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CASE REPORT
Electrophysiology

Broad and narrow complex tachycardia resulting in cardiorespiratory arrest in a child: what is the optimal treatment strategy?

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Background We describe a child with a broad and narrow complex tachycardia causing haemodynamic collapse.

Case summary A 9-year-old girl (weight 26 kg, height 114 cm) with a 5-year history of refractory 'epilepsy' presented with cardiorespiratory arrest and tonic-clonic seizure, witnessed by her mother. Electrocardiogram documented recurrent episodes of simultaneous broad and narrow tachycardias associated with haemodynamic compromise. Diagnostic electrophysiologic study (EPS) confirmed a dual tachycardia mechanism. The challenge in selecting the optimal treatment strategy is discussed. A diagnosis of dual tachycardia was made with catecholaminergic polymorphic ventricular tachycardia (CPVT) and simultaneous focal atrial tachycardia.

Discussion Bidirectional ventricular tachycardia (VT) induced by isoproterenol in this clinical scenario is strongly suggestive of CPVT. Diagnostic EPS can be useful in challenging clinical situations to understand the mechanism of arrhythmias and to tailor the most appropriate treatment strategy. Combination therapy with nadolol and flecainide is highly effective in ventricular arrhythmia control. Implantable cardioverter defibrillator implantation is not without risk in CPVT as there is a potential of electrical storm driven by shock therapy that increases adrenergic drive. Cervical sympathectomy may be considered if further VTs occur in future despite optimum medical therapy.

Keywords CPVT Catecholaminergic polymorphic ventricular tachycardia • ECMO Extra corporeal membrane oxygenator • VT Ventricular tachycardia • EPS Electrophysiological study • Case report

ESC curriculum 5.8 Cardiac ion channel dysfunction • 5.6 Ventricular arrhythmia • 5.5 Supraventricular tachycardia

Learning points

- Avoidance of adrenaline and combination therapy with flecainide and nadolol is effective in catecholaminergic polymorphic ventricular tachycardia (CPVT).
- Isoproterenol-dependant bidirectional ventricular tachycardia and atrial tachycardia can present as dual tachycardia as a presenting feature of CPVT.
- Diagnostic electrophysiologic study is a useful tool in challenging clinical situations to understand mechanism of arrhythmias and to choose the best treatment strategy.

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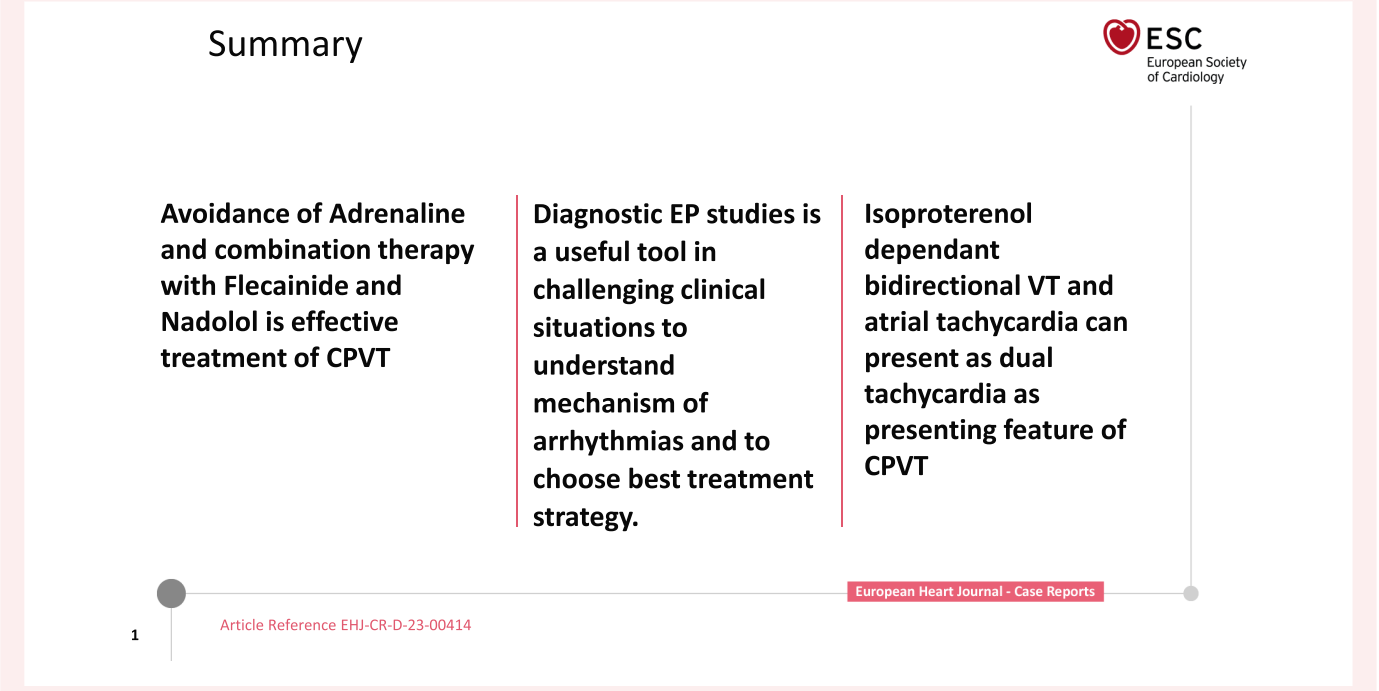
Introduction

