Idiopathic ventricular tachycardia: an overview of subtypes

Idiopathic ventricular tachycardia (VT) conventionally refers to the presence of monomorphic VT in the absence of structural or coronary disease. A notable exception is catecholaminergic polymorphic VT (CPVT). They are generally associated with a good prognosis. There is a reasonable response to arrhythmia-modifying drugs (AMDs) and are often amenable to ablation. There are various subtypes, and a succinct overview of each is now provided.

Outflow tract VT

Right ventricular outflow tract (RVOT) VT has the characteristic appearance of left bundle branch block (LBBB) morphology in V1 and an inferior axis in the frontal plane. Vagal manoeuvres such as adenosine can terminate the dysrhythmia, whereas sympathetic overdrive in the context of exercise or stress, for example, can initiate and perpetuate. Beta-blockers and verapamil are of benefit. A similar tachycardia can be observed in left ventricular outflow tract (LVOT) VT. However, its distinguishing feature is the presence of a S wave in lead I.

Annular VT

These arise from the mitral or tricuspid annulus, and account for 4-7% of idiopathic cases. For mitral annular VT, the ECG pattern is typically right bundle branch block (RBBB) with a S wave in V6. For tricuspid annular VT, the foci generally originate in the septal region, and thus, there is a LBBB pattern with narrower QRS complexes.

Fascicular VT

These arise in the left posterior fascicle. 12-lead electrocardiogram (ECG) would classically show a relatively narrow RBBB morphology and a left superior axis. Intravenous administration of dihydropyridine calcium channel antagonists such as verapamil or diltiazem often suppress this type of VT, whilst adenosine is rarely effective. This is suggestive that the slow inward current may be implicated, possibly via a mechanism of re-entry or delayed after-depolarisations (DADs). Catheter ablation is often curative and should be considered as an early treatment strategy.

Catecholaminergic polymorphic VT

CPVT is a rare form of VT that occurs in children and adolescents. Mutations in ryanodine receptor gene RyR2 or calsequestrin gene CASQ2 are implicated, with both inherited in an autosomal dominant pattern. Patients typically present with syncope or aborted sudden cardiac death (SCD), with a dysrhythmia that is highly reproducible by stress. QTc interval is normal. Typically, exercise results in physiological sinus tachycardia. This can subsequently be

accompanied by ventricular extra-systoles followed by salvos and progression into persistent, polymorphic VT. Classically, it is of a bidirectional appearance with beat-to-beat alternation of the frontal axis. Of note, a similar appearance can also be seen in severe digoxin toxicity. Treatment of choice is beta-blocker therapy and device therapy with an implantable cardioverter-defibrillator (ICD), though sympathectomy does appear to be effective in some cases. Patients should also be provided with strict lifestyle advice to avoid rigorous exercise.

Brugada syndrome

This is a distinct clinical entity resulting from mutations in genes responsible for sodium channels (SCN5A) and calcium channels. It is likely to account for around 50% of all cases of idiopathic ventricular fibrillation (VF), with symptoms occurring predominantly at night. It is more common in South Asian populations. Patients typically have evidence of RBBB and ST elevation in anterior precordial leads, though three types of repolarisation patterns are described. Imaging excludes presence of structural heart disease. Sodium channel blockers, such as flecainide, amjaline or procainamide, can be utilised as pharmacological provocative agents under strict monitoring to assist with diagnosis. ICDs are the only established treatment option in these patients to prevent SCD, though use is controversial in those that are asymptomatic. Research has suggested that evidence of a fragmented QRS complex (i.e. multiple spikes) can be useful to stratify risk in these cohorts.

Torsades de pointes

The underlying mechanism behind Torsades de pointes is a prolonged QT interval. This can be congenital with gene mutations resulting in channelopathies. Examples include Romano-Ward syndrome (autosomal dominant) and Jervell and Lange-Nielsen syndrome (autosomal recessive). The most common cause of acquired long-QT is pharmacotherapy, with various agents implicated such as tricyclic anti-depressants, amiodarone, sotalol, digoxin and lithium. Electrolyte abnormalities such as hypokalaemia and hypocalcaemia are also known precipitants.

In terms of symptoms, patients may present with syncope or seizures. The primary risk is of SCD, likely from degeneration of VT into VF. Categorisation of the disorder can be made based on triggers. LQT1 appears to be precipitated by exercise, particularly swimming, and emotional stress. LQT2 also occurs in the context of emotional stress, or by a sudden loud noise. LQT3 is associated with rest or sleep. LQT1 and 2 are associated with mutations in potassium channels, whilst LQT3 is associated with sodium channels.

Diagnosis relies on assessment of the QT interval as electrophysiological studies are generally not helpful. The QT interval is inherently affected by resting heart rate, and therefore, it is the corrected interval (QTc) that is more informative. Roughly, a duration >460ms for men and >470ms for women is abnormal. A significant prolongation in interval, usually >500ms, can result in the R-on-T phenomenon, whereby ventricular depolarisation, usually a premature

ventricular contraction (PVC), occurs during repolarisation. This can precipitate successive bursts of VT whereby QRS complexes of varying amplitude appear to rotate around the isoelectric baseline, giving the typical twisting appearance of Torsades de pointes. Congenital forms generally have a preceding period of sinus tachycardia, whilst the acquired forms are precipitated by bradycardia or pauses.

It is important to emphasise that management of Torsade de pointes relies on identification of prolonged QTc as the precipitating event. In congenital causes, beta-blockade should be instituted. With acquired cases, any precipitating drugs should be discontinued. A full electrolyte screen is required to address potentially reversible causes. Intravenous magnesium is recommended in providing general myocardial protective effects beyond reduction of dysrhythmic potential. Relevant mechanisms may include coronary vasodilation, reduced catecholaminergic drive and cellular magnesium-calcium interactions restricting calcium deposition in mitochondria. Thus, there is evidence of benefit even in patients with normal serum levels. Careful commencement of isoprenaline and/or temporary pacing are beneficial in preserving heart rate to reduce propensity for dysrhythmia. In patients who presents with congenital, symptomatic Torsades de pointes, ICD therapy is usually indicated after review by a specialist service. These patients can also be treated concomitantly with beta-blockers. Additionally, the device can be utilised to provide overdrive pacing to prevent bradycardia-induced dysrhythmia. Algorithms can also be set to prevent typical pauses post-PVC. The evidence for primary prevention ICD in patients with acquired long-QT and a family history of SCD but no syncope is not established. Patients with congenital forms are advised to avoid strenuous exercise.

Peysh Patel March 2017