Codeine, Dilaudid, Hydromorphone, Oxycodone, Ketorolac, Ibuprofen, Tylenol. Occasionally Fentanyl.
  • Often admitted and come to the ER for severe pain
  • Patients often know what works for them

• Transfusion Therapy
  • Therapeutic vs. Prophylactic (treatment of a complication and for prevention of a complication)
  • Simple vs. Partial Exchange

Health Homes Care Managers can assist in ensuring that the patient comes in for appointments
There are Improvements in care

- Diagnosis by Newborn screening
- Penicillin to prevent infections till age 5
- Folic acid for all
- Special Immunizations-less infections
- Identifying stroke risk- special testing- (Transcranial doppler) TCD
- Transfusions for Stroke prevention and treatment
Improvements

- Better Comprehensive care in pediatrics
- Education of the family
- Communication with school
- Children often are eligible for Insurance
- Parents work closely with the health care team
- Can work with Home Health Care Manager
Hydroxyurea - increases Fetal hemoglobin and decreases complications.
Cure for Sickle Cell

- **Bone Marrow/ Stem Cell Transplant**
- **From a matched sibling who does not have the disease**
- **Over 90% event free survival**\(^1,2\)
- **Available in 18% of patients**\(^3\)
- **Not all patients are candidates**
- **Has a 5-8% failure rate**

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Gene Therapy

1. Cells are removed from patient.
2. In the laboratory, a virus is altered so that it cannot reproduce.
3. A gene is inserted into the virus.
4. The altered virus is mixed with cells from the patient.
5. The cells from the patient become genetically altered.
6. The altered cells are injected into the patient.
7. The genetically altered cells produce the desired protein or hormone.
Current State of the Art

- Curative Therapy
  - PCN
  - Blood Tx
  - Pain Rx
  - Immunizations
  - TCD- Stroke prevention

- Symptomatic care

- Bone Marrow Transplantation
- Gene Therapy

- Hydroxyurea
Sickle cell- a chronic illness

• **Life expectancy** has increased from 15 years in the 70’s to 50 years today. Some live into their 60’s.

• Not enough Adult Centers to transition the patients into

• What happens when they go over to “Adult Care”

We believe that..

- The process of transition is very complicated and stressful for patients and families.
- If the family is connected to a Health Home even before this process, it would be significantly less stressful as there would be help with coordination of care.
Types of transfer from Pediatric care to Adult care

In a survey:

- **Age out**- policies regarding age
- **Drop out**- stop coming for care
- **Forced out**- lack of insurance
- **Move out**- leaves practice to seek care from another provider
- **Hang out**- continue to come for care to the pediatrician
- **Transfer**- patient is asked to go to an Adult provider
- **Transition**- the best way to do this
Scenario

- 18 ½ year old in the Adult ER complaining of pain. Patient last seen at age 17 ½.
- Nobody is sure if he is really in pain and how much pain medication he needs.
- He is unsure and is not able to advocate for himself.
- He waits to get evaluated and gets less medication so the pain has worsened ... meanwhile- he is sent home with minimal pain medications.
- Comes back to the ER the next day.
- The ER personnel ask him, ”Are you here for your narcotic fix?”
Traumatized patient

- Patient refuses to leave the pediatric setting
- He does not know about his Insurance
- No knowledge of the disease
- He has no clue as to what medications he received in the past
- There is no parent to advocate - he is considered an adult
Definition of Transition

- What is transition?- Transition is a process initiated with a child-and parent-focused care and ending with an adult-focused health care.
- Why do we need it?- To prepare the patient with a chronic illness for good life-long care
- What are the needs?- Education, Self advocacy, Preparedness, Availability of good care
- How do we provide this?

What is the role of the Health Home CM?
Important aspects of transition

- Health transition happens simultaneously with transition in other areas as well (-school, college, home)
- Transition proceeds at different rates for different individuals, families (and programs).
- A transition program for adolescents should include not only physical transfer of medical care from one facility and provider to another, but also address the needs of the developing adolescent

Role of the Health Home CM
What are the needs of an Adolescent

• Challenges of high school
• Plans for college or vocational education
• Knowledge of other risky behavior- HIV prevention, Drugs etc.
• Strides toward becoming independent.