A 9-year-old girl (weight 26 kg, height 114 cm) with a 5-year history of refractory 'epilepsy' presented with cardiorespiratory arrest and tonic-clonic seizure, witnessed by her mother. Electrocardiogram documented recurrent episodes of simultaneous broad and narrow tachycardias associated with haemodynamic compromise (*Figure 1*). She had received multiple direct cardioversions (DC) to restore sinus rhythm (*Figure 2*), requiring intubation and ventilation. There was no family history of sudden death. Initial echocardiography showed severe left ventricular impairment and inotropic support with adrenaline infusion was commenced in the intensive care unit. Sustained narrow complex tachycardia was also observed intermittently (*Figure 3*). Despite amiodarone and verapamil, she continued to develop frequent episodes of both broad and narrow complex tachycardias causing haemodynamic compromise, needing cardiopulmonary resuscitation and repeated DC shocks, which were only transiently effective. Myocarditis and Covid-19 infection were excluded on lab testing. A decision was therefore made to proceed to electrophysiologic study (EPS) with extracorporeal membrane oxygenator (ECMO) standby on the 4th day of admission.

At Electrophysiological study, sinus rhythm was observed at baseline, normal atrial-His interval (124 ms), normal His-ventricle interval (46 ms), antegrade decremental conduction, retrograde ventricle-atria (VA) block indicating normal sinus node, and atrial-ventricular node conduction with no evidence of accessory pathway. No tachycardia was inducible with programmed stimulation in the atria or ventricle with three extra stimuli. However, isoproterenol infusion (starting dose 0.03 μg/kg/min) induced repetitive broad and narrow complex tachycardias resembling the clinical arrhythmia (*Figure 4*). Intermittent bidirectional ventricular tachycardia with VA dissociation, His dissociation, and simultaneous atrial tachycardia with intermittent capture beats could be seen (*Figure 4*). This was the result of simultaneous occurrence of a non-sustained bidirectional/polymorphic ventricular tachycardia (VT) (*Figure 4*) and a sustained atrial tachycardia at 130–150 b.p.m. with proximal to distal activation sequence in the coronary sinus (*Figure 5*). Limited atrial mapping suggested a focal left atrial tachycardia mechanism that was dependent on isoproterenol, but because of increasing VT episodes due to isoproterenol infusion, a decision was made not to attempt ablation.

Summary figure



Case summary

A diagnosis of dual tachycardia was made with catecholaminergic polymorphic ventricular tachycardia (CPVT) and simultaneous focal atrial tachycardia. Oral flecainide (2 mg/kg b.i.d.) and nadolol (1 mg/kg o.d.) were commenced after withdrawing inotropic support. No further tachycardia was seen after the combination therapy, and she was extubated within 2 days. She was later discharged after normalization of her left ventricular function with implantable loop recorder monitoring and remained free of arrhythmias after 4 months. Genetic testing did not show a positive variant for the CPVT panel.

