

# RBC Band 3 Protein Reduction in Hereditary Spherocytosis

Content Review: November 2021    Last Update: December 2023

Hereditary spherocytosis is a genetic disorder that causes congenital hemolytic anemia through the production of structurally compromised red blood cells (RBCs). 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.<sup>1</sup>

## Disease Overview

### Prevalence

1/2,000 in individuals of northern European descent<sup>2</sup>

## Genetics

### Genes

*ANK1, EPB42, SLC4A1, SPTA1, SPTB*

### Inheritance

- Autosomal dominant: 75%<sup>1</sup>
- Autosomal recessive: 25%<sup>1</sup>

### Penetrance

Variable

### Structure/Function

- Chromosomal location: 17q21.31
- Provides structure for the red cell cytoskeleton

## Test Interpretation

### Sensitivity/Specificity

Clinical sensitivity: 93%

Analytic sensitivity/specificity: unknown

## 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.

# Results

Test Result	Interpretation
Normal	Normal staining of band 3 protein with EMA does not suggest hereditary spherocytosis
Abnormal <sup>a</sup>	Decreased staining of band 3 protein with EMA provides evidence for hereditary spherocytosis
Equivocal	Insufficient evidence to support or negate a diagnosis of hereditary spherocytosis Unclear whether the level of band 3 protein staining is normal

<sup>a</sup>Three other rare disorders may be associated with a positive result for this test: congenital dyserythropoietic anemia type 2, Southeast Asian ovalocytosis, and hereditary pyropoikilocytosis.

EMA, eosin-5-maleimide

## 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.
2. Zamora EA, Schaefer CA. [Hereditary spherocytosis](#). 2021. In: StatPearls. StatPearls Publishing; 2021.

## Related Information

### [Hemolytic Anemias](#)

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