Sickle Cell Disease
Objectives

- Most common sickle cell complications as well as signs and symptoms associated with them
- Identify appropriate nursing cares and appropriate interventions
- Gain some knowledge of usual medical management of these complications
General Information

- Hereditary disease characterized by abnormality in the structural part of the Hgb gene
- Most common genetic disorder among African Americans
- Estimated over 70,000 individuals affected by sickle cell disease and additional 1500 babies born each year
- The term sickle cell disease includes a number of sickle hemoglobinopathies
- All babies diagnosed through Newborn Screening
- Hemoglobin electrophoresis only accurate test
Traits

Most Common Traits

Sickle Cell Trait
Most Common
No Complications

C Trait
Less Common
No Complications

Thalassemia Traits
Least Common
May have mild anemia
Sickle Cell Disease

Most Common Types

Hgbss Disease
(Sickle Cell Anemia)
Most Common
Complications mild - severe

Hgb SC Disease
2nd most common
Disease course milder
Complications usually milder

Hgb SB 0 Thalassemia
Less common form
Disease course similar to hgbss disease
Complications mild - severe

Hgb SB + Thalassemia
Mildest form
Disease course mild
Complications usually mild
Sickle Cell Database

- Includes all patients – inpatient admits
- Diagnosis, chronic/other problems & surgeries, baseline hgb & retic, IV pain medication(s) and inpatient complications
- On all desktops, need password to access
Sickle Cell Complications

- Unpredictable, can occur at anytime, with minimal to no warning
- Numerous complications, several life threatening
- Most complications cannot be prevented
- Patients have little to no control over occurrence and frequency
- Can have more than one complication at same time
Infection

- #1 cause of death in children with hgbss disease under 5 yrs of age
- Happens because spleen becomes damaged by sickled rbcs → immunocompromised
- Most at risk for pneumococcal
- Types of infection → bacteremia/sepsis, pneumonia, meningitis, osteomyelitis
- Prophylactic penicillin started by 2 months of age
Signs and Symptoms

- Temperature of 38.5 or higher
- Tachycardia, tachypnea
- Lethargy, irritable
- Nuchal rigidity
- ↓ or coarse breath sounds
- Joint swelling or swelling around bony area with fever
- 02 Sats < 95%, high wbc count
Physician to evaluate all patients with fever of 38.5 or higher
Should have septic workup (i.e. cbc, blood cult, UA, CXR)
IV antibiotics → cephalosporin, Nafcillin if patient “appears septic” or if not sensitive to cephalosporin
ID & Hematology consult with positive bacterial infection, ortho consult with ? bone infection
Anemia

- Most children have anemia which is chronic & well compensated for
- Not due to iron deficiency, happens because sickled rbc's die in 14-21 days
- Causes slow growth & development & sexual maturity
- Usual hgb: Hgbss (6-8 gms), Hgbsc (10-12 gms), Hgbsb 0 (7-9 gms), Hgbsb + (11-12)
- Decision to transfuse should be based on child’s clinical condition not just hgb
Splenic Sequestration

- Potentially life threatening – hgb can drop to half its baseline within a few hours
- Blood enters spleen, sickled rbc s block exit →blood pools in spleen→ ↑spleen size
- Most common with hgbss disease, can start as early as 2 months of age & usually not a problem with hgbss disease after 5 yrs of age
- Can continue to be a lifelong problem for hgbsc and hgbssb + thalassemia
Signs and Symptoms

- Palpable spleen
- Abdomen may be distended (some pts have splenomegaly, not same as sequestration)
- May c/o abdominal pain
- Low hgb (than baseline), low plt count
- Irritable and/or lethargy, pale
- Tachycardia
Usual Medical Treatment

- Hematology consult
- IV fluids to prevent hypovolemic shock
- T&C (transfuse if necessary)
- Antibiotics with fever, 02 with sat < 95%
- Transfusion program or splenectomy after several recurrences
Aplastic Crisis

