

## Overview

### Useful For

Identifying defects of red cell enzyme metabolism

Evaluating patients with hemolytic anemia

### Profile Information

Test ID	Reporting Name	Available Separately	Always Performed
EEEV	Erythrocyte Enzyme Interpretation	No	Yes
G6PD	G-6-PD, QN, RBC	Yes	Yes
PK	Pyruvate Kinase, RBC	Yes	Yes
GPI	Glucose Phosphate Isomerase, B	Yes	Yes
HEXK	Hexokinase, B	No	Yes

### Reflex Tests

Test ID	Reporting Name	Available Separately	Always Performed
GLTI	Glutathione, B	No	No
RBCE	Reflexed RBC Enzymes	No	No

### Testing Algorithm

This is a consultative evaluation in which the case will be evaluated at Mayo Clinic Laboratories, the appropriate tests performed at an additional charge, and the results interpreted.

**Note:** RBCE / Reflexed RBC Enzymes, Blood includes: adenylate kinase, phosphofructokinase, phosphoglycerate kinase, triosephosphate isomerase, and pyrimidine 5'nucleotidase.

See [Benign Hematology Evaluation Comparison](#) in Special Instructions.

### Special Instructions

- [Metabolic Hematology Patient Information](#)
- [Benign Hematology Evaluation Comparison](#)

### Method Name

EEEV: Consultative Interpretation

G6PD, GPI, PK, GLTI, HEXK, RBCE: Kinetic Spectrophotometry (KS)

### NY State Available

Yes

**Specimen**
**Specimen Type**

Whole Blood ACD-B

**Specimen Required**
**Container/Tube:** Yellow top (ACD solution B)

**Specimen Volume:** 12 mL

**Collection Instructions:** Do not transfer blood to other containers.

**Forms**

1. [Metabolic Hematology Patient Information](#) (T810) is available in Special Instructions
2. If not ordering electronically, complete, print, and send a [Benign Hematology Test Request Form](#) (T755) with the specimen.

**Specimen Minimum Volume**

5 mL

**Reject Due To**

Gross hemolysis	Reject
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**Specimen Stability Information**

Specimen Type	Temperature	Time	Special Container
Whole Blood ACD-B	Refrigerated	8 days	

**Clinical and Interpretive**
**Clinical Information**

All enzyme defects, including erythrocyte enzyme errors, are inherited; some are sex-linked and located on the X chromosome. Some family members have no hematologic abnormalities, while others have a hemolytic anemia. For a number of RBC enzyme defects (eg, deficiencies of hexokinase, glucose phosphate isomerase, pyruvate kinase), the sole clinical manifestation is hemolytic anemia. Glucose-6-phosphate dehydrogenase deficiency is the most common metabolic error of the red cell and presents with acute hemolytic anemia in response to oxidant stress (eg, drugs, acute infections, fava bean ingestion).

This is a consultative evaluation looking at red cell enzyme defects as the cause for early red cell destruction.

**Reference Values**

Definitive results and an interpretive report will be provided.

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## Interpretation

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

## Cautions

No significant cautionary statements

## Clinical Reference

Beutler E: Glucose-6-phosphate dehydrogenase deficiency and other enzyme abnormalities. In Hematology. Fifth edition. Edited by E Beutler, MA Lichtmann, BS Collier, TJ Kipps. New York, McGraw-Hill Book Company, 1995, pp 564-581

## Performance

### Method Description

Erythrocyte Enzyme Interpretation:

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

Glucose-6-Phosphate Dehydrogenase (G6PD):

G6PD in a hemolysate catalyzes the oxidation of glucose-6-phosphate to 6-phosphogluconate. Concomitantly, nicotinamide adenine dinucleotide phosphate (NADP) is changed to its reduced form (nicotinamide adenine dinucleotide phosphate-oxidase: NADPH), a reaction measured spectrophotometrically. (Beutler E: Red Cell Metabolism: A Manual of Biochemical Methods. Third edition. New York, Grune and Stratton, 1984, pp 68-71)

Pyruvate Kinase:

A red cell hemolysate is incubated with adenosine diphosphate and phosphoenolpyruvate. The amount of pyruvate formed is quantitated by adding lactic dehydrogenase and reduced nicotinamide adenine di-nucleotide and measuring the rate of decrease in absorbance at 340 nm. (Beutler E: Red Cell Metabolism: A Manual of Biochemical Methods. Third edition. New York, Grune and Stratton, 1984, pp 68-71)

Glucose Phosphate Isomerase:

Washed erythrocytes are hemolyzed and the hemolysate is mixed with glucose, adenosine triphosphate (ATP), glucose-6-phosphate dehydrogenase, and nicotinamide adenine dinucleotide phosphate (NADP). The reduction of NADP is measured spectrophotometrically and is proportional to the enzymatic conversion of ATP and glucose to glucose-6-phosphate. (Beutler E: Red Cell Metabolism: A Manual of Biochemical Methods. Third edition. New York, Grune and Stratton, 1984, pp 40-42)

Hexokinase:

Hexokinase (in the presence of magnesium) catalyzes the reaction of ATP and glucose to G-6-P and ADP. In this assay the formation of glucose-6-phosphate (G-6-P) is measured by linking its further oxidation to 6-phosphogluconate (6-PG) to the reduction of NADP through the glucose-6-phosphate dehydrogenase (G-6-PD) reaction. The increase in absorbance which occurs as NADP+ is reduced is measured at 340 nm. (Beutler E: Red cell metabolism: A Manual of Biochemical Methods. Third edition. Grune and Stratton, New York, 1984, pp 38-40)

Glutathione:

Virtually all of the nonprotein sulfhydryl of red cells is in the form of reduced glutathione (GSH). 5,5'-dithiobis (2-nitrobenzoic acid) is a disulfide compound which is readily reduced by sulfhydryl compounds, forming a highly colored yellow anion. The absorbance of this resultant yellow substance is measured by 412 nm and compared to that of a known standard. (Beutler E: Red cell metabolism. In A Manual of Biochemical Methods, Second edition, Grune and Stratton, 1984)

**PDF Report**

No

**Day(s) and Time(s) Test Performed**

Monday through Friday; Varies

**Analytic Time**

2-10 days (not reported Saturday or Sunday)

**Maximum Laboratory Time**

13 days

**Specimen Retention Time**

14 days

**Performing Laboratory Location**

Rochester

**Fees and 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 Regional Manager. For assistance, contact [Customer Service](#).

**Test Classification**

See Individual Test IDs

**CPT Code Information**

82955-G-6-PD

84087-Glucose phosphate isomerase

84220-Pyruvate kinase

82657-Hexokinase

82978-Glutathione (if appropriate)

83915-RBC Enzymes (if appropriate)

**LOINC® Information**



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Test ID	Test Order Name	Order LOINC Value
EEEVP	RBC Enzyme Evaluation	In Process

Result ID	Test Result Name	Result LOINC Value
21065	Erythrocyte Enzyme Interpretation	59466-3
G6PD_	G-6-PD, QN, RBC	32546-4
GPI_	Glucose Phosphate Isomerase, B	44050-3
HEXK_	Hexokinase, B	49216-5
PK_	Pyruvate Kinase, RBC	32552-2