Storm of Short Coupling Polymorphic Ventricular Tachycardia Triggered by Ventricular Ectopy from Distal Purkinje System: Management and Outcome

Alexander Chapurnykh*, Sergey Lakomkin and Vladimir Niznichenko

Department of Surgical Treatment of Complex Cardiac Arrhythmias and Cardiac Pacing. Central Clinical Hospital of the Management Department of the President of the Russian Federation

Abstract: *Background*: Storm of short coupling PVT (SC PVT) is a rear clinical syndrome. Implantable cardioverter-defibrillator and radiofrequency ablation of arrhythmia trigger are optional for life threatening control and freedom from arrhythmia.

Method and Result: This case report describes a 50-year-old woman with normal repolarization duration who survived multiple electrical storms and cardiac arrest related to recurrent SC PVT. Betablocker and amiodarone suppressed storm of PVT and cardioverter defibrillator was implanted. After than radiofrequency ablation of premature ventricular contraction – trigger of PVT was performed.

Result: Radiofrequency ablation revealed high efficacy in long term prevention of recurrence of PVC triggering PVT and VF.

Keywords: Short-couple polymorphic ventricular tachycardia, electrical storm, antiarrhythmic treatment, radiofrequency ablation.

Polymorphic ventricular tachycardia (PVT), characterized by changing or multiform QRS morphology from beat to beat, occasionally associated with premature ventricular beats from distal Purkinje fibers with short coupling R-on-T interval and induces ventricular fibrillation [1, 2]. Repetitive ventricular tachycardia (VT) deteriorated or not in ventricular fibrillation (VF), occurring more than three times within 24-hour each requiring intervention for termination presents VT/VF storm [3]. Storm of short coupling PVT (SC PVT) is rear clinical syndrome, but in most cases, PVT deteriorates into VF [1]. Implantable cardioverter-defibrillator and radiofrequency ablation of arrhythmia trigger are optional for life threatening control and freedom from arrhythmia [1]. We present a clinical case of management and long-term follow-up of patient suffered from SC PVT storm.

CASE PRESENTATION

Fifty years old female was admitted to the hospital 08.07.2014 soon after short episode of syncope. She hasn't had any family history of syncope or sudden cardiac death. Mild arterial hypertension was only associate disease. First episode of syncope patient had 9 years before. At that date Holter ECG monitoring (HM ECG) showed short episode of spontaneous terminated VT degenerated to VF (Picture **1**). Patient was treated by intravenous amiodarone 600 mg. Endocardial electrophysiology investigation didn't found any abnormalities. Genetic testing has not found any SCN5A abnormalities. After all, betablocker bisoprolol was started. For next 8,5 years till last episode she didn't experienced any rhythm disturbances or syncope. Two months before last clinical event patient was referred for angiography of coronary artery because of chest discomfort during exertion. The examination revealed muscular bridge narrowing left anterior descending artery by 50%. After that betablocker bisoprolol was concealed and Ca^{2+} antagonist diltiazem was started.

At time of her admission to the hospital after last episode of syncope physical examination didn't reveal any abnormalities. Transthoracic echocardiography revealed normal left atrial size – 35 mm and volume – 57 ml, left and right ventricular dimensions: left ventricle end diastolic size 44 mm, interventricular septum and posterior wall were 10 mm, left ventricle ejection fraction – 60%; right ventricular outflow tract was 28 mm, mitral and tricuspid regurgitation 1+, systolic pulmonary artery pressure 29 mm Hg. Blood analysis didn't reveal any electrolytes abnormalities.

