

## Produktinformation



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### SZABO-SCANDIC HandelsgmbH

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# **PRODUCT** INFORMATION



## Isobutyryl-L-carnitine-d<sub>3</sub> (chloride)

Item No. 26576

Formal Name:	(R)-3-carboxy-2-(isobutyryloxy)-N,N-dimethyl-N- (methyl-d <sub>3</sub> )propan-1-aminium, monochloride	0
Synonym:	L-Isobutyrylcarnitine-d <sub>3</sub>	Ŭ,
MF:	$C_{11}H_{19}D_3NO_4 \bullet Cl$	0 0
FW:	270.8	Ŭ, J, I
<b>Chemical Purity:</b>	≥98% (IsobutyryI-L-carnitine)	но
Deuterium		• CI-
Incorporation:	≥99% deuterated forms (d <sub>1</sub> -d <sub>3</sub> ); ≤1% d <sub>0</sub>	
Supplied as:	A solid	
Storage:	-20°C	5
Stability:	≥2 years	
Information represents the product specifications. Batch specific analytical results are provided on each certificate of analysis.		

#### Laboratory Procedures

Isobutyryl-L-carnitine-d<sub>3</sub> (chloride) is intended for use as an internal standard for the quantification of isobutyryl-L-carnitine (chloride) (Item No. 17440) 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).

Isobutyryl-L-carnitine-d<sub>2</sub> (chloride) is supplied as a solid. A stock solution may be made by dissolving the isobutyryl-L-carnitine-d<sub>3</sub> (chloride) in the solvent of choice. Isobutyryl-L-carnitine-d<sub>3</sub> (chloride) is soluble in organic solvents such as ethanol, DMSO, and dimethyl formamide (DMF), which should be purged with an inert gas. The solubility of isobutyryl-L-carnitine-d<sub>3</sub> (chloride) in ethanol is approximately 25 mg/ml and approximately 20 mg/ml in DMSO and DMF.

#### Description

Isobutyryl-L-carnitine is a natural 4-carbon acylcarnitine that is involved in fatty acid oxidation and organic acid metabolism.<sup>1</sup> Elevated levels of isobutyryl-L-carnitine are associated with isobutyryl-CoA dehydrogenase deficiency.<sup>2,3</sup>

#### References

- 1. Turer, A.T., Stevens, R.D., Bain, J.R., et al. Metabolomic profiling reveals distinct patterns of myocardial substrate use in humans with coronary artery disease or left ventricular dysfunction during surgical ischemia/reperfusion. Circulation 119(13), 1736-1746 (2009).
- 2. Koeberl, D.D., Young, S.P., Gregersen, N., et al. Rare disorders of metabolism with elevated butyryl- and isobutyryl-carnitine detected by tandem mass spectrometry newborn screening. Pediatr. Res. 54(2), 219-223 (2003).
- 3. Rinaldo, P., Cowan, T.M., and Matern, D. Acylcarnitine profile analysis. Genet. Med. 10(2), 151-156 (2008).

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

#### SAFETY DATA

al 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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