Successful treatment with Flecanide in a patient with Andersen Tawil Syndrome Type I (ATS I) and incessant ventricular arrhythmias

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Introduction:
We report a case of 15 year old girl who presented unexpectedly during anaesthesia for velopharyngoplasty with numerous polymorphic ventricular extrasystoles (VES) and short runs of VT.

Methods:
ECG on admission showed sinus rhythm (SR) with regular time intervals (QTc 380) and prominent U wave.
24 hours ECG showed SR with very frequent VES (45% of all QRS complexes) 3129 bigemini, 3000 couplets, 973 triplets, 1365 short runs of VTs (2.3-12.7 sec, frequency 125-150/min) which increased during periods of rest.
Exercise test revealed decreased VES during maximal exercise (2.9 Watt/kg) and early recovery.
Tilt test showed, during position change from supine to standing, heart rate increased and the amount of VES decreased. VES increased again in supine position.
Echocardiography and cardiac MRT showed a structural and functional normal heart.
Clinical examination revealed mild dysmorphic features known in patients with Andersen-Tawil-Syndrome (ATS) like short stature, micrognathia, hypertelorism, broad nasal bridge.
Genetic analysis revealed a heterozygote mutation in the KCNJ2 Gene (Exon 2c.652C>T, p.Arg21^8Trp).
Family history included unexpected death of grandfather at the age of 36 years and two maternal cousins in their early thirties.

Results:
24 hours ECG under Propanolol 3,4 mg/kg showed slight increase in the duration of VTs otherwise no significant change.
24 hours ECG under Flecaïnide 3,0 mg/kg showed significant reduction of arrhythmias: 12% polymorphic VES of all QRS complexes, 1260 bigemini, 516 couplets, 12 triplets, 16 short runs of VTs (2.7-5.7 sec, frequency 111-140/min) which increased during periods of rest.
24 hours ECG under Flecaïnide 6,5 mg/kgKG showed only 450 VES of two morphologies mainly during periods of rest. No VTs, triplets or couplets.
24 hours ECG after three months treatment with Flecaïnide 6,5 mg/kg showed only 40 VES of two morphologies. No VTs, triplets or couplets.

Conclusion:
ATS I is a disorder of ventricular depolarisation caused by mutation in the KCNJ2 gene. It is characterised by a high burden of difficult to treat ventricular ectopics. In our patient treatment with Flecaïnide  6,5 mg/kg abolished all VT runs.