The patient's 12 lead ECG showed normal sinus rhythm: PQ 160 mc QRS 86 mc, QT 400 mc, QTc 449 mc. Short episode of PVT induced by short coupled ventricular premature beat (VPB) with coupling interval of 240 ms falling early on T wave of the last sinus beat

*Address correspondence to this author at the Department of Surgical Treatment of Complex Cardiac Arrhythmias and Cardiac Pacing. Central Clinical Hospital of the Management Department of the President of the Russian Federation; Tel: +7 495 5300123; Fax: +7 499 1424250;
E-mail: chapurnykh@mail.ru

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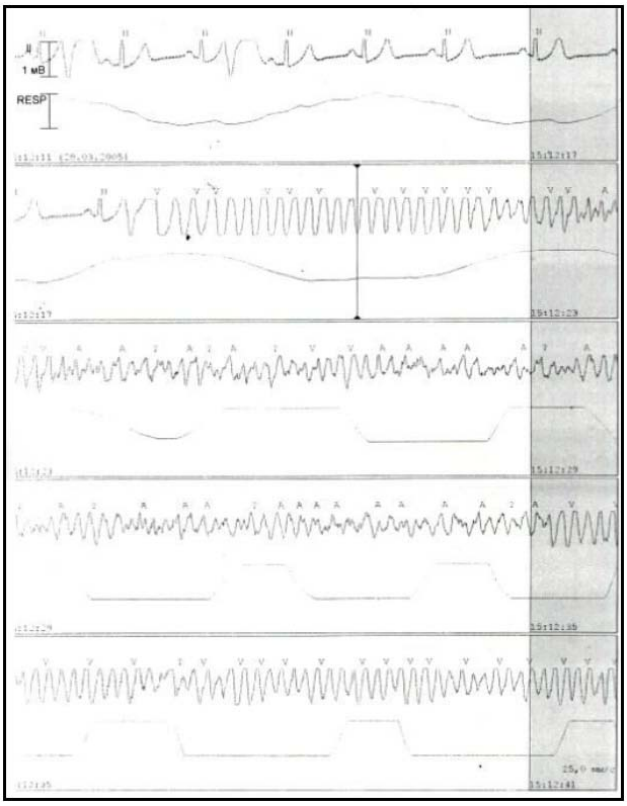
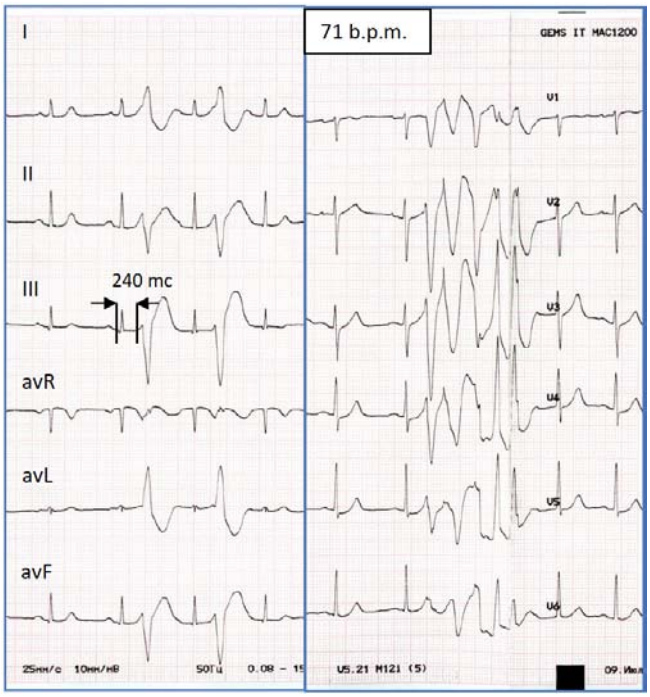


Picture 1: Holter ECG monitor presents short episode of ventricular tachycardia degenerated to ventricular fibrillation.



Picture 2: Twelve lead ECG. Sinus rhythm, 72 beats per minute. Repetitive monomorphic ventricular premature beats (VPB): Left anterior axis, left bundle branch block morphology, QRS 158 mc, transition zone V5. Coupling interval 240 mc. The last VPB in precordial leads induces short episode of polymorphic ventricular tachycardia.

was found. All VPBs were monomorphic, had short coupling interval and left superior axis with left bundle branch block type and V5 transition zone presumed right septal apical origin (Picture **2**).

