



# SZABO SCANDIC

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## Produktinformation



Forschungsprodukte & Biochemikalien



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Diagnostik & molekulare Diagnostik



Laborgeräte & Service

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### Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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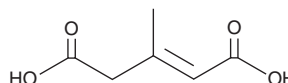
# PRODUCT INFORMATION



## 3-Methylglutaconic Acid

Item No. 34055

<b>CAS Registry No.:</b>	5746-90-7
<b>Formal Name:</b>	3-methyl-2-pentenedioic acid
<b>Synonyms:</b>	$\beta$ -Methylglutaconic Acid, 3-MGA, 3-MGC Acid, NSC 249232
<b>MF:</b>	C <sub>6</sub> H <sub>8</sub> O <sub>4</sub>
<b>FW:</b>	144.1
<b>Purity:</b>	≥95% (mixture of E/Z isomers)
<b>UV/Vis.:</b>	$\lambda_{\text{max}}$ : 213 nm
<b>Supplied as:</b>	A solid
<b>Storage:</b>	-20°C
<b>Stability:</b>	≥2 years



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

### Laboratory Procedures

3-Methylglutaconic acid (3-MGA) is supplied as a solid. A stock solution may be made by dissolving the 3-MGA in the solvent of choice, which should be purged with an inert gas. 3-MGA is soluble in organic solvents such as ethanol, DMSO, and dimethyl formamide. The solubility of 3-MGA in these solvents is approximately 30 mg/ml.

Further dilutions of the stock solution into aqueous buffers or isotonic saline should be made prior to performing biological experiments. Ensure that the residual amount of organic solvent is insignificant, since organic solvents may have physiological effects at low concentrations. Organic solvent-free aqueous solutions of 3-MGA can be prepared by directly dissolving the solid in aqueous buffers. The solubility of 3-MGA in PBS (pH 7.2) is approximately 10 mg/ml. We do not recommend storing the aqueous solution for more than one day.

### Description

3-MGA is a branched-chain organic acid and product of leucine catabolism.<sup>1</sup> 3-MGA accumulates in 3-methylglutaconic aciduria, which is caused by an inborn error of leucine metabolism characterized by 3-MGA-CoA hydratase deficiency, leading to delayed development, hypoglycemia, metabolic acidosis, and neurological impairments.<sup>1,2</sup>

### References

1. Wortmann, S.B., Kluijtmans, L.A., Engelke, U.F.H., *et al.* The 3-methylglutaconic acidurias: What's new? *J. Inherit. Metab. Dis.* **35(1)**, 13-22 (2012).
2. Leipnitz, G., Seminotti, B., Amaral, A.U., *et al.* Induction of oxidative stress by the metabolites accumulating in 3-methylglutaconic aciduria in cerebral cortex of young rats. *Life Sci.* **82(11-12)**, 652-662 (2008).

#### 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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