Discussion

Bidirectional VT induced by isoproterenol in this clinical scenario is strongly suggestive of CPVT, which is a rare but potentially lethal inherited channelopathy affecting 1 in 10 000. Atrial arrhythmias and sinus node dysfunction have been associated with this condition in a case series.[1] In that report, programmed stimulation was able to induce atrial flutter or fibrillation in two of eight patients, and in only one case did isoproterenol induce atrial fibrillation. Our present case is unique

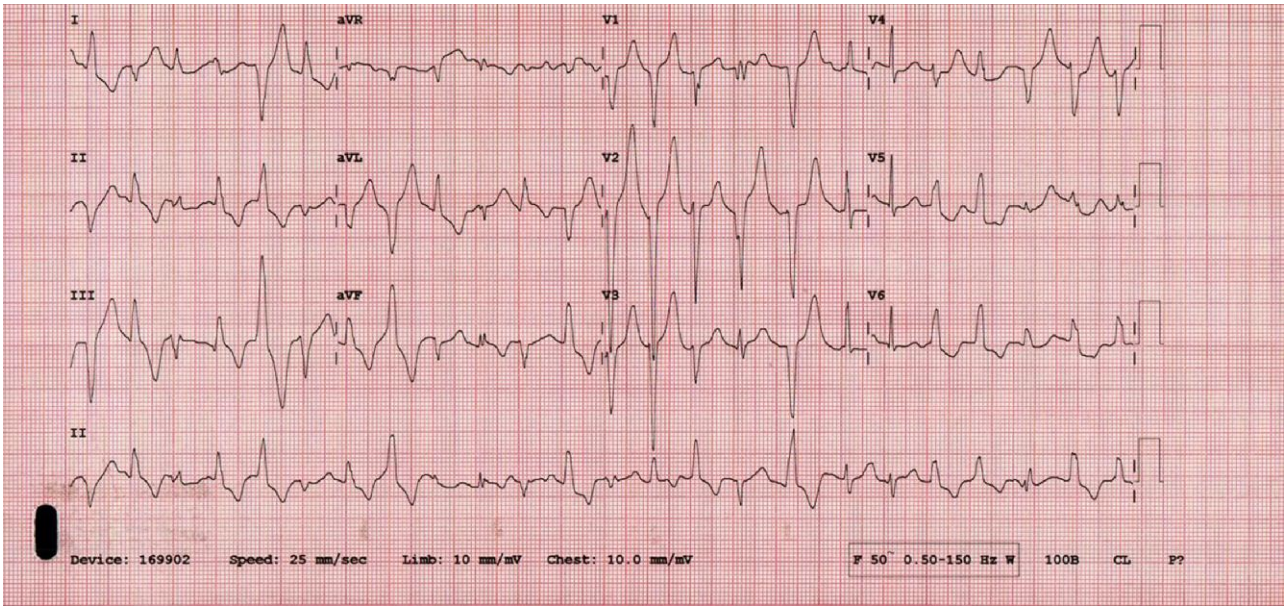


Figure 1 Electrocardiogram showing irregular broad and narrow complex tachycardia with variable QRS axis representing polymorphic ventricular tachycardia.

