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Childhood Activity on Progression in Limb Girdle Muscular Dystrophy 2I.

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Abstract

Limb girdle muscular dystrophy 2I is a slowly progressive muscular dystrophy due to mutations in the Fukutin-related protein (FKRP) gene. Clinicians are frequently asked if physical activity is harmful for pediatric patients with limb girdle muscular dystrophy 2I. The primary objective of this study was to determine if there is a relationship between self-reported childhood activity level and motor function and respiratory function in older children and adults with limb girdle muscular dystrophy 2I. We compared retrospective self-reported middle school activity level and sport participation with age at onset of weakness, 10-meter walk test, and forced vital capacity later in life in 41 participants with FKRP mutations. We found no relationship between activity level in childhood and disease course later in life, suggesting that self-directed physical activity in children with limb girdle muscular dystrophy 2I does not negatively affect disease progression and outcome.

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KEYWORDS: FKRP; dystroglycanopathy; exercise; muscular dystrophies; α -dystroglycan

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