

## CASE REPORT

doi: 10.5455/medarch.2020.74.478-481

MED ARCH. 2020 DEC; 74(6): 478-481

RECEIVED: JUL 21, 2020 | ACCEPTED: SEP 30, 2020

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# Giant Apical Left Ventricular Diverticulum in a Health Teenager - Report of a Case within the Context of a Literature Review

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## ABSTRACT

**Introduction:** Congenital left ventricular diverticulum appears to be a developmental anomaly, an idiopathic dysplasia of left ventricular endocardium and myocardium. No evidence of a viral aetiology was found. **Aim:** We have reviewed the relevant medical literature, outlined the natural history of these left ventricular abnormalities, and discussed options in regard to their management. **Results:** The prognosis of LV outpouchings can vary from benign to catastrophic, depending upon the underlying cause. Accurate diagnosis is required to guide management decisions. High-quality imaging will characterize LV outpouchings well, helping clinicians to better understand the natural history of these conditions and to manage them appropriately. **Conclusion:** We believe that diverticulum can be detected on ECHO when it is performed precisely and carefully. In advanced centers selective computed tomography and LV angiography can be used in some cases to clearly demonstrate the outlet, size, and location of the diverticulum without the need for cardiac tomography or an MRI.

**Keywords:** congenital heart diverticulum, heart aneurysm, transthoracic echocardiography.

## 1. INTRODUCTION

Congenital apical left ventricular diverticulum (LVD), is a rare clinical entity that differs from congenital left ventricular aneurysm (LVA) and occurs with other thoracoabdominal anomalies or as an isolated anomaly. Its clinical presentation is very variable and there is no standard treatment due to its low prevalence (1). Imaging modalities have an important role in the diagnosis of the disease, due to its asymptomatic nature. Various imaging techniques are useful in diagnosis and enable the condition to be differentiated from congenital left aneurysm (2). In this article, we report a case of a 13-year old teenager, absolutely health and without any clinical signs or symptoms, who was referred to our institution as a reason of accidental systolic murmur registered due to school systematic examination. A congenital apical LV diverticulum without any clinical signs and symptoms was diagnosed.

## 2. AIM

We have reviewed the relevant medical literature, outlined the natural history of these left ventricular abnormalities, and discussed options in regard to their management.

## 3. CASE REPORT

A totally health 13-year old boy, without a family history of congenital heart disease was referred to our institution for cardiac evaluation due to a systolic non-common murmur detected from a primary care physician during the systematic school examination. He denied before any exertion intolerance, chest pain, palpitations, dyspnea, syncope or prae syncope. His review of systems and medical history were negative. He had no history of myocardial infarction (MI), arrhythmias, or stroke. His family history was positive for hypertension in his maternal grandparents. However, he and his family denied any cardiac disease, including congenital cardiac disease and sudden death in young adults.

Cardiac examination revealed a regular rhythm with normal S1 and S2. He had a grade 2 out of 6 continuous murmur noted at the apex. There were

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Figure 1. X-ray chest showed normal heart silhouette with slightly prominent bulge at the apex

wall of the chamber. Wall-motion analysis showed that the muscle of the chamber thickened during systole. No thrombus was seen. In addition, the aortic valve was bicuspid without any hemodynamic disturbances. The right ventricle looks smaller with normal contractility (Figure 4, 5). There are no any other morphologic or hemodynamic disturbances and, all time during examination normal rhythm was noted. Holter monitoring demonstrated scant supraventricular extrasystoles with repeat phenomena, and rare ventricular extrasystoles in pairs.

