Adenosine sensitive left ventricular summit ventricular tachycardia in a pre-adolescent: case report

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Background	Sustained forms of tachycardia especially from the left ventricular summit are rare. Adenosine sensitive outflow tachycardias, especially from the left ventricular summit, are rarer still. These arrhythmias may be exercise or stress induced as they are facilitated by catecholamines and characteristically terminate with adenosine, vagal manoeuvres, and beta-blockers. The surface 12-lead electrocardiogram can be used to localize the anatomic site of origin before catheter ablation; however, prediction of the precise origin may still be challenging due to the intimate and complex anatomy of the outflow tracts.
Case summary	A 12-year-old female presented to an emergency room with frequent runs of wide complex tachycardia that terminated with adenosine but would spontaneously reinitiate. After three additional temporary terminations with adenosine and because of an inability to completely eliminate tachycardia, she was started on an esmolol infusion that resulted in an abrupt termination of tachycardia. At follow-up, she reported breakthrough episodes of tachycardia with exercise, especially associated with beta-blocker non-compliance. The rest of her cardiac testing was normal apart from an anomalous right coronary artery origin from the left coronary sinus. Given the increased frequency of symptomatic palpitations and medication non-compliance, she underwent an electrophysiology study. During the study, a ventricular tachycardia was successfully mapped to an epicardial focus at the left ventricle summit and was successfully ablated.
Discussion	The response of this patient's ventricular tachycardia to adenosine suggests a triggered mechanism. To our knowledge, this is the first unambiguous example of left ventricular tachycardia due to cAMP-mediated triggered activity in this age group.
Keywords	Pre-adolescent • Ventricular tachycardia • Adenosine • Triggered activity • Ablation • Case report
ESC curriculum	5.1 Palpitations • 5.6 Ventricular arrhythmia

Learning points

- Adenosine sensitive heart rhythm disorders do not always implicate a supraventricular tachycardia.
- There are technical challenges associated with the ablation of a tachycardia from the left ventricle epicardial summit.
- The risk of coronary artery injury is of concern with epicardial summit ablations, and close monitoring during and after the procedure is very important.
- Ablation of an epicardial focus with a catheter in the endocardium can be safe and effective in a young patient.

Introduction

Left ventricular summit tachycardias are a type of outflow tract tachycardia, and sustained forms of these summit tachycardias are extremely rare. Adenosine sensitive outflow tachycardias, especially from the left ventricular summit, are rarer still, and to our knowledge, this is the first unambiguous example of left ventricular tachycardia due to cAMP-mediated triggered activity in this age group. The principal theme of this case report is to review the approach to diagnosis and treatment of this very rare form of ventricular arrhythmia.