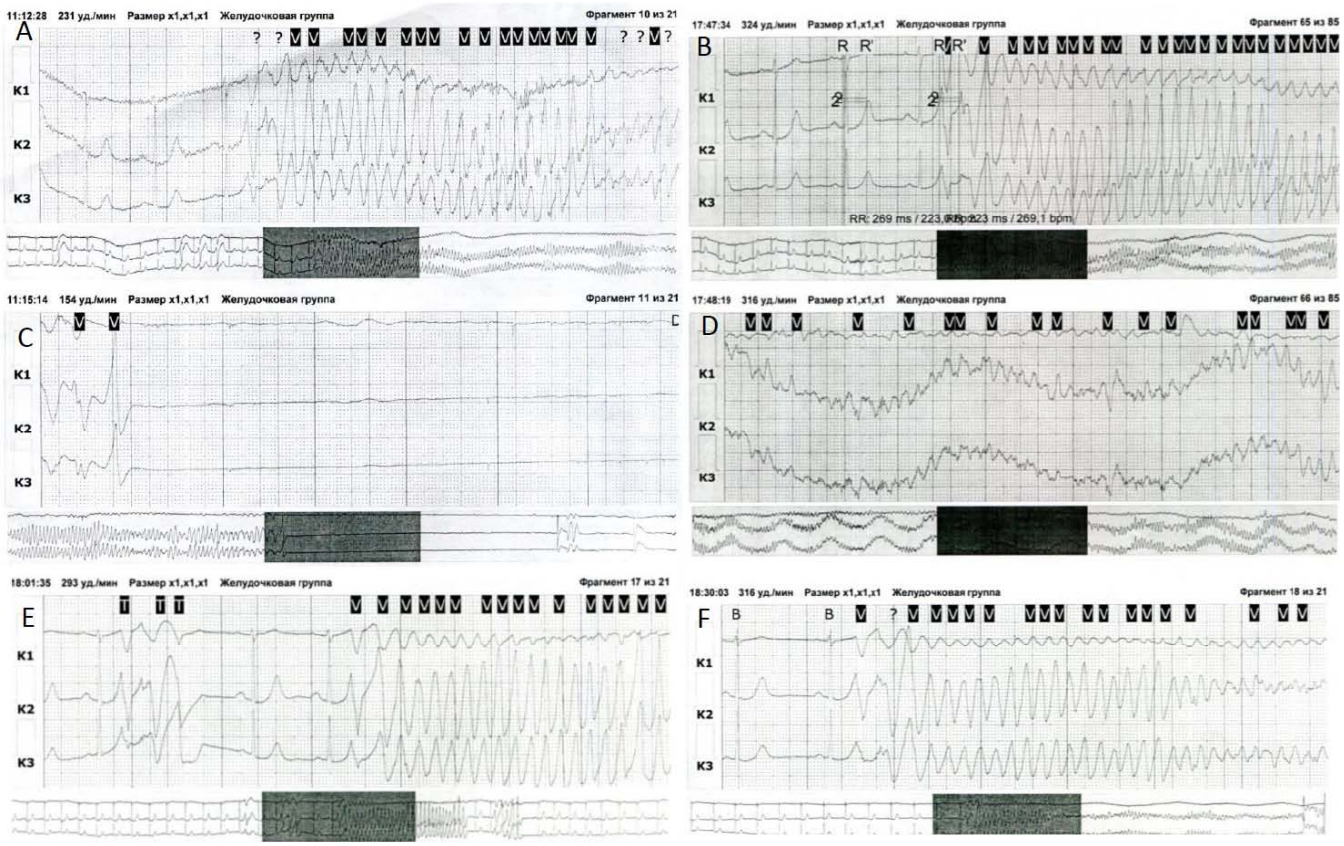
Holter 24-hour monitoring revealed short couple VPBs with coupling interval 230-280 mc, numerous recurrent PVT episodes deteriorating into ventricular fibrillation requiring defibrillation to restore sinus rhythm. All episodes of PVT started with short coupled VPB. Some of them terminated spontaneously with long pause of rhythm (Picture **3**). Recurrent PVT/VF occurred 6 times per one hour and most of them were converted to sinus rhythm with electrical cardioversion.