- RBC production is shut down by parvovirus B19 → hgb to drop
- Usually preceded by URI
Signs and Symptoms

- Pallor, fatigue, lethargy
- May c/o headache
- Hgb usually < 5 gms, retic of 0 (for hgbss disease)
- Tachycardia
- With severe crisis – signs of CHF
Usual Medical Treatment

- Hematology consult
- Depends on severity of crisis
- IV fluids to prevent hypovolemic shock
- T&C – blood on hand or transfuse slowly
- Antibiotics with fever, O2 with sats < 95%
- Isolation for parvovirus
Vaso-Occlusion

- RBCs sickle, stick together → ↓ blood & oxygen to surrounding tissue
- Affected area undergoes hypoxia & infarction → pain and dysfunction
- Area of vaso-occlusion determines sickle cell related complication
- Can have more than one type of vaso-occlusion occurring at same time
Sickle Cell Pain Episode

- Most common type of vaso-occlusion
- Most common sickle cell complication
- Most common reason for hospitalization
- Most common with Hgbss disease
Characteristics

**Duration**
- Length of pain
- Gradual or sudden onset

**Severity**
- Mild to severe
- Hospitalized – moderate to severe pain

**Character**
- Burning, sharp, deep, gnawing, throbbing
- Consistent with each episode
- Migratory
Developmental Factors
- Early as 6 months
- Dactylitis usually 1st type of pain
- Extremity pain – children
- Abdominal, chest, extremity pain – teens

Temporal Factors
- Frequency - which varies among patients
- If common during childhood → will usually continue at same or ↑ rate
- Usually have own established pattern
Precipitating Factors

- Sickle cell pain episodes may be brought on by infection, dehydration, hypoxemia, stress, fatigue, menses, pregnancy, prolonged exposure to heat & cold
- Most pain episodes have no defining precipitator
Sickle Cell Pain Management

- Centered around 3 foci
  1. Adequate analgesia
  2. Sufficient hydration
  3. Identification & intervention of precipitating factors if present
- Goal is to make patient as comfortable as possible
- Most of population undertreated
Pain Assessment

Subjective Data
- Self report !!!!!!
- Assessment tool
- Learned behavior

Objective Data
- No Real data
- Vital signs, hgb, exam not reliable indicators
- Assess for edema, redness, warmth
Pharmacologic Management

- Causes enormous frustration for patient and staff
- Stay on top of pain control (remember pts may have been taking oral meds for a number of days at home)
- MSO4, Dilaudid – ideal choice, avoid Demerol
- Should add Nonsteroidal – Toradol
- Pain team (some pts are followed in pain clinic)
How are analgesics given?

Method

Scheduled
Assured of pain med
Resp depression
Usually not effective

Continuous Drip
Gives constant blood level
Sedation
Resp Depression

PCA
Safest & most effective
Low lockout (10 minutes)
Low basal
Assessing Effectiveness

- Ask patient /family
- Use an effective pain assessment tool and manage pain according to too (i.e., high "pain scale" → change in pain management)
- Be careful of using own judgment
- Look at change in activity level, appetite
Prolonged Pain Episode

**Tolerance**
- After 5-7 days same dose may not be effective
- May c/o increased pain

**Dependence**
- After 5-7 days may have physiological dependence
- Withdrawal symptoms
Weanings

- Should be done with parent and patient knowledge
- Is individualized
- PCA – Patient will usually wean themselves
- May add oral pain med before stopping IV pain med
Other

- Treat narcotic side effects → nausea, itching, constipation
- Hydrate → Initial fluids at 1 ½ maintenance, then maintenance
- Monitor O2 sats (>94%)
- Encourage IS with chest pain, shallow breathing, sedated
- Monitor activity
- Use nonpharmacologic methods (heat, music, distraction)
Acute Chest Syndrome

