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Cardiomyopathy in limb girdle muscular dystrophy R9, FKRP related

Eric M Libell¹, Julia A Richardson¹, Katie L Lutz¹, Benton Y Ng¹, Shelley R H Mockler², Katie M Laubscher², Carrie M Stephan¹, Bridget M Zimmerman³, Erik R Edens⁴, Benjamin E Reinking¹, Katherine D Mathews^{1 5}

Affiliations

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Abstract

Introduction: Reported frequencies of cardiomyopathy in limb girdle muscular dystrophy R9 (LGMDR9) vary. We describe the frequency and age at onset of cardiomyopathy in an LGMDR9 cohort.

Methods: Echocardiograms from 56 subjects (157 echocardiograms) with LGMDR9 were retrospectively reviewed. The cumulative probability of having an abnormal echocardiogram as a function of age was assessed by survival analysis for interval-censored data by genotype. Correlations between cardiac and clinical function were evaluated.

Results: Twenty-five (45%) participants had cardiomyopathy. The median age at first abnormal echocardiogram for subjects homozygous for the c.826C>A variant was 54.2 y compared to 18.1 y for all other fukutin-related protein (FKRP) genotypes ($P < .0001$). There was a weak correlation between ejection fraction and 10-Meter Walk Test speed ($r = 0.25$), but no correlation with forced vital capacity ($r = 0.08$).

Discussion: Cardiomyopathy is prevalent among those with LGMDR9 and occurs later in subjects homozygous for the c.826C>A mutation. These data will help to guide surveillance and management.

Keywords: All neuromuscular disease; FKRP; cardiomyopathy; dystroglycanopathy; limb-girdle muscular dystrophy; muscular dystrophy.

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