Acute PVT/VF were treated with intravenous betablocker betalock with starting dose of 5 mg up to 50 mg per hour and amiodarone 600 mg intravenous per hour. Pharmacotherapy brought to abolish of PVB and PVT/VF after sinus rhythm was decreased up to 50 b.p.m. (Picture **4**). After PVT/VF were successfully treated with antiarrhythmic drugs, cardioverter defibrillator Protecta DR (Medtronic, USA) was implanted.

She was discharged from the hospital on amiodarone 200 mg and betablocker – bisoprolol 5 mg per day. Six month later alveolitis was diagnosed and amiodarone was canceled. For next one year she hadn't had any syncope or palpitation, but HM ECG and 12 lead surface ECG showed premature ventricular contraction with the same morphology and coupling interval as it was before. It became the reason to refer patient for radiofrequency ablation of PVB, potential cause of PVT/VF.

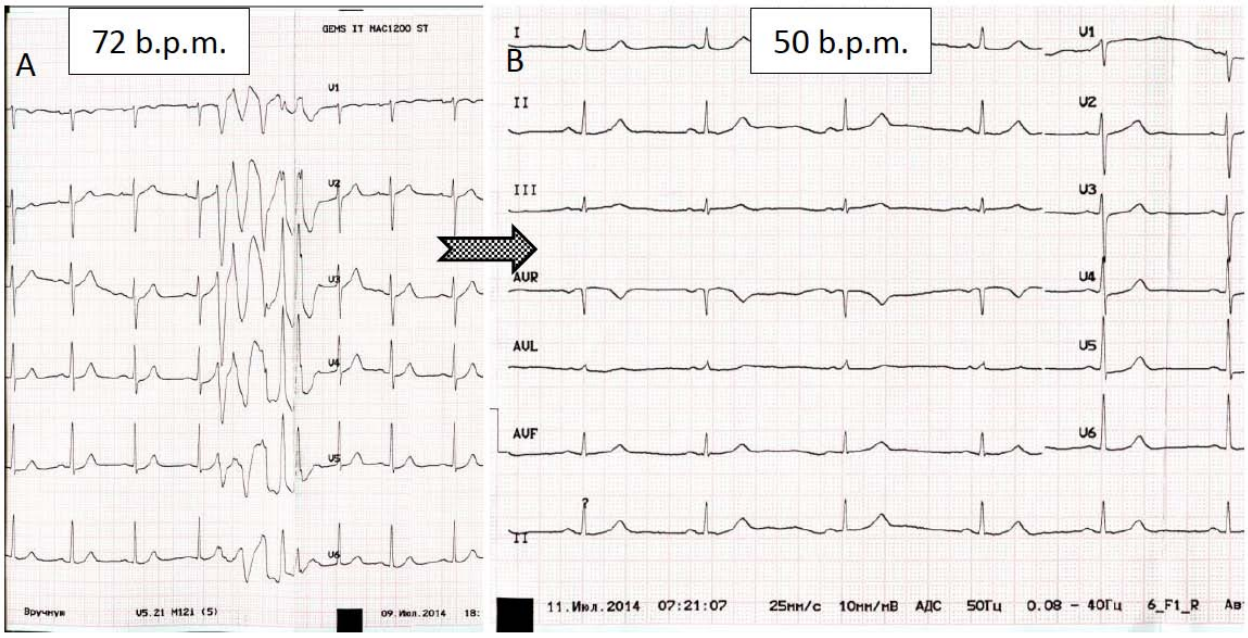
Electrophysiology investigation and PVB RFA was performed using CATRO 3 magnetic navigation system. Coronary sinus, His and right ventricle multipolar electrodes catheter were used. Fast anatomical and activation mapping were performed with ThermoCool 3,5 mm irrigated catheter (Biosence Webster). Interventricular septum was reconstructed. The early activation site of VPB was found in RV inferoseptal localization, 26 ms ahead of the PVB onset surface ECG. Ablation lesions were delivered at the earliest site with radiofrequency energy up to 35 Wt with 47° C. Fluoroscopic and 3D electroanatomical image at the site of the earliest PVB activation are shown in Pictures **5** and **6**, respectively.

