Fibrillin microfibril assembly

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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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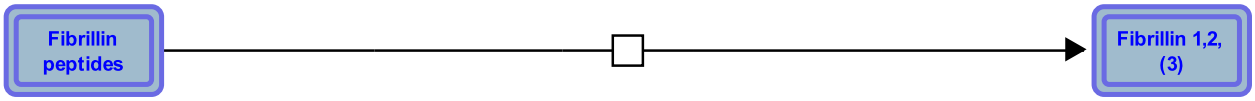
Stable identifier: R-HSA-2129362

Type: transition

Compartments: extracellular region



Fibrillin microfibril assembly is a cell regulated process, independent of tropoelastin. Distinct microfibril populations have been identified, suggesting that the cellular environment plays a role in regulating microfibril fate (Kielty et al. 2002). Fibrillin 1 may undergo limited assembly into dimers or trimers in the secretory pathway (Ashworth et al. 1999, Trask et al. 1999) but the formation of large microfibril polymers is extracellular. Microfibrils assemble close to the cell surface in a process that may require cell surface receptors. Fibrillins interact with several integrins (Sakamoto et al. 1996, Jovanovic et al. 2008) suggesting an assembly mechanism with similarities to fibronectin matrix formation. Heparan sulphate proteoglycans (HSPGs) and chondroitin sulfate containing proteoglycans (CSPGs) have also been proposed to have a role in assembly (Tiedemann et al. 2001). Fibrillin polymerization into fibres further requires the formation of disulfide bonds between fibrillins (Reinhardt et al. 2000), initially via calcium-binding epidermal growth factor domains at the C-terminus (Hubmacher et al. 2008), and transglutaminase cross-links (Kielty et al. 2002).

Literature references

Kielty, CM., Ashworth, J., Wess, TJ., Shuttleworth, CA., Baldock, C., Purslow, PP. et al. (2001). Fibrillin-rich microfibrils of the extracellular matrix: ultrastructure and assembly. *Micron, 32*, 185-200. ↗

Editions

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