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Arrhythmic manifestations in patients with congenital left ventricular aneurysms and diverticula

Haegeli, L M ; Ercin, E ; Wolber, T ; Brunckhorst, C ; Tanner, F C ; Jenni, R ; Lüscher, T F ; Duru, F

Abstract: Congenital left ventricular aneurysms and diverticula (LVA/Ds) are rare cardiac malformations that can be detected using echocardiography or other imaging techniques. Some of these patients present with ventricular arrhythmias. This study investigated clinical characteristics of patients with congenital LVA/D presenting with arrhythmic manifestations. Over the previous 20 years 250 patients were diagnosed to have congenital LVA/D at our institution. Diagnosis was made using echocardiography after exclusion of coronary artery disease, local cardiac inflammatory processes, traumatic causes, or cardiomyopathies. At initial presentation 32 of the 250 patients (13%, average age 45 years, range 25 to 65, 21 men and 11 women) exhibited arrhythmias. At least 2 LVA/Ds were present in 6 of these patients. LVA/Ds were localized at the posterobasal, apical, anteroseptal, and anterolateral walls in 12, 11, 4, and 5 patients, respectively. The most common complaints at presentation were syncope or presyncope in 18 patients and palpitations in 11 patients. One patient had survived sudden cardiac death. Long-term electrocardiographic recordings showed ventricular tachycardia (VT) or ventricular fibrillation in 17 patients (53%). Twelve patients underwent electrophysiologic testing. Nine patients had inducible ventricular tachyarrhythmia, whereas induced tachycardia was similar to that during spontaneous arrhythmia in 7 patients. In conclusion, patients with congenital LVA/Ds who present with arrhythmic manifestations commonly have VT. Electrophysiologic testing can reproduce clinical VT in most of these patients.

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Arrhythmic Manifestations in Patients with Congenital Left Ventricular Aneurysms and Diverticula

Short title: Left Ventricular Aneurysms and Diverticula

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Abstract

Congenital left ventricular aneurysms and diverticula (LVA/D) are rare cardiac malformations, which can be detected using echocardiography or other imaging techniques. Some of these patients present with ventricular arrhythmias. This study investigates the clinical characteristics of patients with congenital LVA/D presenting with arrhythmic manifestations. Over the last 20 years, 250 patients were diagnosed to have a congenital LVA/D at our institution. The diagnosis was made using echocardiography after exclusion of coronary artery disease, local cardiac inflammatory processes, traumatic causes or cardiomyopathies. At initial presentation, 32 of the 250 patients (13%) with an average age of 45 years (range 25 - 65 years) and predominant male gender (21 males and 11 women) exhibited arrhythmias. Two or more LVA/Ds were present in 6 of these patients. The LVA/Ds were localized at the posterobasal, apical, anteroseptal and anterolateral walls in 12, 11, 4 and 5 patients, respectively. The most common complaints at presentation were syncope or presyncope in 18 patients and palpitations in 11 patients. One patient presented with survived sudden cardiac death. Long-term ECG recordings showed ventricular tachycardia (VT) or ventricular fibrillation (VF) in 17 patients (53%). 12 patients underwent electrophysiologic testing. 9 patients had an inducible ventricular tachyarrhythmia, while the induced tachycardia was similar to that during the spontaneous arrhythmia in 7 patients. In conclusion, patients with congenital LVA/Ds who present with arrhythmic manifestations commonly have VT. Electrophysiologic testing can reproduce the clinical VT in most of these patients.

Key words: Diverticula, aneurysm, congenital, ventricular tachycardia, electrophysiologic testing

Cardiac arrhythmias are among the clinical manifestations of patients with congenital LVA/D. Cardiac embolism, wall rupture, infective endocarditis and congestive heart failure also have been described. [1] [2] [3] [4] [5] [6] [7] [8] [9] [10] [11] [12] In this study, we investigated in the setting of a single academic tertiary-care centre the clinical characteristics of patients with congenital LVA/D who had arrhythmic manifestations at initial presentation.

