

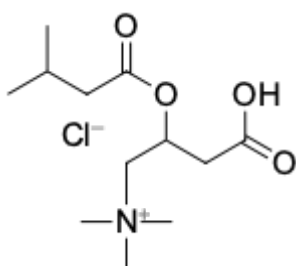
## (C5) Isovalerylcarnitine

<http://www.lumiprobe.com/p/isovalerylcarnitine>

(C5) Isovalerylcarnitine is an analogue of endogenous acylcarnitine, formed in the body as a result of leucine metabolism, and has an isovaleric acid residue as an acyl substituent. The profile of acylcarnitines in blood and tissues is used as a marker of various conditions associated with mitochondrial (and/or peroxisomal) dysfunction.

The product is used primarily as a control for MS/MS.

Isovalerylcarnitine — the main biochemical marker for isovaleric acidemia, which occurs as a result of deficiency of the mitochondrial enzyme isovaleryl-CoA dehydrogenase. Isovalerylcarnitine has been found to be associated with celiac disease and very long chain acyl-CoA dehydrogenase deficiency (VLCAD). Also, the level of isovalerylcarnitine is determined when studying neuralgia, for example, with trigeminal neuralgia.



**Structure of (C5) Isovalerylcarnitine**

### General properties

Appearance:	white solid
Molecular weight:	281.78
CAS number:	31023-24-2 (inner salt); 139144-12-0 (chloride)
Molecular formula:	C <sub>12</sub> H <sub>24</sub> ClNO <sub>4</sub>
IUPAC name:	(2R)-3-Carboxy-N,N,N-trimethyl-2-[(3-methylbutanoyl)oxy]-1-propanaminium
Solubility:	DMF, DMSO, ethanol
Quality control:	NMR <sup>1</sup> H and HPLC-MS (95+%)
Storage conditions:	24 months after receipt at -20°C in the dark. Transportation: at room temperature for up to 3 weeks. Desiccate.
Legal statement:	This Product is offered and sold for research purposes only. It has not been tested for safety and efficacy in food, drug, medical device, cosmetic, commercial or any other use. Supply does not express or imply authorization to use for any other purpose, including, without limitation, in vitro diagnostic purposes, in the manufacture of food or pharmaceutical products, in medical devices or in cosmetic products.