Transition of Care

Sickle Cell Disease

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GOAL

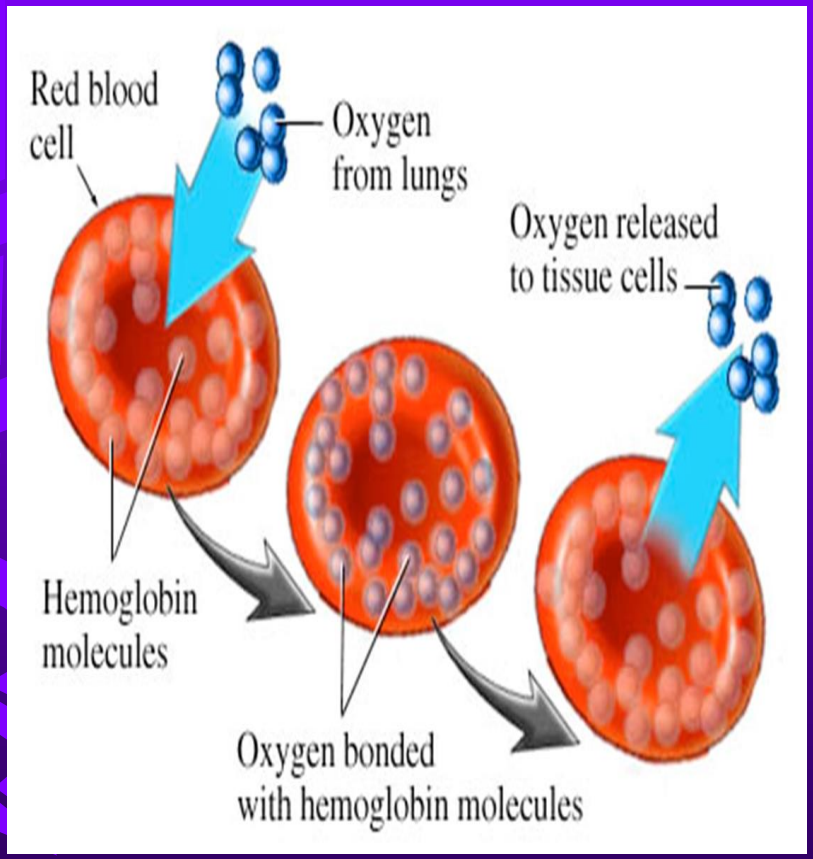
- To enhance the Health Home Care Manager's knowledge of issues that youth and young adults with sickle cell disease may encounter.

