

Mitochondrial Metabolites, Plasma

Test ID: MMPP

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.

Useful for:

Monitoring patients with mitochondrial disorders, organic acidurias, and ketone body disorders

Methods:

Gas Chromatography-Mass Spectrometry (GS-MS)

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

Specimen Requirements:

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.
Minimum Volume:	0.1 mL

Specimen Stability Information:

Specimen Type	Temperature	Time
Plasma	Frozen	7 days

Cautions:

3-hydroxyisobutyric acid can cause a false elevation in the quantitation of 3-hydroxybutyric acid. Patients affected by 3-hydroxyisobutyric aciduria may have falsely elevated 3-hydroxybutyric acid.

Gross elevations of methylmalonic acid may interfere with the quantitation of 3-hydroxyisovaleric and succinic acid. When observed, the report will include a comment indicating presence of interference.

Gross elevations of acetoacetic acid may interfere with the quantification of 3-methyl-2-ketovaleric acid. When observed, the report will include a comment indicating presence of interference.

CPT Code:

82542

Day(s) Performed: Tuesday **Report Available:** 3-9 days

Questions

Contact Melissa Tricker-Klar, Laboratory Technologist Resource Coordinator at 800-533-1710.