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CASE REPORT: CLINICAL CASE

Ventricular Tachycardia Storm in a Young Adult Post-ALCAPA Repair

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ABSTRACT

Anomalous left coronary artery to pulmonary artery (ALCAPA) is a leading cause of pediatric myocardial ischemia. This paper presents a case of a young man presenting with ventricular tachycardia storm 18 years after ALCAPA repair. Clinicians should recognize the risk of ventricular tachycardia in this patient population. (JACC Case Rep 2024;29:102418)

HISTORY OF PRESENTATION

An 18-year-old man with a remote history of anomalous left coronary artery to pulmonary artery (ALCAPA) repair in infancy presented to the emergency department (ED) for recurrent palpitations. His heart rate was 200 beats/min with blood pressure of 73/64 mm Hg. He was diaphoretic and tachycardic on examination. Laboratory data were notable for high-sensitivity cardiac troponin I level of 228 pg/mL (normal ≤20 pg/mL). Electrocardiogram (ECG) showed wide complex tachycardia (WCT) with an atypical right bundle branch block (RBBB) and inferior axis at a rate of 200 beats/min, consistent with monomorphic ventricular tachycardia (MMVT) (**Figure 1A**). The patient was electrically converted to sinus rhythm and admitted to the cardiac care unit (CCU). An implantable cardioverter-defibrillator (ICD) was placed, and he was discharged on metoprolol succinate. Less than 48 hours later, he returned to the ED with ventricular tachycardia (VT) storm with multiple ICD shocks and was readmitted to the CCU.

LEARNING OBJECTIVES

- To recognize the risk of VT in adults despite early and successful ALCAPA repair in infancy.
- To distinguish VT from other forms of wide complex tachycardia.
- To formulate an appropriate surveillance protocol to mitigate the risk of sudden cardiac death in this high-risk patient population.

PAST MEDICAL HISTORY

As a neonate, the patient developed recurrent respiratory distress, ultimately leading to the diagnosis of

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ABBREVIATIONS AND ACRONYMS

AAD = antiarrhythmic drug

ALCAPA = anomalous left coronary artery to pulmonary artery

CCU = cardiac care unit

ECG = electrocardiogram

ED = emergency department

ICD = implantable cardioverter-defibrillator

LV = left ventricular

MMVT = monomorphic ventricular tachycardia

RBBB = right bundle branch block

SVT = supraventricular tachycardia

VT = ventricular tachycardia

WCT = wide complex tachycardia

dilated cardiomyopathy and ALCAPA at 5 weeks of age. He underwent repair via coronary reimplantation and was noted to have a large anterolateral myocardial infarction intraoperatively. The patient was followed by pediatric cardiology growing up, with regular echocardiographic surveillance and Holter monitoring. Left ventricular (LV) systolic function was normal throughout this time.

Nine months prior to the current episode, he presented to the ED with chest pain, and was found to have a non-ST elevation myocardial infarction with a peak high-sensitivity cardiac troponin I level of 2,084 pg/mL (normal ≤20 pg/mL). ECG showed normal sinus rhythm with right axis deviation and incomplete RBBB (**Figure 1B**). Cardiac catheterization showed no anastomotic strictures, kinking, or obstruction of the coronary artery but mildly reduced LV systolic function. He was discharged on guideline-directed medical therapy, including carvedilol.

This event was followed by multiple instances of palpitations over the subsequent months. ECGs were interpreted as sinus tachycardia with RBBB or as supraventricular tachycardia (SVT) with aberrancy in the 190s (tracings not available), requiring cardioversions in the field by paramedics. He was discharged each time after an unremarkable evaluation from the ED.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for a young patient presenting with WCT includes VT, SVT with aberrancy, SVT with antegrade conduction via an accessory pathway, and antidromic atrioventricular reentrant tachycardia.

INVESTIGATIONS

Cardiac magnetic resonance performed prior to ICD placement revealed LV ejection fraction of 42% and late gadolinium enhancement in the anterolateral LV wall (**Figure 2**). After presenting with VT storm, his ICD was interrogated and showed 26 appropriate shocks for VT.