Role of the Health Home CM
Transition issues in any chronic illness

- Patient not educated about disease
- Parents have been involved in care so the patient has never spoken up
- Patient not aware of own medical history
- Patient has other Adolescent issues

Transition issues- specific to Sickle Cell

• Most patients are on Medicaid\(^1\)

• Need to navigate Insurance issues\(^2\)


Transition issues- specific to Sickle Cell

- 30-35% of patients with Sickle cell disease can have Silent strokes- cognitive deficits – impact education and employment ¹,²

- Even patients without Silent strokes have school difficulties, need support in school, probably related to multiple hospitalizations and school absences³

3. VichinskyEP et al, Neuropsychological dysfunction and neuroimaging abnormalities in neurologically intact adults with sickle cell anemia. JAMA. 2010 May 12;303(18):1823-31
Sickle Cell Co-Morbidities

High incidence of co-morbidities

- Silent strokes
- Asthma
- Avascular Necrosis of the hips
- Chronic lung disease
- Cardiac issues
- Renal failure
Care of the patients

- In NY, most children with SCD attend a Comprehensive Sickle cell Center

- Most Adults end up in episodic care in the ER, occasional with an Adult Provider, or with a Primary Care Physician who may or may not be aware of the complications
Why are patients reluctant to transition to Adult Care

• Fear of leaving the provider that they know
• Fear that adult providers may not understand their specific problems
• Fear that they cannot express their needs
• Fear that they don’t know about their disease
• Fear of being treated as adults

Role of Health Home Care Manager - connect with the ones that cover Children and Adults
Re-hospitalizations and Death

- **Brousseau:**

<table>
<thead>
<tr>
<th>Re-Hospitalization</th>
<th>30 day</th>
<th>14 day</th>
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</thead>
<tbody>
<tr>
<td>SCD</td>
<td>33.4%</td>
<td>22.1%</td>
</tr>
<tr>
<td>SCD 18-30</td>
<td>41.1%</td>
<td>28.4%</td>
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</table>

- Re-hospitalizations were also highest for publicly insured patients.

- Quinn et al – in 940 participants followed for 8857 patient-years demonstrated the majority of deaths occurred after 18 years of age and after transfer to an adult provider (85%, 6 of 7).

- The **mean time to death** after transfer to adult was 1.8 years.

- **Role of the HH CM**
Stages of Transition

• Transition should be a gradual process
• Timeline of goals to fulfill
  • Individual assessment of Transition readiness
  • Assessment of needs
  • Provision of the tools necessary
  • Evaluate if the intervention was successful

3 Stage Program

- Transition should begin at age 12
- Early Stage- Ages 10-12
- Middle stage-Ages 13-15
- Late stage-Ages 16-18
Our Transition Activities

- Comprehensive Health Discussions
- Clinical Health Discussions
- Self Management Resources
- Adult Hematologist Introduction
- Other Engagement Activities
Comprehensive Health Discussions

At each stage of transition:

- Medical
- Self Advocacy
- Social issues
- Educational/Vocational
- Independent Living
- Referrals made
- Family Participation
Clinical Health Discussions

**MD and RN**
- Assess knowledge about sickle cell disease
- Discuss ways to avoid common complications
- Pain management techniques and plan
- Genetic counseling
- Give education

**Social Worker/Care Manager**
- Self-assessment by teen
- Parental assessment of the teen’s readiness
- Assess knowledge every time
- Create an action plan with goals
- Promote activities aimed at improving the identified weaknesses
Identify Self management/Create Resources

- Patient creates Medical History Booklet (with help)
- Reviews the Pain management plan with the Doctor/Nurse
- Records important information such as type of sickle cell, baseline hemoglobin, urine results, complications (acute chest syndromes, infections, painful crises, antibodies, strokes, gallstones, retinal and hip problems), surgeries, immunizations, blood phenotype, number of transfusions received, and special tests (Transcranial Doppler ultrasonography, echocardiogram, ophthalmological tests.)
- Reviews insurance information, including eligibility and the application process.
My Health History Passport

Dear Medical Provider/Emergency Room Provider,

My name is ______________________

I have Sickle Cell Disease Type- __________

This book will give you important information about my medical history and the type of pain management that works best for me.