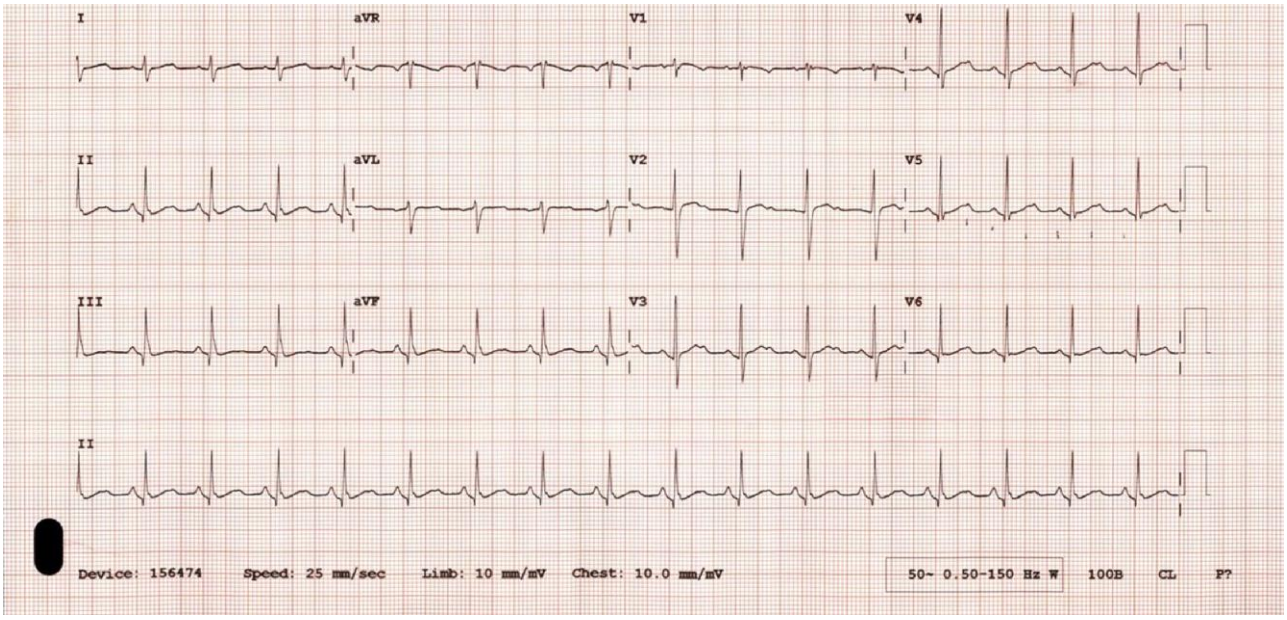


Figure 2 Baseline electrocardiogram showing regular sinus rhythm normal axis, intervals, and within normal limits.

with an ectopic focal atrial tachycardia inducible with isoproterenol in a patient with polymorphic VT, which has not been previously reported to our knowledge. The observation that both ventricular and atrial arrhythmias were initiated at the same time by isoproterenol, but not by programmed stimulation, strongly suggests a common autonomic (adrenergic) trigger in our patient.

Diagnostic EPS can be useful in challenging clinical situations to understand the mechanism of arrhythmias and to tailor the most appropriate treatment strategy. Bidirectional broad complex tachycardias, VA dissociation, His dissociation, and simultaneous atrial tachycardia with intermittent capture beats established the diagnosis of dual tachycardias in this case (*Figure 5*). Provocative testing with isoprenaline can unmask arrhythmias in CPVT[2] and induce ectopic atrial tachycardias. Our patient was genotype negative for the CPVT panel, but the genetic yield for CPVT diagnosis is estimated to be ~65%.[3]

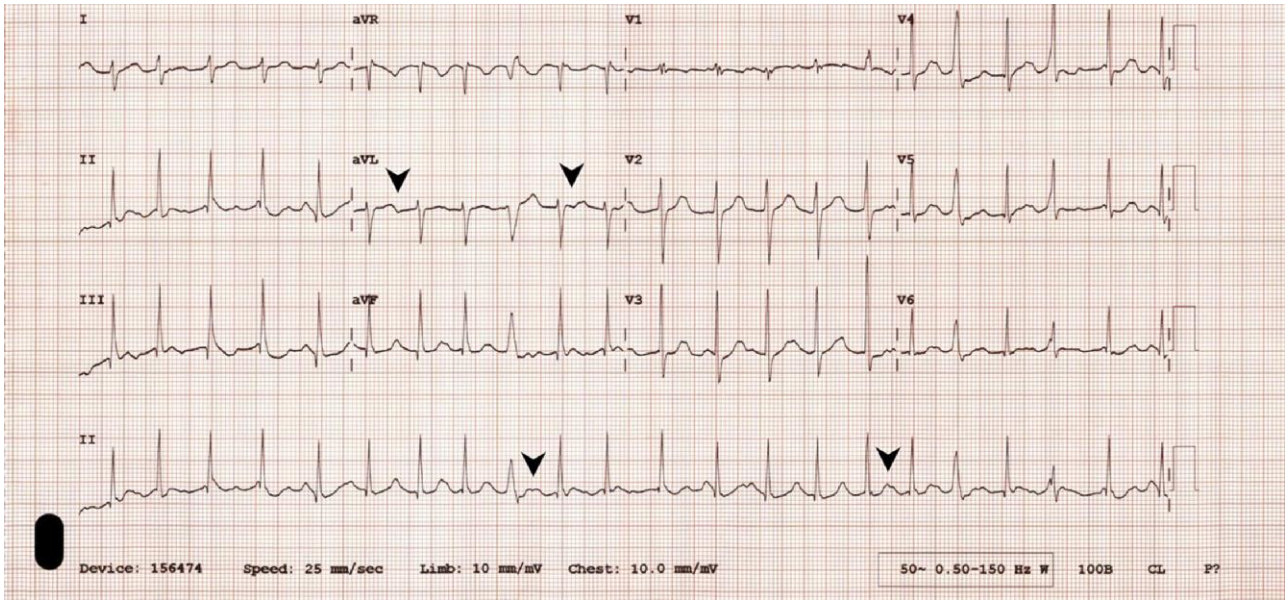


Figure 3 Electrocardiogram showing spontaneous narrow complex tachycardia with slight irregularity in rhythm. Abnormal P waves marked by arrows. Atrial tachycardia as likely mechanism for tachycardia.

