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

Newborn Screening ACT Sheet

FE (HbEE or HbE/ Beta Zero Thalassemia)

EE or Hb E/ β0 Disease

Differential Diagnosis

Hemoglobin FE pattern on newborn screen is highly suggestive of homozygous hemoglobin E or hemoglobin E/beta zero (β0) thalassemia.

Condition Description

A red blood cell disorder characterized on the newborn screen by presence of fetal hemoglobin (F) and hemoglobin E in the absence of hemoglobin A. The hemoglobins are listed in order of the amount of hemoglobin present (F> E).

Take the Following Actions

- **Contact the family to inform them of the screening result;**
- **Evaluate infant, assess for splenomegaly, and do complete blood count (CBC) for Hb, and mean corpuscular volume (MCV) at the initial visit and at six months to differentiate hemoglobin EE from hemoglobin E/beta zero thalassemia;**
- **Repeat newborn screen if second screen has not yet been done;**
- **Contact a pediatric hematologist to determine need for further testing;**
- **Initiate timely confirmatory/diagnostic testing as recommended by consultant; and**
- **Report findings to newborn screening program.**

Diagnostic Evaluation

CBC and MCV. Hemoglobin separation by electrophoresis, isoelectric focusing (IEF), or high performance liquid chromatography (HPLC), which shows FE pattern. DNA studies will usually confirm genotype.

Clinical Expectations

Hemoglobin EE is clinically benign. Individuals with Hb EE are not anemic but have microcytosis and target cells on blood smear. Clinical expression of Hb E/β0 thalassemia is variable. Most individuals with Hb E/β0 thalassemia have moderately severe anemia, hepatosplenomegaly, intermittent jaundice, growth retardation, and overexpansion of the bone marrow. Severely affected individuals require lifelong transfusion, splenectomy, and treatment for iron overload.

Additional Information

American College of Medical Genetics and Genomics – Hemoglobin EE ACT Sheet
Utah Department of Health & Human Services – Hemoglobin C, D, and E Disorders
Kids Health – Thalassemias
U.S. National Library of Medicine, Medline Plus – Beta thalassemia

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