Name of Hematologist: _________________

Hospital: ______________________________

Telephone: ______________________________

If I am incapacitated, please feel free to talk to __________________ about my treatment.

Contact Information:

Name: ________________________________

Date of Birth: __/__/____ Sex: __________

Address: ______________________________

City: ______________ State: __ Zip: ______

Telephone: Home:(__) __-____

Cell: (___) ____-____ Language: __________

Mother’s name: _________________________

Phone Number _________________________

Father’s Name: _________________________

Phone Number _________________________

Next of Kin: ___________________________

Phone number: _________________________

In case of Emergency Contact Name: __________________________

Phone Number: _________________________

Table:

<table>
<thead>
<tr>
<th>Name</th>
<th>Contact Info</th>
</tr>
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<tbody>
<tr>
<td>Primary Care Physician</td>
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<tr>
<td>Hematologist</td>
<td></td>
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<tr>
<td>Hematology nurse</td>
<td></td>
</tr>
<tr>
<td>Hospital</td>
<td></td>
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<tr>
<td>Pharmacy</td>
<td></td>
</tr>
</tbody>
</table>
Diagnosis:  SS  SC  S-β-Thal  SB+Thal  Other
Baseline Hb Electrophoresis:
Hb S __%,  Hb F __%,  Hb A2 __%,
Hb C __%,  Hb A __%
WBC: ___  Hb: ___  Hct: ___  Retic: ___
Labs Date: ________
WBC___  Hb____  Hct____  Retic:_____
Ferritin Level______ (Date:______)
Blood Pressure:___/___ mmHg
Pulse Oximetry___%
Spleen Size_____,  Splenectomy____
Blood type (phenotype):_____________
Antibodies:_______________________
Other Diagnosis:___________________
Comments:_______________________

Penicillin Dose___________________
Folic acid ______________________
Hydroxyurea:_____________________
I started on __________ because of
_______________________________
Current dose:____________________
Other medications:
_______________________________
Any Allergies:

Tonsillectomy:  Date________
Adenoidectomy:  Date________
Splenectomy:  Date________
Cholecystectomy:  Date________
Hip:  Date:______
Other Surgeries:
_______________________________
_______________________________
_______________________________
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<thead>
<tr>
<th>ORGAN INVOLVED</th>
<th>COMPLICATION</th>
<th>Yes/No Date</th>
<th>TEST</th>
<th>DATE</th>
<th>RESULT</th>
<th>DATE</th>
<th>RESULT</th>
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<tbody>
<tr>
<td>Brain</td>
<td>Stroke</td>
<td></td>
<td>CBC</td>
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<td></td>
<td></td>
<td></td>
<td>PULSE OX</td>
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<tr>
<td>Eyes</td>
<td>Sickle Cell Retinopathy</td>
<td></td>
<td>XRAY CHEST</td>
<td></td>
<td></td>
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<td></td>
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<td></td>
<td></td>
<td></td>
<td>EKG</td>
<td></td>
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<tr>
<td>Bones</td>
<td>Infection</td>
<td></td>
<td>ECHO</td>
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<tr>
<td></td>
<td>Aseptic Necrosis</td>
<td></td>
<td>PFT</td>
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<tr>
<td>Skin</td>
<td>Ulcers</td>
<td></td>
<td>TCD</td>
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<tr>
<td>Lung</td>
<td>Pneumonia</td>
<td></td>
<td>MRI BRAIN</td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td></td>
<td>Acute Chest Syndrome</td>
<td></td>
<td>MRA BRAIN</td>
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</table>
Other Engagement Activities

- Patients and their families participate in several activities, such as:
  - Summer camp
  - Monthly teen and parent support group
  - Back-To-School Fair
  - Holiday Party

Designed to promote wellness, enhance advocacy, and maintain a strong Team approach to patient centered care.
Introduction to the Adult Hematologist/Health Care team

• The Adult Hematologist/team comes to a non-emergency pediatric clinical visits

• Readiness to fully transition to the adult clinic should be determined during this discussion.