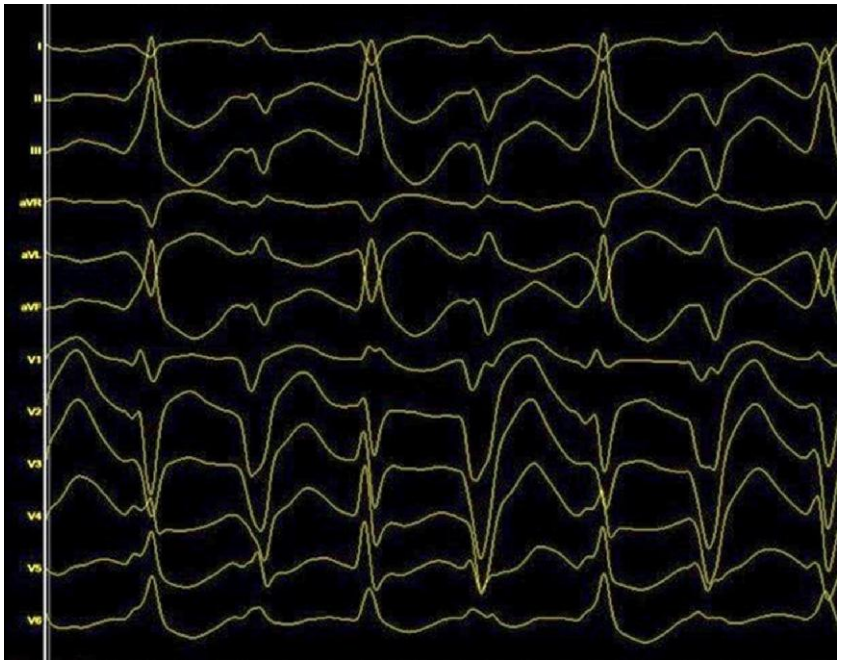


Figure 4 Electrocardiogram during electrophysiologic studies showing bidirectional ventricular tachycardia during low dose isoproterenol infusion, pathognomonic of catecholaminergic polymorphic ventricular tachycardia.

Impairment of left ventricular function in our patient could have been caused by the effects of the cardiorespiratory arrest and repeated external defibrillation shocks. Inotropes such as adrenaline is frequently used for treatment of low cardiac output state and poor ventricular function in the intensive care setting. But, our case illustrates a challenging clinical situation of adrenergically mediated ventricular and atrial arrhythmias, in which adrenaline should be avoided and instead beta blockade is given with ECMO on standby. Our patient's ventricular function did improve following successful rhythm control by flecainide and nadolol combination within 2 days.

Combination therapy with nadolol and flecainide is highly effective in ventricular arrhythmia control for CPVT as shown in our case.[4]

