Emery-Dreifuss Muscular Dystrophy-linked genes and Centronuclear myopathy-linked genes regulate myonuclear movement by distinct mechanisms.

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Abstract
Muscle cells are a syncytium in which the many nuclei are positioned to maximize the distance between adjacent nuclei. Although mispositioned nuclei are correlated with many muscle disorders, it is not known whether this common phenotype is the result of a common mechanism. To answer this question, the expression of genes linked to Emery-Dreifuss Muscular Dystrophy (EDMD) and Centronuclear myopathy (CNM) was disrupted in Drosophila, and the position of the nuclei was evaluated. We found that the genes linked to EDMD and CNM were each necessary to properly position nuclei. However, the specific phenotypes were different. EDMD-linked genes were necessary for the initial separation of nuclei into distinct clusters, suggesting that these factors relieve interactions between nuclei. CNM-linked genes were necessary to maintain the nuclei within clusters as they moved toward the muscle ends, suggesting that these factors were necessary to maintain interactions between nuclei. Together these data suggest that nuclear position is disrupted by distinct mechanisms in EDMD and CNM.

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