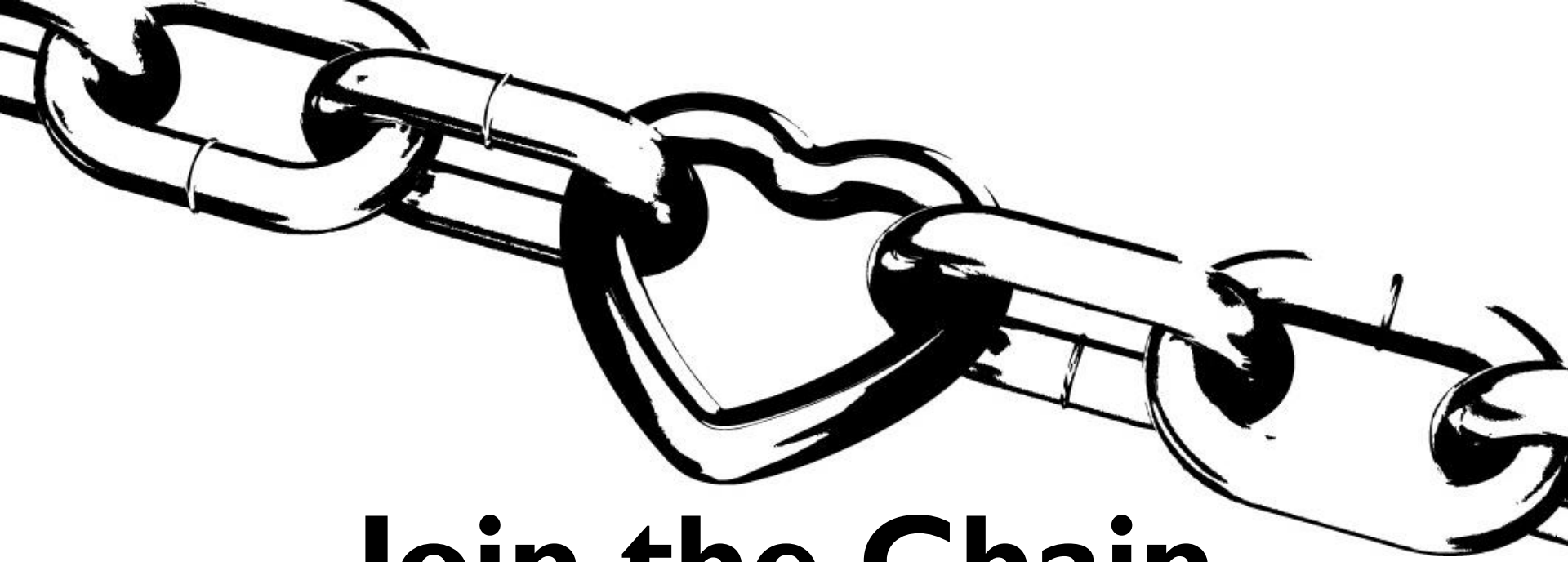

Join the Chain

Stop the Pain of Sickle Cell

Statewide Sickle Cell Conference
April 20, 2012

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Overview of care and management of adult sickle cell patients

Asmar Muhammad, PA-C
IHTC Adult Care

April 20th 2012

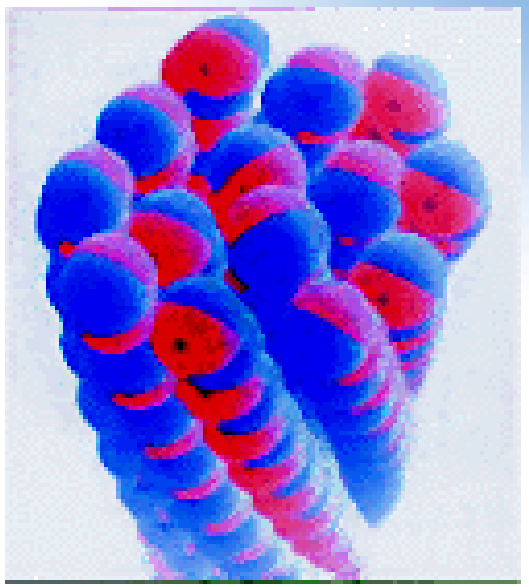
Indiana Hemophilia & Thrombosis Center

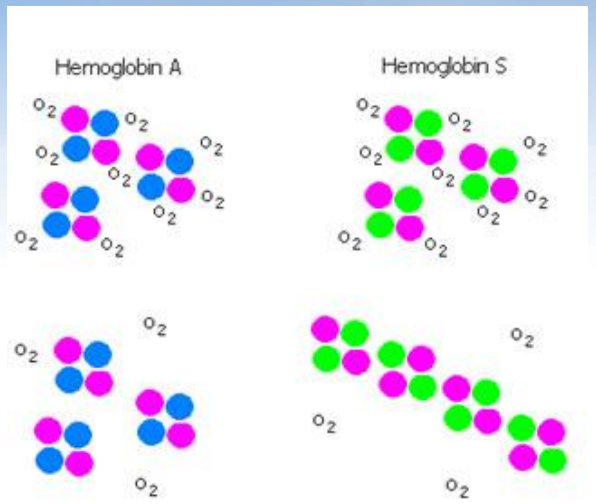
IHTC adult sickle cell population

- Approximately 240 adult patients
- >130 patients with hemoglobin SS disease
- 74 patients on hydroxyurea therapy

Pathophysiology of sickle cell disease.

