Lamins and Lamin-associated Proteins in Gastrointestinal Health and Disease.

Brady GF¹, Kwan R², Bragazzi Cunha J², Elenbaas JS³, Omary MB⁴.

Abstract
The nuclear lamina is a multi-protein lattice composed of A- and B-type lamins and their associated proteins. This protein lattice associates with heterochromatin and integral inner nuclear membrane proteins, providing a link between the genome, nucleoskeleton, and cytoskeleton. In the 1990s, mutations in EMD and LMNA were linked to Emery-Dreifuss muscular dystrophy. Since then, the number of diseases attributed to nuclear lamina defects, including laminopathies and other disorders, has increased to include more than 20 distinct genetic syndromes. Studies of patients and mouse genetic models have indicated the important roles for lamins and their associated proteins in the function of gastrointestinal organs including liver and pancreas. We review the interactions and functions of the lamina in relation to the nuclear envelope and genome, the ways in which its dysfunction is thought to contribute to human disease, and possible avenues for targeted therapies.

KEYWORDS: envelopathies; lipodystrophy; myopathy; neuropathy; nonalcoholic fatty liver disease; nucleoskeleton; progeria

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