Methods

All consecutive patients undergoing transthoracic echocardiography between January 1, 1990 and December 31, 2010 at the University Hospital of Zurich were eligible for this study.

The diagnosis of a congenital LVA/D was made after exclusion of coronary artery disease, local cardiac inflammatory process or traumatic causes, as well as cardiomyopathies. Patients with diverticula or aneurysms in chambers other than the left ventricle were excluded from our analysis.

Among patients having a congenital LVA/D diagnosed by echocardiography the clinical data of the patients with arrhythmic manifestations were analyzed. For the purpose of this study, a patient was considered to have an arrhythmic manifestation, if there were ECG documentations of atrial or ventricular ectopic beats or tachyarrhythmias, or if there were symptoms suggesting an underlying arrhythmia, such as palpitations or presyncope/syncope.

The clinical data were obtained by retrospective review of patient records. Apart from the demographic and clinical data, the findings of transthoracic echocardiograms, 12-lead surface and 24-hour Holter recordings and invasive electrophysiologic studies were analyzed.

The protocol of the standardized ventricular stimulation consisted in the delivery of up to three extrastimuli (with a minimum cycle length of 180ms) at three different basic drive

train cycle lengths (600, 500 and 400ms) and ventricular burst pacing at two right ventricular sites (right ventricular apex and right ventricular outflow tract). Sustained ventricular tachycardia was defined as tachycardia of ventricular origin with a duration of more than 30 seconds or ventricular tachycardia leading to hemodynamic compromise.

Results

A total of 250 patients were diagnosed to have a congenital LVA/D among 93,851 patients (0.26%) who underwent an echocardiographic examination at our institution. Among these patients, 32 (13%) had arrhythmic manifestations at initial presentation and were studied as the study cohort (table 1). 21 of the patients were males (66%). The average age of the study cohort was 45 years (range 25 - 65 years). Two patients had a family history of sudden cardiac death in the absence of previously known heart disease.

The most common complaints at initial presentation were syncopal spells in 18 (56%) and palpitations in 11 (34%) patients. Other symptoms were fatigue, chest pain, dyspnea and tachycardia. One patient presented with survived sudden cardiac death.

The localization of the congenital LVA/D was posterobasal in 12 patients and apical in 11 patients (figure 1). Anteroseptal or anterolateral wall involvement was observed in 9 patients (figure 2). 6 patients had more than one LVA/D.

The LV ejection fraction (EF) of the patient cohort was 57±9% while five patients had a mildly impaired systolic LV function (EF 40-50%). The mean enddiastolic and endsystolic diameters of the LV were 50.7±10.6mm and 36.0±7.3mm, respectively, while four patients had mild dilatation of the LV chamber. A false tendon was present in the LV in four patients. The interventricular septal wall and the LV posterior wall were 10.3±2.1 mm and 9.3±1.8 mm, respectively. Five patients had mitral valve prolapse with mild to moderate mitral

insufficiency. One patient had a bicuspid aortic valve. The right ventricular measurements were normal in all patients.

Seven patients (22%) had a ventricular tachycardia (VT) at initial presentation. One patient had survived sudden cardiac death with documented ventricular fibrillation (VF). One patient had sustained common atrial flutter. Six patients had frequent ventricular ectopic activity. The mean heart rate of the study cohort was 63±12 beats per minute during sinus rhythm. The PQ interval was 182±59ms with four patients having a PQ interval longer than 200ms. The QRS duration was 98±19ms while five patients had QRS duration of more than 120ms. The corrected QT interval was 422±38ms. 29 patients (92%) had a normal axis of the QRS complex and three patients had left axis deviation. One patient had a complete right-bundle branch block and 9 patients had an intraventricular conduction delay. Two patients had signs of left ventricular hypertrophy (LVH). Five patients had repolarization abnormalities (ST segment / T wave changes).

