Current insights into LMNA cardiomyopathies: Existing models and missing LINCs.

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Abstract
The nuclear lamina is a critical structural domain for the maintenance of genomic stability and whole-cell mechanics. Mutations in the LMNA gene, which encodes nuclear A-type lamins lead to the disruption of these key cellular functions, resulting in a number of devastating diseases known as laminopathies. Cardiomyopathy is a common laminopathy and is highly penetrant with poor prognosis. To date, cell mechanical instability and dysregulation of gene expression have been proposed as the main mechanisms driving cardiac dysfunction, and indeed discoveries in these areas have provided some promising leads in terms of therapeutics. However, important questions remain unanswered regarding the role of lamin A dysfunction in the heart, including a potential role for the toxicity of lamin A precursors in LMNA cardiomyopathy, which has yet to be rigorously investigated.

KEYWORDS: LINC complex; LMNA; cardiomyocyte; cardiomyopathy; mechanotransduction; nuclear lamina; prelamin A

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