MANAGEMENT

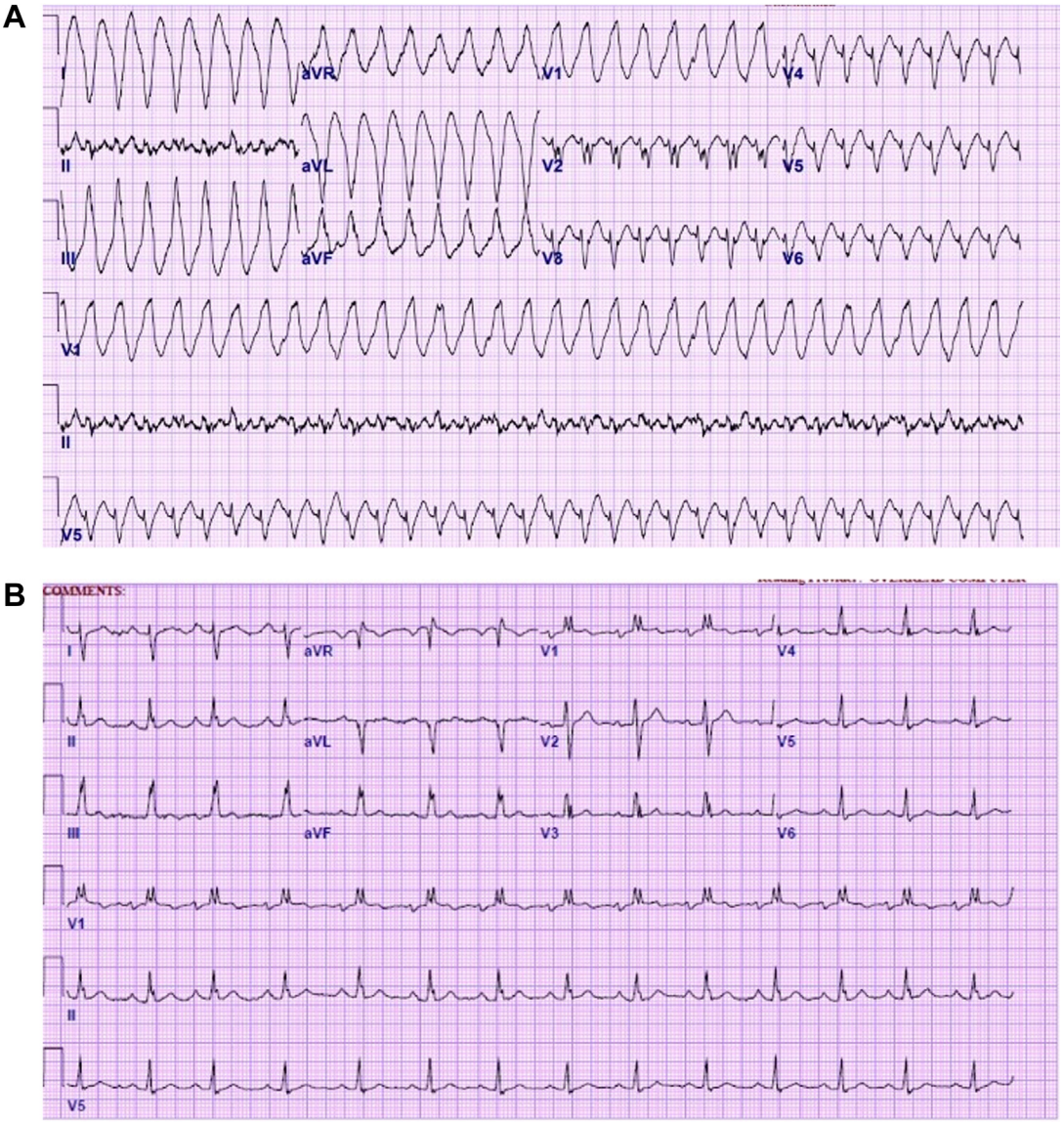
After readmission to the hospital for VT storm, the patient required amiodarone and lidocaine infusions, intubation, and sedation. He underwent endocardial VT ablation. No VT could be induced; however, electroanatomic maps showed scar along the anterolateral left ventricle with local abnormal ventricular activities and decrement-evoked potentials along the basal aspect of the scar (**Figures 3 and 4**). These areas were targeted for ablation with extensive substrate modification. Postprocedure, the patient was extubated, and intravenous antiarrhythmic drugs (AADs) were discontinued. He was discharged on mexiletine and carvedilol.