The start of RF lesion resulted in monomorphic ventricular rhythm acceleration with 92-95% matching to template and advance of the Purkinje potential when

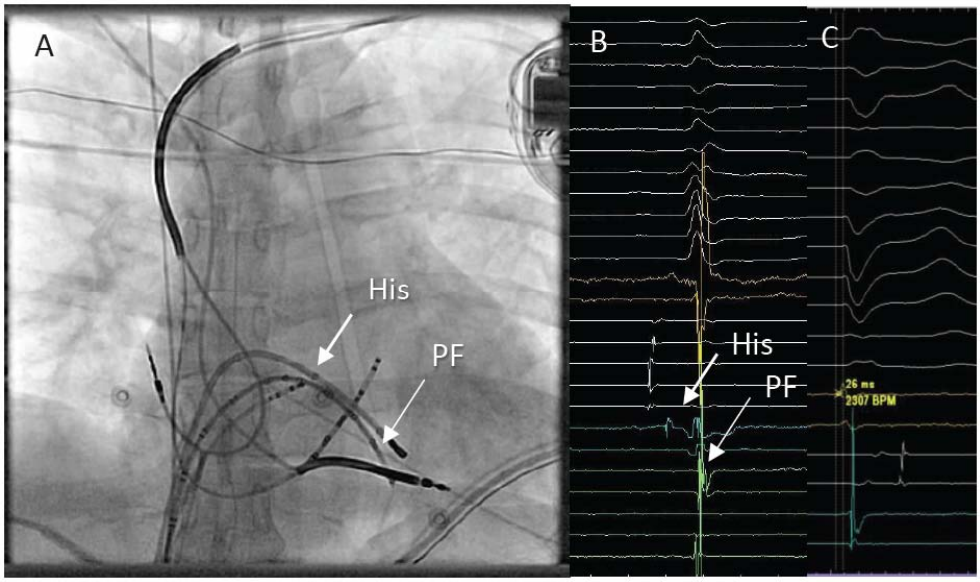


Picture 3: Numerous recurrent PVT episodes degenerating into ventricular fibrillation (A, B, E, F).

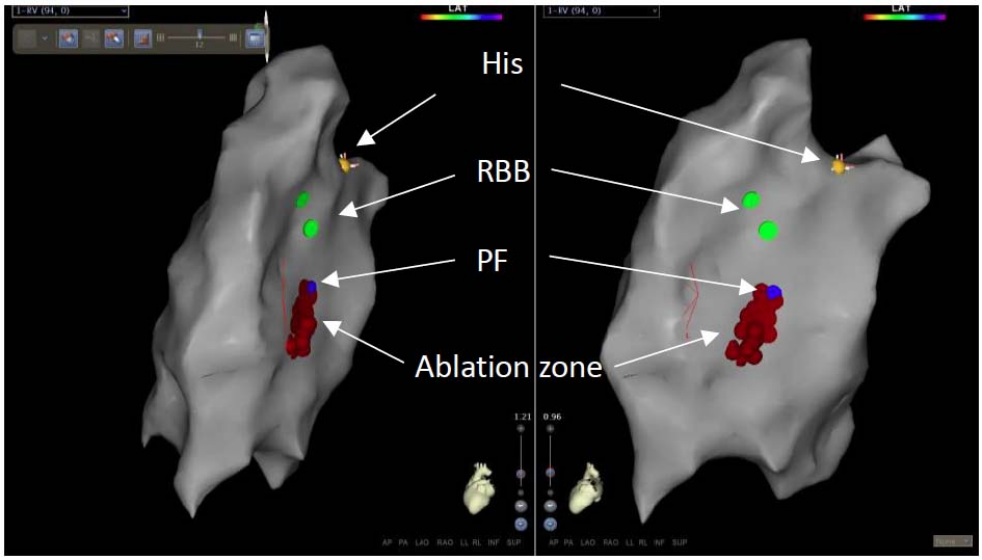
All of them started with short coupled monomorphic VPB with coupling interval of 230-280 mc (A, B, E, F). Some of them terminated spontaneously with long pause of rhythm (C). Episode of VF (D).



