MAYO CLINIC LABORATORIES

# Test Definition: HKC

Hexokinase Enzyme Activity, Blood
(Non-Orderable)

## Overview

### Useful For

**Only orderable as part of a profile**. For more information see:
HAEV1 / Hemolytic Anemia Evaluation, Blood
EEEV1 / Red Blood Cell (RBC) Enzyme Evaluation, Blood

Evaluating individuals with Coombs-negative chronic hemolysis as a part of a profile

### Method Name

Only orderable 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 orderable 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 |
| Fully clotted | Reject |

### Specimen Stability Information

| Specimen Type | Temperature | Time | Special Container |
|---|---|---|---|
| Whole Blood ACD-B | Refrigerated | 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 orderable 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 younger than 12 months.

### 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 HK activity levels to other red blood cell enzyme activity can be very useful. Heterozygous individuals 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 HK activity levels to other red blood cell 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;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(3):388-397

## Performance

### Method Description

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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 to the reduction of nicotinamide adenine dinucleotide phosphate (NADP[+]) through the glucose-6-phosphate dehydrogenase 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; Rab MAE, van Wijk R. Enzymes of the red blood cell. In: Rifai N, Chiu RWK, Young I, Burnham CAD, Wittwer CT, eds. Tietz Textbook of Laboratory Medicine. 7th ed. Elsevier; 2023:chap 78)

### PDF Report
No

### Day(s) Performed
Weekly

### Report Available
5 days

### Specimen Retention Time
28 days

### Performing Laboratory Location
Mayo Clinic Laboratories - Rochester Main Campus

## Fees & Codes

### Fees
- Authorized users can sign in to Test Prices for detailed fee information.
- 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
This test was developed and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. It 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 | Test Result Name | Result LOINC® Value |
|---|---|---|

| | | |
|---|---|---|
| HKCL | Hexokinase, B | 49216-5 |