The Indian Journal of Pediatrics (May 2019) 86(5):480
https://doi.org/10.1007/s12098-019-02863-1

SCIENTIFIC LETTER

A Girl with Palpitations and Periodic Weakness

Debopam Samanta[1]

Received: 18 September 2018 / Accepted: 10 January 2019 / Published online: 18 January 2019

To the Editor: An 11-y-old girl presented with one-year history of episodic 1–2 d of leg weakness provoked by physical exertion and recent onset episodes of palpitation and syncope. Her physical examination was significant for short stature (125 cm, 2.2 centiles), small chin, bilateral clinodactyly, and a mild weakness (4/5) of the bilateral iliopsoas muscles. Laboratory testing revealed a low potassium level during the episodes of acute weakness. An EKG showed prolonged QTc interval and frequent premature ventricular contractions (PVCs) in bigeminal fashion. A multigene panel revealed a known pathogenic variant of *KCNJ2 Arg 218 Trp*, consistent with a diagnosis of Andersen-Tawil syndrome (ATS). Potassium supplementation and acetazolamide provided remarkable improvement in the severity and frequency of her periodic weakness. However, her ventricular ectopy and tachycardia remained unresponsive to several antiarrhythmics such as flecainide, nadolol, and verapamil, and her cardiac symptoms ultimately abated after cardioverter-defibrillator (ICD) implantation.

ATS, the rarest form of periodic paralysis, is characterized by a triad of episodic muscle weakness, ventricular arrhythmias, and dysmorphic features due to pathogenic variants in the *KCNJ2* gene which encodes inward rectifier potassium channel protein Kir 2.1 [1]. Differential diagnosis of episodic flaccid weakness includes hypokalemic, hyperkalemic, and thyrotoxic periodic paralyses. Episodes of acute weakness may respond to the correction of potassium concentration with or without chronic maintenance therapy. Carbonic anhydrase inhibitors have been used as prophylactic agents to reduce the frequency, severity, and the duration of the paralytic episodes [2].Empiric treatment with flecainide can be considered for significant and frequent ventricular arrhythmias in the setting of reduced left ventricular function [3]. Other antiarrhythmics such as beta-blockers, calcium channel blockers, and amiodarone have been used effectively also; however, some antiarrhythmics such as lidocaine, mexiletine, propafenone, and quinidine may exacerbate neuromuscular symptoms and should be used cautiously. Patients should avoid medicines that can prolong QT intervals. ICD implantation may be indicated in severe drug-resistant ventricular arrhythmias, especially if it is associated with syncope, left ventricular dysfunction, or cardiac arrest; however, bidirectional ventricular tachycardia typical for ATS is usually self-terminating and fatal outcome is rare compared to the other Long QT or Brugada syndromes [4].

Compliance with Ethical Standards

Conflict of Interest None.

Source of Funding None.

References

1. Tawil R, Ptacek LJ, Paclakis SG, et al. Andersen's syndrome; potassium-sensitive periodic paralysis, ventricular ectopy, and dysmorphic features. Ann Neurol. 1994;35:326–30.
2. Statland JM, Fontaine B, Hanna MG, et al. Review of the diagnosis and treatment of periodic paralysis. Muscle Nerve. 2018;57:522–30.
3. Miyamoto K, Aiba T, Kimura H, et al. Efficacy and safety of flecainide for ventricular arrhythmias in patients with Andersen-Tawil syndrome with KCNJ2 mutations. Heart Rhythm. 2015;12:596–603.
4. Bienias P, Kostera-Pruszczyk A, Bieganowska K, Miszczak-Knecht M, Pruszczyk P. Should a cardioverter-defibrillator be implanted in an Andersen-Tawil syndrome patient with severe ventricular arrhythmias and syncope? Kardiol Pol (Polish Heart J). 2014;72:755.

✉ Debopam Samanta
dsamanta@uams.edu

[1] Department of Pediatrics, University of Arkansas for Medical Sciences, 1 Children's Way, Little Rock, AR 72202, USA