Test Definition: HKC

Hexokinase, B

Overview

Useful For

The evaluation of individuals with Coombs-negative chronic hemolysis

Method Name

Only available as part of a profile. For more information see:

-HAEV1 / Hemolytic Anemia Evaluation, Blood

-EEEV1 / Red Blood Cell (RBC) Enzyme Evaluation, Blood

Kinetic Spectrophotometry (KS)

NY State Available

Yes

Specimen

Specimen Type

Whole Blood ACD-B

Specimen Required

Only available as part of a profile. For more information see:

-HAEV1 / Hemolytic Anemia Evaluation, Blood

-EEEV1 / Red Blood Cell (RBC) Enzyme Evaluation, Blood

Reject Due To

Gross hemolysis	Reject

Specimen Minimum Volume

1 mL

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Whole Blood ACD-B	Refrigerated (preferred)	20 days	

Clinical & Interpretive

Clinical Information

Hexokinase (HK) is the first enzymatic step in glycolysis, converting glucose to glucose-6-phosphate. Hexokinase deficiency (OMIM 235700) is a rare cause of chronic nonspherocytic hemolytic anemia and its inheritance is autosomal recessive. Clinically significant HK deficiency manifests in early onset anemia with variable severity ranging from mild to severe. Some patients show neurologic impairment of which the mechanism is unclear.

Reference Values

Only available as part of a profile. For more information see:

-HAEV1 / Hemolytic Anemia Evaluation, Blood

-EEEV1 / Red Blood Cell (RBC) Enzyme Evaluation, Blood

> or =12 months: 0.7-1.7 U/g Hb

Reference values have not been established for patients who are less than12 months of age.

Interpretation

Clinical correlation or genetic confirmation may be required to establish hexokinase (HK) deficiency as a cause of hemolytic anemia as the assayed activity level in confirmed cases can vary from markedly decreased to borderline normal levels due to a compensated increase in enzyme by reticulocytes. Comparison of hexokinase activity levels to other RBC enzyme activity can be very useful. Heterozygotes have moderately decreased to low normal HK levels and are expected to be clinically unaffected.

Increased HK activity may be seen when reticulocytes are increased and is not supportive of a diagnosis of HK deficiency.

Cautions

Recent transfusion may mask the patient’s intrinsic enzyme activity and cause unreliable results.

Hexokinase (HK) activity level can vary from markedly decreased to borderline normal levels in affected individuals due to a compensated increase in enzyme by reticulocytes. Comparison of hexokinase activity levels to other RBC enzyme

activity can be very useful.

Clinical Reference

1. Koralkova P, Mojzikova R, van Oirschot B, et al: Molecular characterization of six new cases of red blood cell hexokinase deficiency yields four novel mutations in HK1. Blood Cells Mol Dis. 2016 Jul;59:71-76

2. 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;36:388-397

Performance

Method Description

Hexokinase catalyzes the reaction of adenosine triphosphate (ATP) and glucose to glucose 6-phosphate (G6P) and adenosine diphosphate (ADP). In this assay the formation of G6P is measured by linking its further oxidation to 6-phosphogluconate (6-PG) to the reduction of nicotinamide adenine dinucleotide phosphate (NADP) through the glucose-6-phosphate dehydrogenase (G6PD) reaction. The increase in absorbance, which occurs as NADP(+) is reduced to NADPH, is measured spectrophotometrically at 340 nm on an automated chemistry analyzer.(Beutler E: Red Cell Metabolism: A Manual of Biochemical Methods. 3rd ed. Grune and Stratton; 1984:38-40; van Solinge WW, van Wijk: Enzymes of the red blood cell. In: Rifai N, Horvath AR, Wittwer CT: eds. Tietz Textbook of Clinical Chemistry and Molecular Diagnostics. 6th ed. Elsevier; 2018:chap 30)

PDF Report

No

Specimen Retention Time

28 days

Performing Laboratory Location

Rochester

Fees & Codes

Test Classification

This test was developed, and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. This test has not been cleared or approved by the US Food and Drug Administration.

CPT Code Information

82657

LOINC® Information

Test ID	Test Order Name	Order LOINC Value
HKC	Hexokinase, B	49216-5

Result ID	Reporting Name	LOINC®
HKCL	Hexokinase, B	49216-5