

Reporting Title: Mitochondrial Metabolites, P**Performing Location:** Rochester**Ordering Guidance:**

This test is not the recommended initial screening test for evaluating patients with suspected mitochondrial disorders, organic acidurias, and ketone body disorders. For these purposes, the preferred tests for first-tier assessment are OAU / Organic Acids Screen, Random, Urine; AAQP / Amino Acids, Quantitative, Plasma; and ACRN / Acylcarnitines, Quantitative, Plasma.

Analytes from LAPYP / Lactate Pyruvate Panel, Plasma are included in this test. If ordered together, LAPYP may be canceled.

Specimen Requirements:

Collection Container/Tube:

Preferred: Green top (sodium heparin)

Acceptable: Green top (lithium heparin)

Submission Container/Tube: Plastic vial

Specimen Volume: 0.5 mL

Collection Instructions: Centrifuge and aliquot plasma into plastic vial.

Specimen Minimum Volume:

0.1 mL

Forms:

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

Specimen Type	Temperature	Time	Special Container
Plasma	Frozen	7 days	

Result Codes:

Result ID	Reporting Name	Type	Unit	LOINC®
616819	Interpretation	Alphanumeric		59462-2
616798	Lactic acid	Numeric	nmol/mL	2524-7
616799	2-Hydroxybutyric acid	Numeric	nmol/mL	69843-1

Result ID	Reporting Name	Type	Unit	LOINC®
616800	3-Hydroxybutyric acid	Numeric	nmol/mL	6873-4
616801	Pyruvic acid	Numeric	nmol/mL	32338-6
616802	cis-Aconitic acid	Numeric	nmol/mL	75083-6
616803	Citric acid	Numeric	nmol/mL	15038-3
616804	3-Hydroxypropionic acid	Numeric	nmol/mL	47536-8
616805	3-Hydroxy-2-methylbutyric acid	Numeric	nmol/mL	69789-6
616806	3-Hydroxyisovaleric acid	Numeric	nmol/mL	72450-0
616807	Succinic acid	Numeric	nmol/mL	35871-3
616808	Fumaric acid	Numeric	nmol/mL	75081-0
616809	3-Methylglutaconic acid	Alphanumeric	nmol/mL	33273-4
616810	Malic acid	Numeric	nmol/mL	75068-7
616811	2-Ketobutyric acid	Numeric	nmol/mL	In Process
616812	2-Ketoisovaleric acid	Alphanumeric	nmol/mL	35868-9
616813	Acetoacetic acid	Alphanumeric	nmol/mL	35867-1
616814	3-Methyl-2-ketovaleric acid	Alphanumeric	nmol/mL	35869-7
616815	2-Ketoisocaproic acid	Alphanumeric	nmol/mL	35870-5
616816	2-Methylcitric acid	Numeric	nmol/mL	26904-3
616817	2-Ketoglutaric acid	Numeric	nmol/mL	69803-5
616818	Reviewed by	Alphanumeric		18771-6

LOINC and CPT codes are provided by the performing laboratory.

Supplemental Report:

No

CPT Code Information:

82542

Reference Values:**LACTIC ACID**

< or = 4000.0 nmol/mL

2-HYDROXYBUTYRIC ACID

< or = 124.0 nmol/mL

3-HYDROXYBUTYRIC ACID

< or = 700.0 nmol/mL

PYRUVIC ACID

< or = 350.0 nmol/mL

cis-ACONITIC ACID

< or = 9.0 nmol/mL

CITRIC ACID

< or = 250.0 nmol/mL

3-HYDROXYPROPIONIC ACID

< or = 12.4 nmol/mL

3-HYDROXY-2-METHYLBUTYRIC ACID

< or = 2.5 nmol/mL

3-HYDROXYISOVALERIC ACID

< or = 15.4 nmol/mL

SUCCINIC ACID

< or = 10.0 nmol/mL

FUMARIC ACID

< or = 5.0 nmol/mL

3-METHYLGLUTACONIC ACID

< or = 1.6 nmol/mL

MALIC ACID

< or = 20.0 nmol/mL

2-KETOBUTYRIC ACID

< or = 16.0 nmol/mL

2-KETOISOVALERIC ACID

< or = 35.0 nmol/mL

ACETOACETIC ACID

< or = 350.0 nmol/mL

3-METHYL-2-KETOVALERIC ACID
< or = 70.0 nmol/mL

2-KETOISOCAPROIC ACID
< or = 70.0 nmol/mL

2-METHYLCITRIC ACID
< or = 1.0 nmol/mL

2-KETOGLUTARIC ACID
< or = 40.0 nmol/mL