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

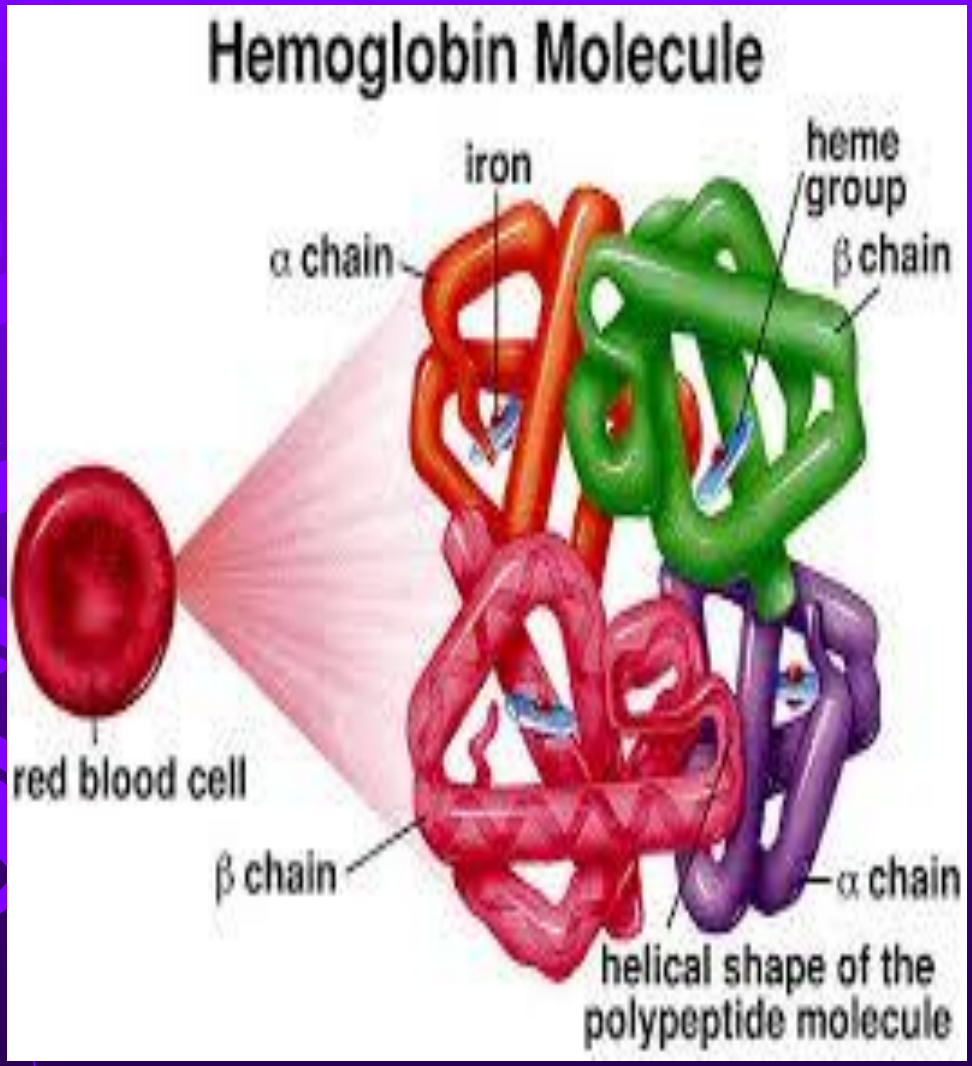
Hemoglobin

- 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



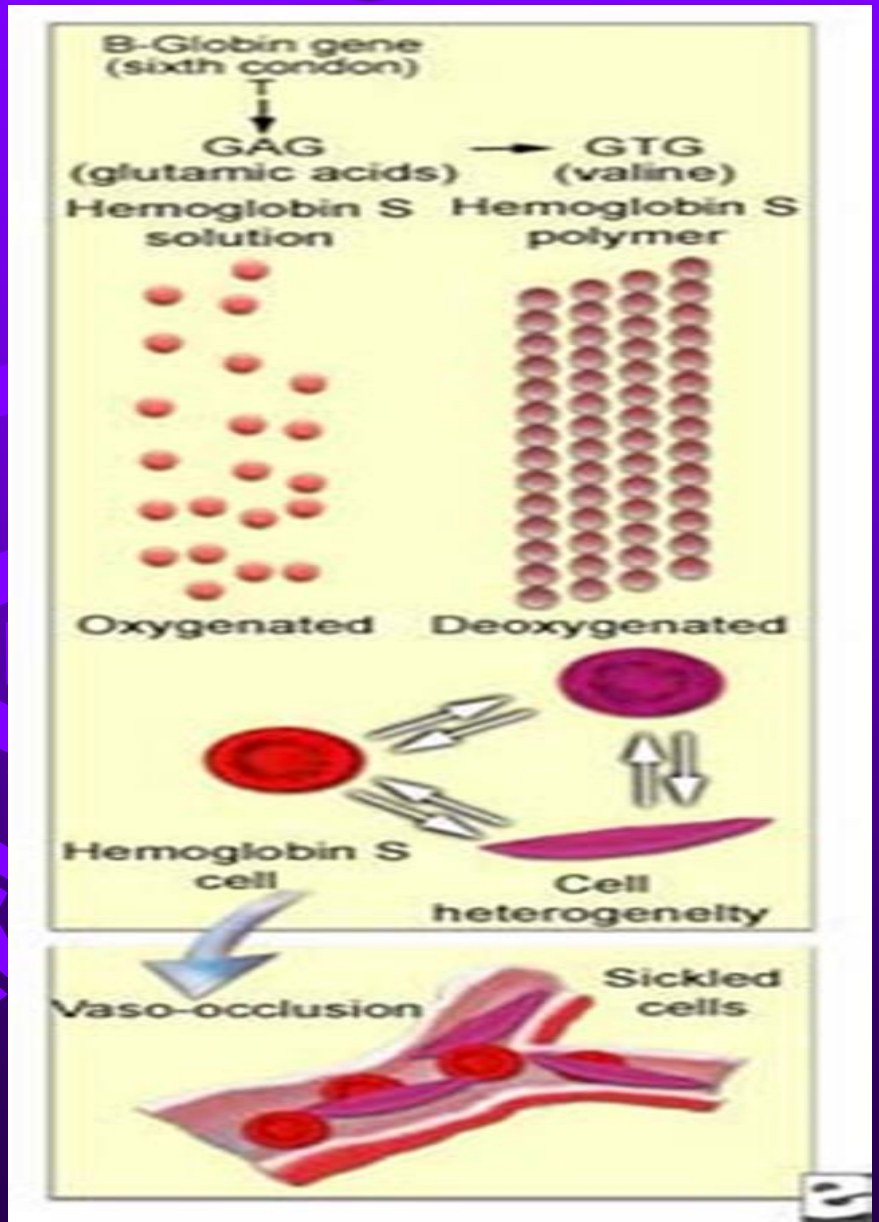
Hemoglobin

- 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

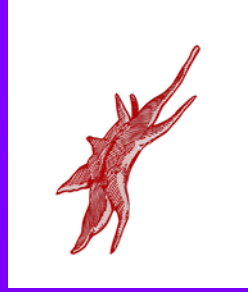


Sickle Hemoglobin

- 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

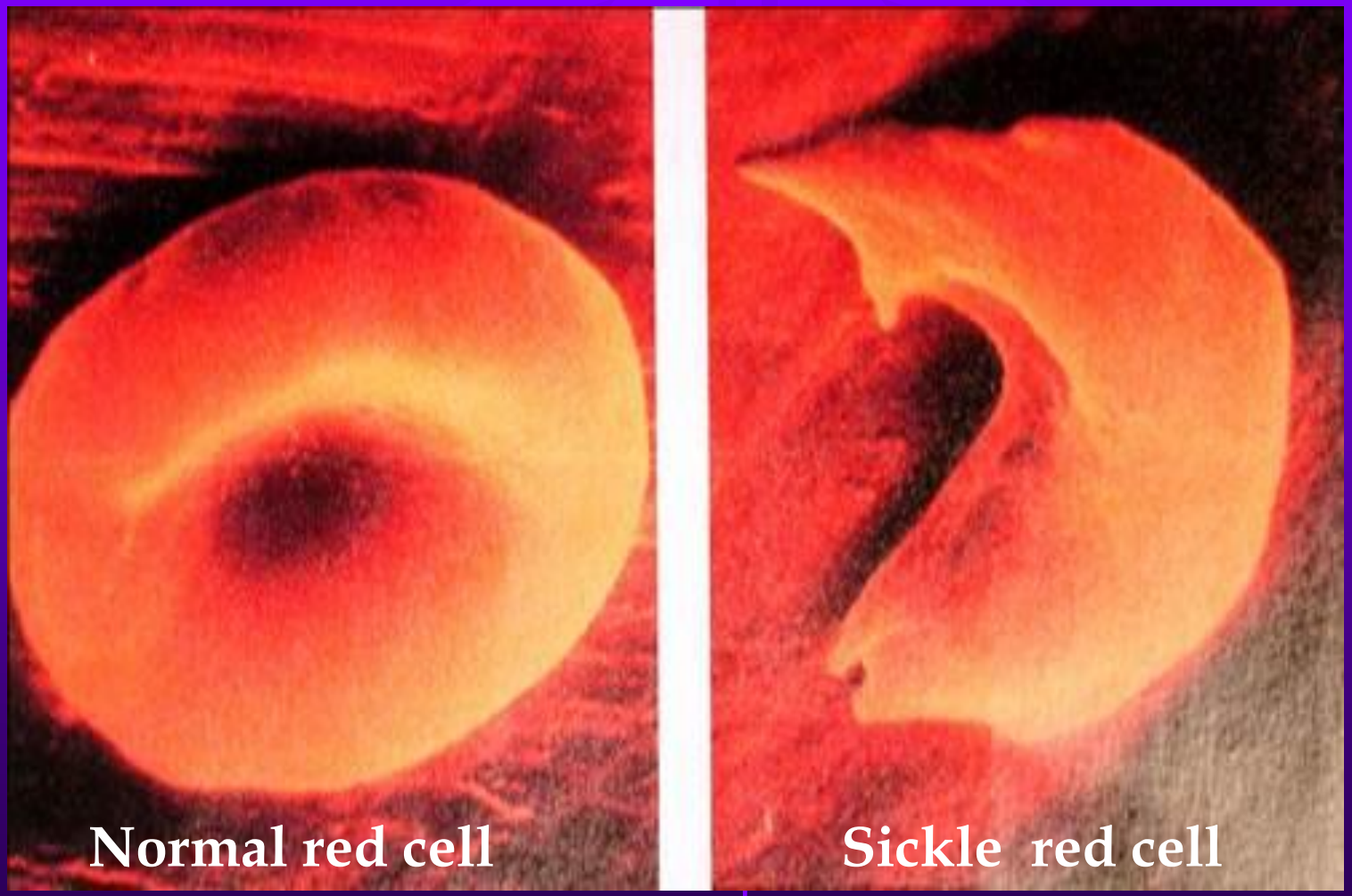


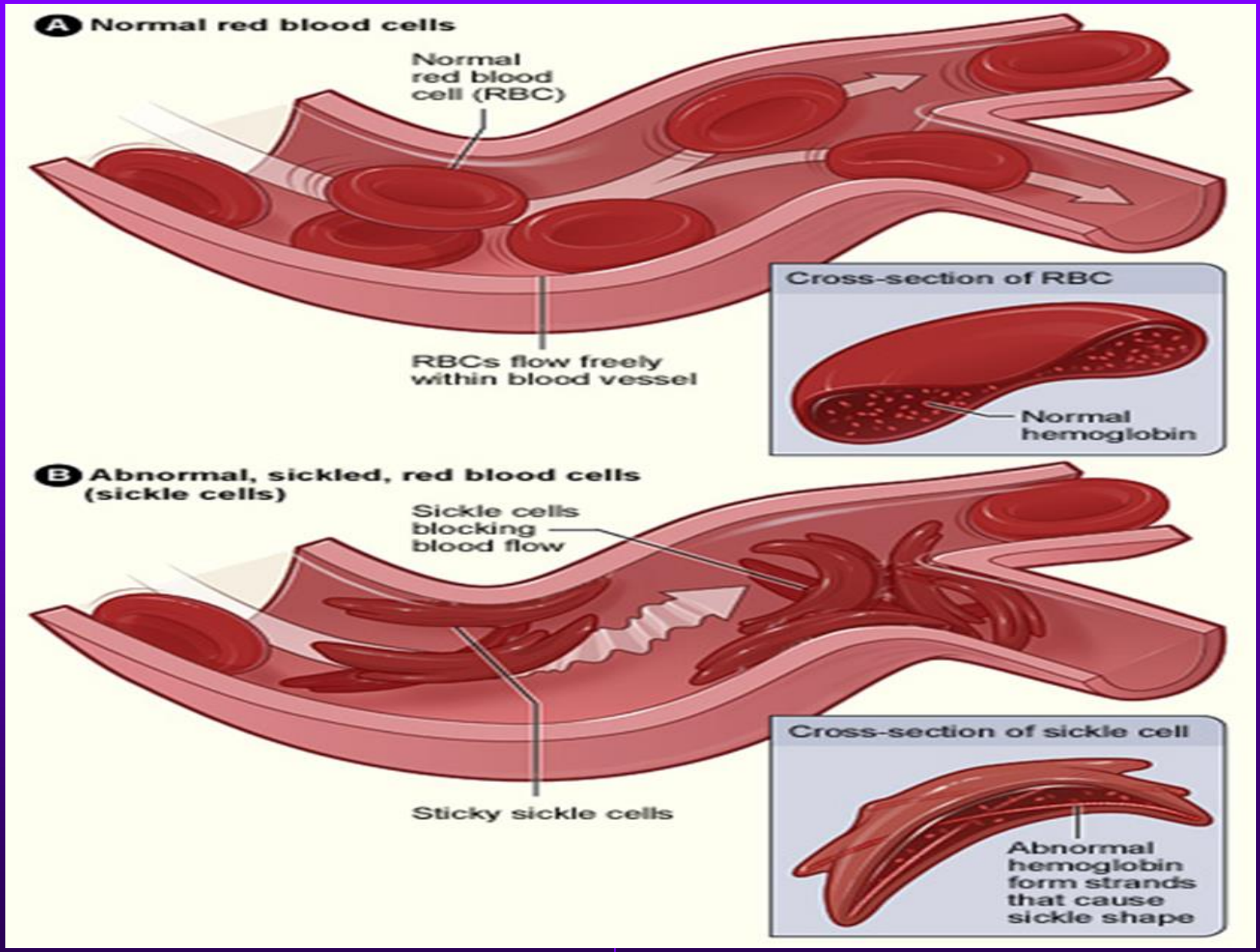
Clinical Complications



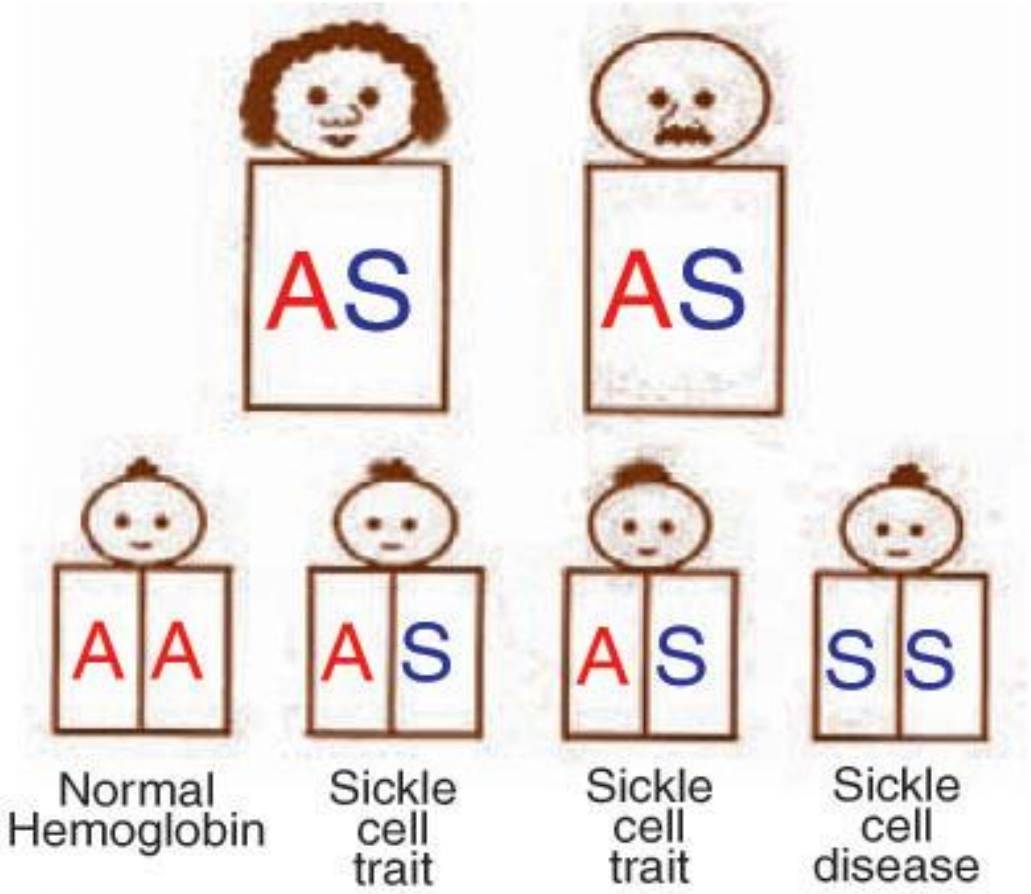
- **Anemia-** because the red cells break down easily and have a reduced life span.
