Sickle Cell Disease

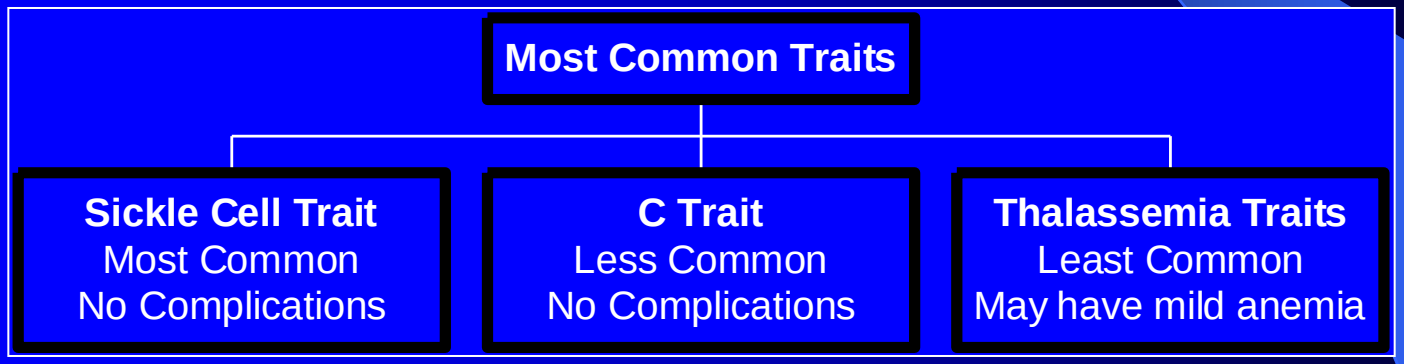
Objectives

- 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

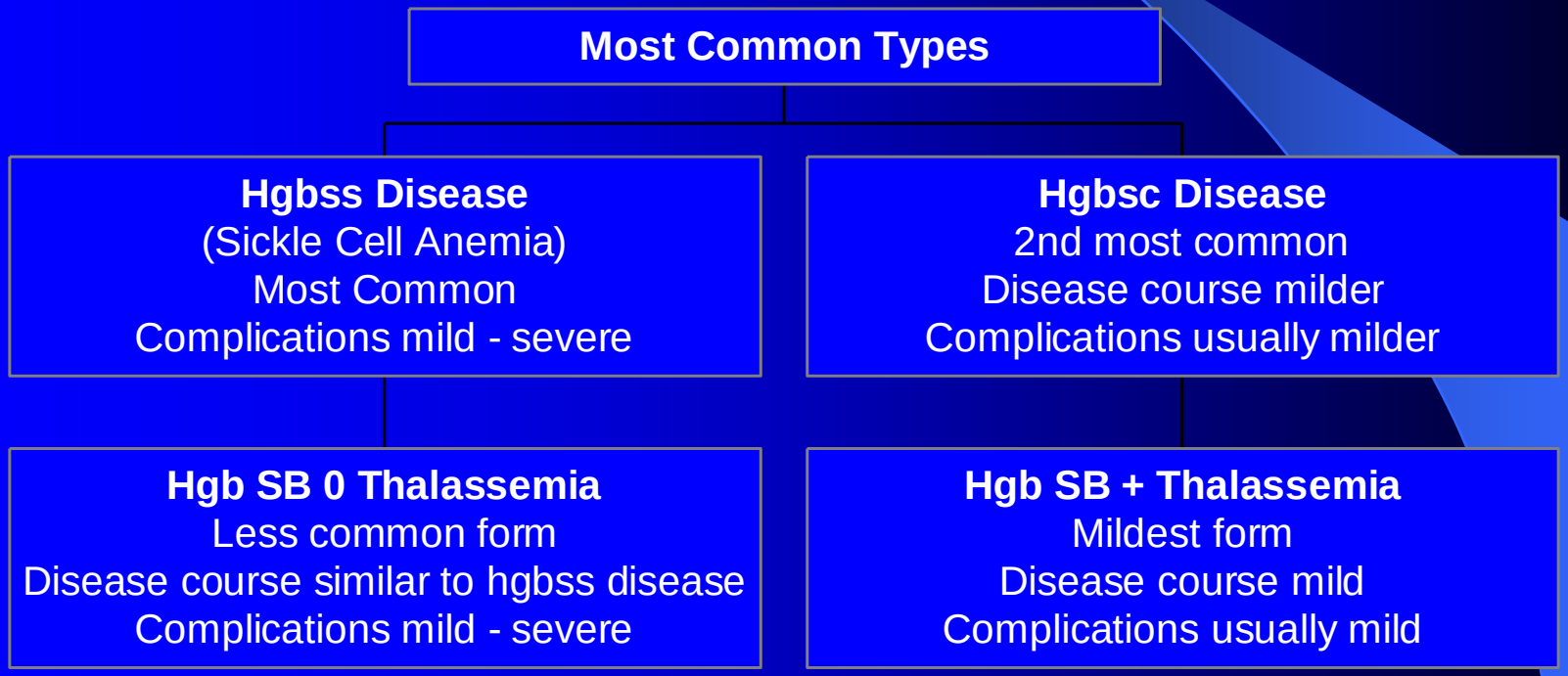
General Information

- 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



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, osteomylitis
- 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

Usual Medical Treatment

- 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 rbcs 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 rbcs 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 hgbsb + 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 sats < 95%
- Transfusion program or splenectomy after several recurrences

Aplastic Crisis

- RBC production is shut down by parvovirus B19 $\rightarrow$ 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, 02 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

