

5.3 (old 3.13.3) Follow-up for Hemoglobin FE

GENERAL INFORMATION: Hemoglobin FE is seen in newborns with either Hb EE, a benign condition, or Hb E/beta⁰ thalassemia, which can be a serious transfusion-dependent disorder. Most infants with FE patterns will prove to be Hb EE.

POLICY: Written notification to parents and physicians will be made utilizing state-approved Parent and Doctor Letters (See Sections 11.1 & 11.2).

PROTOCOL:

Resp. Person	Action
ASC NBS Coord.	<ul style="list-style-type: none"> • Daily Reviews the Headline Case Report for Hb FE cases. • Within 48 Hours notifies the physician of record by phone of the screening result and requests blood specimens to be drawn from the infant and both parents (if possible) for confirmatory testing to determine whether the infant has a significant hemoglobin disorder. Assists in arranging for specimen collection at birth hospital lab or other collection site and sent to the Hemoglobin Reference Lab at Children’s Hospital & Research Center at Oakland (CHRCO). • When confirmatory results are received, only infants with Hb E/beta thalassemia results are referred to a CCS Sickle Cell Disease Center (SCDC). • Sends initial Doctor Letter #15 (FE) along with a copy of <i>Instructions for Collection, Handling, and Mailing Of Confirmatory Blood Specimens</i> (Section 5.8) and a copy of the pamphlet “<i>Why Retest For Hemoglobin E/beta thalassemia</i>”. • Sends <i>Instructions for Collection, Handling, And Mailing Of Confirmatory Blood Specimens (Section 5.8)</i>, as well as the shipping materials (cylinder and GSO label) to lab obtaining confirmatory specimen(s). • Within 2-3 days of first phone call to the primary care provider (PCP), follows up with the PCP to find out if family has been contacted. • After confirming that family has been notified, sends Parent Letter #11 (FE) and the pamphlet “<i>Why Retest For Hemoglobin E/beta thalassemia</i>”.

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	<ul style="list-style-type: none"> • If asked by the PCP to notify the family directly, after the contact is made, sends Parent Letter #12 (FE) and the pamphlet “<i>Why Retest For Hemoglobin E/beta thalassemia</i>”, confirming discussion with the parent and providing information about immediate follow-up care for the infant. • Continues to attempt to call family for notification and confirmatory testing if initially unsuccessful. • After one week of trying to contact the family, if the newborn’s PCP and/or Coordinator is unable to reach the family, sends Parent Letter #2 informing the family to call the newborn’s PCP or Coordinator regarding baby’s test results. Parent Letter #2 shall be sent by regular 1st class mail and a second copy sent by Certified Receipt mail requested to maximize receipt by parent. • If contact with family is still not made after one week of sending Parent Letter #2, makes arrangements for home visit by local health department public health nurse.
Hb Reference Lab	<ul style="list-style-type: none"> • Conducts confirmatory testing on liquid blood specimen(s). • Within 11 working days enters results in SIS and informs the ASC NBS Coordinator of the confirmatory test results (initial analysis) by fax, followed by a hard copy sent to the ASC NBS Coordinator and NBSB Hemoglobin Coordinator. Includes the following results: <ol style="list-style-type: none"> a. Separation of hemoglobins F, A, S, C, D, and E with relative concentrations for each hemoglobin on all specimens by cellulose acetate-citrate agar electrophoresis, isoelectric focusing, high pressure liquid chromatography, and/or DNA analysis as outlined in the Hb Reference Lab NBS vendor agreement scope of work or as approved by the NBSB. b. Hemogram on each suitable specimen which includes hemoglobin, hematocrit, mean corpuscular volume (MCV) and mean corpuscular hemoglobin (MCH). c. Free Erythrocyte Protoporphyrin (FEP) on specimens with microcytic hypochromic anemia. d. Quantitative Hb A2 when necessary to resolve phenotype. • Within 31 calendar days of receipt of specimen(s), enters results in SIS, faxes and mails reports on beta globin DNA analysis if done: <ul style="list-style-type: none"> • To rule out Hb E/beta⁰ thalassemia in the absence of specimens from both parents • to determine the beta thalassemia mutation if E/ beta⁰ thalassemia is confirmed in the infant • To rule out E/ beta⁰ thalassemia in a parent with electrophoresis results of Hb E. • to examine inconsistencies between thin layer isoelectric focusing profiles of newborn and parents.

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ASC NBS Coord.	<ul style="list-style-type: none">• Phones the NBS Hb Coordinator with any confirmatory results that are inconsistent with the NBS results prior to contacting the PCP.• Phones the PCP with the confirmatory test results.• For benign EE no referral to an SCDC is required. Sends follow-up Doctor letter #16a or 17a (EE) and “<i>Hemoglobin E</i>” pamphlet for the family.• Resolves confirmed EE cases in SIS as <u>Disorder</u> and in the “Disease” drop-down list select <u>Homozygous EE</u> with “Reliability” of “Certain”.• Assists with referral of infants with Hb E/beta thalassemia to an SCDC (see Protocol 7.2 <i>Referral to CCS Special Care Centers</i>). Sends follow-up Doctor letter #16b or 17b (E/Beta⁰ Thal.), #16c (FE; DNA pending), with the lab report to the PCP. Includes a copy of <i>Diagnosis And Treatment</i> form to be used for reporting the disease.• For Hb E/beta thalassemia, sends a copy of Doctor letter and lab result to the SCDC.• Resolves Hb E/beta thalassemia case in SIS when <i>Diagnosis & Treatment</i> form has been received, or the SCDC has completed the Hemoglobin Service Report in SIS indicating that infant has been seen and treatment is either initiated or not required. See Case Resolution Protocol 7.30.• Reports any missed cases, lost to follow-up cases or other unusual occurrences of potential significance to NBSB Nurse Consultant/ASC Contract Liaison.• Refers case to Child Protective Services as appropriate, and with approval of NBS Nurse Consultant/Contract Liaison (See 7.1 <i>Referral of Cases to Child Protective Services</i>).
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