• If the team agrees, an appointment with the adult clinic is scheduled and the patient will sign an information release form.
Successful Transition

• Patients is successfully transitioned when they attend two appointments at the adult clinic.
• Follow-up meeting 3-6 months after the transition to discuss the success of the transition; identify barriers; and brainstorm solutions, as needed.
• We started this process in 2010
Sickle Cell Care

- Most rewarding
- You can make a tremendous difference in the lives of these patients
- Help navigating the system
- Create confident, independent adults with the skills to advocate for themselves
ASH

• American Society of Hematology
• Sickle Cell Disease Transition Readiness Assessment Template
Important Questions

• How important is it to you to manage your own health care? Likert scale 0-10
• How confident do you feel about your ability to manage your own health care?
• How confident do you feel about preparing for/changing to an adult doctor before the age of 22?
<table>
<thead>
<tr>
<th>Disease Knowledge</th>
</tr>
</thead>
<tbody>
<tr>
<td>I know what type of sickle cell disease I have.</td>
</tr>
<tr>
<td>I know my medical needs and can explain them to someone.</td>
</tr>
<tr>
<td>I know what a hematologist is and why I go to one.</td>
</tr>
<tr>
<td>I know what to do in case of a medical emergency.</td>
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<tr>
<td>I understand what causes a pain episode.</td>
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<tr>
<td>I understand how drugs, alcohol and tobacco affect sickle cell disease.</td>
</tr>
<tr>
<td>I have friends that I can talk to about sickle cell disease.</td>
</tr>
<tr>
<td>I know about necessary screening exams (echo annually, kidney function annually, retinal exams, etc.).</td>
</tr>
<tr>
<td>I know how to get blood work and x-rays.</td>
</tr>
</tbody>
</table>
# Medication Management

- I know what my medications are for.
- I know the names and doses of my medications.
- I remember to take my medications without my parent reminding me.
- I fill prescriptions before I run out of medications.
- I am aware of what hydroxyurea is and how it prevents sickling of my red blood cells.
- I know how to prevent a pain episode and what to do if I have pain.
### Appointments

- I make my own doctors’ appointments.
- I know how to get medical care when the doctor’s office is closed.
- I fill out my own medical history form.
- I keep track of my own medical information.
- I keep track of my doctors’ and other appointments.
- I make a list of questions before my visit with my doctors.
- I answer questions on my own during medical visits.
- I arrange my own transportation to medical appointments.

### Insurance

- I carry my own insurance card.
- I understand my insurance plan.

### Privacy Information

- I understand how health care privacy changes at age 18, when I am legally an adult.
Sickle Cell Resources

The Sickle Cell Information Center website,
www.scinfo.org

Sickle Cell Education
Sickle Cell and Thalassemia Patients Network (SCTPN)
1139 St. Johns Place
Brooklyn, New York 11213
347-533-8485 or 8486
http://www.sctpn.org/index.html
Email: info@sctpn.org

American Sickle Cell Anemia Association
http://www.ascaa.org/education-material-videos.php

CDC
http://www.cdc.gov/ncbddd/sicklecell/index.html

National Heart, Blood and Lung Institute
http://www.nhlbi.nih.gov/health/topics/topics/sca/

Sickle Cell Slime O Rama
http://sicklecell.starlight.org/

Sickle Cell Kids
http://www.sicklecellkids.org/

Got Transition? National Health Care Transition Center
http://www.gottransition.org

Health Care Transitions: The Institute for Child Health Policy at the University of Florida
http://hctransitions.ichp.ufl.edu/hct-promo

Healthy and Ready to Work National Resource Center
www.hrtw.org

SCINFOR.ORG
Kids As Self Advocated (KASA)
http://www.fvkasa.org

Quick Links to Health Care Transition Resources
Connecticut Department of Public Health Youth with Special Health Care Needs
Sickle Cell Transition

Thank you
Questions?