Lab Dept: Chemistry

Test Name: ACYLCARNITINES, QUANTITATIVE, PLASMA

General Information

Lab Order Codes: ACYP

Synonyms: Glycine conjugates plasma

CPT Codes: 82017 – Acylcarnitines, quantitative, each specimen

Test Includes: Acetylcarnitine (C:2); Propionylcarnitine (C3); Iso-/Butyrylcarnitine (C4); Isovaleryl-/2-Methylbutyrylcarnitine (C5); Hexanoylcarnitine (C6); 3-OH-Hexanyoylcarnitine (C6-OH); Octenoylcarnitine (C8:1), Octanoylcarnitine (C8); Decenoylcarnitine (C10:1); Decanoylcarnitine (C10); Glutarylcarnitine (C5-DC); Dodecenoylcarnitine (C12:1); Dodecanoylcarnitine (C12); 3-OH-Dodecanoylcarnitine (C12-OH); Tetradecadienoylcartinine (C14:2); Tetradecenoylcarnitine (C14:1); Tetradecanoylcarnitine (C14); 3-OH-Tetradecenoylcarnitine (C14:1-OH); 3-OH-Tetradecanoylcarnitine (C14-OH); Hexadecenoylcarnitine (C16:1);Hexadecanoylcarnitine (C16); 3-OH-Hexadecenoylcarnitine(C16:1-OH); 3-OH-Hexadecanoylcarnitine (C16-OH); Linoleylcarnitine (C18:2); Oleylcarnitine (C18:1); Stearoylcarnitine (C18); 3-OH-Linoleylcarnitine (C18:2-OH); 3-OH-Oleylcarnitine (C18:1-OH) levels all reported in nmol/mL.

Logistics

Test Indications: Diagnosis of fatty acid beta-oxidation disorders and several organic acidurias. In general, more than 20 inborn errors of metabolism can be identified using this method. For most of the disorders involving fatty acid beta-oxidation, this is the most informative screening test. Quantitative acylcarnitine analysis can also be used to evaluate the treatment during follow-up of patients with these disorders.

Lab Testing Sections: Chemistry - Sendouts

Referred to: Mayo Medical Laboratories (MML Test: ACRN)

Phone Numbers: MIN Lab: 612-813-6280

STP Lab: 612-651-6550

Test Availability: Daily, 24 hours

Turnaround Time: 2 – 5 days, test performed Monday - Friday

Special Instructions: Include family history, clinical condition (asymptomatic or acute episode), diet, and drug therapy information. Patient's age is required on the request form for processing. See Patient Preparation.

Specimen

Specimen Type: Blood

Container: Green top (Lithium Heparin) tube
Alternate tubes: Lavender (EDTA), Green (Sodium Heparin)

Draw Volume: 0.5 mL (Minimum: 0.2 mL) blood

Processed Volume: 0.1 mL (Minimum: 0.05 mL) plasma

Collection: Routine venipuncture

Special Processing: Lab Staff: Centrifuge specimen, remove plasma aliquot into a screw-capped round bottom plastic vial. Ship and store at frozen temperatures.

Patient Preparation: Draw specimen just prior to next scheduled meal or feeding.

Sample Rejection: Mislabeled or unlabeled specimens

Interpretive

Reference Range:

Age:	Range (nmol/mL):
Acetylcarnitne, C:2:	
1 – 7 days:	2.14 – 15.89
8 days – 7 years:	2.00 – 27.57
≥8 years:	2.00 – 17.83
Acrylylcarnitine C3:1	
1 – 7 days:	<0.04
8 days – 7 years:	<0.05
≥8 years:	<0.07
Propionylcarnitine, C3:	
1 – 7 days:	<0.55

8 days – 7 years:	<1.78
≥8 years:	<0.88
Formiminoglutamate, FIGLU	
1 – 7 days:	<0.43
8 days – 7 years:	<0.08
≥8 years:	<0.14
Iso-/Butyrylcarnitine, C4:	
1 – 7 days:	<0.46
8 days – 7 years:	<1.06
≥8 years:	<0.83
Tiglylcarnitine, C5:1	
1 – 7 days:	<0.05
8 days – 7 years:	<0.09
≥8 years:	<0.11
Isovaleryl-/2-Methylbutyrulcarnitine, C5:	
1 – 7 days:	<0.38
8 days – 7 years:	<0.63
≥8 years:	<0.51
3-OH-iso-/butyrylcarnitine, C4-OH	
1 – 7 days:	<0.13
8 days – 7 years:	<0.51
≥8 years:	<0.18
Hexenoylcarnitine, C6:1	

