

Overview

Useful For

Interpretation of the results for the evaluation of hemolytic anemia

Evaluation of lifelong or inherited hemolytic anemias, including red cell membrane disorders, unstable or abnormal hemoglobin variants, and red cell enzyme disorders

Special Instructions

- [Metabolic Hematology Patient Information](#)

Method Name

Only orderable as part of a profile. For more information see HAEV1 / Hemolytic Anemia Evaluation, Blood.

Medical Interpretation

NY State Available

Yes

Specimen

Specimen Type

Whole Blood ACD-B

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Whole Blood ACD-B	Refrigerated	72 hours	

Clinical & Interpretive

Clinical Information

Hemolytic anemia (HA) is characterized by increased red cell destruction and a decreased red cell life span. Patients usually have decreased hemoglobin concentration, hematocrit, and red blood cell count, but some can have compensated disorders, and symptoms such as reticulocytosis, pigmented gallstones, and decreased haptoglobin are factors that raise clinical suspicion. Blood smear abnormalities may include variable amounts of poikilocytosis including spherocytes, elliptocytes, schistocytes, stomatocytes, echinocytes, polychromasia, basophilic stippling, and target cells. Osmotic fragility can be increased due to the presence of spherocytes. These are all nonspecific features that can be present in both hereditary and acquired hemolytic disorders.

Inherited hemolytic disorders may include red cell membrane disorders, red cell enzyme defects, or abnormalities in the hemoglobin molecule in the red cell. This panel assesses for possible causes of congenital/hereditary causes of hemolytic anemia and does not evaluate for acquired causes. Therefore, the anemia should be lifelong or familial in nature. Examples of acquired HA (which should be excluded prior to ordering this panel) include: autoimmune HA (Coombs-positive HA, Coombs-negative autoimmune HA), cold agglutinin disease, paroxysmal nocturnal hemoglobinuria, paroxysmal cold hemoglobinuria, mechanical hemolysis (aortic stenosis or prosthetic heart valves), disseminated intravascular coagulation/thrombotic microangiopathy, and drug-induced HA.

This consultation evaluates for a hereditary cause of increased red cell destruction and includes testing for red cell membrane disorders, such as hereditary spherocytosis and hereditary pyropoikilocytosis, hemoglobinopathies, and red cell enzyme abnormalities.

This panel is of limited use in patients with a history of recent transfusion and should be ordered as remote a date from transfusion as possible in those patients who are chronically transfused.

Reference Values

Only orderable as part of a profile. For more information see HAEV1 / Hemolytic Anemia Evaluation, Blood.

Definitive results and an interpretive report will be provided.

Interpretation

A hematopathologist expert in these disorders evaluates the case, appropriate tests are performed, and an interpretive report is issued.

Cautions

Recent transfusion may cause unreliable results.

A normal shipping control for osmotic fragility (OF) is necessary to exclude false-positive results due to preanalytical artifact. OF and eosin-5-maleimide (EMA) binding testing will be canceled if no shipping control is received or if the shipping control is abnormal.

This panel is most effectively interpreted in the context of clinical information and the peripheral blood morphology. Fill out the [Metabolic Hematology Patient Information](#) sheet available in Special Instructions to maximize the interpretive capabilities of the panel.

This group of tests should not ordinarily be requested in patients who are likely to have immune hemolytic anemia (HA), such as that due to either warm or cold antibodies or to paroxysmal nocturnal hemoglobinurias. Coombs tests, tests for cold agglutinins, sucrose hemolysis, and Hams and Crosby tests are not part of the HA evaluation. In general, the foregoing tests should have been performed and found to be negative prior to requesting an HA evaluation. Since Wilson disease is another rare cause for acute intermittent hemolysis, testing for Wilson disease also may be appropriate prior to requesting an HA evaluation.

Clinical Reference

1. Steiner LA, Gallagher PG. Erythrocyte disorders in the perinatal period. *Semin Perinatol.* 2007;31(4):254-261. doi:10.1053/j.semperi.2007.05.003
2. Beutler E. Glucose-6-phosphate dehydrogenase deficiency and other enzyme abnormalities. In: Beutler E, Lichtman MA, Coller BS, Kipps TJ, eds. *Hematology.* 5th ed. McGraw-Hill Book Company; 1995: 564-581

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4. King MJ, Garcon L, Hoyer JD, et al. International Council for Standardization in Haematology. ICSH guidelines for the laboratory diagnosis of nonimmune hereditary red cell membrane disorders. *Int J Lab Hematol*. 2015;37(3):304-325. doi:10.1111/ijlh.12335
5. Lux SE. Anatomy of the red cell membrane skeleton: unanswered questions. *Blood*. 2016;127(2):187-199 doi:10.1182/blood-2014-12-512772
6. Gallagher PG. Abnormalities of the erythrocyte membrane. *Pediatr Clin North Am*. 2013;60(6):1349-1362. doi:10.1016/j.pcl.2013.09.001
7. Bianchi P, Fermo E, Vercellati C, et al. Diagnostic power of laboratory tests for hereditary spherocytosis: a comparison study in 150 patients grouped according to molecular and clinical characteristics. *Haematologica*. 2012;97(4):516-523. doi:10.3324/haematol.2011.052845
8. Cappellini MD, Fiorelli G. Glucose-6-phosphate dehydrogenase deficiency. *Lancet*. 2008;371(9606):64-74
9. Glader B. Hereditary hemolytic anemias due to red blood cell enzyme disorders. In: Greer JP, Arber DA, Glader B, et al, eds. *Wintrobe's Clinical Hematology*. 13th ed. Wolters Kluwer/Lippincott, Williams and Wilkins; 2014:728
10. Gallagher PG. Diagnosis and management of rare congenital nonimmune hemolytic disease. *Hematology Am Soc Hematol Educ Program*. 2015; 392-399. doi:10.1182/asheducation-2015.1.39211
11. Koralkova P, van Solinge WW, van Wijk R. Rare hereditary red blood cell enzymopathies associated with hemolytic anemia- pathophysiology, clinical aspects, and laboratory diagnosis. *Int J Lab Hematol*. 2014;36(3):388-397. doi:10.1111/ijlh.12223

Performance

Method Description

A hematopathologist who is an expert in these disorders evaluates the case and an interpretive report is issued.

PDF Report

No

Day(s) Performed

Monday through Friday

Report Available

2 to 10 days

Performing Laboratory Location

Rochester

Fees & Codes

Fees

- Authorized users can sign in to [Test Prices](#) for detailed fee information.

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- Clients without access to Test Prices can contact [Customer Service](#) 24 hours a day, seven days a week.
 - Prospective clients should contact their account representative. For assistance, contact [Customer Service](#).

Test Classification

Not Applicable

CPT Code Information

83020-26

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
HAEVI	Hemolytic Anemia Interpretation	59466-3

Result ID	Test Result Name	Result LOINC® Value
608427	Hemolytic Anemia Interpretation	59466-3
608441	Reviewed By	18771-6