

## Opis choroby \*

### Definicja

A rare subtype of autosomal recessive limb-girdle muscular dystrophy characterized by atrioventricular block resulting in repeated syncope episodes, elevated creatine kinase serum levels and adult-onset of slowly progressive proximal limb skeletal muscle weakness and atrophy. Muscular dystrophic changes observed in muscle biopsy include diameter variability, increased central nuclei, and presence of necrotic and regenerating fibers.

### Dane

Klasyfikacja	Synonimy
Choroba	Autosomal recessive limb-girdle muscular dystrophy-cardiac arrhythmia syndrome LGMD2X Zespół autosomalnej recesywnej dystrofii obręczowo-konczynowej i arytmii serca BVES-related LGMD LGMD type 2X LGMD2X Limb-girdle muscular dystrophy 2X

**Kod ORPHA**  
476084

**Kod OMIM**  
616812

**Kod ICD10**  
G71.0

### Kod ICD11

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### \*Źródło

orphanet