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 one 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 academia

Diagnostic specificity of inborn errors of metabolism via urine acylglycine testing is available only for selected inborn errors of metabolism; it is recommended that urine organic acids (OAU / Organic Acids Screen, Random, Urine) be ordered and assessed simultaneously due to the limited number of metabolites included in this urine acylglycine test.

Lab Testing Sections: Urine/Stool - Sendouts

Referred to: Mayo Clinic Laboratories (MML Test: AGU20)

Phone Numbers: MIN Lab: 612-813-6280

STP Lab: 651-220-6550

Test Availability: Daily, 24 hours

Turnaround Time: 3-6 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. Specimen is stable frozen (preferred) for 416 days, refrigerated for 9 days.
Patient Preparation:	None
Sample Rejection:	Specimens other than urine; warm specimens; mislabeled or unlabeled specimens

Interpretive

Reference Range:	As of Mayo 16, 2024, reference ranges will not be segregated by gender or age. 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 phone number of key contacts who may provide these studies at Mayo Clinic or elsewhere; and a phone number to reach one of the laboratory directors in case the referring physician has additional questions.
Critical Values:	N/A
Limitations:	N/A
Methodology:	Gas Chromatography-Mass Spectrometry (GC-MS) Stable Isotope Dilution Analysis
References:	Mayo Clinic Laboratories May 2024
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 5/16/2024: Updated turnaround time and reference range information. Added specimen stability.