#### 4. DISCUSSION

Left ventricular diverticulum is defined as an out-pouching structure that contains endocardium, myocardium, and pericardium. Diverticula displays normal contraction and it's distinguished by aneurysms, which have fibrous walls and exhibit paradoxical motion (3). LVD are considered to be congenital if there is no history of conditions that have injured the myocardium. In utero viral infection, muscle and connective-tissue defects, and excessive primordial-cell stimulation have been proposed as causes (4). Most LV diverticula are

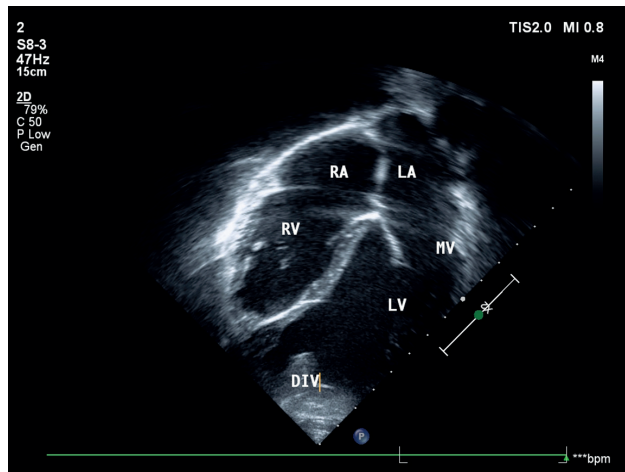
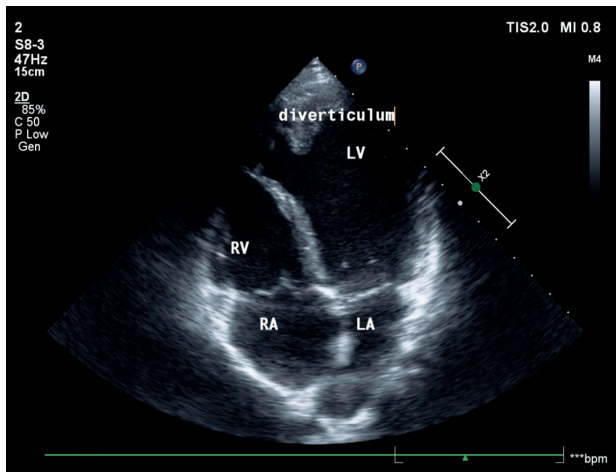


Figure 2 and 3. Giant diverticulum in the inferior-apical/apical-lateral walls of the left ventricle. Two-D four chamber view in systole - arrow (Figure 2) and in diastole (Figure 3) presented active participation of a giant diverticulum during the heart contraction in the apical-lateral walls of the left ventricle. Abbreviation: LA- left atrium, RA - right atrium, LV - left ventricle, RA - right ventricle, MV - mitral valve, DIV - diverticulum

no S3, S4 or click. He had 2 plus pulses bilaterally in both upper and lower extremities. An electrocardiogram showed normal sinus rhythm with poor precordial R-wave and with no conduction or repolarization abnormalities.

Chest radiography revealed normal heart silhouette with a prominent localized bulge at the apex of left ventricle (Figure 1). An echocardiogram showed normal cardiac connections, normal left ventricular size and function, dimensions, and wall thickness. The window to evaluate left ventricular apex was poor and careful examination confirmed a diverticulum diameter 4.5 x 4.3 cm, in the inferoapical/apical-lateral walls of the left ventricle (LV) (Figure 2, 3). By color-Doppler systolic inflow and diastolic outflow was noted, being continuously with other part of the left ventricle. Myocardium with prominent trabeculation was seen along the lateral

found in the apex. An outpouching can occur in a weaker area of the ventricular wall in the first 2 or 3 weeks of embryonic life (5). When LVD is associated with midline thoracoabdominal congenital abnormalities, diaphragmatic and sternal defects, and partial absence of the inferoapical pericardium, the condition is called Cantrell syndrome (6). In our case we think that etiology of LVD is congenital due to negative history of viral infection during the pregnancy, negative history of myocardial infarction, penetrating injuries or family history of connective-tissue disorders.

