Newborn Screening ACT Sheet
[FSC]
Hemoglobin SC Disease (HbSC)

Differential Diagnosis: Hemoglobin SC disease most likely.

Condition Description: A red cell disorder characterized by the presence of fetal hemoglobin (F) and hemoglobins S and C in the absence of Hb A. The hemoglobins are listed in order of the amount of hemoglobin present (F>S>C). This result is different from FAS which is consistent with sickle carrier.

You Should Take the Following Actions:

- Contact the family to inform them of the screening result.
- Consult a specialist in hemoglobinopathies; refer if needed.
- Evaluate infant and assess for splenomegaly.
- Initiate timely confirmatory/diagnostic testing as recommended by consultant.
- Initiate treatment as recommended by the consultant.
- Educate parents/caregivers regarding the risk of sepsis, the need for urgent evaluation for fever of ≥38.5° C (101° F) and signs and symptoms of splenic sequestration.
- Report findings to state newborn screening program.

Confirmation of Diagnosis: Hemoglobin separation by electrophoresis, isoelectric focusing or HPLC showing FSC. Family or DNA studies may be used to confirm genotype.

Clinical Expectations: Newborn infants are usually well. Hemolytic anemia and vaso-occlusive complications develop during infancy or early childhood. Complications include life-threatening infection, splenic sequestration, pneumonia, acute chest syndrome, pain episodes, aplastic crisis, dactylitis, priapism and stroke. Comprehensive care including family education, immunizations, prophylactic penicillin and prompt treatment of acute illness reduces morbidity and mortality.

Additional Information:
(Click on the name to take you to the website. Complete URLs are listed in the Appendix)

Grady Comprehensive Sickle Cell Center

Management and Therapy of Sickle Cell Disease

Sickle Cell Disease in Children and Adolescents: Diagnosis, Guidelines for Comprehensive Care, and Care Paths and Protocols for Management of Acute and Chronic Complications

American Academy of Pediatrics

Sickle Cell Disease Association

Referral *(local, state, regional and national)**:***

Comprehensive Sickle Cell Center Directory

Sickle Cell Information Center

(Funded in part through MCHB/HRSA/HHS grant #U22MC03957)

APPENDIX: Resources with Full URL Addresses

Additional Information:

Grady Comprehensive Sickle Cell Center http://scinfo.org/hemoglb.htm#SICKLE%20HEMOGLOBINS

Management and Therapy of Sickle Cell Disease
http://www.nhlbi.nih.gov/health/prof/blood/sickle/index.htm

Sickle Cell Disease in Children and Adolescents: Diagnosis, Guidelines for Comprehensive Care, and Care Paths and Protocols for Management of Acute and Chronic Complications.
http://www.dshs.state.tx.us/newborn/pdf/sedona02.pdf

American Academy of Pediatrics http://pediatrics.aappublications.org/cgi/content/full/109/3/526

Sickle Cell Disease Association http://www.sicklecelldisease.org/

Referral *(local, state, regional and national)**:***

Comprehensive Sickle Cell Center Directory http://www.rhofed.com/sickle/index.htm

Sickle Cell Information Center http://www.scinfo.org/clinics.htm

(Funded in part through MCHB/HRSA/HHS grant #U22MC03957)