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Effects of metformin on congenital muscular dystrophy type 1A disease progression in mice: a gender impact study.

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Abstract

Congenital muscular dystrophy with laminin α2 chain-deficiency (LAMA2-CMD) is a severe muscle disorder with complex underlying pathogenesis. We have previously employed profiling techniques to elucidate molecular patterns and demonstrated significant metabolic impairment in skeletal muscle from LAMA2-CMD patients and mouse models. Thus, we hypothesize that skeletal muscle metabolism may be a promising pharmacological target to improve muscle function in LAMA2-CMD. Here, we have investigated whether the multifunctional medication metformin could be used to reduce disease in the dy^{2J}/dy^{2J} mouse model of LAMA2-CMD. First, we show gender disparity for several pathological hallmarks of LAMA2-CMD. Second, we demonstrate that metformin treatment significantly increases weight gain and energy efficiency, enhances muscle function and improves skeletal muscle histology in female dy^{2J}/dy^{2J} mice (and to a lesser extent in dy^{2J}/dy^{2J} males). Thus, our current data suggest that metformin may be a potential future supportive treatment that improves many of the pathological characteristics of LAMA2-CMD.

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