

Is the Advanced Cardiac Life Support Algorithm Always Correct? A Case of Catecholaminergic Polymorphic Ventricular Tachycardia

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ABSTRACT

A 19-year-old male with catecholaminergic polymorphic ventricular tachycardia (CPVT) presented with outside-of-hospital cardiac arrest in electrical storm. Ventricular arrhythmias persisted while following the Advanced Cardiac Life Support (ACLS) algorithm using epinephrine boluses. Upon deviation from the ACLS algorithm and use of beta blockers, ventricular arrhythmias ceased and patient was successfully stabilized. Treatment of CPVT electrical storm is challenging due to limited guidelines. Antiarrhythmic agents such as beta blockers and flecainide are

among the primary treatment options. This case report highlights a unique example of how deviation from traditional ACLS may be necessary to best treat CPVT storm.

Keywords: Catecholaminergic polymorphic ventricular tachycardia, ventricular arrhythmia, electrical storm, ACLS

INTRODUCTION

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare inherited arrhythmogenic disorder linked to mutations in the cardiac ryanodine receptor (RYR2) and cardiac calsequestrin 2 (CASQ2) [1,2]. CPVT is characterized by adrenergic-induced bidirectional and polymorphic ventricular tachycardia (VT) [1,2]. Due to mutations in these receptors, the sarcoplasmic reticulum inappropriately leaks calcium leading to a large amount of calcium in the cytosol which can predispose patients with CPVT to ventricular arrhythmias due to delayed afterdepolarizations [1,3].

CPVT usually presents as syncope or sudden cardiac death in children with structurally normal hearts at an average age of 8 years old [1,2]. In patients with CPVT, adrenergic stimulation, such as emotional stress or exercise, results in intracellular calcium mishandling and ventricular arrhythmias [1]. The mortality rate of untreated CPVT is reportedly as high as 31% by the age of 30 years old [1].

The resting electrocardiogram (ECG) findings include a normal QTc, which is an important distinction, as it differentiates polymorphic VTs arising from congenital long QT syndrome from CPVT [1]. However, as heart rate increases, these patients will develop monomorphic premature ventricular beats then transition into bidirectional VT, polymorphic VT, and ultimately ventricular fibrillation (VF) [1,2]. The reverse sequence in arrhythmias occur as the heart rate downtrends [1].

Ambulatory management of CPVT is focused on lifelong beta blockers (BBs), notably nadolol. BBs are usually effective for the prevention of recurrent arrhythmias, although there have been reports of recurrence of cardiac events/ incomplete prevention [4,5]. An implantable cardioverter defibrillator (ICD) should be considered in these patients who continue to have arrhythmias despite BB usage [6]. However, ICDs come with a potential to induce electrical storms due to inappropriate shocks. An alternative antiarrhythmic agent, flecainide, has shown promise, in conjunction with conventional drug therapy, to reduce exercise-induced ventricular arrhythmias in patients with CPVT [6,7]. Specifically, flecainide directly blocks the RYR2 channels, thus preventing calcium release preventing ventricular arrhythmias [6,7]. A potential treatment to consider in refractory cases to BBs is left cardiac sympathetic denervation, in which the left stellate ganglion (T1) and thoracic ganglia (T2-T4) are resected, interrupting norepinephrine release, thus providing an anti-fibrillatory effect [2]. However, this technique has side effects and requires an experienced surgeon [1,2]. Recommended lifestyle medications include limiting competitive sports, strenuous exercise, and stressful situations [2].

Treatment of patients with CPVT storm is challenging because of limited guidelines. Consistent with treatment for prevention, BBs are first line during acute electrical storm. Intravenous BBs can be combined with IV procainamide as well. Additionally, deep sedation and mechanical circulatory support (MCS) should be considered when electrical storm is refractory to IV

antiarrhythmics [3]. According to the current 2020 Adult Basic Life Support (ACLS) algorithm, the first drug of choice is epinephrine for a patient in VF/pulseless VT [8]. However, in patients with CPVT, epinephrine can rather be arrhythmogenic. Here, we present a case of VT storm successfully treated by deviating from the ACLS algorithm.