1 – 7 days:	<0.12
8 days – 7 years:	<0.10
≥8 years:	<0.15
Hexanoylcarnitine, C6:	
1 – 7 days:	<0.14
8 days – 7 years:	<0.23
≥8 years:	<0.17
3-OH-isovalerylcarnitine, C5-OH	
1 – 7 days:	<0.08
8 days – 7 years:	<0.12
≥8 years:	<0.10
Benzoylcarnitine	
1 – 7 days:	<0.13
8 days – 7 years:	<0.07
≥8 years:	<0.10
Heptanoylcarnitine, C7	
1 – 7 days:	<0.05
8 days – 7 years:	<0.05
≥8 years:	<0.06
3-OH-Hexanoylcarnitine, C6-OH:	
1 – 7 days:	<0.08
8 days – 7 years:	<0.19
≥8 years:	<0.09

Phenylacetylcarnitine	
1 – 7 days:	<0.15
8 days – 7 years:	<0.22
≥8 years:	<0.29
Salicylcarnitine	
1 – 7 days:	<0.08
8 days – 7 years:	<0.09
≥8 years:	<0.09
Octenoylcarnitine, C8:1:	
1 – 7 days:	<0.48
8 days – 7 years:	<0.91
≥8 years:	<0.88
Octanoylcarnitine, C8:	
1 – 7 days:	<0.19
8 days – 7 years:	<0.45
≥8 years:	<0.78
Malonylcarnitine, C3-DC	
1 – 7 days:	<0.09
8 days – 7 years:	<0.14
≥8 years:	<0.26
Decadienoylcarnitine, C10:2	
1 – 7 days:	<0.11
8 days – 7 years:	<0.12

≥8 years:	<0.26
Decenoylcarnitne, C10:1	
1 – 7 days:	<0.25
8 days – 7 years:	<0.46
≥8 years:	<0.47
Decanoylcarnitine, C10:	
1 – 7 days:	<0.27
8 days – 7 years:	<0.91
≥8 years:	<0.88
Methylmalonyl-/succinylcarnitine, C4-DC	
1 – 7 days:	<0.05
8 days – 7 years:	<0.05
≥8 years:	<0.05
3-OH-decenoylcarnitine, C10-1-OH	
1 – 7 days:	<0.12
8 days – 7 years:	<0.12
≥8 years:	<0.13
Glutarylcarnitine, C5-DC:	
1 – 7 days:	<0.06
8 days – 7 years:	<0.10
≥8 years:	<0.11
Dodecenoylcarnitine, C12:1:	
1 – 7 days:	<0.19

8 days – 7 years:	<0.37
≥8 years:	<0.35
Dodecanoylcarnitine, C12:	
1 – 7 days:	<0.18
8 days – 7 years:	<0.35
≥8 years:	<0.26
3-Methylglutarylcarnitine, C6-DC	
1 – 7 days:	<0.28
8 days – 7 years:	<0.21
≥8 years:	<0.43
3-OH-dodecenoylcarnitine, C12:1-OH	
1 – 7 days:	<0.11
8 days – 7 years:	<0.10
≥8 years:	<0.13
3-OH-Dodecanoylcarnitine, C12-OH:	
1 – 7 days:	<0.06
8 days – 7 years:	<0.09
≥8 years:	<0.08
Tetradecadienoylcarnitine, C14:2:	
1 – 7 days:	<0.09
8 days – 7 years:	<0.13
≥8 years:	<0.18
Tetradecenoylcarnitine, C14:1:	

