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**Lab Dept:** Urine/Stool

**Test Name:** ACYLGLYCINES, QUANTITATIVE, URINE

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**General Information**

**Lab Order Codes:** ACYU

**Synonyms:** Glycine conjugates urine

**CPT Codes:** 82542 – Column chromatography, includes mass spectrometry, if performed, non-drug analytes, not elsewhere specified, qualitative or quantitative, each specimen

**Test Includes:** The following acylglycines reported in mg/g 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), n-Acetylglycine, n-Propionylglycine, 3-Methylcrotonylglycine, n-Tiglylglycine, 3-Methylglutaconic Acid

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**Logistics**

**Test Indications:** Useful for diagnosis and monitoring for patients affected with 1 of the following inborn errors of metabolism:|

Fatty Acid Oxidation Disorders

- Glutaric acidemia type II
- Medium-chain 3-ketoacyl-CoA thiolase (MCKAT) deficiency
- Medium-chain acyl-CoA dehydrogenase (MCAD) deficiency
- Short chain acyl-CoA dehydrogenase (SCAD) deficiency

Organic Acidurias

- 2-Methyl-3-hydroxybutyryl-CoA dehydrogenase (2M3HBD) deficiency
- 2-Methylbutyryl-CoA dehydrogenase deficiency
- 3-Methylcrotonyl-CoA carboxylase deficiency
- 3-Methylglutaconyl-CoA-hydratase deficiency
- Aminoacylase 1 deficiency
- Beta-ketothiolase deficiency
- Ethylmalonic encephalopathy
- Glutaryl-CoA dehydrogenase deficiency
- Isobutyryl-CoA dehydrogenase (IBD) deficiency
- Isovaleryl-CoA dehydrogenase deficiency
- Multiple carboxylase deficiency
- Propionic acidemia

**Lab Testing Sections:** Urine/Stool - Sendouts

**Referred to:** Mayo Medical Laboratories (MML Test: AGU20)

**Phone Numbers:** MIN Lab: 612-813-6280  
STP Lab: 651-220-6550

**Test Availability:** Daily, 24 hours

**Turnaround Time:** 5 – 10 days, test performed Monday, Wednesday, Friday

**Special Instructions:** Please include family history, clinical conditions (asymptomatic or acute episode), diet, and drug therapy information.

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### ***Specimen***

**Specimen Type:** Urine, random

**Container:** Leak-proof urine container

**Draw Volume:** Entire specimen

**Processed Volume:** 10 mL (Minimum: 4 mL) urine

**Collection:** Routine urine collection, no preservative

**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.

Note: If insufficient volume is obtained, submit as much specimen as possible in a single container; Mayo will determine if volume is sufficient for testing.

**Patient Preparation:** None

**Sample Rejection:** Specimens other than urine; warm specimens; mislabeled or unlabeled specimens

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### ***Interpretive***

**Reference Range:**

Reference ranges will accompany the report, reported in mg/g Creat. Ranges include pediatric through adult values. See Mayo website for full table.

**Interpretation:** When abnormal results are detected, a detailed interpretation is given, including an overview of the results and of their significance; a correlation to available clinical information; elements of differential diagnosis; recommendations for additional biochemical testing and in vitro confirmatory studies (enzyme assay, molecular analysis); name and number of key contacts who may provide these studies or answer questions.

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<b>Critical Values:</b>	N/A
<b>Limitations:</b>	N/A
<b>Methodology:</b>	Gas Chromatography-Mass Spectrometry (GC-MS) Stable Isotope Dilution Analysis
<b>References:</b>	<a href="#">Mayo Clinic Laboratories</a> February 2021
<b>Updates:</b>	6/23/2010: Updated recommended volume, previously listed as 5 mL. 11/30/2010: Units change, previously listed as mcg/mg Cr. 1/26/2016: CPT updates 2/11/2021: Mayo added five additional constituents, ref range update