Severe congenital RYR1-associated myopathy complicated with atrial tachycardia and sinus node dysfunction: a case report.

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Abstract

BACKGROUND: Cardiac arrhythmias are sometimes encountered in patients with hereditary myopathies and muscular dystrophies. Description of arrhythmias in myopathies and muscular dystrophies is very important, because arrhythmias have a strong impact on the outcomes for these patients and are potentially treatable.

CASE PRESENTATION: A girl with severe congenital RYR1-related myopathy exhibited atrial tachycardia and sinus node dysfunction during infancy. She was born after uncomplicated caesarian delivery. She showed no breathing, complete ophthalmoplegia, complete bulbar paralysis, complete facial muscle paralysis, and extreme floppiness. At 5 months old, she developed persistent tachycardia around 200-210 beats per minutes. Holter monitoring revealed ectopic atrial tachycardia during tachyarrhythmia and occasional sinus pauses with junctional escape beats. Propranolol effectively alleviated tachyarrhythmia but was discontinued due to increased frequency and duration of the sinus pauses that led to bradyarrhythmia. There was no evidence of structural heart diseases or heart failure. The arrhythmia gradually resolved spontaneously and at 11 months old, she showed complete sinus rhythm.

CONCLUSIONS: Although supraventricular arrhythmia is sometimes encountered in congenital myopathies, this is the first report of cardiac arrhythmia requiring drug intervention in RYR1-associated myopathy.

KEYWORDS: Atrial tachycardia; Congenital myopathy; RYR1; Sinus node dysfunction

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