CLINICAL CASE

A 19-year-old male with medical history notable for single chamber ICD implant for CPVT with remote cardiac arrest in childhood presented with out-of-hospital cardiac arrest. In the field, the patient was intubated and defibrillated for VF. Upon arrival to the hospital, he developed multiple episodes of bidirectional VT degenerating to VF requiring defibrillation [Fig.1].

He was admitted to the cardiovascular intensive care unit with VT storm, requiring extensive CPR with chest compressions and numerous external shocks, ultimately requiring MCS for support. He was initiated on intravenous amiodarone, lidocaine, and procainamide. During the ACLS algorithm with chest compressions and intermittent epinephrine boluses, it became apparent that the arrhythmia burden worsened after epinephrine. In addition, with resuming chest compressions immediately after defibrillation, it was clear that a narrow complex rhythm degenerated back into VT and VF during continued chest compressions prior to the next scheduled pulse check [Fig. 2].

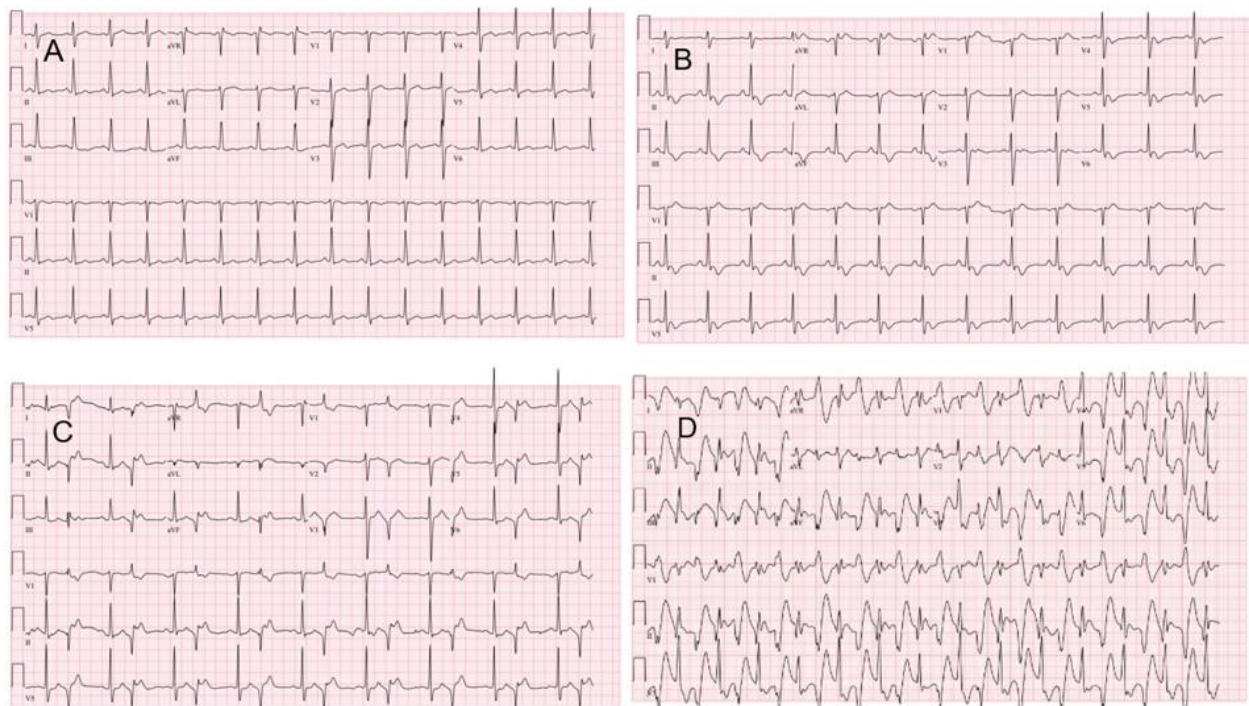
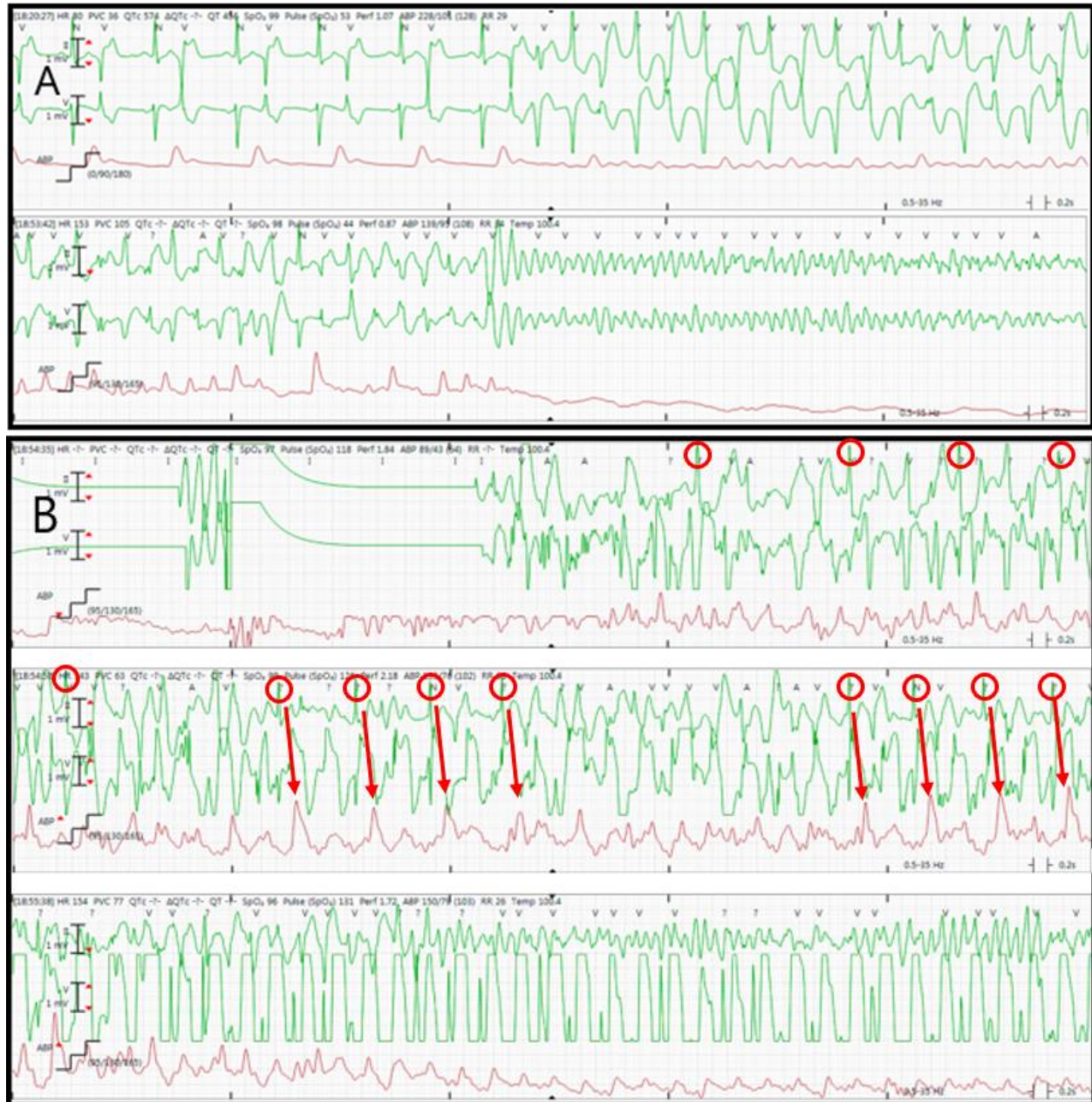


Fig. 1: 12-lead electrocardiograms from admission. (A) Presenting ECG with sinus rhythm. (B) Repeat ECG with sinus rhythm and T wave inversion in inferior and lateral leads. (C) Repeat ECG with frequent short-coupled premature ventricular contractions in a pattern of bigeminy. (D) Final ECG with beat-to-beat alternation of frontal QRS axis consistent with bidirectional ventricular tachycardia.

