

Overview**Useful For**

Confirming cases of suspected methemoglobin reductase (cytochrome b5 reductase) deficiency

Functional studies in families with methemoglobin reductase (cytochrome b5 reductase) deficiency

Method Name

Kinetic Spectrophotometry (KS)

NY State Available

Yes

Specimen**Specimen Type**

Whole Blood ACD-B

Specimen Required**Container/Tube:**

Preferred: Yellow top (ACD)

Acceptable: Lavender top (EDTA)

Specimen Volume: 6 mL

Collection Instructions: Do not transfer blood to other containers.

Additional Information: Patient's age is required.

Forms

If not ordering electronically, complete, print, and send a [Benign Hematology Test Request Form](#) (T755) with the specimen.

Specimen Minimum Volume

1 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	22 days	

Clinical and Interpretive

Clinical Information

Methemoglobin reductase, also called "diaphorase," and more properly called cytochrome b5 reductase, is the enzyme within the erythrocyte that maintains hemoglobin in the reduced (non-methemoglobin) state.

Persons who are heterozygous for methemoglobin reductase mutations have no clinical or laboratory abnormalities, are not cyanotic, and have normal methemoglobin concentrations in their blood. However, they hold an increased risk for more severely symptomatic acute episodes of methemoglobinemia with exposure to inducing agents.

Persons who are homozygous for methemoglobin reductase mutations have normal arterial oxygen saturation but have varying quantities of methemoglobin in their blood, generally 15% to 20%, and are quite cyanotic. Paradoxically, homozygotes typically have normal blood counts; the condition only rarely causes polycythemia. The presence of methemoglobin shifts the hemoglobin-O₂ dissociation curve to the right, so that although the transport of oxygen is diminished, the delivery of oxygen to tissues is normal. Because of the chronicity, the homozygous condition is usually compensated and therefore quite benign, but may cause concern to parents of affected children, be a cosmetic embarrassment to the children, and alarm the attending physician. The cyanosis may be treated with methylene blue.

Reference Values

> or =12 months: 6.6-13.3 U/g Hb

Reference values have not been established for patients who are <12 months of age.

Interpretation

Methemoglobin reductase (cytochrome b5 reductase) activity in neonates (0-6 weeks) is normally 60% of the normal adult value. Adult values are attained by 2 to 3 months of age.

Heterozygotes have results slightly lower than the reference range. Homozygotes demonstrate little to no methemoglobin reductase activity and increased levels of methemoglobin.

Cautions

Individuals who are glucose-6-phosphate-dehydrogenase (G-6-PD) deficient are not candidates for methylene blue therapy. Administration of methylene blue to such persons will cause hemolysis or methemoglobin formation.

Clinical Reference

Beutler E: Methemoglobinemia and other causes of cyanosis. In Williams Hematology. Sixth edition. Edited by E Beutler, M Lichtman, WJ Williams, TJ Kipps. New York, McGraw-Hill Book Company, 2001, p 611

Performance

Method Description

Methemoglobin reductase (cytochrome b5 reductase) catalyzes the NADH-linked reduction of several substrates, including ferricyanide. The activity at 30 degrees C is followed spectrophotometrically by measuring the oxidation of NADH at 340 nm.(Fairbanks VF, Klee GG: Biochemical aspects of hematology. In Tietz Textbook of Clinical Chemistry. Third edition. Edited by CA Burtis, ER Ashwood. Philadelphia, WB Saunders Company, 1999, pp 1647-1648)

PDF Report

No

Day(s) and Time(s) Test Performed

Tuesday, Thursday

Analytic Time

5 days (not reported Saturday or Sunday)

Maximum Laboratory Time

6 days

Specimen Retention Time

7 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

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 U.S. Food and Drug Administration.

CPT Code Information

82657

LOINC® Information

Test ID	Test Order Name	Order LOINC Value
METR	Methemoglobin Reductase, B	32703-1

Result ID	Test Result Name	Result LOINC Value
9322	Methemoglobin Reductase, B	32703-1