

# ALDH5A1 dehydrogenates SUCCSA to SUCCA

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## Introduction

Reactome is open-source, open access, manually curated and peer-reviewed pathway database. Pathway annotations are authored by expert biologists, in collaboration with Reactome editorial staff and cross-referenced to many bioinformatics databases. A system of evidence tracking ensures that all assertions are backed up by the primary literature. Reactome is used by clinicians, geneticists, genomics researchers, and molecular biologists to interpret the results of high-throughput experimental studies, by bioinformaticians seeking to develop novel algorithms for mining knowledge from genomic studies, and by systems biologists building predictive models of normal and disease variant pathways.

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## Literature references

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- Fabregat, A., Jupe, S., Matthews, L., Sidiropoulos, K., Gillespie, M., Garapati, P. et al. (2018). The Reactome Pathway Knowledgebase. *Nucleic Acids Res*, 46, D649-D655. [↗](#)
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Reactome database release: 88

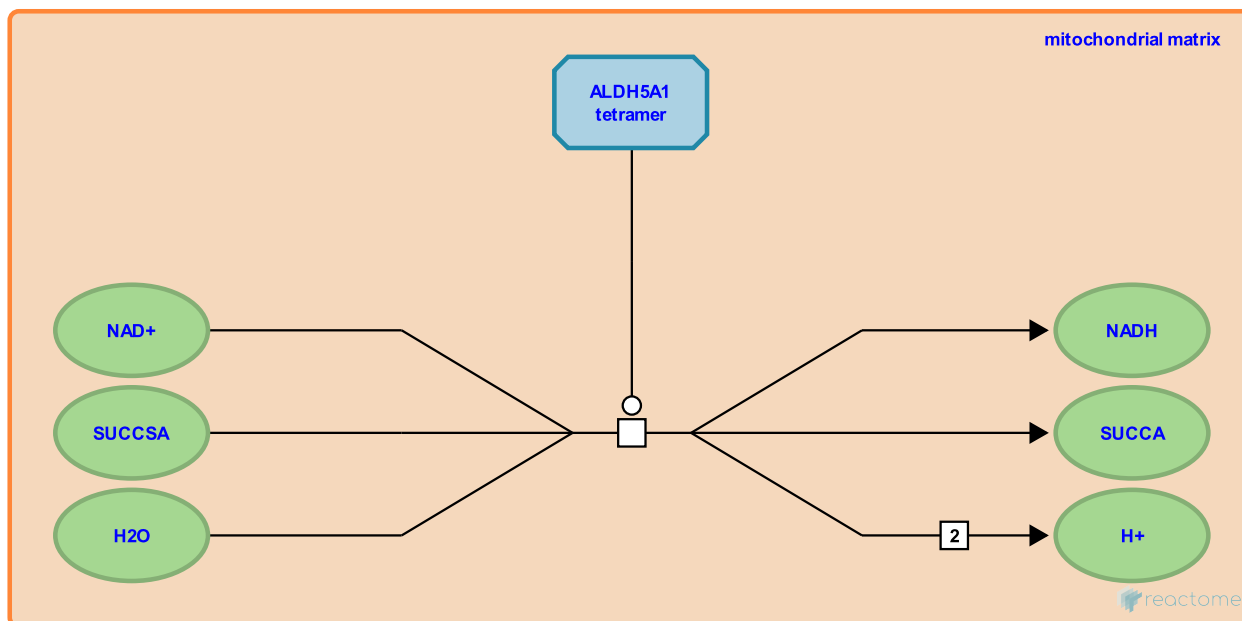
This document contains 1 reaction ([see Table of Contents](#))

## ALDH5A1 dehydrogenates SUCCSA to SUCCA [↗](#)

**Stable identifier:** R-HSA-888548

**Type:** transition

**Compartments:** mitochondrial matrix



Mitochondrial succinate semialdehyde dehydrogenase (ALDH5A1) tetramer catalyzes the reaction of succinate semialdehyde (SUCCSA), H<sub>2</sub>O, and NAD<sup>+</sup> to form succinate (SUCCA) and NADH + 2 H<sup>+</sup> (Kim et al. 2009). Mutations that inactivate this gene cause 4-hydroxybutyric aciduria (succinic semialdehyde dehydrogenase deficiency) (Chambliss et al. 1998).

### Literature references

Chambliss, KL., Gibson, KM., Hinson, DD., Trettel, F., Jakobs, C., Malaspina, P. et al. (1998). Two exon-skipping mutations as the molecular basis of succinic semialdehyde dehydrogenase deficiency (4-hydroxybutyric aciduria). *Am. J. Hum. Genet.*, 63, 399-408. [↗](#)

Kim, KJ., Park, SY., Lee, S., Kim, YG., Kwon, OS., Park, BJ. et al. (2009). Redox-switch modulation of human SSADH by dynamic catalytic loop. *EMBO J*, 28, 959-68. [↗](#)

### Editions

2008-11-27	Reviewed	Restituto, S.
2010-06-30	Authored	Mahajan, SS.
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