



Laboratory Service Manual

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-Hexanoylcarnitine (C6-OH); Octenoylcarnitine (C8:1), Octanoylcarnitine (C8); Decenoylcarnitine (C10:1); Decanoylcarnitine (C10); Glutaryl carnitine (C5-DC); Dodecenoylcarnitine (C12:1); Dodecanoylcarnitine (C12); 3-OH-Dodecanoylcarnitine (C12-OH); Tetradecadienoylcarnitine (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); Oleoylcarnitine (C18:1); Stearoylcarnitine (C18); 3-OH-Linoleylcarnitine (C18:2-OH); 3-OH-Oleoylcarnitine (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#: 82413)

Phone Numbers:

Minneapolis: 612-813-6280

Saint Paul: 651-220-6550

Test Availability: Daily, 24 hours

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



Laboratory Service Manual

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. Obtain green top Sodium (Na) Heparin tube from the laboratory.

Specimen

Specimen Type: Blood

Container: Green top (Na Heparin) top tube

Draw Volume: 0.5 mL (Minimum: 0.3 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: None

Sample Rejection: Specimens other than Na Heparin plasma, mislabeled or unlabeled specimens, warm specimens

Interpretive

Reference Range:

Age:	Range (nmol/mL):
Acetylcarnitine, C:2:	
1 – 7 days:	2.14 – 15.89
8 days – 7 years:	2.00 – 27.57
≥8 years:	2.00 – 17.83
Propionylcarnitine, C3:	
1 – 7 days:	<0.55
8 days – 7 years:	<1.78
≥8 years:	<0.88
Iso-/Butyrylcarnitine, C4:	



Laboratory Service Manual

1 – 7 days:	<0.46
8 days – 7 years:	<1.06
≥8 years:	<0.83
Isovaleryl-/2-Methylbutyrylcarnitine, C5:	
1 – 7 days:	<0.38
8 days – 7 years:	<0.63
≥8 years:	<0.51
Hexanoylcarnitine, C6:	
1 – 7 days:	<0.14
8 days – 7 years:	<0.23
≥8 years:	<0.17
3-OH-Hexanoylcarnitine, C6-OH:	
1 – 7 days:	<0.08
8 days – 7 years:	<0.19
≥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



Laboratory Service Manual

Decenoylcarnitine, 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
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-OH-Dodecanoylcarnitine, C12-OH:	
1 – 7 days:	<0.06
8 days – 7 years:	<0.09



Laboratory Service Manual

≥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
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



Laboratory Service Manual

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
Linoleylcarnitine, C18:2:	
1 – 7 days:	<0.12
8 days – 7 years:	<0.31
≥8 years:	<0.24
Oleylcarnitine, C18:1:	
1 – 7 days:	<0.25
8 days – 7 years:	<0.45
≥8 years:	<0.39
Stearoylcarnitine, C18:	



Laboratory Service Manual

1 – 7 days:	<0.10
8 days – 7 years:	<0.12
≥8 years:	<0.14
3-OH-Linoleylcarnitine, C18:2-OH:	
1 – 7 days:	<0.04
8 days – 7 years:	<0.06
≥8 years:	<0.06
3-OH-Oleylcarnitine, C18:1-OH:	
1 – 7 days:	<0.03
8 days – 7 years:	<0.04
≥8 years:	<0.06

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 acyl-CoA dehydrogenase deficiency, the calculation of ratios between different acylcarnitine species provides a discriminant factor to overcome such problems. Where applicable, the calculation of such ratios will be incorporated in the routine acylcarnitine analysis.

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.

Informative profiles may also not be detected in some disorders where the accumulation of diagnostic acylcarnitines 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.

In some cases, additional methods of higher specificity and sensitivity such as in vitro enzyme assays or molecular genetic testing are advisable as opposed to stressing the patient's metabolism (e.g., fasting test) prior to repeating acylcarnitine analysis.

Methodology: Electrospray Tandem Mass Spectrometry (MS/MS)

References: [Mayo Medical Laboratory Website](#) (February 2010)



Laboratory Service Manual