During clinical investigation with multiple 12-lead resting and 24-hour Holter recordings, spontaneously occurring VTs could be documented in 17 patients (53%). 13 of these VTs were sustained. 9 patients had a monomorphic VT. One of these patients and four others had a polymorphic VT documented. Among 10 patients who had 12-lead ECGs during the VT, 8 of them (80%) had a right-bundle branch and the remaining two had left-bundle branch morphology during the tachycardia. One patient presented with VF and could be resuscitated successfully. Two patients had supraventricular tachycardia and one patient had typical atrial flutter during investigations.

In the study cohort, 12 patients underwent electrophysiologic study. Prior to electrophysiologic testing, 9 of these patients had a VT (monomorphic VT in five and polymorphic VT in four) documented during investigations. The remaining three patients

were tested for further evaluation of syncope or palpitations. During electrophysiologic testing, 9 patients had inducible VT/VF. Among them, 7 patients had an inducible VT with the tachycardia morphology similar to that during the spontaneous arrhythmia (figure 3). Six of these patients had VTs with a surface morphology corresponding to the anatomical location of the LVA/D. Endocardial mapping was performed in three patients showing arrhythmia exit sites corresponding to the LVA/D location. One patient who presented with palpitations had an inducible atrioventricular nodal reentrant tachycardia.

Discussion

Congenital LVA/Ds are rare findings in cardiac patients that are detectable by various imaging techniques, most commonly during an echocardiography. Most of such patients report no symptoms related to this condition. However, some patients may present with arrhythmic manifestations. In this study, we report for the first time, a single tertiary-care centre experience with a large cohort of patients. Overall, only 13% of the patients with LVA/Ds presented with arrhythmic manifestations associated with this condition.

The 32 patients with an arrhythmic manifestation at initial presentation had clinical characteristics that were not different than the rest of the total cohort of 250 patients with a congenital LVA/D. In these patients, syncopal spells and palpitations were the most common symptoms. More than 90% of the documented arrhythmias were ventricular in nature. Specifically, sustained VT was the most common documented arrhythmia. The vast majority of these patients had spontaneous or induced VT morphologies corresponding to the anatomical location of the LVA/D. The posterobasal and the apical walls of the left ventricle were the most common sites of LVA/Ds. Reproducible inducibility of the monomorphic VTs using programmed stimulation suggests reentry as the underlying mechanism of these arrhythmias.

So far, only a few cases of congenital LVA/D with documented VT have been reported in the literature. Maloy et al. described a 26-year-old woman with an apical LVA and a recurrent VT. [7] Fellows et al. published two patients with survived sudden cardiac death and one patient with non-sustained VT who presented with syncope. [13] Shen et. al. have recently described a 24-year-old male with sustained VT who had a posterobasal LVD. [8] Electrophysiologic testing could also reproduce the clinical VT in this patient. Santamaria et al. documented recurrent monomorphic VT with right-bundle branch block morphology and superior axis in a 38-year-old woman who had a large posteroseptal LVD. [10] Finally, Sierra et al. published a 30-year-old woman with a congenital basal LVD and sustained monomorphic VT. [11]

Abnormal electrophysiological findings, i.e. 1st degree atrioventricular block, bundle-branch block and intraventricular conduction delays, repolarization abnormalities among others, could be documented in the majority of our patients with congenital LVA/Ds. This finding is in accordance with those of Ohlow et al. who reported abnormal ECGs in 71 of their 125 (57%) patients with this disorder. [12] In our cohort, no surface ECG parameters during sinus rhythm could be identified that were specific for congenital LVA/Ds. Nevertheless, ECG abnormalities raise a clinical suspicion for the presence of an underlying cardiac abnormality in patients presenting with arrhythmias, a finding that should necessitate further cardiac imaging during the diagnostic workup.

Our study reports the largest patient series with spontaneously occurring and inducible sustained VT in patients with congenital LVA/D. Given the high incidence of reproducible clinical VT during electrophysiologic testing, this invasive tool should be considered in the diagnostic evaluation of patients with congenital LVA/D who present with arrhythmias.

Limitations: Even though the study describes the largest cohort of patients with congenital LVA/D and ventricular arrhythmias to date, the number of patients remains small.