- **Vaso Occlusion-** Blockage of blood vessels and inflammation within the blood vessels causing complications

Electron microscope



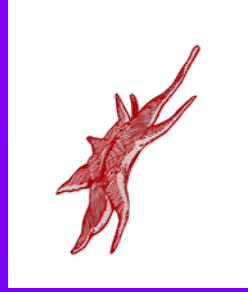

A Normal red blood cells
Normal red blood cell (RBC)
RBCs flow freely within blood vessel
Cross-section of RBC
Normal hemoglobin
B Abnormal, sickled, red blood cells (sickle cells)
Sickle cells blocking blood flow
Sticky sickle cells
Cross-section of sickle cell
Abnormal hemoglobin form strands that cause sickle shape

Genetic Inheritance

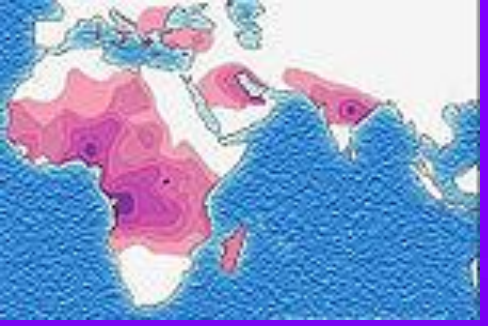


Here, both parents carry S trait.

Types of Sickle cell disease



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

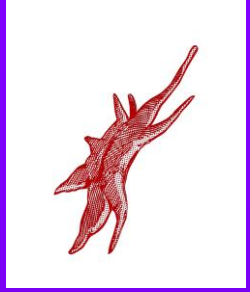


Epidemiology

- 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%)

- 1.Hassell KL. Population estimates of sicklecell disease in the U.S.Am J Prev Med. 2010;38(suppl 4):S512–S521
- Quinn CT, Rogers ZR, Buchanan GR. Survival of children with sickle cell disease. Blood 2004;103(11):4023–4027

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

- Platt OS, Brambilla DJ, Rosse WF, et al. Mortality in sickle cell disease. Life expectancy and risk factors for early death. N Engl J Med 1994;330:1639–1644

