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Format: AbstractMuscle Nerve. 2017 Feb 22. doi: 10.1002/mus.25629. [Epub ahead of print]

Electrical impedance myography (EIM) in individuals with COL6 and LAMA2 congenital muscular dystrophy: a cross-sectional and two-year analysis.

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Abstract

INTRODUCTION: Electrical impedance myography (EIM) is a non-invasive electrophysiological technique that characterizes muscle properties through bioimpedance. We compared EIM measurements to function, strength, and disease severity in a population with congenital muscular dystrophy (CMD).

METHODS: Forty-one patients with CMD, either collagen 6 related disorders (COL6-RD) ($n = 21$) or laminin alpha 2-related disorders (LAMA2-RD) ($n = 20$) and 21 healthy pediatric controls underwent 2 yearly EIM exams. In the CMD cohorts, EIM was compared to functional and strength measurements.

RESULTS: Both CMD cohorts exhibited change over time and had correlation with disease severity. The 50 kHz phase correlated well with function and strength in the COL6-RD cohort but not in the LAMA2-RD cohort.

DISCUSSION: EIM is a potentially useful measure in clinical studies with CMD because of its sensitivity to change over a 1-year period and correlation with disease severity. For COL6-RD, there were also functional and strength correlations. This article is protected by copyright. All rights reserved.

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KEYWORDS: COL6 related dystrophies; LAMA2 related dystrophies; congenital muscular dystrophy; electrical impedance myography; neuromuscular disorders

PMID: 28224647 DOI: [10.1002/mus.25629](https://doi.org/10.1002/mus.25629)

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