MAYO CLINIC LABORATORIES

Test Definition: Q10

Coenzyme Q10, Reduced and Total, Plasma

Overview

Useful For

Diagnosis of primary coenzyme Q10 (CoQ10) deficiencies in some patients who are not supplemented with CoQ10

Diagnosis of CoQ10 deficiency in mitochondrial disorders

Monitoring CoQ10 status during treatment of various degenerative conditions, including Parkinson and Alzheimer diseases

This test is **not useful** for distinguishing primary CoQ10 deficiencies from acquired CoQ10 deficiencies.

Genetics Test Information

This test is appropriate for the diagnosis of secondary coenzyme Q10 (CoQ10) deficiency and for some patients with primary CoQ10 deficiency who are not supplemented with CoQ10. It is also used to monitor CoQ10 status in patients with mitochondrial cytopathies, patients receiving statin therapy, or during treatment of various degenerative conditions including Parkinson and Alzheimer diseases.

Method Name

High-Performance Liquid Chromatography (HPLC) with Electrochemical Detection

NY State Available

Yes

Specimen

Specimen Type

Plasma Heparin

Ordering Guidance

This test provides both reduced and total coenzyme Q10 (CoQ10). For assessment of total only, order TQ10 / Coenzyme Q10, Total, Plasma.

The level of oxidized CoQ10 is affected in specimens with even slight amounts of hemolysis; however, the total Q10 level remains constant. Hemolyzed specimens can be analyzed for total Q10 using TQ10 / Coenzyme Q10, Total, Plasma.

The most reliable test for the diagnosis of primary defects in ubiquinone (ie, CoQ10) biosynthesis is direct measurement of CoQ10 in muscle.

Shipping Instructions

If possible, **do not** send other tests ordered on same vial of plasma. In doing so, the other tests may have increased

turnaround time due to the strict frozen criteria of this assay.

Necessary Information

Patient's age is required.

Specimen Required

Patient Preparation: Fasting (8 hours)
Collection Container/Tube: Green top (lithium or sodium heparin)
Submission Container/Tube: Plastic vial
Specimen Volume: 0.5 mL
Collection Instructions:
1. Immediately after collection, place specimen on wet ice. Maintain on wet ice and process within 3 hours of collection.
2. Centrifuge, aliquot plasma into plastic vial, and freeze immediately.

Forms

If not ordering electronically, complete, print, and send a Biochemical Genetics Test Request (T798) with the specimen.

Specimen Minimum Volume

0.3 mL

Reject Due To

Gross hemolysis	Reject
Gross lipemia	Reject
Gross icterus	OK

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Plasma Heparin	Frozen (preferred)	14 days	
	Refrigerated	8 hours	

Clinical & Interpretive

Clinical Information

Coenzyme Q10 (CoQ10) is an essential cofactor in the mitochondrial respiratory chain responsible for oxidative phosphorylation where it functions as an electron carrier and acts as an antioxidant. It is found in all cell membranes and is carried by lipoproteins in the circulation. Approximately 60% of CoQ10 is associated with low-density lipoprotein (LDL), 25% with high-density lipoprotein, and 15% with other lipoproteins. CoQ10 is present in the body in both the reduced and oxidized forms, with the antioxidant activity of CoQ10 dependent on both its concentration and its reduction-oxidation (redox) status.

CoQ10 deficiencies, which are clinically and genetically diverse, can occur due to defects in genes involved in the biosynthesis of ubiquinone (primary CoQ10 deficiency) or due to other causes, such as mitochondrial disorders

(secondary or CoQ10 deficiency).

Five major clinical phenotypes of CoQ10 deficiency have been described:
-Encephalomyopathy (elevated serum creatine kinase [CK], recurrent myoglobinuria, lactic acidosis)
-Cerebellar ataxia and atrophy (neuropathy, hypogonadism)
-Severe multisystemic infant form (nystagmus, optic atrophy, sensorineural hearing loss, dystonia, rapidly progressing nephropathy)
-Nephropathy, steroid resistant nephrotic syndrome leading to end stage kidney disease
-Isolated myopathy (exercise intolerance, fatigue, elevated serum CK)

Treatment with CoQ10 in patients with mitochondrial cytopathies can improve mitochondrial respiration in both brain and skeletal muscle.