The terms "congenital aneurysm" and "congenital diverticulum" of the heart have been used interchangeably, but some authors define them differently and make a distinction. Patients with congenital LVD are usually asymptomatic, and their differential diagnoses include

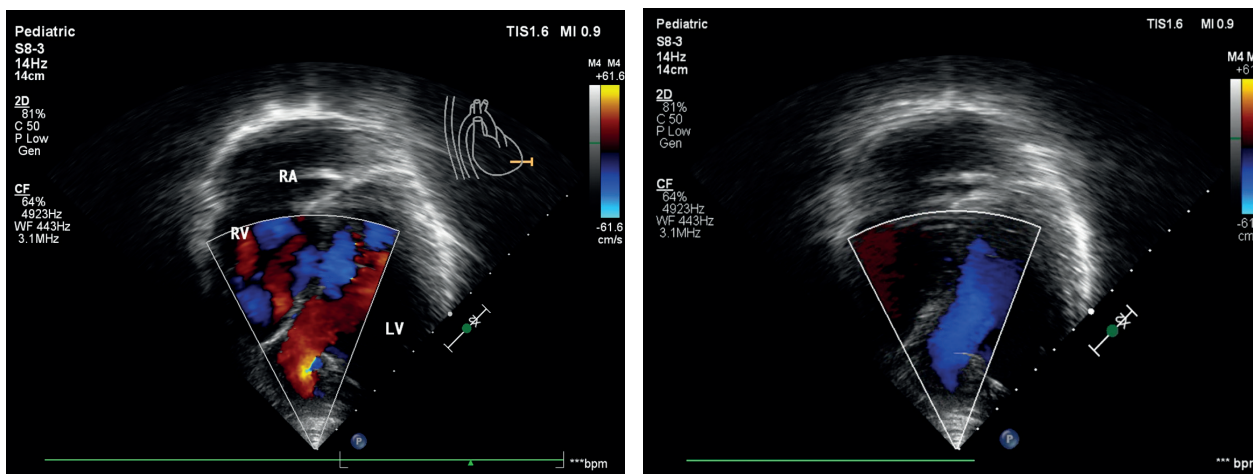


Figure 4 and 5. Colour Doppler imaging of left ventricular diverticulum showed filling of the diverticle in systole – red color (Figure 4) and discharge in diastole - blue color (Figure 5). Abbreviation: RA – right atrium, LV – left ventricle, RA – right ventricle

the left ventricular (LV) aneurysm (1, 7). The former has a large LV neck that connects to a fibrous wall along with a paradoxical motion. In contrast, most cases involving the latter have a narrow neck with a muscular wall and a synchronous contraction with the LV. This makes it difficult to clinically diagnose the disease (2, 8).

Left ventricular diverticula are most often detected incidentally during imaging procedures performed for other indications. Clinically, mostly are asymptomatic. Some of them may cause systemic embolization, heart failure, valvular regurgitation, ventricular wall rupture, ventricular tachycardia, or sudden cardiac death (8). Although, congenital LVD are usually asymptomatic, on rare occasions, they can cause arrhythmia, chest pain, cardiac rupture, and sudden death. Diagnosis is established by imaging studies such as two-dimensional or three-dimensional echocardiography, angiography, and magnetic resonance imaging. It can be used to diagnose diverticula, visualizing the structural changes and accompanying abnormalities (9). Diagnosis can be made after exclusion of coronary artery disease, local or systemic inflammation or traumatic causes as well as cardiomyopathies (10).

It may be difficult to diagnose congenital LVD since they are asymptomatic. Patients with LVD may have a normal chest radiograph, or cardiomegaly, or protrusion may be seen at the region of the diverticulum. In our case we noted a prominent localized bulge at the apex of left ventricle. The electrocardiographic abnormalities include LV hypertrophy with or without strain pattern, and nonspecific ST-segment elevation in the precordial leads. We registered totally normal electrocardiogram with non-important 24-hours Holter disturbances. Two-dimensional and Doppler echocardiography are sensitive methods for detecting a diverticulum in the interventricular septum, LV wall, subvalvular area, or congenital LV diverticulum (11-13).

The case of our patient shows that the diagnosis of congenital LV diverticulum is facilitated only by transthoracic echocardiography. We noted a diverticulum diameter 4.5 x 4.3 cm, in the inferoapical/apical-lateral walls of the left ventricle. The patient's normal coronary

arteries, the exclusion of previous acute myocardial infarction, the excluding of hypertrophic cardiomyopathy upon transthoracic echocardiography, age of patient, clinical course and our exclusion of other possible causes in the clinical presentation enabled us to deduce that the LVD in our case was probably of congenital origin. This was further supported by normal LV contractility, good LV function, the presence of chordae tendineae extending from the LV wall and from the anterolateral and posteromedial papillary muscles.

