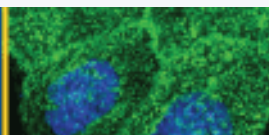


Mice deficient in small leucine-rich proteoglycans: novel *in vivo* models for osteoporosis, osteoarthritis, Ehlers-Danlos syndrome, muscular dystrophy, and corneal.

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# Mice deficient in small leucine-rich proteoglycans: novel *in vivo* models for osteoporosis, osteoarthritis, Ehlers-Danlos syndrome, muscular dystrophy, and corneal diseases

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[Laurent Ameye, Marian F. Young](#)

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## Abstract

Small leucine-rich proteoglycans (SLRPs) are extracellular molecules that bind to TGF $\beta$ s and collagens and other matrix molecules. *In vitro*, SLRPs were shown to regulate collagen fibrillogenesis, a process essential in development, tissue repair, and metastasis. To better understand their functions *in vivo*, mice deficient in one or two of the four most prominent and widely expressed SLRPs (biglycan, decorin, fibromodulin, and lumican) were recently generated. All four SLRP deficiencies result in the formation of abnormal collagen fibrils. Taken together, the collagen phenotypes demonstrate a cooperative, sequential, timely orchestrated action of the SLRPs that altogether shape the architecture and mechanical properties of the collagen matrix. In addition, SLRP-deficient mice develop a wide array of diseases (osteoporosis, osteoarthritis, muscular dystrophy, Ehlers-Danlos syndrome, and corneal diseases), most of them resulting primarily from an abnormal collagen fibrillogenesis. The development of these diseases by SLRP-deficient mice suggests that mutations in SLRPs may be part of undiagnosed predisposing genetic factors for these diseases. Although the distinct phenotypes developed by the different singly deficient mice point to distinct *in vivo* function for each SLRP, the analysis of the double-deficient mice also demonstrates the existence of rescuing/compensation mechanisms, indicating some functional overlap

within the SLRP family.

**Keywords:** *Key words:* [biglycan/collagen/decorin/fibromodulin/lumican](#)

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