Picture 4: Pharmacotherapy with betalock and amiodarone brought to abolish of PVB and PVT/VF after sinus rhythm was decreased up to 50 b.p.m. A – before antiarrhythmic drugs, B – after betalock and amiodarone infusion.



Picture 5: A – X-Ray of four catheters for mapping of earliest activation of PVB (His – His bundle; PF – Purkinje Fibers); B – surface and endocardial electrograms; C – spontaneous clinical VPB occurred 26 ms before surface ECG QRS. Initial sharp potential (A) presents a peripheral Purkinje component during sinus rhythm in the site of VPB onset (C).



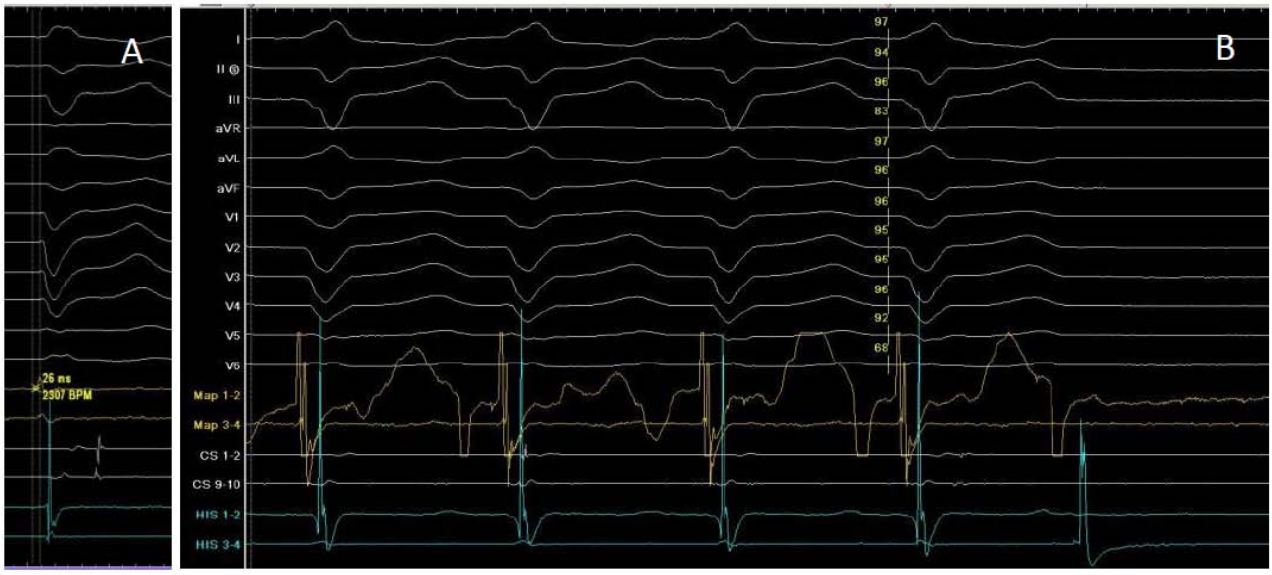
Picture 6: Fast anatomical mapping reconstruction of interventricular septum; 3D electroanatomical image shows localization of His potential, right bundle branch (RBB) and Purkinje Fibers (PF) - culprit of PVB. Activation mapping suggests the earliest signal in the lower part of interventricular septum in the right ventricle. Ablation zone indicates site of successful PVB ablation.

accelerated ventricular rhythm occurred. Termination of ventricular rhythm and PVB was happened in 5 second of energy delivery (Picture **7**). Thirty minutes follow up period didn't reveal any ectopy's and procedure was finished.

