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Sickle Cell Anemia: Chemistry and Clinical Association

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Parkland College - Chemistry 101

Signs and Symptoms
- Abdominal pain
- Breathlessness
- Skin ulcers
- Fatigue
- Paleness
- Lower leg ulcers
- Chest pain
- Frequent urination
- Painful and prolonged erection (priapism)
- Bone pain
- Delayed growth and puberty
- Fever
- Rapid heart rate
- Jaundice
- Excessive thirst
- Poor eyesight/blindness
- Strokes
- Skin ulcers

Pathophysiology
Sickle cell disease (SCD) is a genetic disorder that results from a mutation in the globular protein hemoglobin. The spherical surface of a normal hemoglobin cell contains polar amino acid residues. On the other hand, the internal complex contains nonpolar residues. The surface polarity contributes to a hydrophilic quality thus being water, or aqueous, soluble.

The quaternary structure of hemoglobin is formed by the attachment of two α subunits containing 141 amino acids and two β subunits containing 146 amino acids.

Diagnosis
- Complete blood count (CBC)
- Hemoglobin electrophoresis
- Bilirubin
- CT scan or MRI
- Serum creatinine
- Serum potassium
- White blood cell count (WBC)
- Urinary casts in urine

Prognosis
The main causes of death in sickle cell patients are infection and/or organ failure. The understanding of sickle cell disease has greatly improved with the advancement of technology. As a result, the life expectancy has risen from between the ages of 20-40 years to well over 50 years of age. Moreover, Platt et al. (1994) indicated that 50% of sickle cell patients lived into their fifth decade.

Causes
- Autosomal recessive inheritance
  - If both parents pass the trait to the child sickle cell disease will result
  - If only one parent passes the trait the child will exhibit the sickle cell trait and may exhibit some symptomology
- Two people with the sickle cell trait present the following statistics:
  - 25% chance of delivering a baby with normal hemoglobin
  - 50% chance of delivering a baby with a carrier of the sickle cell trait
  - 25% chance of delivering a baby with sickle cell anemia/disease

Most commonly affects African Americans and Hispanics
One is born with SCD but doesn’t develop symptoms until between the ages of 4-6 months.

Common Treatments
- Bone marrow transplant
- Stem cell transplant
- Symptom management
  - Antibiotics
  - Analgesics (typically intravenous or oral)
  - Hydroxyurea (stimulates production of fetal hemoglobin)
- Blood transfusions
- Dialysis
- Supplemental oxygen
- Nitric oxide
- Rehydration

Potential Complications
- Acute chest syndrome
- Blindness/vision impairment
- Brain and nervous system impairment/symptoms
- Erectile dysfunction
- Hemolytic crisis
- Joint destruction
- Splenic dysfunction
- Splenic sequestration syndrome
- Hand-foot syndrome
- Osteonecrosis

Prevention
- Genetic counseling to determine if either parent carries the sickle cell trait
- Perinatal testing (during pregnancy)
- Adequate fluid intake, oxygen consumption, and treating infections
- Regular physicals (every 3-6 months)
- Vaccinations
- Avoid:
  - Stress
  - High altitudes
  - Tobacco
  - Nonpressurized flights
  - Drugs

Disclosure
The presenter has obtained an Associate degree in General Science from Parkland College in 2006, an Associate degree in Applied Science from Parkland College in 2007, and a Bachelor of Science in Nursing degree from Illinois State University in 2011. She holds a Registered Nurse license in the states of Illinois and Michigan.