Moreover, the true prevalence of arrhythmias in our patient population with congenital LVA/Ds has likely been underestimated as event recording could not be performed in each of the patients. Furthermore, the study has inherent limitations due to the retrospective nature of its analysis, and therefore, the findings must be interpreted with caution.

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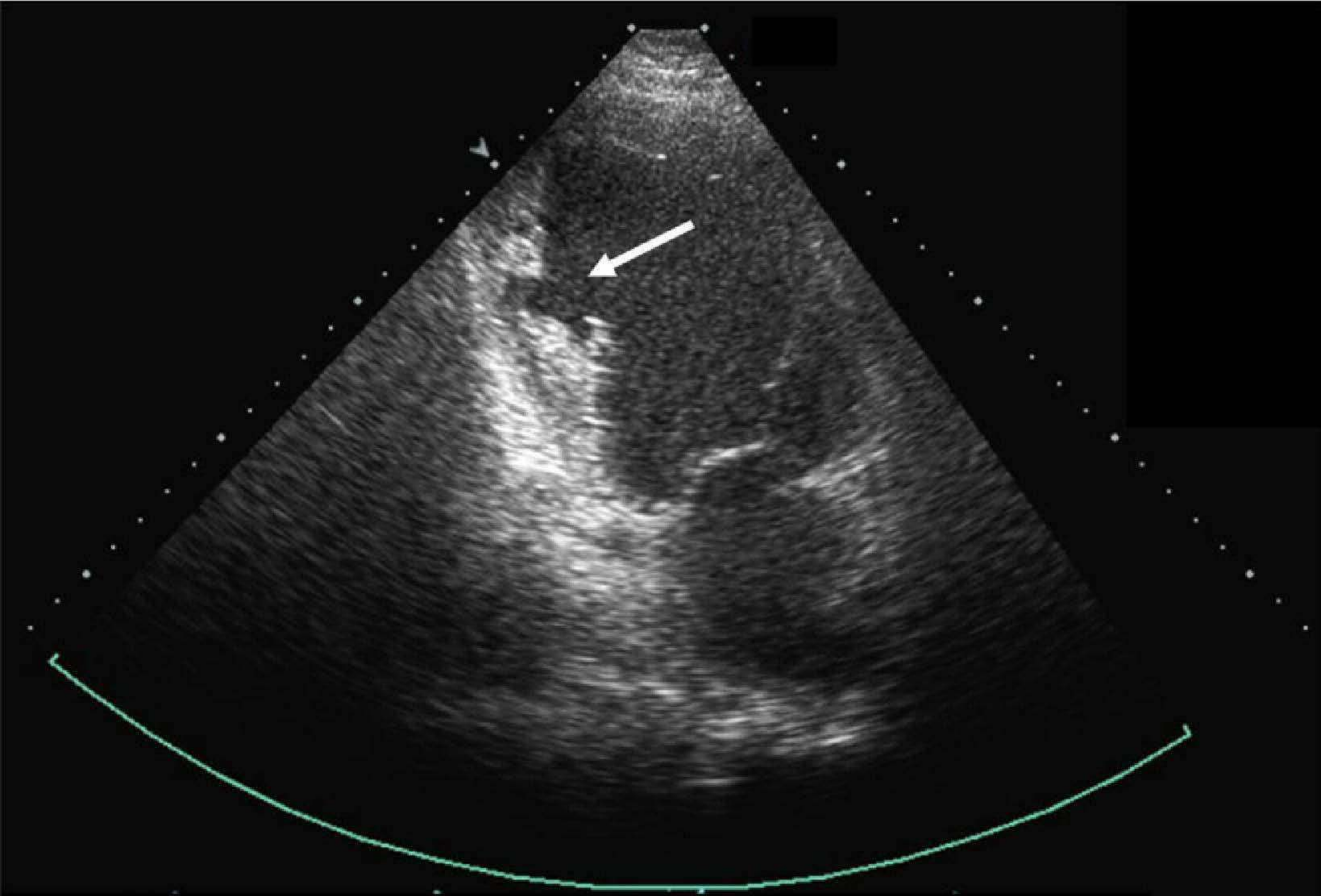
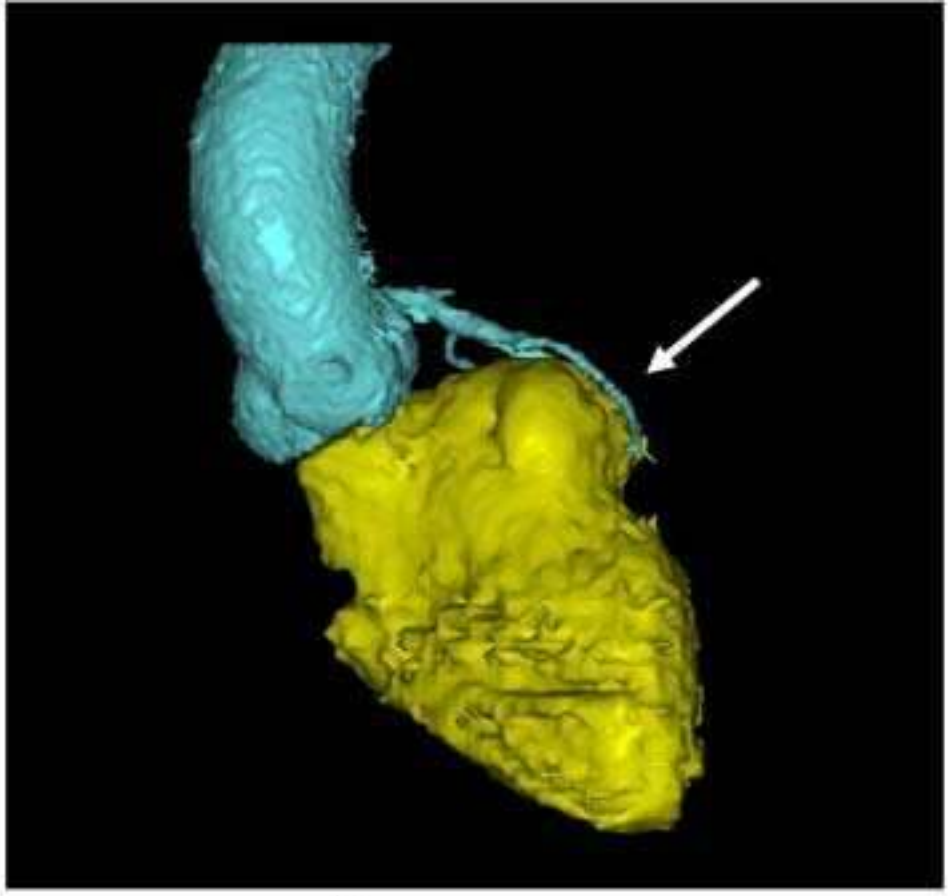
Figure legends

