

ACT SHEET FOR POSITIVE NEWBORN SCREENING RESULT (FSC) HEMOGLOBIN SC DISEASE (HbSC Disease)

Disease Category: Hemoglobinopathy

Meaning of the Screening Result: Hemoglobin FSC pattern on newborn screen is highly suggestive of hemoglobin SC disease.

YOU SHOULD TAKE THE FOLLOWING ACTIONS:

- Contact a specialist in hemoglobinopathies for consultation 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).
- Initiation of penicillin prophylaxis (Pen VK 125mg po bid) is often recommended.
- 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 through the centers at Children's Hospital (402) 955-3950 and UNMC/Nebraska Medical Center (402) 559-7257.

Condition Description: Individuals with hemoglobin S-C disease are compound heterozygotes for the genes for hemoglobin S and hemoglobin C.

Clinical Expectations: Potential clinical problems include moderate 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)

