TEXAS Health and Human Services | Texas Department of State Health Services

Newborn Screening ACT Sheet

[FSA] Hemoglobin S/Beta+ Thalassemia (Hb Sβ+ Disease)

Differential Diagnosis

Hemoglobin FSC pattern on newborn screening is highly suggestive of sickle beta plus thalassemia. The hemoglobins are listed in order (F>S>A) of the amount of hemoglobin present.
This result is different from FAS, which is consistent with sickle cell trait.

Condition Description

A red blood cell disorder characterized by the presence of hemoglobin S and hemoglobin A. Individuals with sickle beta+ thalassemia (Hb Sβ+thal), a form of sickle cell disease, are compound heterozygotes for a hemoglobin S and a beta-thalassemia variant that produces hemoglobin A in the beta-globin genes.

Take the Following Actions

- Contact the family to inform them of the screening result;
- Consult with a pediatric hematologist (See attached list.);
- Perform physical exam on infant;
- Repeat newborn screen if second screen has not yet been done;
- Initiate treatment as recommended by the consultant;
- Educate parents/caregivers regarding the risk of sepsis, the need for urgent evaluation if fever of ≥ 101.5° F, or signs and symptoms of splenic sequestration; and
- Report findings to newborn screening program.

Confirmation of Diagnosis

Hemoglobin separation by electrophoresis, isoelectric focusing, or HPLC showing FSA. Family or DNA studies may be used to confirm genotype. β+thalassemia may not be detected by newborn screening DNA testing. Clinicians may choose to obtain further molecular diagnostic studies as indicated.

Clinical Expectations

Infants are usually normal at birth. Potential complications include life-threatening infection, splenic sequestration, acute chest syndrome, pain episodes, aplastic crisis, dactylitis, priapism, and stroke. Comprehensive care, including family education, immunizations, and prompt treatment of acute illness, reduces morbidity and mortality. Patients with Hb Sβ+thal have variable presentation due to wide variation in the amount of hemoglobin A produced by beta-thalassemia mutations.

Additional Information

Sickle Cell Disease Association of America
Centers for Disease Control and Prevention - Sickle Cell Disease
DSHS Sickle Cell Disease
National Institute of Health: Evidence-Based Management of Sickle Cell Disease
2019 sickle cell disease guidelines by the American Society of Hematology

Revised: 5/23/2024