- Deoxygenated sickle cells are rigid because they contain linear polymers of hemoglobin S (Hb S); polymerization is highly concentration dependent.
- Dilution of Hb S by a nonsickling hemoglobin such as fetal hemoglobin (Hb F) would be expected to lead ultimately to a decrease in the frequency of painful crises.





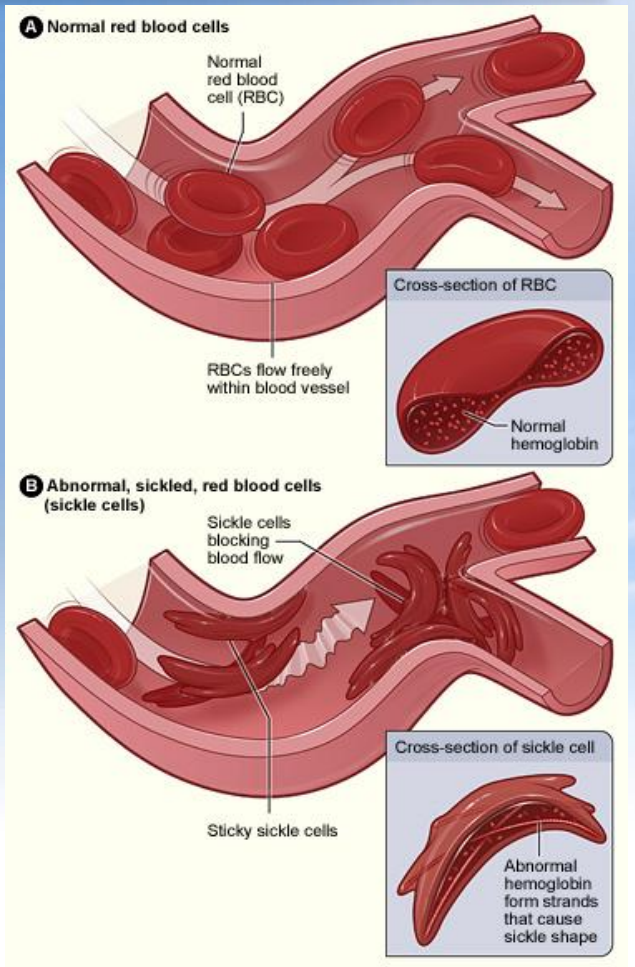
Vaso-occlusion

Hemoglobin S polymerizes on deoxygenation.

Blood flow is obstructed, depriving tissues and organs of oxygen.

Oxygen deprivation (hypoxia) can cause severe pain (the sickle cell crisis).

Over time, it leads to ongoing and progressive destruction of the various organs and tissues throughout the body.



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Common Acute and Chronic problems in adult sickle cell patients

Chronic Problems

- Chronic pain
  - Bone infarcts, tissue hypoxia
- Priapism
- Leg ulcers
- Iron overload secondary to blood transfusion
- Endocrine disorders
- Retinopathy (SC)
- Renal complications
- End organ damage
- Pulmonary artery hypertension (PAH)
- Depression

Acute problems

- Acute Pain
- Acute chest syndrome
- SOB
- Priapism

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Acute chest syndrome

- A medical emergency/admission!!!
- Symptoms
  - Fever
  - Chest pain
  - Pulmonary infiltrate on CXR
  - Pneumonia like presentation
- Treatment
  - Antibiotics
  - Oxygen support
  - Simple transfusion/Red cell exchanges

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Priapism

- Sustained painful erections
- Mean age of onset; 12 y/o, 89 % will experience one episode
- Occlusion causes obstruction of venous drainage from penis.
- Stuttering priapism:
  - Few minutes-3 hours
  - Resolve spontaneously
  - Novel treatment
- Prolonged: More than 3 hours; A medical emergency!!!!
- Treatment
  - IV hydration and analgesia
  - Penile aspiration if no resolution within an hour after fluids
  - Simple blood transfusion or red cell exchanges for chronic
  - Sudafed
- Urology follow-up

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Leg ulcers

- Effects 8- 50 % of patients.
- Tissue hypoxia from vaso-occlusion due to sickling
- Stages 1-4 (full thickness skin loss and necrosis of subcutaneous tissue)
- Painful, Indolent and intractable
- More common in males
- Certain phenotypes may be more susceptible; high hemoglobin, stroke, PAH increased SBP and elevated LDH

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Leg ulcers: Prevention, Treatment and management.

