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April 2016

Hemoglobinopathy Subcommittee
Newborn Screening Program
State of Wisconsin

Re: Barts Hemoglobin

Dear Colleague:

Your patient was recently found to have the presence of Fetal (F), Adult (A) and Barts hemoglobin on newborn screen. Fetal hemoglobin is composed of 2 alpha chains and 2 gamma chains. When there are an insufficient number of alpha chains made, 4 gamma chains combine to form Barts hemoglobin ($gamma_4$). The presence of Barts hemoglobin indicates that your patient likely has a form of ***alpha thalassemia*** (deficient production of alpha chains).

There are 4 α genes and therefore 4 possible clinical forms of α thalassemia. The severity of red cell changes associated with α thalassemia vary from:

- α thal silent carrier (no anemia) = 1α gene deleted
- α thal trait: mild hypochromic, microcytic anemia = 2 α genes deleted
- Hemoglobin H disease: moderately severe anemia with hemoglobin 6-9 g/dL = 3 α genes deleted
- Severe newborn hydrops fetalis associated with high levels of Barts hemoglobin = 4 α genes deleted

The presence of fetal, adult and Barts hemoglobin in your patient indicates the deletion of one, two, or (much less commonly) three α genes. As noted above, deletion of two α genes is associated with a mild hypochromic microcytic anemia. Such patients will have this present as a lifelong characteristic. Anemia due to α thal trait will **not** respond to iron therapy. Therefore, if one finds a mild hypochromic microcytic anemia on routine hemoglobin testing in a child who had Barts hemoglobin, ***we recommend a treatment trial of iron, 4 mg/kg/d of elemental iron, followed by a recheck of the CBC in 4 weeks. If the hemoglobin is normal on follow-up testing, the diagnosis of iron deficiency is confirmed.***

Note: Iron deficiency remains the most common cause of hypochromic, microcytic anemia and often coexists with thalassemia trait in infants who have a poor dietary intake of iron.

If there is no response to oral iron, there is no need for repeated courses of iron because the hypochromic, microcytic anemia found in a child with a newborn screen positive for Barts hemoglobin is most often due to α thalassemia trait. We recommend you ***strongly consider pediatric hematology consultation for patient/family education and counseling.*** More severe anemia (hemoglobin 6-9 gm/dL) suggests hemoglobin H disease and should be evaluated by a hematologist.

We hope this brief explanation of Barts hemoglobin is useful in your assessment of this child's health. If you have questions, please contact one of the hematologists listed below who are members of the Wisconsin Department of Public Health advisory group for hemoglobinopathy screening.

Sincerely,

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