

ACT SHEET FOR POSITIVE NEWBORN SCREENING RESULT (FS) SICKLE CELL ANEMIA (Hgb. SS Disease or Hgb. S/Beta Zero Thalassemia)

Disease Category: Hemoglobinopathies

Meaning of the Screening Result: Hemoglobin FS pattern on newborn screen is highly suggestive of sickle cell anemia or sickle beta zero thalassemia.

YOU SHOULD TAKE THE FOLLOWING ACTIONS:

- Contact a specialist in hemoglobinopathies for consultation or referral on diagnostic evaluation and management.
- Contact the family to inform them of the screening result.
- Evaluate the infant, examine for splenomegaly, draw a complete blood count with reticulocyte count and confirmatory test (hemoglobin electrophoresis)
- Initiate penicillin (PenVK 125 mg po bid) prophylaxis.
- Educate parents to have the infant seen and evaluated for sepsis when temperature is 101° Fahrenheit (38° Celcius) or greater.
- Report findings to Nebraska Newborn Screening Program.

Pediatric specialists in hemoglobinopathies are available at centers at Children's Hospital (402) 955-3950 and UNMC/Nebraska Medical Center (402) 559-7257.

Condition Description: Sickle cell anemia caused by inheritance of the hemoglobin S gene from both parents. Individuals with sickle beta zero thalassemia are compound heterozygotes with genes for hemoglobin S and beta zero thalassemia.

Clinical Expectations: Potential clinical problems include severe hemolytic anemia, life-threatening infection, episodes of pain, and organ damage and organ failure. Comprehensive care including prophylactic penicillin, recommended immunizations, family education and prompt treatment of acute illness reduces morbidity and mortality.

Confirmation of Diagnosis: Diagnosis is confirmed by hemoglobin electrophoresis and parental or DNA studies as indicated.

Additional information:

- Acute and Chronic Complications - www.tdh.state.tx.us/newborn/sc_guide.htm
- Grady Comprehensive Sickle Cell Center Web Site - www.scinfo.org
- Management and Therapy of Sickle Cell Disease - www.nhlbi.nih.gov/health/prof/blood/sickle/index.htm
- Sickle Cell Disease Association - www.sicklecelldisease.org
- Hemoglobinopathy podcasts [http://feeds.feedburner.com/Pediatric bytes](http://feeds.feedburner.com/Pediatric%20bytes)