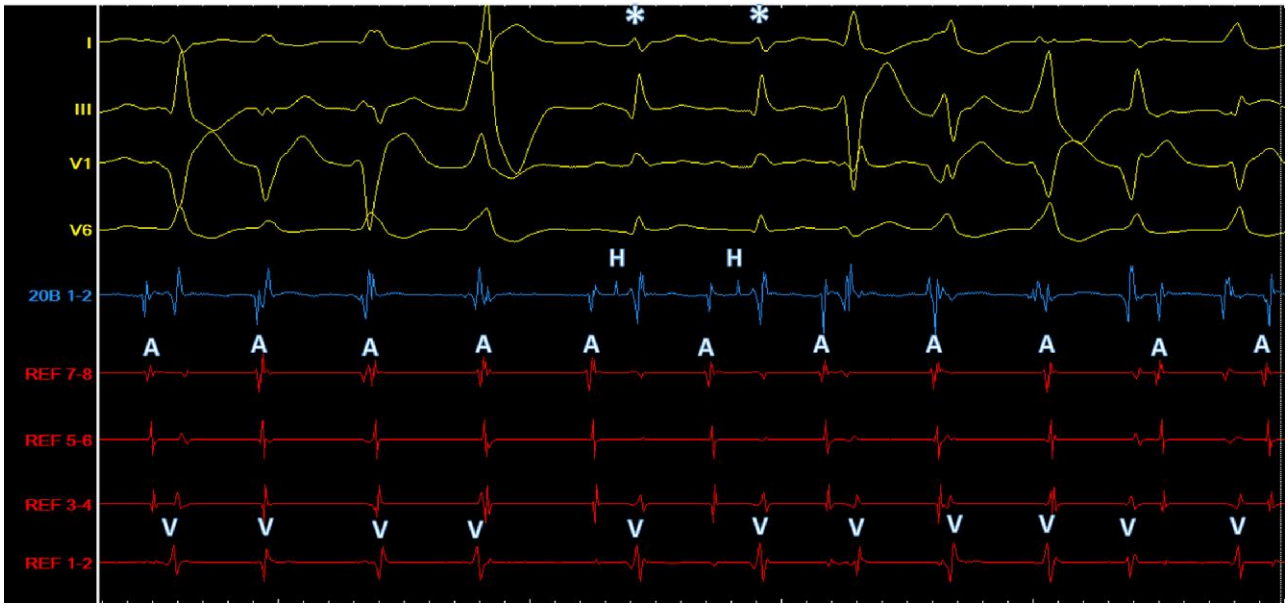


Figure 5 Electrophysiologic studies showing onset of bidirectional ventricular tachycardia, VA dissociation, His dissociation, and simultaneous atrial tachycardia. Surface electrocardiogram (Leads I, III, V1, and V6) shown in yellow. Intracardiac electrogram from His/right ventricle catheter displayed in blue. Coronary sinus signals displayed as red, and complexes marked as * indicates narrow capture beats. Markers A, H, and V indicate atrial, His, and ventricular depolarizations, respectively.

Their antiarrhythmic properties for focal atrial tachycardia were also helpful for this patient. Implantable cardioverter defibrillator implantation is not without risk in CPVT[5] as there is a potential of electrical storm driven by shock therapy that increases adrenergic drive. Catheter ablation has been reported in drug refractory atrial tachycardia in CPVT in an adult patient,[6] but our case was complicated by the occurrence of frequent compromising polymorphic VTs and it was felt too risky to perform detailed biatrial mapping and ablation. We therefore gave an opportunity to the use of antiarrhythmic drugs to control both arrhythmias. Cervical sympathectomy[7] may be considered if further VTs occur in future despite optimum medical therapy.

Lead author biography

Dr Shankar N. Sadagopan practices as Consultant Paediatric Cardiologist and Electrophysiologist at University Hospitals Southampton NHS Trust.

Consent: Informed consent obtained from child's mother for publication of case report.

Conflict of interest: None declared.

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Data availability

The data underlying this article are available in the article.

References

1. Sumitomo N, Sakurada H, Taniguchi K, Matsumura M, Abe O, Miyashita M, *et al.* Association of atrial arrhythmia and sinus node dysfunction in patients with catecholaminergic polymorphic ventricular tachycardia. *Circ J* 2007;**71**:1606–1609.
2. Obeyesekere MN, Klein GJ, Modi S, Leong-Sit P, Gula LJ, Yee R, *et al.* How to perform and interpret provocative testing for the diagnosis of Brugada syndrome, long-QT syndrome, and catecholaminergic polymorphic ventricular tachycardia. *Circ Arrhythm Electrophysiol* 2011;**4**:958–964.
3. Wilde AAM, Semsarian C, Márquez MF, Sepehri Shamloo A, Ackerman MJ, Ashley EA, *et al.* European Heart Rhythm Association (EHRA)/Heart Rhythm Society (HRS)/Asia Pacific Heart Rhythm Society (APHRS)/Latin American Heart Rhythm Society (LAHRS) expert consensus statement on the state of genetic testing for cardiac diseases. *Heart Rhythm* 2022;**19**:e1–e60.
4. Claudio B, Alice M, Daniel S. A focus on pharmacological management of catecholaminergic polymorphic ventricular tachycardia. *Mini Rev Med Chem* 2018;**18**:476–482.
5. Priori SG, Wilde AA, Horie M, Cho Y, Behr ER, Berul C, *et al.* HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes: document endorsed by HRS, EHRA, and APHRS in May 2013 and by ACCF, AHA, PACES, and AEPC in June 2013. *Heart Rhythm* 2013;**10**:1932–1963.
6. Kawada S, Morita H, Watanabe A, Ito H. Radiofrequency catheter ablation for drug-refractory atrial tachyarrhythmias in a patient with catecholaminergic polymorphic ventricular tachycardia: a case report. *J Cardiol Cases* 2018;**19**:36–39.
7. Schwartz PJ, Ackerman MJ. Cardiac sympathetic denervation in the prevention of genetically mediated life-threatening ventricular arrhythmias. *Eur Heart J* 2022;**43**:2096–2102.