1 – 7 days:	<0.16
8 days – 7 years:	<0.35
≥8 years:	<0.24
Tetradecanoylcarnitine, C14	
1 – 7 days:	<0.11
8 days – 7 years:	<0.15
≥8 years:	<0.12
Octanedioylcarnitine, C8-DC	
1 – 7 days:	<0.25
8 days – 7 years:	<0.19
≥8 years:	<0.19
3-OH-Tetradecenoylcarnitine, C14:1-OH:	
1 – 7 days:	<0.06
8 days – 7 years:	<0.18
≥8 years:	<0.13
3-OH-Tetradecanoylcarnitine, C14-OH:	
1 – 7 days:	<0.04
8 days – 7 years:	<0.05
≥8 years:	<0.08
Hexadecenoylcarnitine, C16:1:	
1 – 7 days:	<0.15
8 days – 7 years:	<0.21
≥8 years:	<0.10

Hexadecanoylcarnitine, C16:	
1 – 7 days:	<0.36
8 days – 7 years:	<0.52
≥8 years:	<0.23
3-OH-Hexadecenoylcarnitine, C16:1-OH:	
1 – 7 days:	<0.78
8 days – 7 years:	<0.36
≥8 years:	<0.06
3-OH-Hexadecanoylcarnitine, C16-OH:	
1 – 7 days:	<0.10
8 days – 7 years:	<0.07
≥8 years:	<0.06
Octadecadienoylcarnitine, C18:2 (Linoleylcarnitine, C18:2)	
1 – 7 days:	<0.12
8 days – 7 years:	<0.31
≥8 years:	<0.24
Octadecenoylcarnitine, C18:1 (Oleylcarnitine, C18:1)	
1 – 7 days:	<0.25
8 days – 7 years:	<0.45
≥8 years:	<0.39
Octadecanoylcarnitine, C18 (Stearolycarnitine, C18)	
1 – 7 days:	<0.10
8 days – 7 years:	<0.12

≥8 years:	<0.14
Dodecanediolylcarnitine, C12-DC	
1 – 7 days:	<0.10
8 days – 7 years:	<0.04
≥8 years:	<0.04
3-OH-octadecadienoylcarnitine, C18:2-OH (3-OH-Linoleylcarnitine, C18:2-OH)	
1 – 7 days:	<0.04
8 days – 7 years:	<0.06
≥8 years:	<0.06
3-OH-octadecenoylcarnitine C18:1-OH (3-OH-Oleylcarnitine, C18:1-OH)	
1 – 7 days:	<0.03
8 days – 7 years:	<0.04
≥8 years:	<0.06
3-OH-octadecanoylcarnitine, C18-OH	
1 – 7 days:	<0.03
8 days – 7 years:	<0.05
≥8 years:	<0.03
Interpretation: An interpretive report is provided. The individual quantitative results support the interpretation of the acylcarnitine profile, but are not diagnostic by themselves. The interpretation is based on the pattern recognition. Abnormal results are not sufficient to conclusively establish a diagnosis of a particular disease. To verify a preliminary diagnosis based on an acylcarnitine analysis, independent biochemical or molecular genetic analyses are required.	

Critical Values: N/A

Limitations:

In a few instances, false-negative results occur in the analysis of acylcarnitine profiles. Patients with carnitine deficiency may not exhibit abnormally high acylcarnitine concentrations. For some disorders, such as medium-chain ayl-CoA dehydrogenase deficiency, the calculation of ratios between different acylcarnitine species provides a discriminate factor to overcome such problems. Where applicable, the calculation of such ratios will be incorporated in the routine acylcarnitine analysis. Informative profiles may also not be detected in some disorders where the accumulation of diagnostic acylcarnitnes is a reflection of the residual activity of the defective enzyme, the dietary load of precursors, and the anabolic/catabolic and treatment status of a patient.

Patients with carnitine deficiency may not exhibit abnormally high acylcarnitine concentrations. If the results are indicative for carnitine deficiency, the interpretation will include a remark that this limits the diagnostic value of the test and repeat analysis may be considered following carnitine supplementation.

Follow-up testing such as in vitro enzyme assays or molecular genetic testing may be recommended following abnormal acylcarnitine results. It is not advisable to intentionally stress the patient's metabolism (e.g., fasting test) prior to specimen collection for acylcarnitine analysis.

Methodology:

Flow-injection Analysis Tandem Mass Spectrometry (FIA-MS/MS)

References:

Mayo Medical Laboratory Website (February 2017)

Updates:

12/8/2015: 19 new analytes added.
2/15/2017: Tube update.