The patient was maintained on bisoprolol 5 mg per day. Follow up period over 4 years with Holter ECG monitoring twice per year, regular ECG recordings and doctor appointments didn't reveal any ventricular rhythm disturbances. The effectiveness of radiofrequency ablation in treatment and prevention of short coupling PVT and VTVF storm in clinical case was confirmed by the device memory.

DISCUSSION

Polymorphic VT has a continuously changing QRS configuration from beat to beat indicating a changing ventricular activation sequence [4]. Polymorphic VT storm is rare clinical syndrome and occurred in 7% of all ventricular arrhythmia storms defined as three or more separate episodes of sustained VT within 24 h, each requiring termination by an intervention [4, 5].



Picture 7: The start of RF lesion resulted in monomorphic ventricular rhythm acceleration with 92-95% matching to template and advance of the Purkinje potential when accelerated ventricular rhythm occurred. Termination of ventricular rhythm and PVB was happened in 5 second of energy delivery.

Polymorphic VT in absence of QT interval prolongation is commonly associated with signs or symptoms of recurrent myocardial ischemia [6], hypertrophic cardiomyopathy or acute myocarditis [7]. Another cause of PVT is catecholaminergic PVT [8].

The case presentation shows step wise approach to management of recurrent SC PVT. In this clinical case PVT degenerated into VF and presented a storm of ventricular arrhythmias terminated by electric cardiac current shock and suppressed by antiarrhythmic drugs. Patient presenting idiopathic PVT/VF. Twelve lead ECG was normal. There were no repolarization abnormalities or QT prolongation. But it is known that approximately 25% of long QT syndrome patients have a normal corrected QT (QTc) [9]. Variability of QTc in the range of 410-470 ms may be observed among both LQTS carriers and noncarriers [10]. Polymorphic VT in the clinical case started from monomorphic PVB didn't follow by long-short-long phenomenon.

Ventricular tachycardia presented rather torsade de pointes PVT than bidirectional VT and was not associated with physical or emotional exertion. Short coupling PVT was only diagnosis of this tachyarrhythmia.

Electrical storm in SC PVT occurs unpredictably at rest without any sign. The prevalence of this lethal arrhythmia remains unknown.

Calcium channel leakage resulting from RyR2 gene mutations and an abnormal His-Purkinje system are most possible cause of this arrhythmia. Mutations in the RyR2 gene, coding for catecholaminergic polymorphic ventricular tachycardia (CPVT), have been recently reported among familial cases of SC PVT [11, 12]. Contrasting with CPVT, SC PVT occurs at rest, and calcium channel leakage at rest may contribute to occurrence of SC PVT. In patients with short coupled PVBs, speculated M. Scheiman, calcium waves can result in delayed after depolarization, which lead to reentrant arrhythmias due to defective gate function. It could explain the short coupling interval as well as focal nature of the disease process [13]. Calcium waves have been shown to occur in normal Purkinje cell even without electrical stimulation, originate at cell borders and can propagate through the full extent of Pukinje cell aggregate [14]. These studies show that Purkinje cells are capable of generating both automatic and triggered rhythm. Calcium antagonist verapamil should be considered for suppress or prevention of electrical storm in SC PVT (class IIa recommendation, level of evidence B) [1]. But not the only one. Some studies suggested that the IKr blocker nifekalant may suppress arrhythmia inducibility by prolonging repolarisation, arguing against a causal role of EADs [15,16]. Nifekalant has a dose-dependent IKr channel–blocking effect in the ventricles, sinoatrial node, and atrioventricular node, without affecting the L-type calcium channel current, sodium current, or inward rectifier potassium current. Another, but not pure IKr channel blocker, is amiodarone, multichannel drug with potassium channel blocker effect. From the other hand, b-blockade of both b1- and b2-receptors remains an

important treatment, which can reduce the risk of recurrent VT and VF by more than 50% [17], likely by increasing the threshold required for fibrillation [18]. Ventricular fibrillation storm was successfully treated with amiodarone in combination with betablocker betaloc. Abolish of PVT and VF storm was associated with decrease of heart rhythm (Picture **4**).