Figure 1: Transthoracic echocardiography of a 62-year-old male (patient no. 26) who had syncope and polymorphic VT at presentation showing a posterobasal congenital LVD (arrow) on an apical two-chamber view.

Figure 2: *Panel A:* Three-dimensional computerized tomography imaging of a 28-year-old male (patient no. 31) who had repetitive monomorphic VTs at initial presentation showing a basal anterolateral congenital LVA (arrow). *Panel B:* 12-lead surface ECG of the monomorphic VT of the same patient at a rate of 180 per minute (right-bundle branch pattern, inferior axis and negative deflections in I and aVL) which correlated to the location of the congenital LVA. Furthermore, endocardial mapping identified the exit site of the VT at the border of the LVA.

Figure 3: 12-lead surface ECG of a 35-year-old male (patient no. 14) who had palpitations at initial presentation showing a spontaneous sustained monomorphic VT at a rate of 192 per minute (right-bundle branch pattern, inferior axis). The patient had a posterobasal congenital LVD on transthoracic echocardiography corresponding to the origin of the VT. The same tachycardia could be reproducibly induced during electrophysiologic testing.





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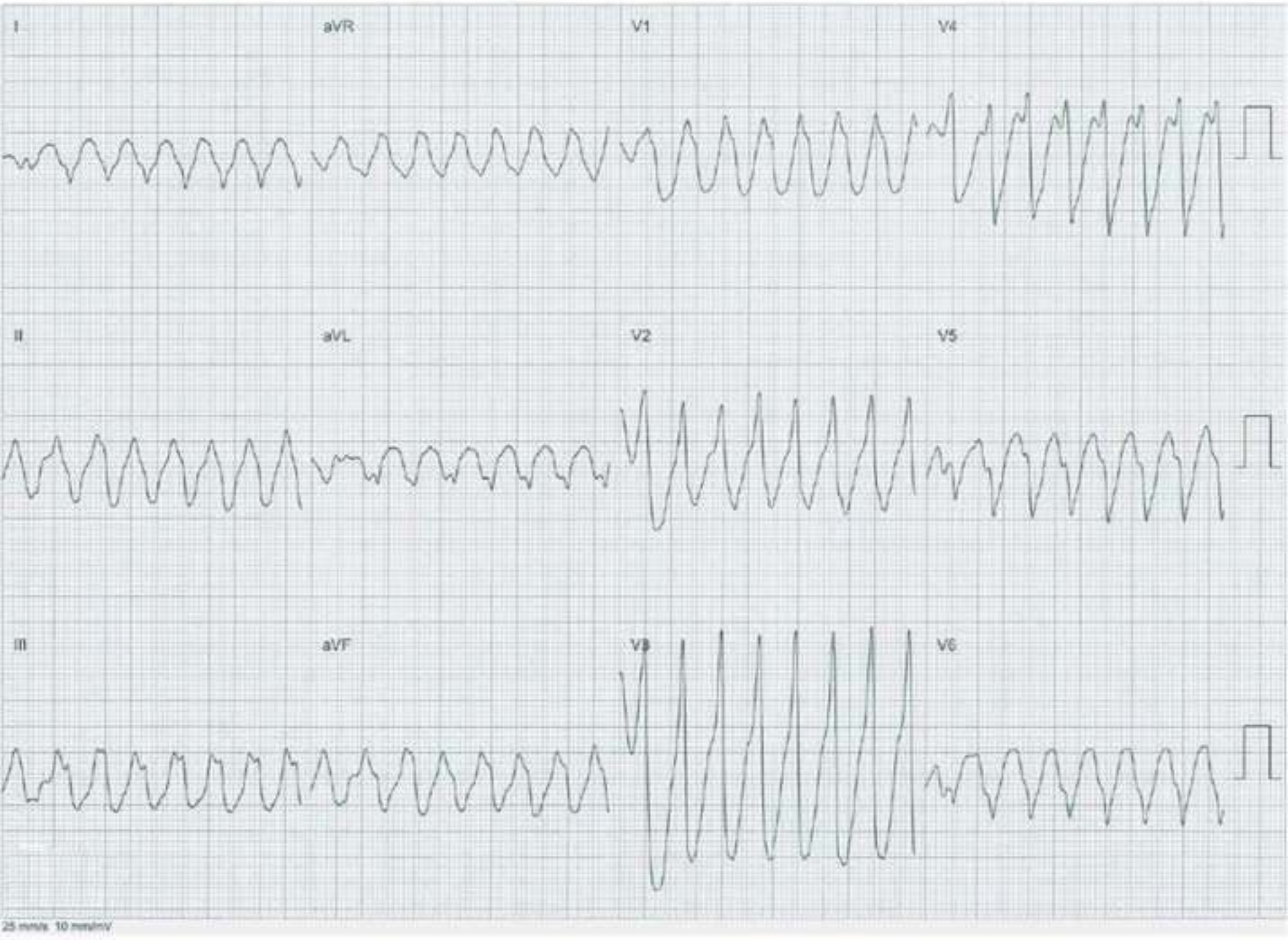

