NEBRASKA
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DEPT. OF HEALTH AND HUMAN SERVICES

ACT SHEET FOR POSITIVE NEWBORN SCREENING RESULT (FSA)
SICKLE CELL BETA PLUS THALASSEMIA (HB Sß+)

Meaning of the Screening Result: Hemoglobin FSA pattern on newborn screen is **highly suggestive of sickle beta plus thalassemia.**

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, and draw a complete blood count with reticulocyte count and repeat newborn screen to confirm FSA.**
- **Initiate penicillin prophylaxis (Pen VK 125mg po bid) in consultation with hematologist.**
- **Educate parents to have the infant seen and evaluated for sepsis when temperature is 101°Farenheit (38°Celsius) or greater.**
- **Report findings** to Nebraska Newborn Screening Program.

Condition Description: Individuals with sickle beta plus thalassemia are compound heterozygotes for the genes for hemoglobin S and beta plus thalassemia.

Clinical Expectations: Potential clinical problems include **mild hemolytic anemia, life-threatening infection, episodes of pain, and organ damage and organ failure.** Prompt treatment of infection and splenic sequestration is associated with decreased mortality in the first three years of life**.** It is strongly recommended that these children be followed by specialists in hemoglobinopathies as part of coordinated with their medical home. Ongoing specific health maintenance improves survival and reduces complications throughout life.

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.

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