- Second most common type of vaso-occlusion
- Vaso-occlusion in lungs → hypoxia → 02 sats to drop → sickling worsens and a vicious cycle ensues
- Requires immediate attention – life threatening
- Determining etiology can be difficult
- Patients with asthma at ↑ risk
Signs and Symptoms

- 02 sats < 95% - usually first sign
- Symptoms of acute resp infection
- May c/o chest, abd or back pain
- Tachycardia, tachypneic, pleuritic pain
- Abnormal CXR (may be normal in beginning)
- Decreased breath sounds
Usual Medical Treatment

- Depends on severity
- Early intervention key to survival

**Mild Episode**
Hematology consult, 02, CXR, ? Blood gases, antibiotics, IV fluids, bronchodilator, simple prbc transfusion

**Severe Episode**
Hematology consult, 02, CXR, blood gases, IV fluids, modified or full exchange transfusion, ? intubate, bronchodilator, ? steroids
Stroke

- Vaso-occlusion in brain, **medical emergency**
- Usually no pain
- Occurs in about 7% of sickle cell population
- Can be reversible if symptoms caught early
Signs and Symptoms

- S&S - depends on area of brain affected
- Usually subtle
- May include asymmetry of face, paralysis of one side or extremity, seizure, altered level of consciousness, sudden change in behavior, speech impairment
Usual Medical Treatment

- Acute infarct – made by MRI (CT scan will usually show bleed or old infarct)
- Even if MRI normal, treatment may be started based on symptoms
- Hematology Consult
- Exchange transfusion
- Anticonvulsants with seizures
- 02, IV fluids
- PT, OT, Speech therapy if needed
- Placed on chronic transfusion program indefinitely
- Long term – chelation therapy
Priapism

- Painful penile erection → trapped sickled rbc
- Common in age 6-20 yrs
- May have inability to void
- Can lead to impotency
Usual Medical Treatment

- Hematology consult
- Urology consult – May need foley or surgery
- May want to avoid cold/warm packs
- Adequate analgesics
- Simple or exchange transfusion
- Lupron therapy
Other Problems

- Sickle cell disease can bring about chronic problems
- Usually occur over a period of time
- Can cause other types of pain
- Should be included as part of assessment

**in other words not all pain is sickle cell pain!**
Cholelithiasis

Gallstones due to hemolytic anemia → ↑ production of bilirubin

Starts at about 5 yrs of age

S&S – abd discomfort after fatty food, N/V, jaundiced sclera

Diagnosed by ultrasound

Cholecystectomy recommended
Renal Problems

Vaso-occlusion occurs in kidneys → loss of urinary concentration ability → ↑urine output → dehydration

Usually have difficulty “holding their urine”, have nocturia

Need to stay well hydrated, dehydration is precipitant of sickle cell pain
Bone Problems

Changes in bony structure with continued vaso-occlusion → chronic pain in adults

Avascular Necrosis – ↓ blood supply to hip → bony head of femur infarcts → continual hip pain (different from sickle cell pain & limp (Usually not reversible w/o intervention - Ortho consult (which results in nonweight bearing and/or surgery)
Tonsil & Adenoid hypertropy

Tonsils and/or adenoids become enlarged

S&S – snoring, mouth breathing, large tonsils, ↓02 sats

ENT consult recommended → T&A
# Treatments – Long Term

<table>
<thead>
<tr>
<th>Hydroxyurea</th>
<th>Transfusion Program</th>
<th>Bone Marrow Transplant</th>
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<tbody>
<tr>
<td>- Chemo drug → ↑ fetal hgb</td>
<td>- Stroke, frequent pain, acute chest splenic sequestration</td>
<td>- Only cure</td>
</tr>
<tr>
<td>- Used to ↓ # of pain episodes &amp; acute chest</td>
<td>- ↓ Amount sickling hgb</td>
<td>- Severe disease &amp; complications</td>
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<tr>
<td>- Can cause ↓ in hgb, wbc, plts</td>
<td>- Can cause Fe overload</td>
<td>- HLA matched sibling</td>
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