The code team elected to cease epinephrine boluses and perform an early pulse check after the next defibrillation. With this, sinus rhythm with a pulse was achieved after the next

defibrillation, and the patient transiently maintained normal sinus rhythm, but again degenerated into VF. The decision was made to pursue VA ECMO. With MCS, the patient's medical regimen was consolidated to procainamide and BB as hemodynamics tolerated. He remained electrically quiescent for the remainder of the hospitalization.

Ultimately, the patient was decannulated and extubated. His home nadolol and flecainide were resumed. The patient later revealed he had stopped taking his home medications, leading to his arrest. At outpatient follow-up, he was doing well and surgical sympathectomy was discussed as a future management option.



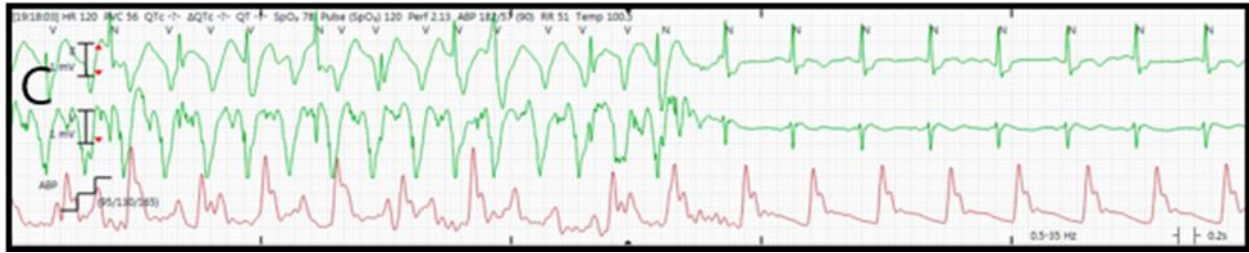


Fig. 2: (A) Telemetry tracing with two vectors showing ventricular bigeminy and then bidirectional VT which further degenerates into VF, along with the corresponding arterial waveform in red. (B) Additional tracing shows attempted defibrillation followed by chest compressions. There appears to be a narrow complex rhythm after defibrillation that corresponds to the arterial waveform (narrow complexes highlighted with red circles and corresponding arterial wave form highlighted with red arrows), but this degenerates back into VF during chest compression. (C) Final tracing shows return of a narrow complex rhythm after subsequent defibrillation. This was recognized and early pulse check was performed, which revealed a perfusing sinus rhythm.

DISCUSSION

This case highlights an example when deviation from traditional ACLS protocol is necessary. Electrical storm, due to CPVT, is characterized by adrenergic-induced ventricular arrhythmias. This patient was initially treated following the ACLS algorithm with persistence of ventricular arrhythmias. Chest compressions and defibrillation resulted in increased adrenergic tone paradoxically creating a cycle of worsening electrical storm. As such, the team deviated from the algorithm by withholding additional epinephrine boluses and administered IV BB and procainamide, with return of sinus rhythm. Additionally, an early pulse check immediately after defibrillation appeared to help prevent the return to VT. Algorithms provide a guideline for medical practice, however, this case provides a unique example of how deviation from traditional ACLS may be necessary to best treat CPVT storm.

CONCLUSIONS

CPVT is a rare inherited disorder that can cause cardiac arrest from ventricular arrhythmias in the setting of elevated catecholamines. During resuscitative efforts of VT storm, avoidance of epinephrine and use of IV antiarrhythmics are best steps of management in ACLS.

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