



Laboratory Service Manual

Lab Dept: Urine/Stool

Test Name: ACYLGLYCINES, QUANTITATIVE, URINE

General Information

Lab Order Codes: ACYU

Synonyms: Glycine conjugates urine

CPT Codes: 82544 – Column chromatography, stable isotope dilution, multiple analytes, quantitative, single stationary and mobile phase

Test Includes: The following acylglycines reported in ug/mg Creatinine: Ethylmalonic Acid, 2-Methylsuccinic Acid, Glutaric Acid, Isobutyrylglycine, n-Butyrylglycine, 2-Methylbutyrylglycine, Isovalerylglycine, n-Hexanoylglycine, n-Octanoylglycine, 3-Phenylpropionylglycine, Suberylglycine, trans-Cinnamoylglycine, Dodecanedioic Acid (12:0), Tetradecanedioic Acid (14:0), Hexadecanedioic Acid (16:0).

Logistics

Test Indications: Useful for biochemical diagnosis of selected inborn errors of metabolism (see below) by quantitative determination of target urinary metabolites that are present in amounts below the detection limit of routine organic acid analysis. Acylglycine analysis is the method of choice, in urine, for the biochemical evaluation of asymptomatic patients affected with 1 of the following inborn errors of metabolism:

- Short chain acyl-CoA dehydrogenase (SCAD) deficiency
- Functional SCAD deficiency (G625A, C611T variants)
- Medium-chain acyl-CoA dehydrogenase (MCAD) deficiency
- Medium-chain 3-ketoacyl-CoA thiolase (MCKAT) deficiency
- Electron transfer flavoprotein (ETF) deficiency (Glutaric acidemia type 2)
- ETF: ubiquinone oxidoreductase (ETF-QO) deficiency
- (Glutaric acidemia type 2)
- Riboflavin-responsive multiple acyl-CoA dehydrogenase deficiency
- Ethylmalonic encephalopathy
- 2-Methylbutyryl-CoA dehydrogenase deficiency
- Isovaleryl-CoA dehydrogenase deficiency
- Glutaryl-CoA dehydrogenase deficiency

Lab Testing Sections: Urine/Stool - Sendouts

Referred to: Mayo Medical Laboratories (MML Test# 81249)

Phone Numbers:

Minneapolis: 612-813-6280



Laboratory Service Manual

Saint Paul: 651-220-6550

Test Availability: Daily, 24 hours

Turnaround Time: 3 – 14 days, test performed Monday and Thursday

Special Instructions: N/A

Specimen

Specimen Type: Urine, random

Container: Leak-proof urine container

Draw Volume: Entire specimen

Processed Volume: 5.0 mL (Minimum: 3.0 mL) urine

Collection: Routine urine collection

Special Processing: Lab Staff: Mix random urine sample well. Remove aliquot into a plastic, 13 mL urine tube. Store and ship at frozen temperatures. Forward promptly.

Patient Preparation: None

Sample Rejection: Specimens other than urine, Warm specimens, mislabeled specimens

Interpretive

Reference Range:

Acylglycine:	Range in ug/mg Creatinine:
Ethylmalonic Acid:	0.5 – 20.2
2-Methylsuccinic Acid	0.4 – 13.8
Glutaric Acid:	0.6 – 15.2
Isobutyrylglycine:	0.00 – 11.0
n-Butyrylglycine:	0.1 – 2.1
2-Methylbutyrylglycine:	0.3 – 7.5
Isovalerylglycine:	0.3 – 14.3
n-Hexanoylglycine:	0.2 – 1.9



Laboratory Service Manual

n-Octanoylglycine:	0.1 – 2.1
3-Phenylpropionylglycine:	0.00 – 1.1
Suberylglycine:	0.00 – 11.0
trans-Cinnamoylglycine:	0.2 – 14.7
Dodecanedioic Acid (12:0):	0.0 – 1.1
Tetradecanedioic Acid (14:0):	0.00 – 1.0
Hexadecanedioic Acid (16:0):	0.00 – 1.0

Critical Values:

N/A

Limitations:

Due to a limited number of metabolites included in the acylglycine analysis, it is recommended that an [Organic Acid Screen, Urine](#) also be performed concurrently.

Methodology:

Gas Chromatography-Mass Spectrometry (GC-MS) Stable Isotope Dilution Analysis

Contraindications:

N/A

References:

[Mayo Medical Laboratory Website](#) June 2007