Newborn Screening ACT Sheet
[FAS]
Sickle Cell Carrier (HbAS)

Differential Diagnosis: Sickle Cell Carrier. The hemoglobins are listed in order of the amount of hemoglobin present (F>A>S). This result is different from FS which is consistent with sickle cell anemia or sickle cell beta zero thalassemia (HbS B^0), or FSA which is consistent with sickle beta-plus thalassemia.

Condition Description: Generally benign genetic carrier state (trait) characterized by the presence of fetal hemoglobin (F), and hemoglobin A and S.

YOU SHOULD TAKE THE FOLLOWING ACTIONS:

- Contact the family to inform them of the screening result to offer education and reassurance that infants and young children do not have clinical problems related to the carrier state for hemoglobin S.
- Repeat screen or confirm result by alternate assay.
  - Order hemoglobin profile analysis (usually performed by electrophoresis).
- Offer family members referral for hemoglobin disorder testing and genetic counseling.
- Report findings to state newborn screening program.

Diagnostic Evaluation: Hemoglobin separation by electrophoresis, isoelectric focusing or high performance liquid chromatography (HPLC) shows FAS. DNA studies may be used to confirm genotype. Sickledex is not adequate for confirmation of the diagnosis.

Clinical Considerations: Newborn infants are usually normal. Prognosis is good, with a normal life expectancy. Carriers are at risk for having children affected by sickle cell disease. Older children and adults may have hematuria. Splenic infarction and an increased risk of sudden death associated with severe hypoxia, extreme physical exertion and dehydration have been reported.

Additional Information:

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 of America

Referral (local, state, regional and national):

Testing
Clinical Services
Comprehensive Sickle Cell Center Directory
Sickle Cell Information Center
Find Genetic Services

ACMG
American College of Medical Genetics and Genomics
Translating Genes Into Health®

LOCAL RESOURCES: Insert State newborn screening program web site links

State Resource site *(insert state newborn screening program website information)*

Name	
URL	
Comments	

Local Resource Site *(insert local and regional newborn screening website information)*

Name	
URL	
Comments	

APPENDIX: Resources with Full URL Addresses

Additional Information:

Grady Comprehensive Sickle Cell Center
http://www.scinfo.org/

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 of America
http://www.sicklecelldisease.org/

Referral (local, state, regional and national):

Testing
http://www.ncbi.nlm.nih.gov/sites/GeneTests/lab/clinical_disease_id/2028?db=genetests&country=United%20States

Clinical Services

Comprehensive Sickle Cell Center Directory
http://www.scinfo.org/index.php?option=com_content&view=article&id=197&Itemid=34

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

Find Genetic Services
http://www.acmg.net/GIS/Disclaimer.aspx

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