Summary figure
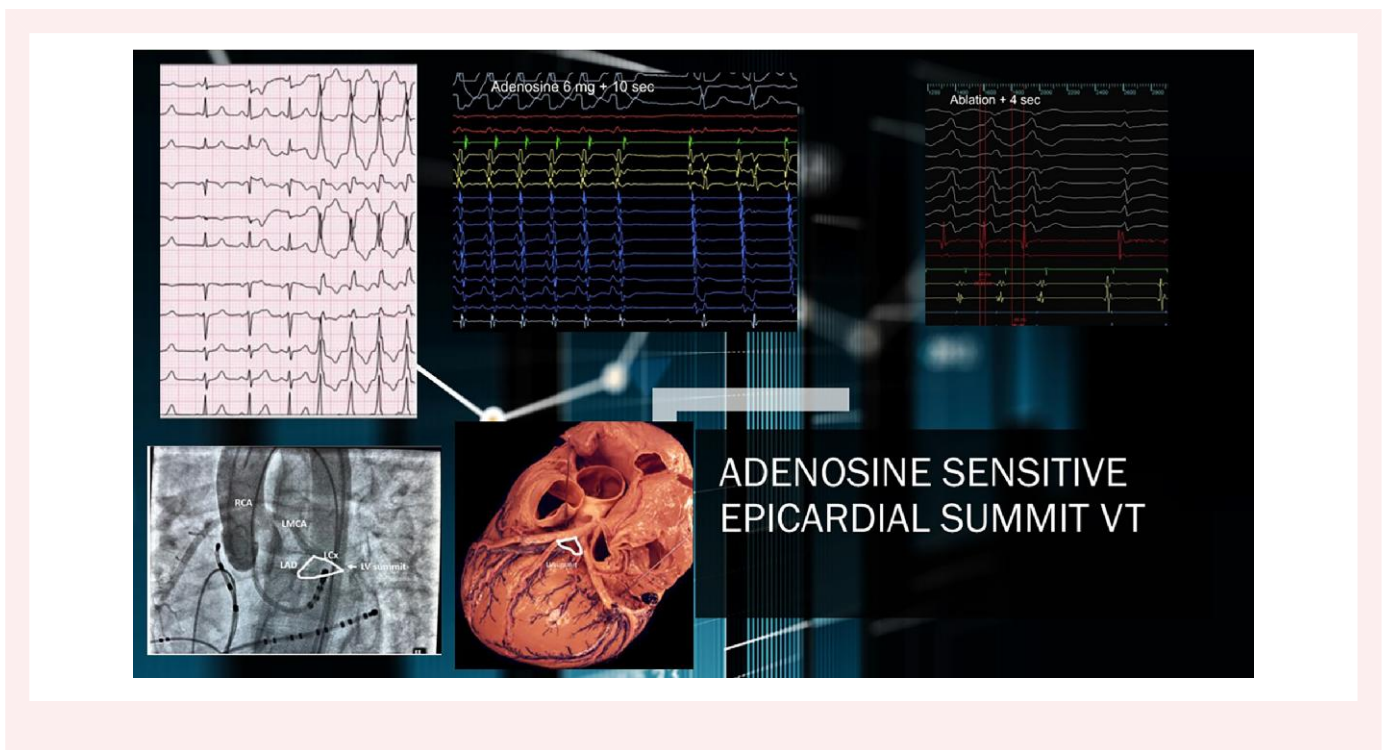
Case summary

A 12-year-old, 32 kg female presented to a local emergency room with frequent runs of wide complex tachycardia at 215 b.p.m. that terminated with adenosine (Figure 1). Apart from diaphoresis and tachycardia at presentation, her physical examination was normal. Due to the patient's age and absence of previous cardiac history, the tachycardia was presumed to be supraventricular tachycardia (SVT) with aberration. The tachycardia recurred within 2–3 min and was again terminated with adenosine. Two additional episodes responded to adenosine within several minutes; however, sustained tachycardia spontaneously recurred (see Supplementary material online, Figure S1), and was terminated with an esmolol infusion (up to 150 µg/kg/min).

Past medical history was significant for intermittent sudden onset and offset palpitations for the 12 months prior to presentation. The episodes were getting longer in duration and frequency necessitating medical attention. There was no other significant medical, surgical, or family history, and the patient was not on any medications.

The patient's resting electrocardiogram (ECG) was normal, and echocardiography and contrast enhanced cardiac magnetic resonance imaging (CMRI) showed normal biventricular structure and function. The CMRI also revealed an anomalous right coronary artery originating from the left coronary sinus.

She was admitted to the hospital, transitioned to Metoprolol-XR 25 mg daily and was discharged 48 h after admission. At follow-up, she reported breakthrough episodes of tachycardia with exercise and acknowledged medication non-compliance. Given the increased frequency of symptomatic palpitations and non-compliance, she underwent an electrophysiology study under moderate sedation with propofol after the beta-blocker was discontinued for 7 days.

During the electrophysiologic study, a wide complex tachycardia spontaneously occurred, which was consistent with ventricular tachycardia (VT). The tachycardia had a negative HV interval, and there was ventriculoatrial dissociation. An identical tachycardia was also induced with atrial burst pacing and atrial extrastimuli (Figure 2). The tachycardia cycle length was 240 ms, and the surface ECG showed R waves across the precordium and an inferior axis. The QRS duration was 157 ms with a maximum deflection index of 56%, suggestive of a possible epicardial site of origin. Administration of adenosine abruptly terminated VT (Figure 3).