CoQ10 has been implicated in other disease processes, including diabetes, neurodegenerative conditions such as Parkinson and Alzheimer diseases, as well as in aging and oxidative stress. CoQ10 may also play a role in hydroxymethylglutaryl-CoA reductase inhibitor (statin) therapy and may be relevant to statin-induced myalgia. Additionally, the redox status of CoQ10 may be a useful early marker for the detection of oxidative LDL modification.

Reference Values

COENZYME Q10 (CoQ10) REDUCED
<18 years: 320-1376 mcg/L
> or =18 years: 415-1480 mcg/L

CoQ10 TOTAL
<18 years: 320-1558 mcg/L
> or =18 years: 433-1532 mcg/L

CoQ10 % REDUCED
<18 years: 93-100%
> or =18 years: 92-98%

Miles MV, Horn PS, Tang PH, et al. Age-related changes in plasma coenzyme Q10 concentrations and redox state in apparently healthy children and adults. Clin Chim Acta. 2004;34:139-144

Interpretation

Abnormal results are reported with a detailed interpretation including an overview of the results and their significance, a correlation to available clinical information provided with the specimen, differential diagnosis, and recommendations for additional testing when indicated and available.

Cautions

Coenzyme Q10 is sensitive to specimen handling and transport temperature. Failure to follow the specimen handling and transportation recommendations may lead to false-positive results.

Clinical Reference

1. Salviati L, Trevisson E, Agosto C, Doimo M, Navas P. Primary coenzyme Q10 deficiency overview. In: Adam MP, Mirzaa GM, Pagon RA, et al. eds. GeneReviews [Internet]. University of Washington, Seattle; 2017. Updated June 8, 2023.

Accessed November 1, 2023. Available at www.ncbi.nlm.nih.gov/books/NBK410087/
2. Desbats MA, Lunardi G, Doimo M, Trevisson E, Salviati L. Genetic bases and clinical manifestations of coenzyme Q10 (CoQ 10) deficiency. J Inherit Metab Dis. 2015;38(1):145-56. doi:10.1007/s10545-014-9749-9
3. Littarru GP, Tiano L. Clinical aspects of coenzyme Q10: An update. Nutrition. 2010;26:250-254
4. Hargreaves I, Heaton RA, Mantle D. Disorders of human coenzyme Q10 metabolism: An overview. Int J Mol Sci. 2020;21(18):6695. doi:10.3390/ijms21186695
5. Banach M, Serban C, Ursoniu S, et al. Statin therapy and plasma coenzyme Q10 concentrations-A systematic review and meta-analysis of placebo-controlled trials. Pharmacol Res. 2015;99:329-336. doi:10.1016/j.phrs.2015.07.008
6. Emmanuele V, Lopez LC, Berardo A, et al. Heterogeneity of coenzyme Q10 deficiency: patient study and literature review. Arch Neurol. 2012;69(8):978-983. doi:10.1001/archneurol.2012.206

Performance

Method Description

Coenzyme Q10 (CoQ10) is extracted from plasma with cold 1-propanol containing coenzyme Q9 as an internal standard. An aliquot of the lipid extract is fractionated by high-performance liquid chromatography (HPLC). The separated coenzyme Q10 reduced and coenzyme Q10 oxidized are quantified by measurement of selective coulometric response emerging from the chromatographic column.(Tang PH, Miles MV, DeGrauw A, Hershey A, Pesce A. HPLC analysis of reduced and oxidized coenzyme Q[10] in human plasma. Clin Chem. 2001;47[2]:256-265; Claessens AJ, Yeung CK, Risler LJ, Phillips BR, Himmelfarb J, Shen DD. Rapid and sensitive analysis of reduced and oxidized coenzyme Q10 in human plasma by ultra performance liquid chromatography-tandem mass spectrometry and application to studies in healthy human subjects. Ann Clin Biochem. 2016;53[Pt 2]:265-273. doi:10.1177/0004563215593097)

PDF Report

No

Day(s) Performed

Monday through Friday

Report Available

3 to 5 days

Specimen Retention Time

1 month

Performing Laboratory Location

Rochester

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

82542

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
Q10	Coenzyme Q10, Reduced and Total, P	In Process

Result ID	Test Result Name	Result LOINC® Value
87853	CoQ10 reduced	81157-0
30091	CoQ10 Total	27923-2
30092	CoQ10 % reduced	81156-2
30159	Interpretation	59462-2