

Newborn Screening ACT Sheet

[Hemoglobin FAS]

Hemoglobin A/S

Differential Diagnosis: Hemoglobin S/Beta Plus (β^+) Thalassemia

Condition Description: Hemoglobin A/S is an inherited red blood cell disorder characterized by abnormal hemoglobin production. It is due to genetic changes in the beta hemoglobin chain. It is generally an asymptomatic genetic carrier state although adults may develop sickling and hypoxia at high altitude, severe dehydration, or extreme exertion.

You Should Take the Following Actions:

- Inform the family of the newborn screening result.
- Ascertain clinical status (newborns are expected to be asymptomatic).
- Evaluate the newborn.
- Provide family with basic information about sickle cell trait and sickle cell disease.
- Initiate confirmatory diagnostic testing and management as recommended by genetic counselor.
- Refer for genetic counseling.

- Report final diagnostic outcome to newborn screening program.

Diagnostic Evaluation: The hemoglobins are listed in order of the amount of hemoglobin present (F>A>S). [Isoelectric focusing, high performance liquid chromatography \(HPLC\) or capillary zone electrophoresis](#) is used to confirm the newborn screening result. [Complete blood count:](#) the CBC, red blood cell count, and reticulocyte count are normal.

Clinical Considerations: Newborns are asymptomatic with normal blood counts and have a normal life expectancy. Older children and adults may develop hematuria. Splenic infarction may occur with exertion and dehydration at high altitude. An increased risk of sudden death has been reported associated with severe hypoxia, extreme physical exertion, and dehydration. In adults there is an increased risk of chronic kidney disease and thromboembolism. Solubility testing (Sickledex) should not be used to confirm the diagnosis. Iron supplements should be avoided unless iron deficiency is documented.

Additional Information:

[How to Communicate Newborn Screening Results](#)

[Gene Reviews](#)

[Medline Plus](#)

[Condition Information for Families- HRSA Newborn Screening Clearinghouse](#)

[Sickle Cell Disease Association of America](#)

[Clinicaltrials.gov](#)

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

[Find A Hematologist \(Filter by Pediatric Hematology-Oncology\)](#)

[Find a Genetics Clinic Directory](#)

[Genetic Testing Registry](#)

[National Alliance of Sickle Cell Centers](#)

Local Resources *(Insert Local Website Links)*
State Resource Site *(Insert Website Information)*

Name	
URL	
Comments	

Local Resource Site *(Insert Website Information)*

Name	
URL	
Comments	

Appendix *(Resources with Full URL Addresses)*

Additional Information

How to Communicate Newborn Screening Results

- <https://bit.ly/NBSResultsHRSA>

Gene Reviews

- <https://www.ncbi.nlm.nih.gov/books/NBK1377/>

Medline Plus

- <https://medlineplus.gov/genetics/condition/sickle-cell-disease/>

Condition Information for Families-HRSA Newborn Screening Clearinghouse

- <https://newbornscreening.hrsa.gov/conditions/sickle-cell-trait>

Sickle Cell Disease Association of America

- <https://www.sicklecelldisease.org/>

Clinicaltrials.gov

- <https://clinicaltrials.gov/>

Referral (local, state, regional and national)

Find A Hematologist (Filter by Pediatric Hematology-Oncology)

- <https://www.hematology.org/education/patients/find-a-hematologist>

Find a Genetics Clinic Directory

- <https://clinics.acmg.net>

Genetic Testing Registry

- <https://www.ncbi.nlm.nih.gov/gtr/>

National Alliance of Sickle Cell Centers

- <https://sicklecellcenters.org/>