To map the origin of the tachycardia, the left ventricle (LV) was accessed retrogradely from the aorta with a 4 mm-tip small curve deflectable catheter (Blazer, Boston Scientific, Natick, MA). Electroanatomical mapping identified the earliest activation site within the region of the LV summit (−40 ms). A coronary angiogram showed that the ablation catheter tip was positioned at the LV summit, just below the bifurcation of the left anterior descending and left circumflex coronary arteries, within 3 mm of the left circumflex artery (Figure 4). Radiofrequency energy (35 W) was delivered for 60 s using an irrigated catheter with abrupt termination of VT (Figure 5). Following ablation VT was no longer inducible. A coronary arteriogram performed 30 min after the final energy application was unchanged.
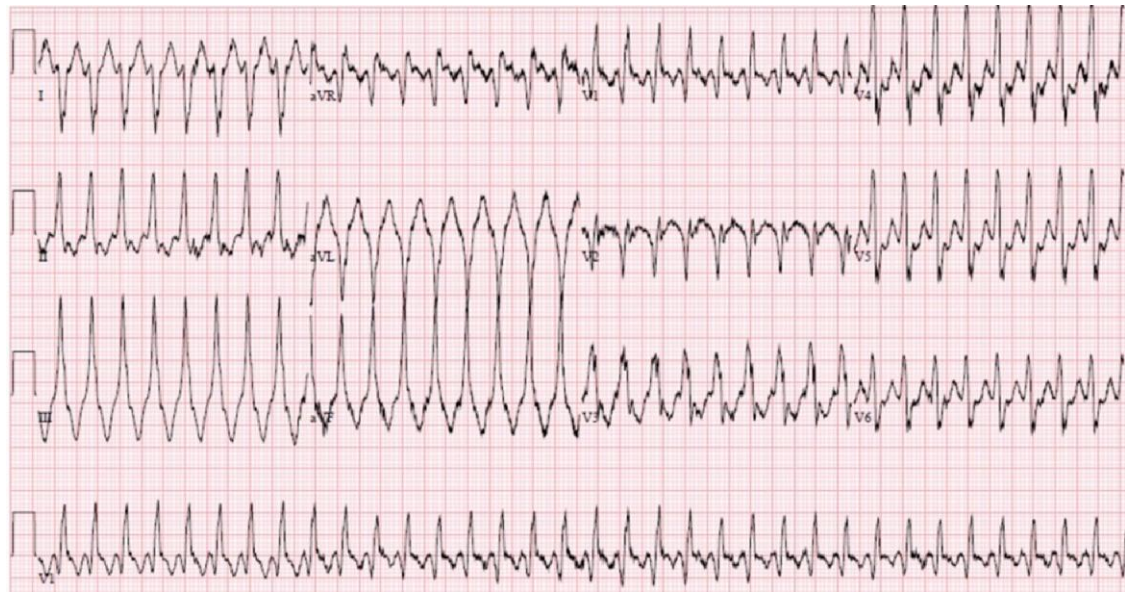
Figure 1 Surface 12-lead ECG depicting a wide complex tachycardia.

Figure 2 Intracardiac tracings and surface ECG tracings showing initiation of VT with atrial extrastimuli.

The patient was discharged home on a 30-day external event monitor and has remained free of VT for over two years post-ablation.

Discussion

Given the patient's prepubescent age and response to adenosine, one of the initial considerations would be SVT with aberrancy. These would include atrioventricular (AV) node re-entry with a bundle branch aberrancy or a bystander accessory pathway conduction or AV reciprocating tachycardia either orthodromic reciprocating tachycardia with a bundle branch aberrancy or an antidromic tachycardia.[1] Unusual types of antidromic tachycardia associated with nodofascicular/nodoventricular pathways are another consideration as well as decremental AV pathways such as atriofascicular connections. In a wide complex tachycardia including an atriofascicular connection, adenosine may terminate

Figure 3 Intracardiac and surface ECG tracings showing termination of VT with adenosine (6 mg).
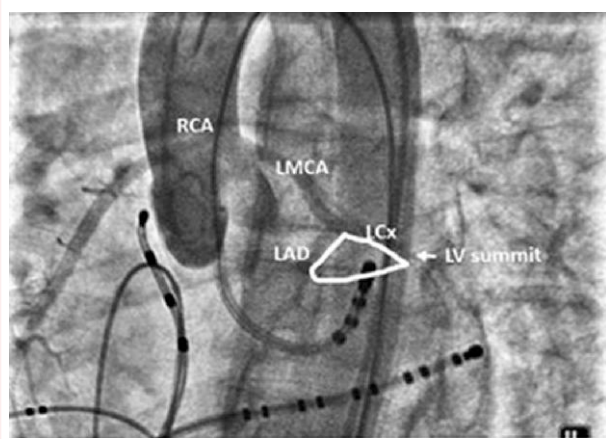

Figure 4 Fluoroscopic image in the left oblique projection with the ablation catheter noted 3 mm away from the left circumflex artery. The LV summit is depicted within the white border. Also noted is the anomalous origin of the right coronary artery from the left sinus. LMCA, left main coronary artery; LCx, left circumflex artery; LAD, left anterior descending artery; RCA, right coronary artery; LV, left ventricle.

the tachycardia with either conduction block in the accessory pathway or AV node. Ventricular tachycardias, including fascicular tachycardias, are less common in this age group, however have been noted and may be sensitive to adenosine. The ECG morphology of the tachycardia however is inconsistent with a bundle branch aberrancy, atriofascicular or a fascicular ventricular tachycardia based on a monophasic right bundle configuration in lead V_1, and positive precordial concordance. An underlying channelopathy would be unlikely based on a normal QT interval, presentation of monomorphic VT, and response to adenosine.

