Clinical Features:

History

- Resuscitated from cardiac arrest, or syncope compatible with tachyarrhythmia especially related to physical activity, or acute emotion, in the presence of an unremarkable ECG (e.g. normal QT interval), and in the absence of structural heart or coronary artery disease
- Presentation usually during childhood and adolescence: 60% by the age of 20 years, 80% by the age of 40 years.

Family history

- Unexplained premature sudden cardiac death among immediate family members, in the context of physical activity or emotion.

Associated Arrhythmias:

- Ventricular fibrillation or polymorphic VT
- Bidirectional VT
- Non-sustained VT
- Atrial arrhythmia on exercise, most commonly atrial fibrillation

ECG characteristics:

- Normal resting ECG
- Bidirectional VT on exercise (but not always reproducible)
Other Cardiac Investigations:

Exclusion of structural heart disease

- Echocardiography, consider MRI, CT scan or coronary angiography

Exercise testing

- Exercise stress may induce ventricular arrhythmia once heart rates of greater than 120 – 130bpm are achieved.
- Isolated ventricular premature beats, non-sustained or sustained atrial and ventricular arrhythmia may occur.
- Bidirectional VT is the typical pattern of CPVT-related arrhythmia, but VF or polymorphic VT may also occur.

Adrenaline infusion

- The sensitivity of adrenaline testing is undetermined, therefore it is not currently recommended.

* QT interval is corrected for heart rate using Bazett’s formula: QTc=QT/√RR, measured in msecs.

Genetics:
Counseling and consent is mandatory before testing noting the complexities that arise if a variant of unknown significance (VUS) is found. All patients should be discussed with colleagues in Clinical Genetics, within an MDT meeting if possible, prior to testing

Diagnostic Criteria:

- Exercise-, emotion- or catecholamine-induced ventricular arrhythmia in the presence of a structurally normal heart and normal ECG.

Or

- Presence of pathogenic mutation

References:

2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death, 8.4 Catecholaminergic polymorphic ventricular tachycardia, page 45


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