Death was because of

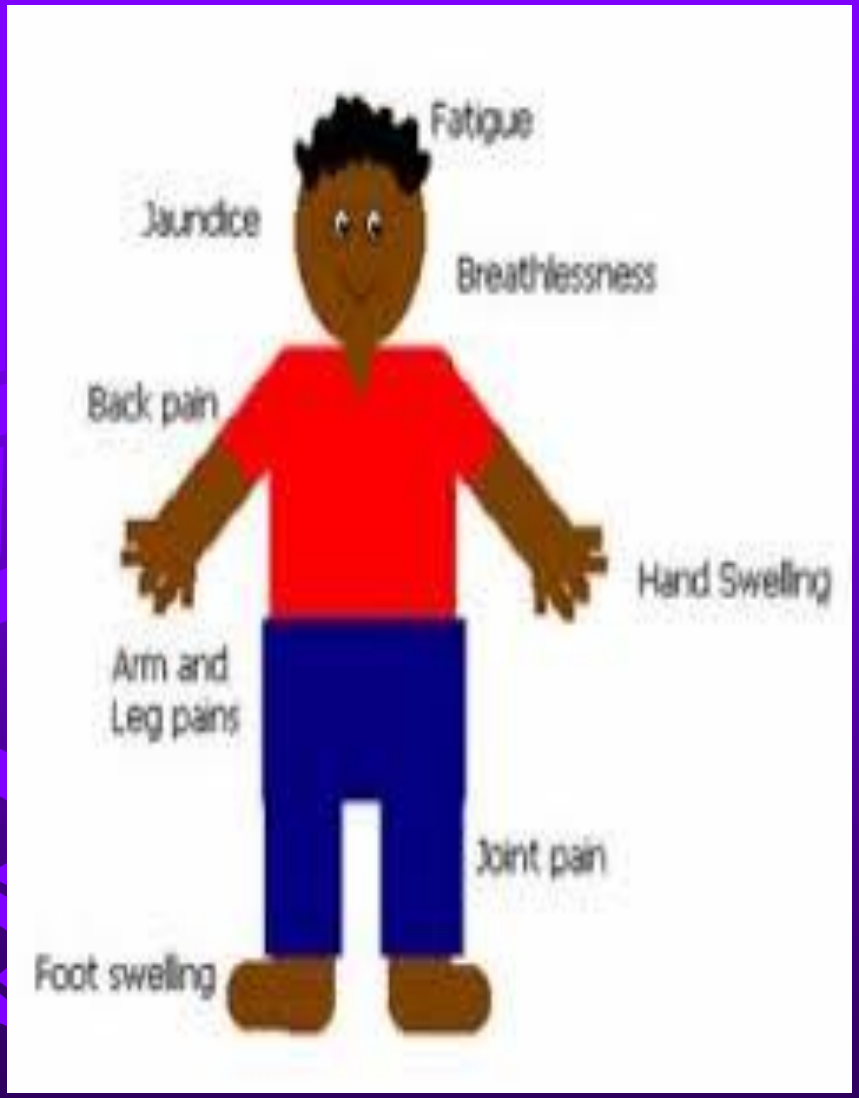
- Sepsis or Infection- sudden onset of fever, infection which spread rapidly
- Splenic Sequestration crises- Sudden enlargement of the spleen causing a sudden drop in blood pressure

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

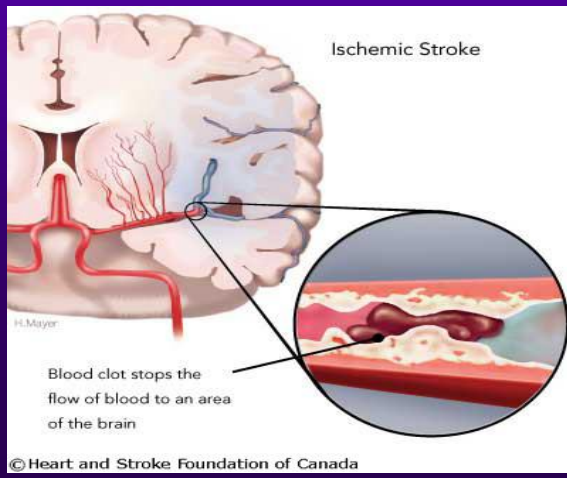
PAIN

- 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

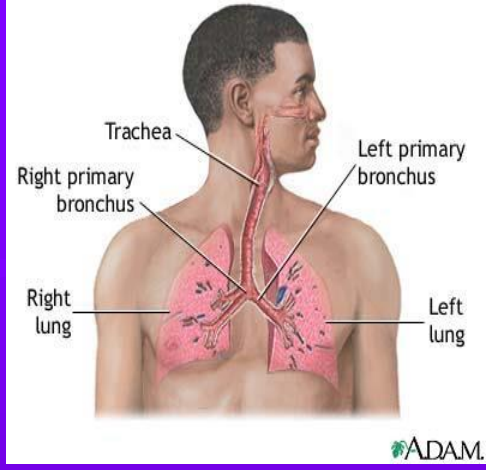
- **Infection**- Sepsis , Meningitis
- Acute Abdominal Crisis
  - Abdominal pain and distention
- **Stroke**
  - Occurs in 11% of patients, Peak at 6-9yrs
  - Subclinical Stroke in 30% of patients
  - Patient may have no symptoms



Clinical Features

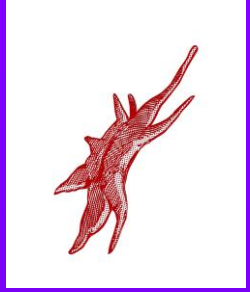
- **Splenic Sequestration Crisis**
  - Rapid accumulation of blood in the spleen
  - Seen in patients < 3 years of age
  - Abdominal distention, enlarged spleen and shock.
  - High mortality
  - Also seen in adults
- **Aplastic Crisis**
  - Temporary stoppage of red cell production often caused by parvovirus B19.
  - Have pallor, fatigue and rapid heart rate

Clinical Features

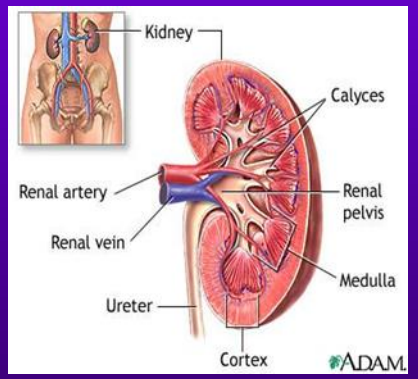


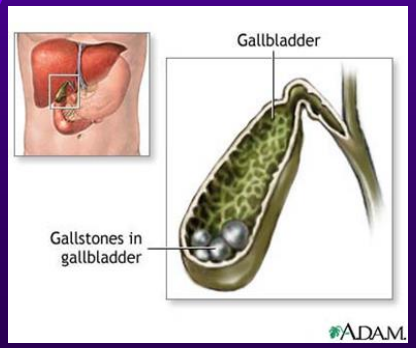
- **Acute Chest Syndrome (ACS)**
  - Classified by new pulmonary infiltrates associated with cough, breathlessness and chest pain.
  - May be severe, attributed to approximately 25% of deaths in patients with SCD.
  - **Often the cause of death in young adults**
  - **Requires hospitalization and transfusions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

