

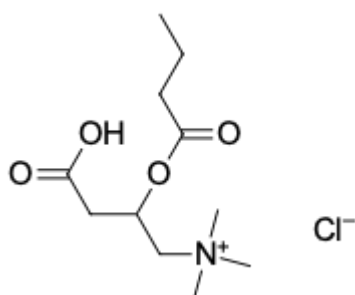
## (C4) Butyrylcarnitine

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

Butyrylcarnitine, also known as (3R)-3-(butyryloxy)-4-(trimethylammonio)butanoate or L-carnitine butyryl ester, is classified as a member of the acylcarnitines. Acylcarnitines are organic compounds containing a fatty acid with the carboxylic acid attached to carnitine through an ester bond. Butyrylcarnitine may be used as an analytical reference standard for the separation and identification of underivatized butyryl-L-carnitine in human plasma samples using high-performance liquid chromatography coupled to tandem mass spectrometry (HPLC-MS/MS).

Butyrylcarnitine can be used to study its role in energy production and its metabolic effects, to understand metabolic diseases and conditions associated with mitochondrial dysfunction. Butyrylcarnitine is a major biological marker of short-chain acyl-CoA dehydrogenase deficiency, glutaric acidemia type II and isobutyrylglycinuria, and ethylmalonic encephalopathy.

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



**Structure of (C4) Butyrylcarnitine**

### General properties

Appearance: white solid

Molecular weight: 267.75

CAS number: 162067-50-7 (chloride), 25576-40-3 (inner salt)

Molecular formula:  $C_{11}H_{22}ClNO_4$

Solubility: DMSO, DMF, ethanol

Quality control: NMR  $^1H$  and HPLC-MS (95+%)

Storage conditions: 24 months after receipt at  $-20^{\circ}C$  in the dark. Transportation: at room temperature for up to 3 weeks. Desiccate.

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