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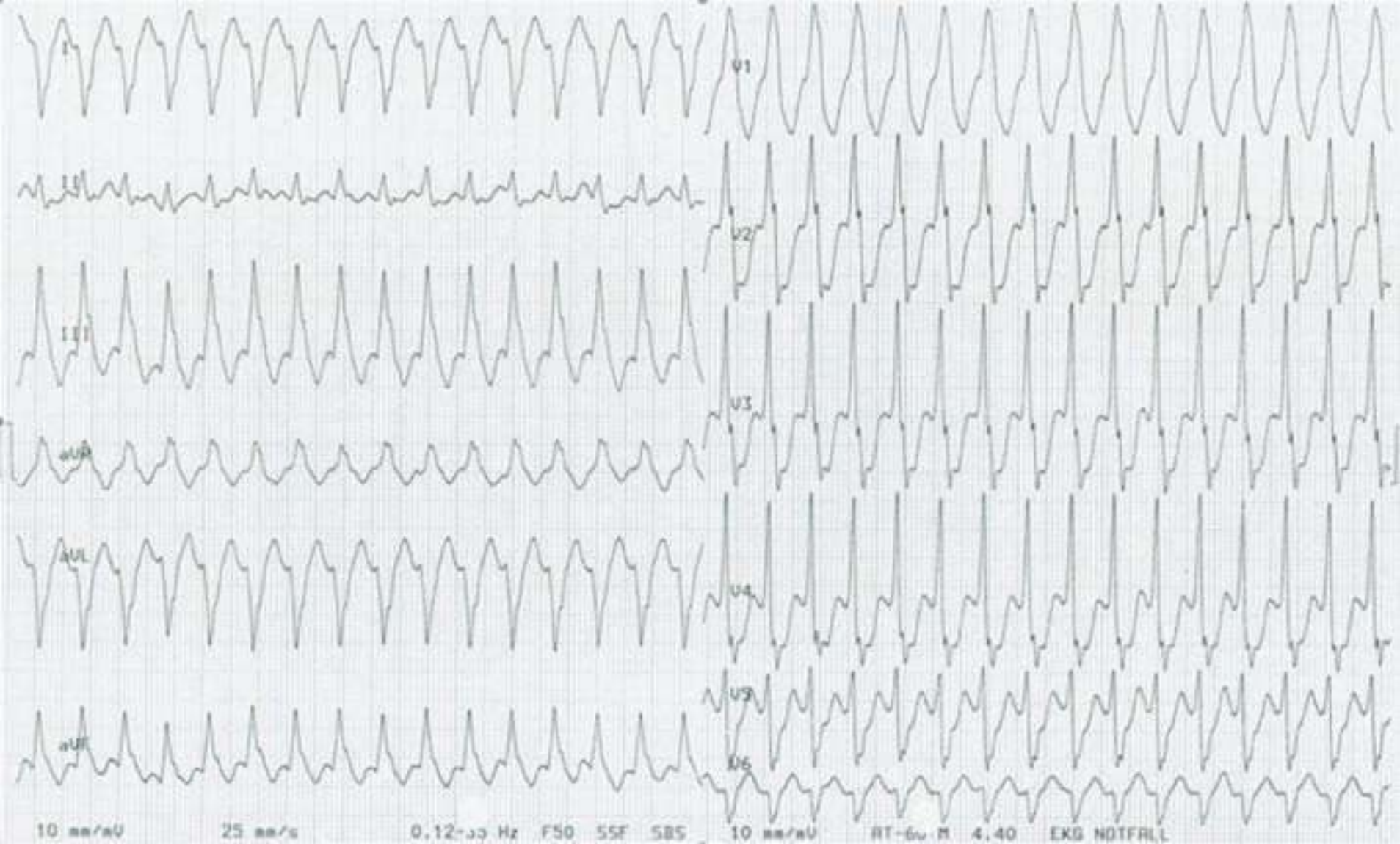
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