NEBRASKA
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DEPT. OF HEALTH AND HUMAN SERVICES

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

Meaning of the Screening Result: Hemoglobin FS pattern on newborn screen is highly suggestive of **sickle cell SS disease 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 repeat newborn screen to confirm FS**
- **Initiate penicillin (PenVK 125 mg po bid) prophylaxis in consultation with hematologist.**
- **Educate parents to have the infant seen and evaluated for sepsis when temperature is 101° Fahrenheit (38° Celsius) or greater.**
- **Report findings** to Nebraska Newborn Screening Program.

Condition Description: Sickle cell SS disease is 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. 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 at centers at Children's Hospital (402) 955-3950 and UNMC/Nebraska Medical Center (402) 559-7257.

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