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Regulation of Fibrosis in Muscular Dystrophy

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Highlights

- Muscular dystrophy pathology creates an imbalance between myogenic and fibrogenic pathways, and leads to progressive fibrotic replacement of skeletal muscle.
- A primary signaling molecule promoting fibrosis is TGFβ, but many additional factors including myostatin, hypoxia, and mechanical stiffness contribute to the fibrotic accumulation.
- Fibrosis compromises the ability for muscle satellite cells to coordinate with inflammatory cells and fibro/adipogenic progenitors to mediate regeneration.
- A major consequence of fibrosis is that it is a barrier to efficient cell based therapies for muscle disease.
- Fibrosis results from increased myofibroblast extracellular matrix production. Cross-linking and strain on the matrix reduces protease mediated extracellular matrix degradation and enhances stiffness to support fibrotic progression.

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