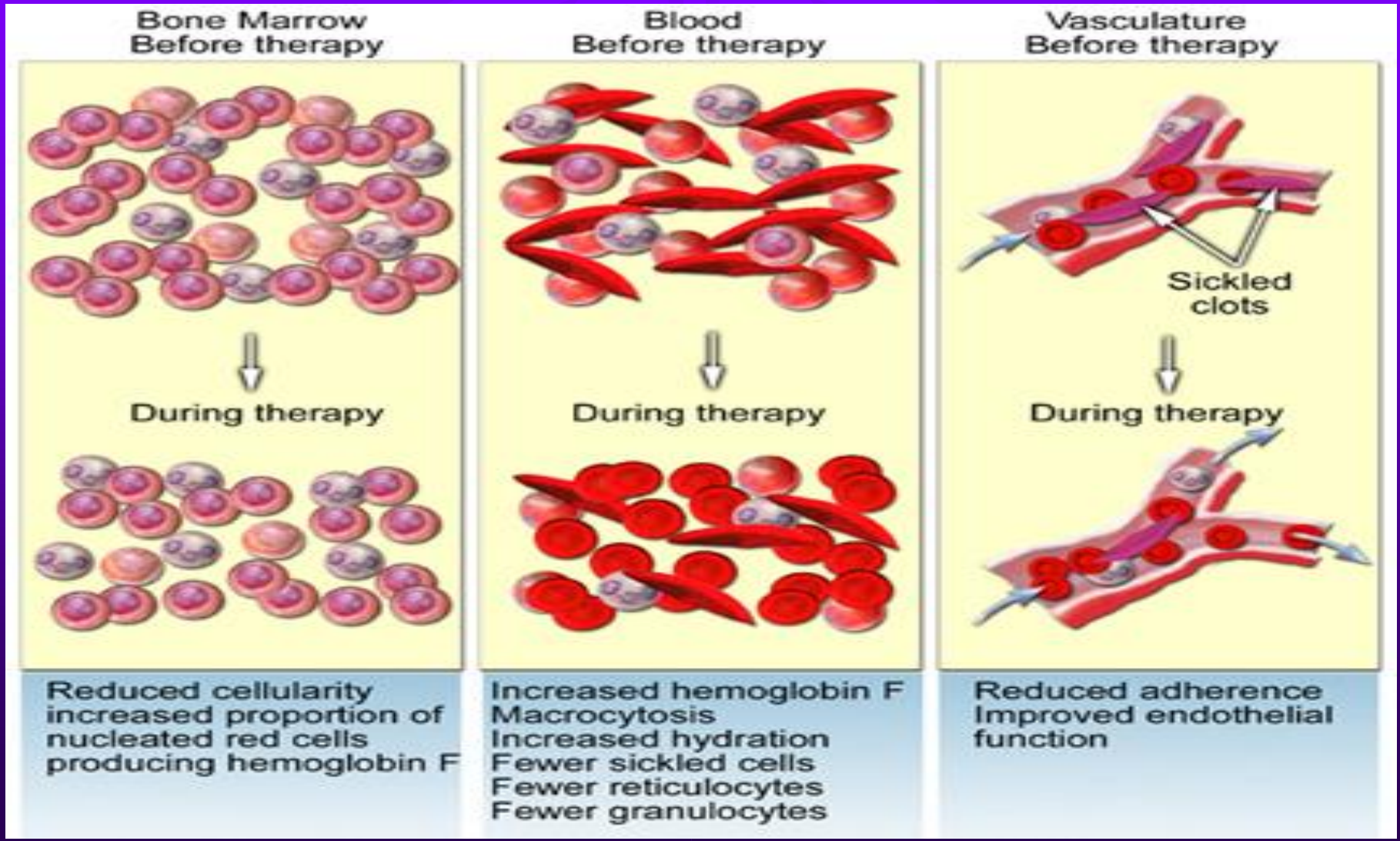
There are Improvements in care

- 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

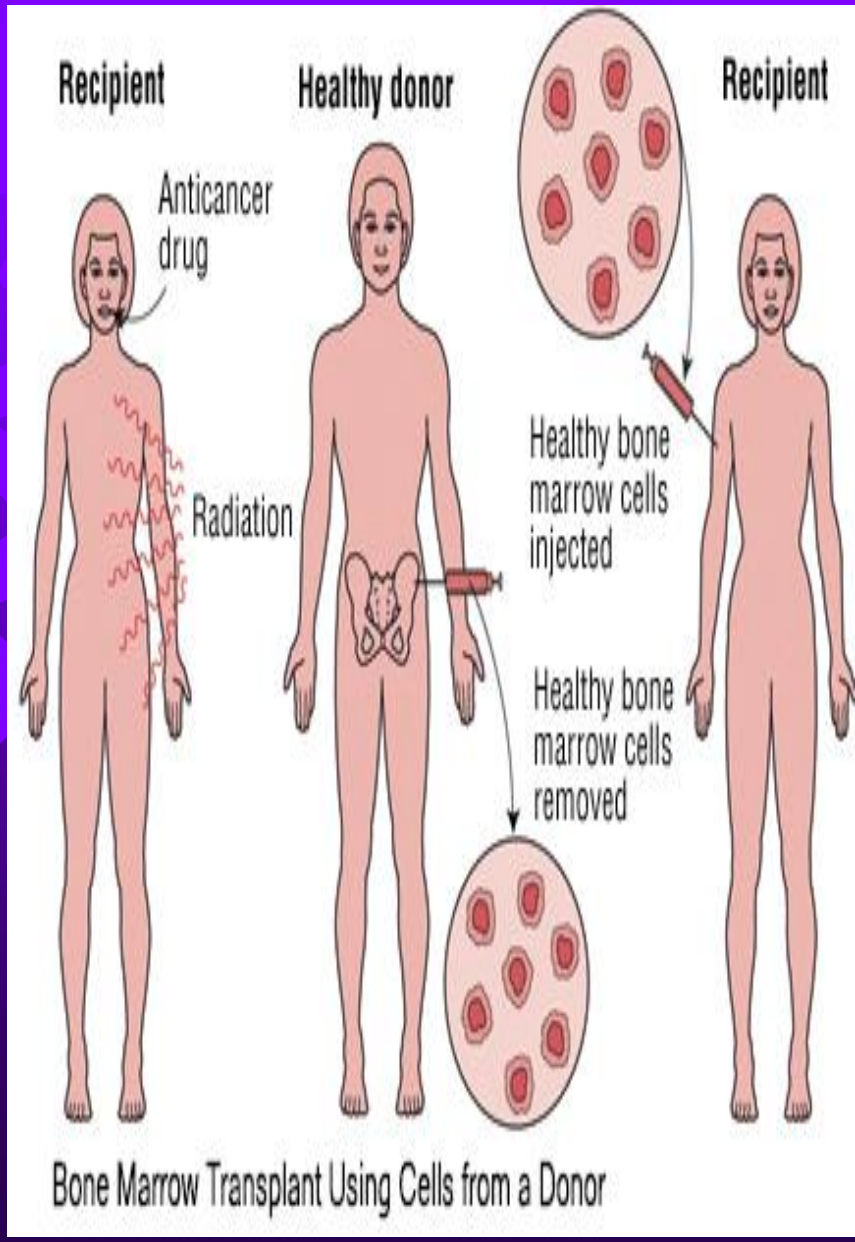
- Better Comprehensive care in pediatrics
- Education of the family
- Communication with school
- Children often are eligible for Insurance
- Parents work closely with the health care team
- **Can work with Home Health Care Manager**