The severity of the clinical manifestations of congenital LVD varies widely from one patient to the other, and includes supraventricular and ventricular arrhythmias, heart failure, peripheral embolism, endocarditis, cardiac rupture, tamponade, or even sudden death (14,15).

Mode of treatment has to be individually tailored and depends on clinical presentation, accompanying abnormalities and possible complications. Options include surgical resection, especially in symptomatic patients, anticoagulation after systemic embolization. Treatment via anti-arrhythmic medical therapy, surgical resection, or radiofrequency ablation depends from many factors. In some cases, treatment should begin as soon as possible after diagnosis because of the risk for arrhythmia, thrombosis, ventricular fibrillation, and sudden death. Transthoracic echocardiography is often preferred as method of choice because it is non-invasive. In our case, echocardiography showed position, inflow and contractility of the diverticulum but also we can present more clearly its localization, size, and area around the congenital LVD. If we had not examined the echocardiography results so carefully, we might have suggested further clinical monitoring of the patient and attributed the existing murmur to pulmonary or aortic obstruction. Thus, the patient might have died before the surgical intervention (14, 15). All non-invasive and minimally invasive techniques are useful in the diagnosis of LV outpouchings. In our case, transthoracic echocardiography and computed tomography best defined the anatomy of the LV and showed the myocardial contraction in the wall of the outpouching (15, 16).

In case with severe forms of arrhythmias or symptomatic ventricular tachycardias radiofrequency ablation or implantation of a cardioverter defibrillator, occasionally combined with class I or III antiarrhythmic drugs. Because of the usually benign course of congenital left ventricular diverticula in the adulthood, most of them can be managed conservatively (17). Because of absent clinical signs and all examinations were normal we started just with Aspirin 100mg daily with no any physical restriction.

The natural history of LVD has not been systematically studied. The major complications of LVA include thrombosis, embolism, ventricular arrhythmia, congestive heart failure, refractory angina (possibly from altered hemodynamic levels), and, rarely, cardiac rupture. The true incidence of these complications is not known because of the rarity of LVD. Early investigators who performed autopsy studies concluded that the natural history of LVA and LVD was poor, with mortality rates as high as 80%. More recently, however, investigators have found 5-year mortality rates of 10% to 50%, depending upon outpouching size. Data from the Coronary Artery Surgery Study (CASS) registry showed that survival in patients with LVA was related to age, LV function, and the clinical severity of heart failure, rather than to the presence of the aneurysm itself (18, 19).

## 5. CONCLUSION

We believe that diverticulum can be detected on ECHO when it is performed precisely and carefully. In advanced centers selective computed tomography and LV angiography can be used in some cases to clearly demonstrate the outlet, size, and location of the diverticulum without the need for cardiac tomography or an MRI.

- **Abbreviation:** LA- left atrium, RA – right atrium, LV – left ventricle, RA – right ventricle, ALV – apex of left ventricle.
- **Acknowledgments:** The authors wish to thank the family involved in this study for their collaboration and Prof Dr Orhan Uzun in the editing and critical approval of this manuscript.
- **Ethical approval:** This report was approved by the Ethical Committee at University Clinical Centre of Kosovo in Prishtina, Number 1704, date 27 of November 2019, and both parents provided written informed consent.
- **Patients Consent Form:** Written informed consent was obtained from patient's parents for publication of this case report and accompanying images.
- **Author contributions:** Each author gave substantial contribution to the conception, in design of in the acquisition, analysis and interpretation of data for the article. Each author had role in drafting the article and revising it critically for important intellectual content. All the authors have accepted responsibility for the entire content of this submitted manuscript and approved submission.
- **Declaration of Interest:** The authors report no conflicts of interest.
- **Financial support and sponsorship:** The authors received no financial support for the research and/or authorship of this article.

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