

PRODUCT INFORMATION



2-Methylbutyryl-L-carnitine-d₃ (chloride)

Item No. 26572

Formal Name: (2R)-3-carboxy-N,N-dimethyl-N-(methyl-d₃)-2-((2-methylbutanoyl)oxy)propan-1-aminium, monochloride

Synonyms: CAR 5:0-d₃, C5:0 Carnitine-d₃, L-Carnitine 2-methylbutyryl ester-d₃, L-Carnitine 2-methylbutyryl ester-d₃, 2-Methylbutyrylcarnitine-d₃, 2-Methylbutyrylcarnitine-d₃

MF: C₁₂H₂₁D₃NO₄ • Cl

FW: 284.8

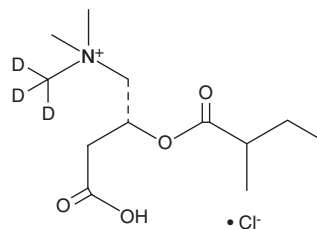
Chemical Purity: ≥98% (2-Methylbutyryl-L-carnitine)

Deuterium Incorporation: ≥99% deuterated forms (d₁-d₃); ≤1% d₀

Supplied as: A solution in ethanol

Storage: -20°C

Stability: ≥2 years



Information represents the product specifications. Batch specific analytical results are provided on each certificate of analysis.

Laboratory Procedures

2-Methylbutyryl-L-carnitine-d₃ (chloride) is intended for use as an internal standard for the quantification of 2-methylbutyryl-L-carnitine by GC- or LC-MS. The accuracy of the sample weight in this vial is between 5% over and 2% under the amount shown on the vial. If better precision is required, the deuterated standard should be quantitated against a more precisely weighed unlabeled standard by constructing a standard curve of peak intensity ratios (deuterated versus unlabeled).

2-Methylbutyryl-L-carnitine-d₃ (chloride) is supplied as a solution in ethanol. To change the solvent, simply evaporate the ethanol under a gentle stream of nitrogen and immediately add the solvent of choice. Solvents such as DMSO and dimethyl formamide purged with an inert gas can be used. The solubility of 2-methylbutyryl-L-carnitine-d₃ (chloride) in these solvents is approximately 20 and 15 mg/ml, respectively.

Description

2-Methylbutyryl-carnitine is a naturally occurring acylcarnitine that is produced via L-isoleucine metabolism.¹ Plasma levels of 2-methylbutyryl-carnitine are elevated in patients with non-alcoholic steatohepatitis (NASH).² Elevated levels of 2-methylbutyryl-carnitine are associated with 2-methylbutyryl-CoA dehydrogenase deficiency (2-MBCDD), also known as short/branched chain acyl-CoA dehydrogenase (SBCAD) deficiency.³

References

1. Gibson, K.M., Burlingame, T.G., Hogema, B., *et al.* 2-Methylbutyryl-coenzyme A dehydrogenase deficiency: A new inborn error of L-isoleucine metabolism. *Pediatr. Res.* **47(6)**, 830-833 (2000).
2. Kalhan, S.C., Guo, L., Edmison, J., *et al.* Plasma metabolomic profile in nonalcoholic fatty liver disease. *Metabolism* **60(3)**, 404-413 (2011).
3. Van Calcar, S.C., Baker, M.W., Williams, P., *et al.* Prevalence and mutation analysis of short/branched chain acyl-CoA dehydrogenase deficiency (SBCADD) detected on newborn screening in Wisconsin. *Mol. Genet. Metab.* **110(1-2)**, 111-115 (2013).

WARNING

THIS PRODUCT IS FOR RESEARCH ONLY - NOT FOR HUMAN OR VETERINARY DIAGNOSTIC OR THERAPEUTIC USE.

SAFETY DATA

This material should be considered hazardous until further information becomes available. Do not ingest, inhale, get in eyes, on skin, or on clothing. Wash thoroughly after handling. Before use, the user must review the complete Safety Data Sheet, which has been sent via email to your institution.

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