The LV summit of the heart is the region of myocardium located between the left anterior descending coronary artery (superior to the first septal perforating branch) and the left circumflex coronary artery.[2,3] It is bounded posteriorly by the right ventricular outflow

tract and is superior to the aortic portion of the LV ostium (see Supplementary material online, Figure S2). The great cardiac vein bisects the LV summit into an accessible inferior lateral area and a superior medial inaccessible region.[2,3] Ablation of ventricular arrhythmias originating from this region may be attempted from the posterior right ventricular outflow tract, the left and right coronary cusps, the LV myocardium below the left cusp, from under the right/left aortic cusp, from within branches of the coronary sinus, or via an epicardial subxiphoid approach.[3–6] Recently, Futyma et al.[5] have also described ablation of a summit VT from a left pulmonary cusp. In fact, due to the challenging nature of ablations in this region, detailed mapping both from the endocardial and epicardial myocardium may be required to determine the most effective and safe site for radiofrequency application.[6] In some cases, a surgical approach may be required to dissect epicardial fat and directly apply ablative energy to this region. When possible, mapping and ablation from the aortic sinus and endocardium are preferable to mapping in the coronary sinus branches, where the ability to deliver a sufficient power as well as proximity to the epicardial coronary branches may be limiting factors for success.[3,5,6]

Mapping VT origin in our case was facilitated by its incessant nature and haemodynamic stability. Despite the VT having some characteristics of an epicardial focus, we were able to achieve successful ablation from the endocardial surface, possibly because of the relatively thin ventricular wall in a child compared to an adult. This differentiates it from the challenges with ablating a summit VT from the endocardium described in the earlier case series on adult patients.[3–6] Due to the proximity of the left coronary arteries to sites of ventricular arrhythmia origin within the superior LV summit, the risk of serious coronary artery injury is a major concern, particularly in a 12-year-old patient. To this end, avoidance of radiofrequency ablation within 5 mm of a coronary artery is recommended.[4] In this report, ablation with an irrigated catheter within a 3 mm proximity to the left circumflex artery was a concern. The operators were careful to monitor for ST-segment changes during the application of radiofrequency and performed a repeat coronary angiogram before the end of the case. The patient was observed overnight for clinical signs of coronary artery injury.

The 12-lead ECG during VT had characteristics of an LV summit origin, with a right bundle branch block morphology and monophasic R wave across the precordial leads, a monophasic R pattern in all inferior leads with late notching noted in lead III, a QS pattern in leads aVR and aVL, and S waves in lead I.[7] The R wave in III/II and S wave aVL/aVR amplitude ratios were positive, suggesting a site of origin within the accessible area.[8]

Figure 5 Intracardiac recordings of signals on the ablation catheter 40 ms before the QRS onset at the site of successful ablation, with termination of tachycardia 4 s after the onset of radiofrequency ablation.

Idiopathic ventricular arrhythmias commonly originate from the right and left ventricular outflow tractin adults with structurally normal hearts. Most of these arrhythmias manifest as frequent premature ventricular contractions. The mechanism for these arrhythmias is largely conjectural because their short, transient, and irregular pattern does not permit rigorous and reproducible mechanistic evaluation, as is applicable to the study of sustained VT. However, sustained forms of tachycardia from the outflow tract are relatively rare, particularly from the LV. Nearly all data on LV summit arrhythmias and mechanism are derived from adults. The unique aspects of this case include the unusual presentation of sustained LV summit VT in a pre-adolescent and the response of the patient's VT to adenosine, suggesting a triggered mechanism. Unlike supraventricular tissue, where the action of adenosine is via activation of inward rectifying potassium current, I_{KAdo} is not expressed in ventricular myocardium.[9,10] Therefore, the action of adenosine, in these tachycardias, is in fact, directly on the ventricular myocardium. Under conditions of cAMP stimulation, adenosine attenuates levels of adenylyl cyclase thereby terminating cAMP-mediated triggered activity. Due to its high specificity, termination of VT with adenosine, as was noted in our case, is considered to be diagnostic of cAMP-mediated triggered activity.[11,12]

To our knowledge, this is the first unambiguous example of left ventricular VT due to cAMP-mediated triggered activity in this age group.

