Improving Reproducibility of Phenotypic Assessments in the DyW Mouse Model of Laminin-α2 Related Congenital Muscular Dystrophy.

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Abstract
Laminin-α2 related Congenital Muscular Dystrophy (LAMA2-CMD) is a progressive muscle disease caused by partial or complete deficiency of laminin-211, a skeletal muscle extracellular matrix protein. In the last decade, basic science research has queried underlying disease mechanisms in existing LAMA2-CMD murine models and identified possible clinical targets and pharmacological interventions. Experimental rigor in preclinical studies is critical to efficiently and accurately quantify both negative and positive results, degree of efficiency of potential therapeutics and determine whether to move a compound forward for additional preclinical testing. In this review, we compare published available data measured to assess three common parameters in the widely used mouse model DyW, that mimics LAMA2-CMD, we quantify variability and analyse its possible sources. Finally, on the basis of this analysis, we suggest standard set of assessments and the use of available standardized protocols, to reduce variability of outcomes in the future and to improve the value of preclinical research.

KEYWORDS: LAMA2-CMD; LAMA2-RD; MDC1A; mouse models; preclinical

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