DISCUSSION

ALCAPA is a congenital heart defect wherein the left coronary artery originates from the pulmonary trunk instead of the aorta.[1] Without treatment, 90% of infants die in the first year of life. There are a scarcity of data on long-term survival outcomes after pediatric ALCAPA repair. However, various cohort studies show that ALCAPA has a favorable prognosis for mortality with early repair, with survival ranging from 86% to 100% at 10 years.[2] Late sequelae postrepair includes mitral regurgitation, ischemic cardiomyopathy, scar-mediated arrythmias, and sudden cardiac death. Life-long monitoring is recommended due to the risk of late complications, which remains largely unquantified by large-scale studies.[3]

This case highlights the importance of the past medical history in risk stratification. He had a late ALCAPA repair as demonstrated by the presence of a large anterolateral myocardial infarction during the original surgery (and persistent scar in the same territory by cardiac magnetic resonance 18 years later). Because myocardial scar is a well-recognized substrate for arrhythmogenic sequelae and sudden cardiac death, lifelong surveillance was warranted. Palpitations in this patient should have been aggressively evaluated, particularly after a non-ST-segment elevation myocardial infarction. His multiple presentations of WCT, with associated hemodynamic instability, were unfortunately dismissed as benign due to his young age, a form of anchoring

FIGURE 1 **Electrocardiograms**

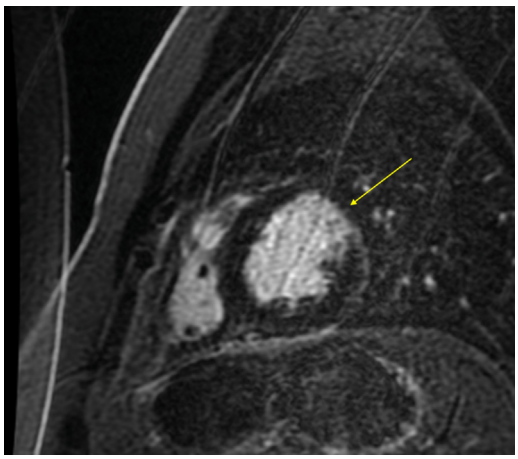


(A) Monomorphic ventricular tachycardia with atypical right bundle branch block, right inferior axis, and atrioventricular dissociation (best seen in V_1), with initial dominant R in aVR, monophasic R waves in V_1 and aVR, and rS in V_2 to V_6. (B) Sinus rhythm and incomplete right bundle branch block.

bias, and his baseline RBBB. Had his past medical history been taken into more careful consideration, the diagnosis of VT could have been made much sooner.

His past medical history aside, the ECG demonstrating WCT is most consistent with VT. Several features of the ECG help distinguish between VT and SVT with aberrancy in the setting of a WCT. In this case, factors favoring VT includes regular rate, wide QRS interval >140 milliseconds in an atypical RBBB, right inferior axis, and atrioventricular dissociation, with initial dominant R in

