

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



Diagnostics

Sickle-Turbidity Test (Sickledex) finger stick

Possibility of 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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