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

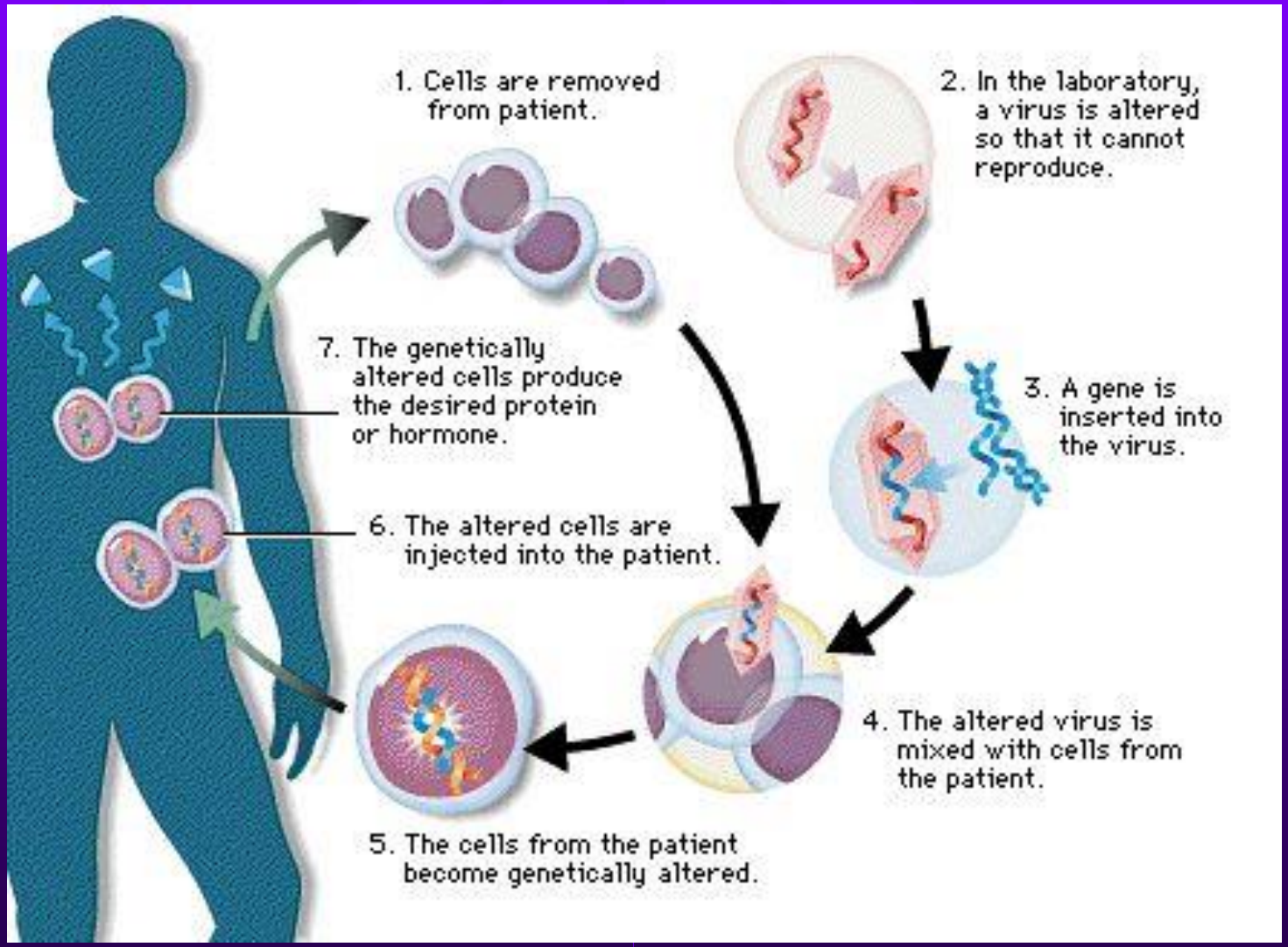


1. Locatelli F, Kabbara N, et al. Outcome of patients with hemoglobinopathies given either cord blood or bone marrow transplantation from an HLA-identical sibling. Blood. 2013;122(6):1072-1078.