- No real prevention.
  - Providers should be aware of some causes such as Infection, trauma, recurrent LE edema, IV access and dry skin.
- Protect ankles with orthosis.
- Wet to dry dressing.
- Longer than 6 months
  - Transfusion, Unna boot, zinc sulfate, hyperbaric O2.
  - Zinc is important in wound healing; check for deficiency
  - Grafts with exchange transfusion.
  - Opioid analgesics.
  - Regranex/Procuren
  - Anecdotal modalities; Herbs and magic
  - Encourage frequent elevation

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Iron overload

- Iron overload: Multiple blood transfusions for anemia.
- Blood transfusions contain extra iron and the body has no way of removing excess iron.
- Long-term iron chelation:
  - Slows the accumulation of excess iron in the liver
  - Cardiomyopathy
  - Renal failure

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Iron overload: Diagnostics test and treatment

- **Serum Ferritin**
- **Liver biopsy:** Gold standard
- **R2-MRI** (also known as **FerriScan**): Replacing liver biopsy for detecting liver iron load.
- Treatment
  - Desferal (deferoximine)SC or IV
  - Exjade (deferisirox)Oral

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Endocrine Deficiencies; Zinc, testosterone and vitamin-D

- Vitamin-D deficiency: 98 % of SCD population
  - Lab monitoring: 3-4 months
  - Treatment: Oral supplementation
- Testosterone deficiency (Hypogonadism)
  - Lab monitoring: Annually
  - Treatment: Oral supplement or Topical gels
- Zinc deficiency
  - Lab monitoring: Annually
  - Treatment: Oral supplementation

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Maintenance regimen for adults with sickle cell

- Oral or transdermal pain management
- Folic acid supplement
- Hydroxyurea (Hydrea) (Droxia)
- Blood transfusions:
  - Acute issues; hypoxemia, priapism and acute chest syndrome
  - Scheduled automated red cell exchanges; Preservation of organ function and leg ulcer healing.

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Pain management

- NSAID's
- Tylenol
- Moderate strength opioids: Hydrocodone
- Adjunctive measures: Heat, relaxation
- Chronic pain:
  - Short acting opioids: Breakthrough pain
  - Long acting opioids: Q 8-12 hours
- IV pain management in clinic or inpatient
- Referral to pain management services

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Hydroxyurea

- Increases fetal hemoglobin which can make red cells act more normal.
- Decreases intensity and frequency of pain episodes. Priapism , acute chest etc…
- Weight based dosing
- Labs are monitored regularly
- Side effects: Myelosuppression, GI upset, dermatologic changes
- Reversible
- Efficacy corresponds with compliance

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Health Maintenance/Surveillance

- Periodic lab monitoring for renal and liver function.
- Pulmonary hypertensive screening.
- Provide periodic immunizations.
- Encourage annual ophthalmologic and fundoscopic evaluation especially in patients with hemoglobin SC disease.
- Strongly encourage all patients to enroll in a primary care service.

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IHTC Outpatient Clinic

- Hours of operation 8:30 am- 4:30 pm
- Infusion: Maximum for 4 patients.
- Staffed by RN's PA's and supervising M.D.
- Patients are typically treated 1-3 days before admission or resolution of pain.
- Admissions are managed by the IHTC hospitalist.

IHTC Support services

- **Physical therapy:**
  - Evaluation and treatment of musculoskeletal issues
  - Coordination with the orthopedic service
- **Vocational rehabilitation:**
  - Job placement
  - Job training
    - Locate education funding
- **Social services:**
  - Psychosocial support
    - Insurance issues
    - Medication assistance
    - Housing
    - Transportation assistance

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Community outreach/Patient advocacy

- New born screening program: (Kisha Braswell)
- Sickle cell information booth at Indiana Black Expo Summer celebration.
- Annual “Sickle Cell-Abration” concert each fall to raise public awareness
- Blood drives

IHTC adult hematology staff

- Anne Greist M.D.
- Chirag Amin M.D.
- Randy Riley M.D. (Hospitalist)

- Asmar Muhammad PA-C
- JoAnne Coulter, NP
- Kimber Blackwell PA-C

Prognosis

- Dramatic improvement in 30 years
- 90 % of all phenotypes will survive past 20 y/o
- Patients are now living in there 7^{th} and 8^{th} decades
- Decrease risk of early death associated with acute complications

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Challenges

- Improving quality of life with longer life span
- Persistent drug seeking stigma
- Geriatric issues in SCD patients is not well studied
- Fear of hydroxyurea
- Improved world wide treatment
- Continued research

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Thank you and have a happy Friday!!!