Dermatan sulfate biosynthesis

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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


Reactome database release: 77

This document contains 1 pathway and 4 reactions (see Table of Contents)
Dermatan sulfate biosynthesis

Stable identifier: R-HSA-2022923

Dermatan sulfate (DS) consists of N-acetylgalactosamine (GalNAc) residues alternating in glycosidic linkages with glucuronic acid (GlcA) or iduronic acid (IdoA) residues. As with CS, GalNAc residues can be sulfated in CS chains but also the uronic acid residues may be substituted with sulfate at the 2- and 4- positions. The steps below outline the synthesis of a simple DS chain (Silbert & Sugumaran 2002).

Literature references


Editions

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Dermatan-sulfate epimerase (DSE) converts chondroitin sulfate (CS) to dermatan sulfate (DS)

**Location:** Dermatan sulfate biosynthesis

**Stable identifier:** R-HSA-2022052

**Type:** transition

**Compartments:** Golgi membrane, Golgi lumen

The glucuronate (GlcA) moiety of chondroitin sulfate (CS) can undergo C-5 epimerization to change into an iduronic acid (IdoA) moiety, thus changing the polymer composition and creating dermatan sulfate (DS). The GlcA of unsulfated chondroitin chains can also undergo this C-5 epimerization to produce dermatan. Dermatan-sulfate epimerase (DSE) mediates these reactions (Tiedemann et al. 2001). More recently, a single homologue of DSE, dermatan sulfate epimerase-like (DSEL), has been determined to possess epimerase activity (Pacheco et al. 2009). DSEL is genetically associated with type II bipolar disorder (Goossens et al. 2003).

**Followed by:** DSPGs are secreted, Dermatan sulfate can be further sulfated on position 2 of iduronate, CHST14 transfers SO4(2-) to GalNAc in dermatan or DS

**Literature references**


**Editions**

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**Dermatan sulfate can be further sulfated on position 2 of iduronate**

**Location:** Dermatan sulfate biosynthesis

**Stable identifier:** R-HSA-2022061

**Type:** transition

**Compartments:** Golgi membrane, Golgi lumen

Uronyl 2-sulfotransferase (UST) catalyzes the transfer of sulfate from PAPS to position 2 of iduronyl residues in dermatan sulfate (Kobayashi et al. 1999).

**Preceded by:** Dermatan-sulfate epimerase (DSE) converts chondroitin sulfate (CS) to dermatan sulfate (DS)

**Followed by:** CHST14 transfers SO4(2-) to GalNAc in dermatan or DS, DSPGs are secreted

**Literature references**


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https://reactome.org
**CHST14 transfers SO4(2-) to GalNAc in dermatan or DS**

**Location:** Dermatan sulfate biosynthesis

**Stable identifier:** R-HSA-2022063

**Type:** transition

**Compartments:** Golgi membrane, Golgi lumen

Important functional domains in dermatan or dermatan sulfate (DS) are generated by the action of an epimerase (which converts D-glucuronic acid into its epimer L-iduronic acid) together with 4-O-sulfation. These domains are named 4-O-sulfated iduronic acid blocks (Pachebo et al. 2009). Carbohydrate sulfotransferase 14 (CHST14) (Evers et al. 2001) mediates the transfer of sulfate to position 4 of another N-acetylgalactosamine (GalNAc) residue of D2,4(S)2-PG (sulfated on position 2 of IdoA and position 4 of GalNAc) to produce a further sulfated product D2,4,4(S)3-PG (sulfated on another GalNAc in addition to the ones above).

**Preceded by:** Dermatan sulfate can be further sulfated on position 2 of iduronate, Dermatan-sulfate epimerase (DSE) converts chondroitin sulfate (CS) to dermatan sulfate (DS)

**Followed by:** DSPGs are secreted

**Literature references**


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DSPGs are secreted

**Location:** Dermatan sulfate biosynthesis

**Stable identifier:** R-HSA-2022065

**Type:** omitted

**Compartments:** Golgi lumen, extracellular region

Various forms of dermatan sulfate are excreted from the cell once formed. The mechanism of transport is unknown but most likely involves the trans-golgi network (Silbert & Sugumaran 2002).

**Preceded by:** Dermatan-sulfate epimerase (DSE) converts chondroitin sulfate (CS) to dermatan sulfate (DS), Dermatan sulfate can be further sulfated on position 2 of iduronate, CHST14 transfers SO4(2-) to GalNAc in dermatan or DS

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