ASS1 tetramer:NMRAL1 dimer:NADPH transforms L-Asp and L-Cit to ARSUA

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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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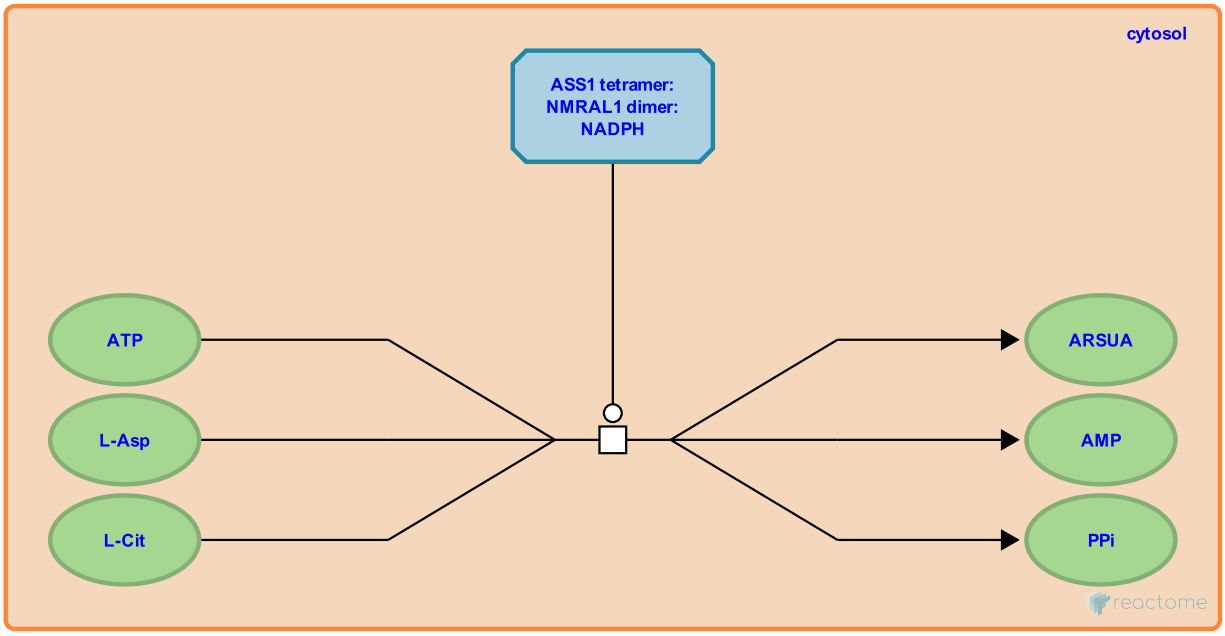
Stable identifier: R-HSA-70577

Type: transition

Compartments: cytosol



Cytosolic argininosuccinate synthase (ASS1 tetramer) catalyzes the reaction of aspartate (L-Asp), citrulline (L-Cit), and ATP to form argininosuccinate (ARSUA), AMP, and pyrophosphate (PPi). The function of the human enzyme in vivo is inferred from the hypercitrullinemia observed in patients with defective forms of the enzyme (e.g., Engel et al. 2009). The enzyme is active as a homotetramer (O'Brien 1980; Karlberg et al. 2008) and binds NmrA-like family domain-containing protein 1 (NMRAL1). NMRAL1 is a redox sensor protein that can undergo restructuring and subcellular redistribution in response to changes in intracellular NADPH/NADP+ levels. Under normal NADPH levels, it can form an asymmetrical dimer with one subunit occupied by one NADPH molecule, hiding the binding site for ASS1 thus impairing its activity and reducing the production of nitric oxide (Zheng et al. 2007, Zhao et al. 2008).

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Editions

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