

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

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 repeat newborn screen to confirm FSC.**
- **Initiate penicillin prophylaxis (Pen VK 125 mg po bid) in consultation with hematologist.**
- **Educate parents to have the infant seen and evaluated for sepsis, when the baby's temperature is 101°Fahrenheit (38° Celsius) or greater.**
- **Report findings** to Nebraska Newborn Screening Program.

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. It is strongly recommended that these children be followed by specialists in hemoglobinopathies as part of coordinated care with their medical home.

Confirmation of Diagnosis: Confirmation may be done by **submitting a repeat dried blood spot filter paper specimen.** Parental or DNA studies may be done as indicated.

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