# Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)

#### Introduction

CPVT is a condition that is characterized by exercise or emotion related fainting or cardiac arrest. Patients with CPVT usually have a normal resting ECG, but abnormalities present themselves with exercise or emotional stress. Swimming is uniquely associated with CPVT and type 1 Long QT Syndrome <sup>1, 2</sup>.

Catecholaminergic polymorphic ventricular tachycardia is inherited, with other affected family members in 30% of cases, with genetic testing yielding a causative mutation in the majority<sup>46</sup>.



### Diagnosis

Abnormal heart rhythms are often detected in individuals who undergo exercise testing and Holter monitoring <sup>3-6</sup>. Infusions of adrenaline can also unmask key abnormal heart rhythms that help to diagnose CPVT (Figure) <sup>3, 5, 7, 8</sup>.

# Treatment

Patients are treated with beta-blockers, drugs that block the adrenaline effect on the heart. An ICD is considered depending on the severity of presentation and family history  $\frac{4}{9}, \frac{6}{9}, \frac{9}{10}$ .

# **Selected References**

- 1. Choi G, Kopplin LJ, Tester DJ, Will ML, Haglund CM, Ackerman MJ. Spectrum and frequency of cardiac channel defects in swimming-triggered arrhythmia syndromes. Circulation 2004;110:2119-24.
- Ackerman MJ, Tester DJ, Porter CJ. Swimming, a gene-specific arrhythmogenic trigger for inherited long QT syndrome. Mayo Clin Proc 1999 Nov;74:1088-94.
- 3. Krahn AD, Healey JS, Chauhan V, et al. Systematic assessment of patients with unexplained cardiac arrest: Cardiac Arrest Survivors With Preserved Ejection Fraction Registry (CASPER). Circulation 2009;120:278-85.
- 4. Napolitano C, Priori SG. Diagnosis and treatment of catecholaminergic polymorphic ventricular tachycardia. Heart Rhythm 2007;4:675-8.
- 5. Krahn AD, Gollob M, Yee R, et al. Diagnosis of unexplained cardiac arrest: role of adrenaline and procainamide infusion. Circulation 2005;112:2228-34.
- 6. Priori SG, Napolitano C, Memmi M, et al. Clinical and molecular characterization of patients with catecholaminergic polymorphic ventricular tachycardia. Circulation 2002 Jul 2;106:69-74.
- 7. Krahn AD, Subbiah RN, Klein GJ, et al. Diagnosis and Outcome of 15 Canadian Families with Catecholaminergic Polymorphic Ventricular Tachycardia. Heart Rhythm 2008;5:P01-80.
- 8. Krahn AD, Simpson CS, Parkash R, et al. Utilization of a national network for rapid response to the Medtronic Fidelis lead advisory: the Canadian Heart Rhythm Society Device Advisory Committee. Heart Rhythm 2009;6:474-7.
- 9. Fisher JD, Krikler D, Hallidie-Smith KA. Familial polymorphic ventricular arrhythmias: a quarter century of successful medical treatment based on serial exercise-pharmacologic testing. J Am Coll Cardiol 1999 Dec;34:2015-22.
- Leenhardt A, Lucet V, Denjoy I, Grau F, Ngoc DD, Coumel P. Catecholaminergic polymorphic ventricular tachycardia in children. A 7-year follow-up of 21 patients. Circulation 1995 Mar 1;91:1512-9.