**Lab Dept:** Urine/Stool  

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

### 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  

### 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.

**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

**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.
Critical Values: N/A

Limitations: N/A

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

References: Mayo Clinic Laboratories February 2021

Updates: 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