

DCN Gene

Subjects: **Genetics & Heredity**

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Decorin

genes

1. Normal Function

The *DCN* gene provides instructions for making a protein called decorin. This protein is a component of the extracellular matrix, which is the intricate lattice of proteins and other molecules that forms in the spaces between cells. Decorin is found in the extracellular matrix of a variety of connective tissues, including skin, tendon, bone, and cartilage. Connective tissues support the body's joints and organs.

Decorin is involved in the organization of proteins called collagens. Collagens strengthen and support connective tissues throughout the body. Collagens also play an important role in the cornea, which is the clear outer covering of the eye. Bundles of collagen called fibrils must be strictly organized for the cornea to be transparent. Decorin ensures that these collagen fibrils are uniformly sized and regularly spaced.

Researchers have proposed several additional functions for decorin. This protein likely helps regulate cell growth and division, the attachment of cells to one another (cell adhesion), and the self-destruction of cells (apoptosis). Studies suggest that decorin plays a role in the formation of new blood vessels (angiogenesis), wound healing, bone development, inflammation, and preventing the growth of cancerous tumors. Decorin also regulates the activity of several growth factors, including transforming growth factor-beta (TGF β). These growth factors control a diverse range of processes important for cell growth.

2. Health Conditions Related to Genetic Changes

2.1 Congenital Stromal Corneal Dystrophy

Several mutations in the *DCN* gene have been identified in families with congenital stromal corneal dystrophy. Each of the known mutations leads to the production of an abnormally short version of the decorin protein. This abnormal protein interferes with the organization of collagen fibrils in the cornea. As poorly arranged collagen fibrils accumulate, the cornea becomes cloudy. These corneal changes lead to a loss of sharp vision (reduced visual acuity) and other eye abnormalities related to visual impairment.

3. Other Names for This Gene

- bone proteoglycan II
- decorin proteoglycan
- dermatan sulphate proteoglycans II
- DSPG2
- PG40
- PGII
- PGS2
- PGS2_HUMAN
- proteoglycan core protein
- SLRR1B
- small leucine-rich protein 1B

References

1. Bredrup C, Knappskog PM, Majewski J, Rødahl E, Boman H. Congenital stromaldystrophy of the cornea caused by a mutation in the decorin gene. Invest Ophthalmol Vis Sci. 2005 Feb;46(2):420-6.
2. Danielson KG, Fazzio A, Cohen I, Cannizzaro LA, Eichstetter I, Iozzo RV. The human decorin gene: intron-exon organization, discovery of two alternatively spliced exons in the 5' untranslated region, and mapping of the gene to chromosome 12q23. Genomics. 1993 Jan;15(1):146-60.
3. Rødahl E, Van Ginderdeuren R, Knappskog PM, Bredrup C, Boman H. A second decorin frame shift mutation in a family with congenital stromal corneal dystrophy. Am J Ophthalmol. 2006 Sep;142(3):520-1.
4. Rühland C, Schönherr E, Robenek H, Hansen U, Iozzo RV, Bruckner P, Seidler DG. The glycosaminoglycan chain of decorin plays an important role in collagen fibril formation at the early stages of fibrillogenesis. FEBS J. 2007 Aug;274(16):4246-55.

5. Zhang G, Chen S, Goldoni S, Calder BW, Simpson HC, Owens RT, McQuillan DJ, Young MF, Iozzo RV, Birk DE. Genetic evidence for the coordinated regulation of collagen fibrillogenesis in the cornea by decorin and biglycan. *J Biol Chem*. 2009 Mar 27;284(13):8888-97. doi: 10.1074/jbc.M806590200.
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