Cardiac manifestations of congenital LMNA-related muscular dystrophy in children: three case reports and recommendations for care.


Abstract
Skeletal and cardiac muscle laminopathies, caused by mutations in the lamin A/C gene, have a clinical spectrum from congenital LMNA-related muscular dystrophy to later-onset Emery-Dreifuss muscular dystrophy, limb girdle muscular dystrophy, and dilated cardiomyopathy. Although cardiac involvement is observed at all ages, it has only been well described in adults. We present the evolution of cardiac disease in three children with congenital muscular dystrophy presentation of LMNA-related muscular dystrophy. In this series, atrial arrhythmia was the presenting cardiac finding in all three patients. Heart failure developed up to 5 years later. Symptoms of right heart failure, including diarrhoea and peripheral oedema, preceded a rapid decline in left ventricular ejection fraction. Recommendations for cardiac surveillance and management in these patients are made.

KEYWORDS: Lamin A/C LMNA; arrhythmia; automatic implantable cardiac defibrillator; cardiomyopathy; congenital muscular dystrophy

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