The common algorithm for acute management of VF storm with amiodarone and betablockers was adopted by Cardiological Society in 2006 [19]. Taking in account that antiarrhythmic drugs are not eliminate the risk of PVT/VF recurrence, implantable cardioverter defibrillator (ICD) was implanted [1]. Torsade de pointes (TdP) meaning “twisting of the points”, first described by Dessertenne, is a potentially life threatening form of polymorphic ventricular tachycardia characterized by changing amplitude of the complexes with characteristic feature of beat-to-beat varying QRS morphology twisting around the isoelectric baseline [20]. Late, in 1994, Leenhardt A *et al*. reported 14 cases were selected among patients with no structural heart disease after severe syncopal attacks due to documented ventricular tachyarrhythmias initially qualified as VF. The cases presented with syncope related to typical torsade de pointes with no evidence long QT syndrome and extremely short coupling interval of the first beat or the isolated premature beats (245 +/- 28 ms) in contrast to the long coupling interval of the first beat in the classic TdP [21].

Ashida K. *et al*. first reported successful ablation of right ventricular outflow tract (RVOT) triggers in a patient with recurrent VF episodes. [22]. Premature ventricular contraction was reproduced by pace-mapping at the septal RVOT and it seems to be not short coupled PVC. Kusano *et al*. [23] showed the efficacy of catheter ablation of short-coupled PVBs and polymorphous VT originated from the right ventricular outflow tract. Haïssaguerre *et al*. 2002 [2] then revealed that the PVC triggering VF could more commonly arise from either the right or the left Purkinje system, and as well but less common either from ventricular muscle. The interval from the Purkinje potential to myocardial activation in sinus rhythm was 11±5 ms, suggesting that recordings were obtained from the distal Purkinje fibers. The Purkinje potential was recorded 10 to 150 ms before the ventricular premature beat. They noted that the trigger PVB was either monomorphic or pleomorphic and usually was localized to the Purkinje system. Ablation of the Purkinje potentials resulted in a very high rate of elimination of PVB of 89%.

Radiofrequency catheter ablation is highly effective in the treatment of critically timed PVBs that are responsible for triggering VF in structurally normal heart. Electrophysiology mapping and RFA of these PVBs shows high effectiveness at eliminating or reducing of VF episodes and may be life-saving in cases of electrical storm. In cases of recurrence of PVT triggered by PVBs despite drug therapy, catheter ablation should be strongly considered. The ablation target is the PVB initiating TdP [1]. Mapping can be performed by conventional electrophysiology techniques taking two or four intracardiac catheters to identify the earliest site of ventricular activation and Purkinje fibers which precede PVB onset. Fast electroanatomical mapping and 3D reconstruction helps to pointing out the site of triggering PVB and subsequent successful ablation. Activation mapping performs for matching paced ventricular contraction to native PVB triggering VF especially in case of insufficient PVB frequency at the time of the EP study to allow prompt mapping to maximize success of ablation procedure.

CONCLUSION

The short-coupled variant of PVT triggering VF in the clinical case responsible for sudden death in the absence of any evidence of structural heart disease and acute illness. We have found efficacy of class III antiarrhythmic drug amiodarone in combination with betablocker in treatment of electrical storm. Implantation of ICD strongly suggested in this SC PVT. Radiofrequency ablation revealed high efficacy in long term prevention of recurrence of PVC triggering PVT and VF.

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