

# BBOX1:AscH-:Fe2+ dimer dioxygenates TEABT and 2OG to form CAR and SUCCA

D'Eustachio, P.

European Bioinformatics Institute, New York University Langone Medical Center, Ontario Institute for Cancer Research, Oregon Health and Science University.

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https://reactome.org

## 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

- Fabregat, A., Sidiropoulos, K., Viteri, G., Forner, O., Marin-Garcia, P., Arnau, V. et al. (2017). Reactome pathway analysis: a high-performance in-memory approach. *BMC bioinformatics*, 18, 142.
- Sidiropoulos, K., Viteri, G., Sevilla, C., Jupe, S., Webber, M., Orlic-Milacic, M. et al. (2017). Reactome enhanced pathway visualization. *Bioinformatics*, 33, 3461-3467.
- 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.
- Fabregat, A., Korninger, F., Viteri, G., Sidiropoulos, K., Marin-Garcia, P., Ping, P. et al. (2018). Reactome graph database: Efficient access to complex pathway data. *PLoS computational biology, 14*, e1005968.

Reactome database release: 88

This document contains 1 reaction (see Table of Contents)

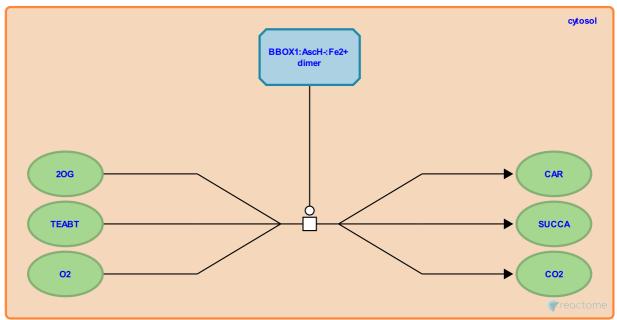
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# BBOX1:AscH-:Fe2+ dimer dioxygenates TEABT and 20G to form CAR and SUCCA 7

**Stable identifier:** R-HSA-71261

Type: transition

**Compartments:** cytosol



Cytosolic gamma-butyrobetaine hydroxylase dimer (BBOX1), a dioxygenase, catalyzes the reaction of oxygen, 4-trimethylammoniobutanoate (TEABT), and 2-oxoglutarate (2OG)to form CO2, succinate (SUCCA), and carnitine (CAR) (Lindstedt and Nordin 1984; Tars et al. 2010; Vaz et al. 1998).

## Literature references

Nordin, I., Lindstedt, S. (1984). Multiple forms of gamma-butyrobetaine hydroxylase (EC 1.14.11.1). *Biochem J*, 223, 119-27.

Kuka, J., Kotelovica, S., Rumnieks, J., Viksna, A., Tars, K., Kazaks, A. et al. (2010). Crystal structure of human gamma-butyrobetaine hydroxylase. *Biochem Biophys Res Commun*, 398, 634-9.

Vaz, FM., Ijlst, L., van Gool, S., Ofman, R. (1998). Carnitine biosynthesis: identification of the cDNA encoding human gamma-butyrobetaine hydroxylase. *Biochem Biophys Res Commun*, 250, 506-10. *▶* 

### **Editions**

2022-07-19 Revised D'Eustachio, P.