Clinical presentation and management of a patient with catecholamine sensitive polymorphic ventricular tachycardia (CPVT) - 2006

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1. Clinical presentation

The patient was a 17-year-old boy with height 169 cm and body weight 45kg. He has had 8 syncopal episodes since age 7. Most of these spells were triggered by exercise, exertion or emotional distress. Four episodes occurred during or immediately after climbing stairs. During the spells, the patient looked pale and had irregular pulses, urinary incontinence or vomiting, preceded by dizziness and chest discomfort. The patient was physically weak since his early childhood and caught cold very frequently. He rarely involved in physical exercises, became tired easily, and complained precordial chest pain occasionally. The heart rate (HR) was 50-60 bpm at rest, 38-50 bpm during sleep, 70-80bpm on standing and easily went above 100bpm with walking. The physical examination was otherwise unremarkable. The blood pressure was 120/70mmHg. The echocardiogram showed normal heart structures. The brain CT scan was normal. The ECG was normal at rest（Fig.1）with QTc of 0.35 sec. and frequent premature ventricular beats after climbing 4 flights of stairs while on metoprolol (37.5mg, Tid, Fig.2). The premature ventricular beats resolved completely within 2 minutes of rest（fig.3）. The ECG showed frequent premature ventricular contractions and non-sustained ventricular tachycardia with bi-directional and/or polymorphic ventricular tachycardia during the exercise test （Fig.4）. The blood concentration of norepinephrine was 1.45 pM/ml (normal range 0.51-3.26), and epinephrine, 0.52 pM/ml (normal range 0.05-1.39). One of his 2 siblings had sudden death at age of 9 and the other was alive and well.

2. Management

The patient was treated with atenolol (25 mg bid) when he was 8 years old and he had no syncope for 5 years. Then, he had 2 episodes of syncope at age 13. The dosage of atenolol was increased to 37.5mg/25mg/25mg. One year ago, atenolol was changed to metoprolol (37.5mg, tid). In May 2006, he had syncope again while climbing stairs and regained consciousness in about 1-2 min. Due to the insufficient protective effect of beta blockers, verapamil was tried from July 21 to July 26 with dosage of 240mg，once daily. On the second day of verapamil administration, significantly enlarged U waves（fig. 5）、monomorphic VT（fig. 6）and polymorphic/ bidirectional VT （fig7）were observed. From July 27 on, the patient was restarted back on metoprolol (37.5mg, tid). On Sept 14, he had another syncopal episode after climbing stairs. Up to now, he has been Metoprolol (extended release tablet), and the effect is under observation. The genotyping study for the patient is being undertaken.

4. Questions：

1) Can the diagnosis of CPVT be established?

2) What other therapies can be considered in addition to beta-blockers and ICD the patient can not afford ICD?

3) The catecholamine concentration in blood was normal for this patient. What do we expect of the catecholamine concentrations in CPVT patients in general?

4) What is the possible mechanism for the U wave enlargement and frequent p/BiVT during verapamil administration?