FIGURE 2 Cardiac Magnetic Resonance

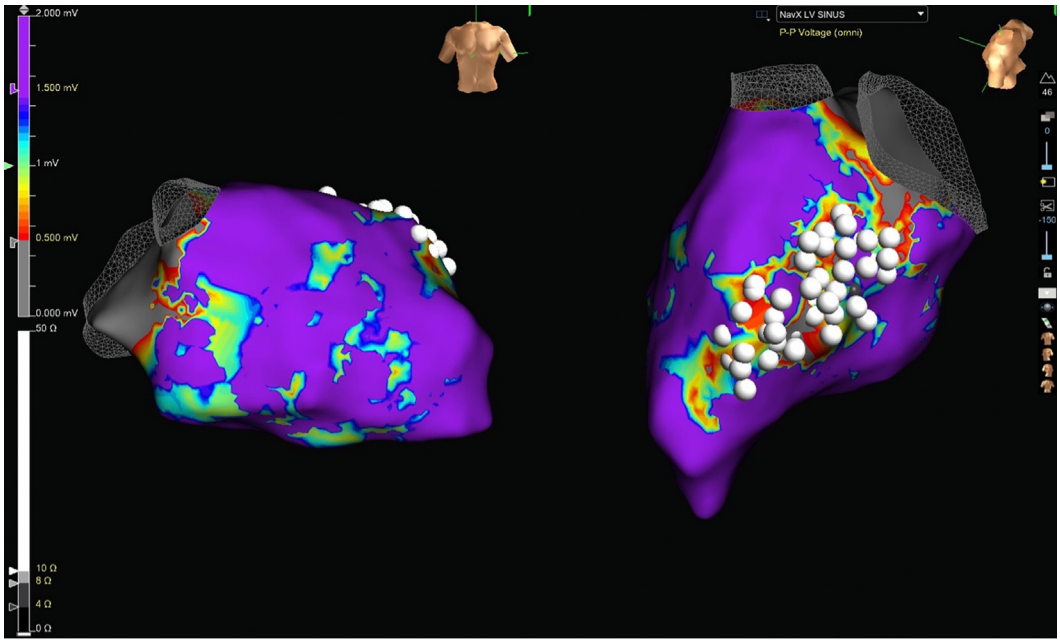


Late gadolinium enhancement is seen in the anterolateral wall of the left ventricle (yellow arrow).

aVR, monophasic R waves in V_1 and aVR, and rS in V_2 to V_6.

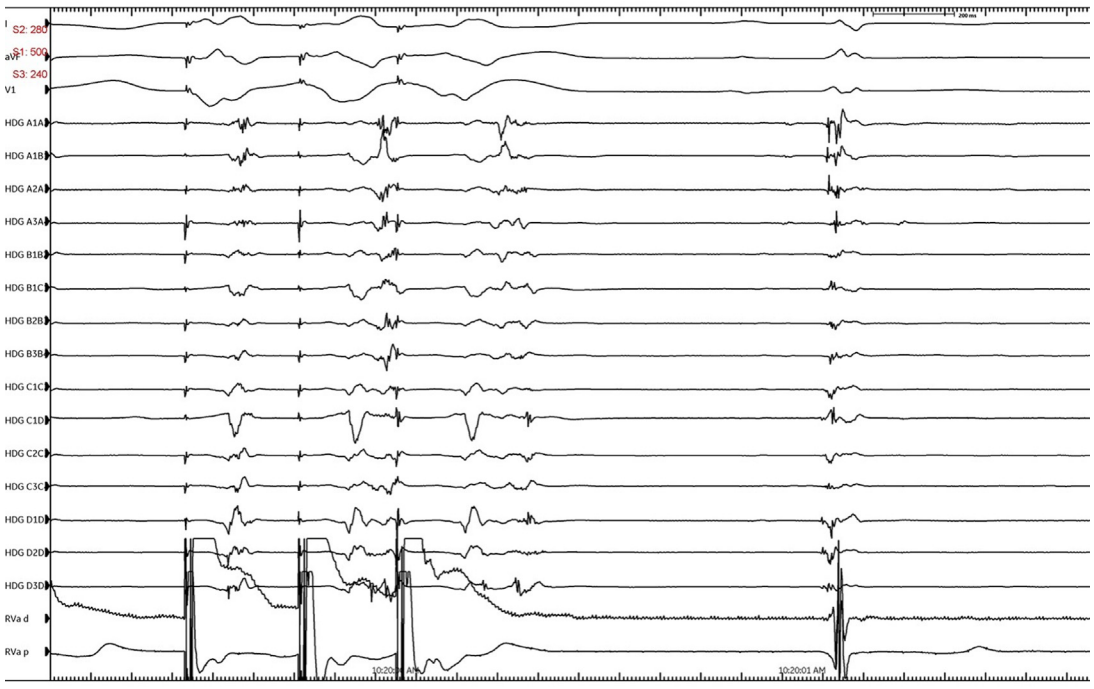
Per the American College of Cardiology/American Heart Association guidelines, the initial management of sustained MMVT in patients with structural heart disease includes direct current cardioversion (Class I) and intravenous AADs such as intravenous procainamide (Class IIa), amiodarone, or sotalol (Class IIb), along with sedation and anesthesia.[4] VT occurring after coronary surgery warrants imaging to exclude any anatomic abnormalities (eg, anastomotic stricture, kinking of the vessel). In this case, this was accomplished with coronary angiography and cardiac computed tomography. Management of VT in young patients can be particularly challenging because the toxicity associated with many AADs is not ideal for this age group. VT refractory to medications and antitachycardia pacing has a Class I recommendation for catheter ablation. We favored an aggressive approach in this young patient in VT storm with substantial myocardial scar, opting for

