

## 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<sup>0</sup>), 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.

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#### ***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.
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**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](#)

Disclaimer: This guideline is designed primarily as an educational resource for clinicians to help them provide quality medical care. It should not be considered inclusive of all proper procedures and tests or exclusive of other procedures and tests that are reasonably directed to obtaining the same results. Adherence to this guideline does not necessarily ensure a successful medical outcome. In determining the propriety of any specific procedure or test, the clinician should apply his or her own professional judgment to the specific clinical circumstances presented by the individual patient or specimen. Clinicians are encouraged to document the reasons for the use of a particular procedure or test, whether or not it is in conformance with this guideline. Clinicians also are advised to take notice of the date this guideline was adopted, and to consider other medical and scientific information that become available after that date.

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*LOCAL RESOURCES:* Insert State newborn screening program web site links

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

Name	<input type="text"/>
URL	<input type="text"/>
Comments	<input type="text"/>

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

Name	<input type="text"/>
URL	<input type="text"/>
Comments	<input type="text"/>

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](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](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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