Conclusion

Adenosine sensitive ventricular tachycardias are highly uncommon, however, due to its high specificity, termination of VT with adenosine is considered to be diagnostic of cAMP-mediated triggered activity.[10] A successful ablation of an epicardial summit VT was performed from the endocardial surface of the LV.

Lead author biography

Dr Ferns is a paediatric and adult congenital electrophysiologist with extensive experience with complex congenital ablations and device implantations. On a national level, Dr Ferns is a renowned clinical scientist and has published more than 50 peer-reviewed papers and given multiple invited talks at scientific sessions. She also serves on the Research Committee for the Heart Rhythm Society and the Pediatric Congenital Electrophysiology Society. In addition to her own career successes, she has taken an active role in mentoring several young female physicians, and prioritizes teaching fellows, residents, medical students, and nurses as part of her practice.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports*.

Data availability

The data underlying this article will be shared on reasonable request to the corresponding author.

References

1. Brugada J, Katritsis DG, Arbelo E, Arribas F, Bax JJ, Blomström-Lundqvist C, *et al.* 2019 ESC Guidelines for the management of patients with supraventricular tachycardia. The task force for the management of patients with supraventricular tachycardia of the European Society of Cardiology (ESC): developed in collaboration with the Association for European Paediatric and Congenital Cardiology (AEPC). *Eur Heart J* 2020;**41**:655–720.
2. Altmann D, Knecht S, Sticherling C, Ammann P, Osswald S, Kuhne M. Ventricular tachycardia originating from the "Bermuda Triangle". *Cardiovasc Med* 2013;**16**:208–210.
3. Enriquez A, Malavassi F, Saenz LC, Supple G, Santangeli P, Marchlinski FE, *et al.* How to map and ablate left ventricular summit arrhythmias. *Heart Rhythm* 2017;**14**:141–148.
4. Liang Z, Liu X, Li X, Zhang T, Ren X, Wu Y, *et al.* Ventricular arrhythmias ablated successfully in the subvalvular interleaflet triangle between the right and left coronary cusps: electrophysiological characteristics and catheter ablation. *Heart Rhythm* 2021;**18**:2148–2157.
5. Futyma P, Moroka K, Derndorfer M, Kollias G, Martinek M, Pürerfellner H. Left pulmonary cusp ablation of refractory ventricular arrhythmia originating from the inaccessible summit. *Europace* 2019;**21**:1253.
6. Iijima K, Chinushi M, Furushima H, Aizawa Y. Epicardial and endocardial mapping determine most successful site of ablation for ventricular tachyarrhythmias originating from left ventricular summit. *Europace* 2012;**14**:911–912.
7. Lin YN, Xu J, Pan YQ, Zhang T, Ren X, Wu Y, *et al.* An electrocardiographic sign of idiopathic ventricular tachycardia ablatable from the distal great cardiac vein. *Heart Rhythm* 2020;**17**:905–914.
8. Yamada T, McElderry HT, Doppalapudi H, Okada T, Murakami Y, Yoshida Y, *et al.* Idiopathic ventricular arrhythmias originating from the left ventricular summit: anatomic concepts relevant to ablation. *Circ Arrhythm Electrophysiol* 2010;**3**:616–623.
9. Lerman BB, Ip JE, Shah BK, Thomas G, Liu CF, Ciaccio EJ, *et al.* Mechanism-specific effects of adenosine on ventricular tachycardia. *J Cardiovasc Electrophysiol* 2014;**25**:1350–1358.
10. Lerman BB. Mechanism of outflow tract tachycardia. *Heart Rhythm* 2007;**4**:973–976.
11. Lerman BB. Ventricular tachycardia: mechanistic insights derived from adenosine. *Circ Arrhythm Electrophysiol* 2015;**8**:483–491.
12. Lerman BB, Belardinelli L, West GA, Berne RM, DiMarco JP. Adenosine-sensitive ventricular tachycardia: evidence suggesting cyclic AMP-mediated triggered activity. *Circulation* 1986;**74**:270–280.