2. Bernaudin F, Socie G, Kuentz M, et al. Long-term results of related myeloablative stem-cell transplantation to cure sickle cell disease. Blood. 2007;110(7):2749-2756

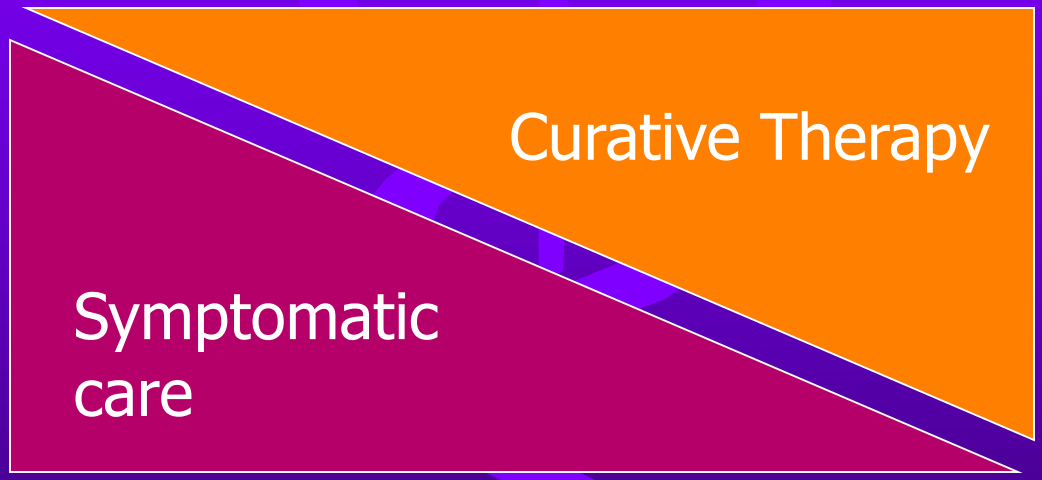
3. Mentzer W, Heller S, Pearle P, Hackney E, Vichinsky E. Availability of related donors for bone marrow transplantation in sickle cell anemia. Am J Pediatr Hematol Oncol. 1994;16(1):27-29.

Gene Therapy



Current State of the Art

- PCN
- Blood Tx
- Pain Rx



- Bone Marrow Transplantation
- Gene Therapy

Immunizations

TCD- Stroke prevention

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"

Quinn CT, Rogers ZR, Buchanan GR. Survival of children with sickle cell disease. Blood 2004;103(11):4023–4027

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

Treadwell M, Telfair J, Gibson RW, et al. Transition from pediatric to adult care in sickle cell disease: establishing evidence-based practice and directions for research. Am J Hematol 2011; 86:116.

Transition issues- specific to Sickle Cell

- Most patients are on Medicaid[1]
- Need to navigate Insurance issues[2]

- 1. Brousseau DC, Owens PL, Mosso AL,Panepinto JA, Steiner CA. Acute care utilization and rehospitalizations for sickle cell disease.JAMA. 2010;303(13):1288–1294
- 2. Telfair, J., Ehiri, J. E., Loosier, P. S., & Baskin, M. L. (2004). Transition to adult care for adolescents with sickle cell disease: Results of a national survey. *International Journal of Adolescent Medicine and Health, 16*(1), 47-64.

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 absences[3]

- 1. Francoise Bernaudin, Impact of early transcranial Doppler screening and intensive therapy on cerebral vasculopathy outcome in a newborn sickle cell anemia cohort Blood 2011;117(4):1130-40
- 2. Quinn CT1, Rogers ZR, McCavit TL, Buchanan GR. Improved survival of children and adolescents with sickle cell disease. Blood. 2010 Apr 29;115(17):3447-52. doi: 10.1182/blood-2009-07-233700.
- **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**:

Re-Hospitalization	30 day	14 day
SCD	33.4%	22.1%
SCD 18-30	41.1%	28.4%

- 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**

- Brousseau DC, Owens PL, Mosso AL,Panepinto JA, Steiner CA. Acute care utilization and rehospitalizations for sickle cell disease.JAMA. 2010;303(13):1288–1294
- Quinn CT, Rogers ZR, Buchanan GR. Survivalof children with sickle cell disease.Blood.2004;103(11):4023–4027

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

Treadwell M, Johnson S, Sisler I, et al. Self-efficacy and readiness for transition from pediatric to adult care in sickle cell disease. Int J Adolesc Med Health 2016; 28:381.

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.

Medical Summary Booklet

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:_____

	Name	Contact Info
Primary Care Physician		
Hematologist		
Hematology nurse		
Hospital		
Pharmacy		

Diagnosis: ¤ SS ¤ SC ¤ S-B⁰Thal ¤ 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:

_____

_____

_____

_____

_____

_____

_____

Acute Chest Syndrome:

_____

_____

_____

Vaso Occlusive Crisis requiring admission

_____

_____

_____

Transfusion History:

Number Of Transfusions: _____

Antibodies: _____

OTHER:

_____

_____

ORGAN INVOLVED	COMPLICATION	Yes/No Date
Brain	Stroke Headache	
Eyes	Sickle Cell Retinopathy	
Bones	Infection Aseptic Necrosis	
Skin	Ulcers	
Lung	Pneumonia Acute Chest Syndrome	

TEST	DATE	RESULT	DATE	RESUL
CBC				
PULSE OX				
XRAY CHEST				
EKG				
ECHO				
PFT				
TCD				
MRI BRAIN				
MRA BRAIN				

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
- http://www.hematology.org/Advocacy/Policy-News/2016/5581.aspx

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?

Disease Knowledge
I know what type of sickle cell disease I have.
I know my medical needs and can explain them to someone.
I know what a hematologist is and why I go to one.
I know what to do in case of a medical emergency.
I understand what causes a pain episode.
I understand how drugs, alcohol and tobacco affect sickle cell disease.
I have friends that I can talk to about sickle cell disease.
I know about necessary screening exams (echo annually, kidney function annually, retinal exams, etc.).
I know how to get blood work and x-rays.

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/health-topics/topics/sca/

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

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

Quick Links to Health Care Transition Resources
Connecticut Department of
Public Health Youth with Special
Health Care Needs
http://www.ct.gov/dph/cwp/view.asp?a=3138&q=432684

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

Sickle Cell Transition

Thank you

Questions?