**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.

The development of Reactome is supported by grants from the US National Institutes of Health (P41 HG003751), University of Toronto (CFREF Medicine by Design), European Union (EU STRP, EMI-CD), and the European Molecular Biology Laboratory (EBI Industry program).

**Literature references**


Reactome database release: 68

This document contains 1 pathway and 2 reactions (see Table of Contents)
The activation of arylsulfatases

Stable identifier: R-HSA-1663150

Sulfatase activity requires a unique posttranslational modification (PTM) of a catalytic cysteine residue into a formylglycine. This modification is impaired in patients with multiple sulfatase deficiency (MSD) due to defects in the SUMF1 (sulfatase-modifying factor 1) gene responsible for this PTM. SUMF2 can inhibit the activity of SUMF1 thereby providing a mechanism for the regulation of sulfatase activation (Ghosh 2007, Diez-Roux & Ballabio 2005).

Literature references


Editions

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**SUMF1 mediates the oxidation of cysteine to formylglycine, producing active arylsulfatases**

**Location:** The activation of arylsulfatases

**Stable identifier:** R-HSA-1614362

**Type:** transition

**Compartments:** endoplasmic reticulum lumen

The sulfatase-modifying factor 1 (SUMF1, also called C-alpha-formylglycine-generating enzyme, FGE) (Preusser-Kunze et al. 2005, Cosma et al. 2003, Landgrebe et al. 2003) oxidises the critical cysteine residue in arylsulfatases to an active site 3-oxoalanine residue thus conferring sulfatase activity (Roeser et al. 2006). Defects in SUMF1 cause multiple sulfatase deficiency (MSD) (MIM:272200), an impairment of arylsulfatase activity due to defective post-translational modification of the cysteine residue (Cosma et al. 2003, Dierks et al, 2003). This post-translational modification is thought to be highly conserved in eukaryotes (Selmer et al. 1996, von Figura et al. 1998). SUMF1 is active as either a monomer or a homodimer. A monomer is described in this reaction.

**Literature references**


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Sulfatase-modifying factor 2 (SUMF2, also called C-alpha-formylglycine-generating enzyme 2, pFGE) is the parologue of SUMF1. While SUMF1 can modify a critical residue on arylsulfatases to confer activity to them, SUMF2 lacks this ability (Mariappan et al. 2005) and instead, SUMF2 can inhibit the action of SUMF1 by dimerising with it (Zito et al. 2005). SUMF2 can interact with sulfatases with and without SUMF1 (Zito et al. 2005).

**Literature references**


**Editions**

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