Polymorphic Ventricular Tachycardia/Ventricular Fibrillation and Sudden Cardiac Death in the Normal Heart

Ashok J. Shah, MD[a,*], Meleze Hocini, MD[b], Arnaud Denis, MD[b], Nicolas Derval, MD[b], Frederic Sacher, MD[b], Pierre Jais, MD[b], Michel Haissaguerre, MD[b,*]

KEYWORDS

• Polymorphic ventricular tachycardia • Ventricular fibrillation • Sudden cardiac death
• Electrical storm • Primary electrical disease • Normal heart • Catheter mapping • Ablation

KEY POINTS

- Polymorphic ventricular tachycardia (PMVT) and ventricular fibrillation (VF), due to primary electrical diseases or idiopathic VF, are responsible for about 10% of sudden cardiac deaths (SCDs).
- Despite multimodality secondary prevention of SCD including the avoidance of provocative agents and circumstances, up to 20% of patients experience recurrent episodes of PMVT/VF or electrical storms.
- PMVT/VF is largely triggered by premature ventricular ectopic beats (PVBs), which occur frequently in isolation around the period of electrical storm.
- In the majority of patients with primary PMVT/VF, the triggering PVBs can be mapped and localized to the peripheral Purkinje system of both ventricles.
- Right ventricular outflow tract is the most likely site of origin of culprit PVBs in Brugada syndrome.

INTRODUCTION

Polymorphic ventricular tachycardia (PMVT) and ventricular fibrillation (VF) are the most life-threatening, complex, cardiac rhythm disorders.[1] Referring to heart as normal in a patient with sudden cardiac death (SCD) sounds antithetical. However, normal conventionally refers to its structure and vascular supply examined using techniques like echocardiogram and angiogram and in the era of advanced imaging, possibly also cardiac computed tomography (CT), MRI and PET.

Although cardiac asystole has increasingly become the more common arrhythmic cause of SCD events, PMVT and VF are still associated with a substantial number of them.[2–4] About 90% of SCD events occur in a diseased heart, the most common being the coronary artery disease.[5,6] Coronary arterial abnormalities, dilated and hypertrophic cardiomyopathies, infiltrative disorders, and valvular or congenital cardiac disorder are not found in the remaining about 10% events where the heart is considered structurally normal.[6] It is rare to have spontaneous PMVT/VF in the absence of a known structural or electrical cardiac abnormality. An arrhythmic event resulting from an unknown cause is labeled as idiopathic. Primary electrical abnormalities result from mutated genes encoding proteins involved in handling ion movements in the heart.

[a] Cardio Vascular Services, South Consulting Suites, Peel Health Campus, 110 Lakes Road, Mandurah, Western Australia 6210, Australia; [b] Department of Electrophysiology and Cardiac Pacing, Hôpital Cardiologique du Haut Lévêque, Avenue de Magellan, Pessac Cedex 33604, France
* Corresponding author.
E-mail addresses: drashahep@gmail.com; michel.haissaguerre@chu-bordeaux.fr

Card Electrophysiol Clin 8 (2016) 581–591
http://dx.doi.org/10.1016/j.ccep.2016.04.007
1877-9182/16/$ – see front matter

Because they manifest as electrocardiographic abnormalities but do not cause alterations in gross cardiac structure, the heart is regarded as normal. These disorders include long QT syndrome, Brugada syndrome, catecholaminergic polymorphic VT, short QT syndrome, and early repolarization syndrome.[7] However, Brugada syndrome is associated with epicardial late potential strongly suggestive of underlying structural changes.[8]

This article presents a clinical review of the electrophysiological management of PMVT/VF in a structurally normal heart (Table 1).

OVERVIEW OF MANAGEMENT

PMVT and VF are rapidly lethal arrhythmias. Implantable cardiac defibrillator (ICD) implantation is unequivocally the gold standard, first-line treatment in patients who have survived SCD (secondary prevention) and in the majority of subjects at high-risk of SCD (primary prevention).[7] ICD prolongs survival without influencing the arrhythmogenic substrate.[7] Antiarrhythmic drugs like beta-blockers, quinidine, mexilitine, and flecainide (table) complement ICDs in secondary prevention of PMVT/VF of different etiologies.[7] Drugs are also recommended as the first-line treatment for primary prevention in some forms of SCD.[7] Similarly, surgical sympathetic denervation has also been advocated and proven beneficial in secondary prevention of some cardiac electrical disorders.[7]

Despite multimodality management of SCD including the avoidance of provocative agents and circumstances, up to 20% of patients experience recurrent episodes of PMVT/VF or electrical storms. Such a situation acutely increases the mortality. In the survivors, storm deteriorates the quality of physical life and has an adverse impact psychologically from multiple device therapies delivered within a short period of time.[9] Drugs like isoproterenol, amiodarone, beta-blockers, and lidocaine have been used in life-threatening arrhythmias with variable efficacy and inconsistent benefits.

Catheter ablation of PMVT/VF has been undertaken as a life-saving measure when drugs and, fail to control incessant runs of arrhythmias. The premature ventricular beats (PVBs) triggering storm recur in isolation thereby serving as tangible targets for catheter mapping and ablation of these unmappable arrhythmias. The electrophysiological characteristics of PMVT/VF and its ectopic triggers have been described in various electrical substrates of structurally normal heart in the following sections.

Table 1
Primary electrical diseases

Disorder	Drug
Long QT syndrome	Beta blockers mexilitene in LQT3
Brugada syndrome	Quinidine
Catecholarminergic polymorphic ventricular tachycardia	Nadolol, verapmail, flecainide
Abnormal early repolarization syndrome	Quinidine
Idiopathic VF	Quinidine

MAPPING AND ABLATION OF POLYMORPHIC VENTRICULAR TACHYCARDIA/VENTRICULAR FIBRILLATION

Invasive catheter mapping of PMVT/VF targeting culprit PVBs gets facilitated and ablation yields best results when it is undertaken as close to the storm as possible. If the procedure is delayed, rarity or absence of spontaneous culprit PVCs will reduce the long-term clinical success rate of pace-map based procedure in the absence of any known and reliable provocative agent. Patients with few monomorphic PVBs have a high probability of successful ablation in contrast to those with pleomorphic PVBs arising from a wider area. In addition to being a life-saving therapy, it may provide cure at best or reduce arrhythmia burden at least, if successful.

The best approach is to ablate when the clinical PVC load is high around the time of frequent episodes of VF. Otherwise, there is no reliable method to provoke the clinical PVCs. The authors have anecdotally used atrial pacing, class I antiarrhythmic drugs, isoproterenol, Ca++, pacing induced sinus pause to provoke clinical PVC individualy, but the results are not consistent.

Frequent PMVT/VF needs to be controlled to allow mapping of clinical PVCs, requiring individually beta-blockers in catecholergic arrhythmias, verapamil in short coupled ectopies and IVF, or isoproterenol in acute storm due to abnormal early repolarization/Brugada syndrome/idiopathic VF.

Idiopathic Ventricular Fibrillation

Aizawa and colleagues[10] first reported successful suppression of electrical storms after ablation of premature ventricular beats located in the posterolateral wall of the left ventricle. Haissaguerre and colleagues[11] described ablation of