Nursing Study Guide - Sickle Cell Anemia Cheat Sheet

by helpmee via cheatography.com/148061/cs/32233/

Pathology

Most common hemoglobinopathies
- RBCs do not carry the normal Hgb but instead carry a less effective type

Severe chronic blood disorder that occurs 1/2,000 per year in U.S

Glutamic acid is replaced w/ valine in Hgb molecule → elongated RBC that is rigid & sickle w/ a shortened life spa

Significant anemia may occur when RBCs sickle

When cells sickle, blood becomes more viscous b/c cells clump together & prevent normal blood flow to the tissues of that area since their shape cannot pass through the smaller capillaries & venules → vaso-occlusive process leads to local tissue hypoxia → ischemia → infarction & pain crisis

Hemolysis occurs following sickling

Etiology / Risk Factors

Inherited autosomal recessive pattern	African (1 in 400, 8% carry trait), Mediterranean, Middle Eastern, & Indian descent
Passed on when both parents have gene or trait	May be triggered by stress or traumatic event
25% risk of Hgb SS, 25% Hgb AA, 50% Hgb AS	Infection, fever, acidosis, dehydration, physical exertion, excessive cold exposure, hypoxia
Hgb AS = carrier & usually have only minimal health problems	-

Complications

Cardiomegaly, fxn murmur	Pulmonary HTN, Restrictive lung disease
Retinopathy	Cholelithiasis & gallstones
Jaundice, hepatomegaly	Functional Asplenia
Chronic leg ulcers	MODs common in adulthood
Stroke	Sepsis
Delayed G&D & puberty	Organs

Signs & Symptoms

Infants asymptomatic until 3-4 months d/t Hgb F protection (later half of 1st year of life)

Pain crisis, recurrent pain episodes (vaso-occlusive) - ↑ tachycardia & tachypnea → more sickling - Most common in joints (hot, swollen)	Acute Chest Syndrome (ACS) - Cell clumping in lungs - ↓ gas exchange → hypoxia, wheezing, cough, chest pain, fever → more sickling
Aplastic crisis (profound anemia)	Dactylitis (hand-foot syndrome), aseptic infarction
Pale mucous membranes	Easily fatigued w/ poor appetite
↓ BP d/t severe anemia or ↑ BP d/t SC nephropathy	Acute abdominal pain (most common) d/t sludging & splenomegaly

Signs & Symptoms (cont)

Sequestration crisis / Splenic sequestration - Pooling of blood in liver & spleen w/ ↓ blood volume & shock	Bacterial meningitis or sepsis
	Bone infarction

Nursing Interventions

Immunizations & ABX to ↓ risk of infection	Tx underlying cause (infection)
H-O-P to it! → Hydration, O2, Pain relief	O2 during episodes of crisis to prevent further sickling
↑ fluids to promote hemodilution, 150 mL/kg/day or double maintenance w/ hypotonic, D5W or D5 w/ 0.25% NS	Adequate pain management helps ↓ stress; always believe pain level
NO PRN pain meds, use fix dose, can use w/ non-pharmacologic techniques	Assess pain w/ the right pain tool & look for complications of pain
Assess for S/S of ineffective tissue perfusion	Avoid sudden temp change (cooling mattress for fever)
Cluster care	Quiet environment & privacy

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Diagnostics

Sickle-Turbidity Test (Sickledex) finger stick

> Possibility or SCA or SC trait

Hgb Electrophoresis

> Dx, only accurate test for SCA

Hgb

> ~6-9 mg/dL (normal 11-15 in infant)
> Significantly lower w/ splenic sequestration, ACS, or aplastic crisis

Reticulocyte Count

> ↑ greatly

Peripheral Blood Smear

> Presence of sickle-shaped cells & target cells

Platelet Count, Erythrocyte Sedimentation Rate,

> ↑

LFTs

> ↑ bilirubin

X-Ray Studies or Scans

> Determine extent of organ or tissue damage d/t vaso-occlusion

K+

> ↑ d/t hemolysis of RBCs after transfusion

Pulmonary Infiltrate

> W/ ACS

Collaborative Care & Meds

Stem cell transplant	Splenectomy
Prophylactic ABX	Immunizations
Analgesics	O2
IV fluids	PRBCs
Hydroxyurea	

Teaching

Family support as they often feel guilty or responsible	Promote wireless communication w/ NP for collab & coaching
Disease process, complications, genetics, testing for carrier status	Regular health maintenance visits, immunizations, PCN, coping, adequate fluids
Avoid temp fluctuations, overexertion, & stress	Need 24-hr access to facility that specializes in SCA

Report & Seek Immediate Medical Attention:
- Suspected pain crisis
- Febrile illness
- Pale, listlessness, ↑ fatigue
- Unusual headache, loss of feeling, sudden weakness (stroke)
- Sudden vision changes
- Cough, SOB, chest pain (ACS)
- Limp or swollen joints
- Painful erection that won't go down (priapism)
- Symmetric swelling of hands & feet in (dactylitis)

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