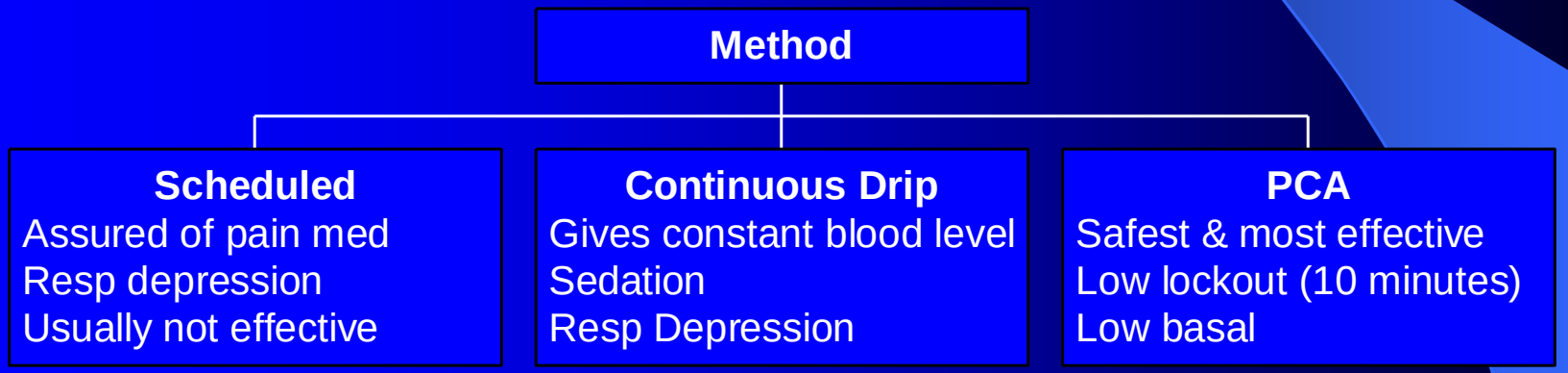
Objective Data

- ***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?



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

Weaning

- 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 02 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 indefintely
- Long term – chelation therapy

Priapism

- Painful penile erection → trapped sickled rbcs
- 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, juandiced 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

Hydroxyurea	**Transfusion Program**	**Bone Marrow Transplant**
•Chemo drug → ↑ fetal hgb •Used to ↓ # of pain episodes & acute chest •Can cause ↓ in hgb, wbc, plts •↑ life expectancy	•Stroke, frequent pain, acute chest splenic sequestration • ↓Amount sickling hgb •Can cause Fe overload	•Only cure •Severe disease & complications •HLA matched sibling