Fibrosis development in early-onset muscular dystrophies: Mechanisms and translational implications.

Serrano AL, Muñoz-Cánoves P.

Abstract

Duchenne muscular dystrophy (DMD) is one of the most devastating neuromuscular genetic diseases caused by the absence of dystrophin. The continuous episodes of muscle degeneration and regeneration in dystrophic muscle are accompanied by chronic inflammation and fibrosis deposition, which exacerbate disease progression. Thus, in addition of investigating strategies to cure the primary defect by gene/cell therapeutic strategies, increasing efforts are being placed on identifying the causes of the substitution of muscle by non-functional fibrotic tissue in DMD, aiming to attenuate its severity. Congenital muscular dystrophies (CMDs) are early-onset diseases in which muscle fibrosis is also present. Here we review the emerging findings on the mechanisms that underlie fibrogenesis in muscular dystrophies, and potential anti-fibrotic treatments.

KEYWORDS: CMD; DMD; Fibrosis; Inflammation; Muscular dystrophy; Regeneration; Skeletal muscle

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