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- Trockeneiszuschlag
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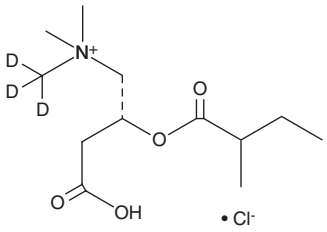
PRODUCT INFORMATION

2-Methylbutyryl-L-carnitine-d_3 (chloride)

Item No. 26572

Formal Name: (2R)-3-carboxy-N,N-dimethyl-N-(methyl-d_3)-2-((2-methylbutanoyl)oxy)propan-1-aminium, monochloride
Synonyms: 2-Methylbutyroylcarnitine-d_3, 2-Methylbutyrylcarnitine-d_3
MF: $C_{12}H_{21}D_3NO_4 \bullet Cl$
FW: 284.8
Chemical Purity: ≥98% (2-Methylbutyryl-L-carnitine)
Deuterium Incorporation: ≥99% deuterated forms (d_1-d_3); ≤1% d_0
Supplied as: A solid
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-d_3 (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-d_3 (chloride) is supplied as a solid. A stock solution may be made by dissolving the 2-methylbutyryl-L-carnitine-d_3 (chloride) in the solvent of choice. 2-Methylbutyryl-L-carnitine-d_3 (chloride) is soluble in organic solvents such as ethanol, DMSO, and dimethyl formamide, which should be purged with an inert gas. The solubility of 2-methylbutyryl-L-carnitine-d_3 (chloride) in these solvents is approximately 25, 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).

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