FIGURE 3 Electroanatomic Voltage Map



Voltage maps (NavX) during sinus rhythm at standard scar settings (0.5-1.5 mV) with AP (left) and LAO cranial (right) views showing areas of low voltage along basal superior aspect and along the entire anterolateral left ventricular wall. AP = anterior-posterior; LAO = left anterior oblique.

FIGURE 4 **Intracardiac Electrograms**



Decremental behavior during pacing seen with premature beats signifies slow conduction and area of interest for ablation.

ablation during his second admission to the CCU in lieu of trialing him on AADs.

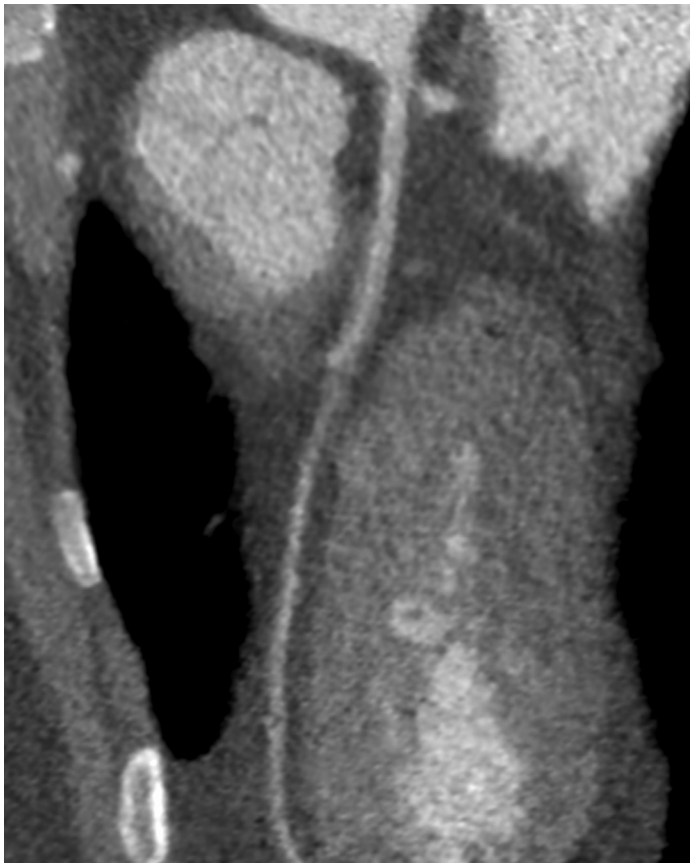
FOLLOW-UP

At 1-month follow-up, the patient had SVT but no VT recurrence as confirmed by ICD interrogations. Cardiac computed tomography showed no stricture, kinking, or obstruction of the reimplanted left coronary artery with normalized LV ejection fraction (Figure 5).

CONCLUSIONS

We describe a case of a young adult man with a history of ALCAPA repair presenting with VT storm mediated by LV scar 18 years after his original surgery. Ischemic insults and resultant myocardial scars sustained as an infant became the substrate for VT. Patients with a history of repaired congenital heart disease who present with WCT warrant a high index of suspicion for VT despite their age. Managing VT in young patients can be difficult due to the long-term adverse effects of many AADs. When VT is resistant to medical therapy, catheter ablation becomes an important option. There is currently a lack of consensus regarding the optimal surveillance strategy for VT in young patients post-ALCAPA repair, suggesting a need for the inclusion of ALCAPA surveillance strategies in future expert consensus guidelines.

FIGURE 5 Cardiac Computed Tomography



There was no kinking or strictures of the reimplanted left coronary artery.

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