

Effects of flecainide therapy on arrhythmias and inappropriate shocks in children with catecholaminergic polymorphic ventricular tachycardia

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Purpose. Current recommendations for therapy of catecholaminergic polymorphic ventricular tachycardia (CPVT) include β -blockade and implantable cardioverter defibrillators (ICD). Patients may experience recurrent arrhythmias, ICD shocks and sudden death despite optimal medical therapy. It was recently shown that flecainide directly targets the molecular defect in CPVT by inhibiting premature Ca^{++} release. This study evaluated the efficacy and safety of flecainide in our CPVT children.

Methods. We collected data from the genotype-positive CPVT children started on flecainide in our tertiary pediatric cardiac center.

Results. The first case is a boy who was diagnosed with CPVT at 3 years of age. He was managed with nadolol (75 mg/m² BSA daily) with no further occurrence of syncope. At age 11, we noted bigeminal ventricular premature beats and couplets during exercise testing and one episode of sustained ventricular tachycardia on Holter recording despite nadolol titrated to maximum tolerable dose, so that an ICD was implanted. A few months later, inappropriate shocks were delivered due to episodes of sinus tachycardia. Holter monitor showed multiple episodes of sinus tachycardia and non-sustained ventricular tachycardia. Flecainide (100 mg = 4 mg/kg BW daily) was added to the β -blocker regimen. At 18-months follow-up, no ICD shock occurred and repetitive Holter recordings and exercise testings excluded sinus tachycardia and arrhythmias. The second case is a girl who was diagnosed with CPVT at 8 years of age. At age 13, we noted bigeminal ventricular premature beats and couplets during exercise testing and on Holter recording despite nadolol titrated to maximum tolerable dose. Flecainide (100 mg = 2 mg/kg BW daily) was added to the β -blocker regimen. At 6-months follow-up, the patient has complete suppression of exercise-induced ventricular arrhythmias and Holter recordings also excluded arrhythmias.

Conclusions. Recent experimental evidence suggested that flecainide can suppress CPVT. The presented cases support that novel strategy and show that flecainide is effective in preventing exercise-induced ventricular arrhythmias and also inappropriate shocks in children with CPVT.