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

Newborn Screening ACT Sheet

FC (HbCC Disease or HbC/ Beta Zero Thalassemia)

HbC/ β0 Disease

Differential Diagnosis

Homozygous hemoglobin C, hemoglobin C/beta zero (β0) thalassemia, or hereditary persistence of fetal hemoglobin (Hb C/HPFH).

Condition Description

An uncommon red blood cell disorder characterized by presence of fetal hemoglobin (F) and hemoglobin C in the absence of hemoglobin A. The hemoglobins are listed in order of the amount of hemoglobin present (F>C).

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) and mean corpuscular volume (MCV);**
- **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, or high performance liquid chromatography (HPLC), shows FC pattern. DNA studies are used to confirm genotype.

Clinical Considerations

Infant is usually normal at birth. Hemoglobin CC is associated with a mild hemolytic anemia. Aplastic crises and gallstones may occur. Individuals with hemoglobin C/beta zero have a more severe anemia, splenomegaly, and rarely, bone changes. C-HPFH is clinically mild.

Additional Information

DSHS Hemoglobin C Disease
Medscape – Hemoglobin C Disease
Utah Department of Health & Human Services - Hemoglobin C, D, and E Disorders

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