RBC Band 3 Protein Reduction in Hereditary Spherocytosis

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Hereditary spherocytosis is a common genetic disorder that causes congenital hemolytic anemia through the production of structurally compromised spherical red blood cells (spherocytes). The severity of the disease varies widely; most cases are classified as moderate and present with symptoms such as anemia, jaundice, and splenomegaly. Depending on the level of severity, a diagnosis of hereditary spherocytosis can rule out other causes of hemolytic anemia.[1]

Hereditary spherocytosis is diagnosed based on family history and clinical features, along with laboratory tests. This test assesses for band 3 (solute carrier family 4 member 1 [SLC4A1]) protein, the most abundant transmembrane protein in human RBCs. Eosin-5-maleimide (EMA) dye is used to mark band 3 on intact RBCs. A reduction of fluorescence intensity will be observed in hereditary spherocytosis. A reduction in fluorescence intensity may also be observed in other rare disorders, specifically congenital dyserythropoietic anemia type II, Southeast Asian ovalocytosis, and hereditary pyropoikilocytosis.

Featured ARUP Testing

RBC Band 3 Protein Reduction in Hereditary Spherocytosis 2008460

Method: Qualitative Flow Cytometry

Use to confirm a diagnosis of hereditary spherocytosis when hemolytic anemia and spherocytes are present.

Test Interpretation

Sensitivity/Specificity

Clinical sensitivity: 93%

Analytic sensitivity/specificity: unknown

Results

Test Result	Interpretation
Normal	Normal staining of band 3 protein with EMA Does not suggest hereditary spherocytosis
Abnormal	Decreased staining of band 3 protein with EMA Provides evidence for hereditary spherocytosis (or more rarely, dyserythropoietic anemia type II, Southeast Asian ovalocytosis, or hereditary pyropoikilocytosis)
Equivocal	Unclear whether the level of band 3 protein staining with EMA is normal Insufficient evidence to support or negate a diagnosis of hereditary spherocytosis

Limitations

Recent transfusion (within the last three months) may affect test results; correlation with clinical and other laboratory findings is strongly recommended.

References

1. Narla J, Mohandas N. Red cell membrane disorders. *Int J Lab Hematol*. 2017;39 Suppl 1:47-52.