

Congenital ventricular diverticulum and MI – Diagnostic challenges and implications



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The use of modern cardiac imaging techniques suggests that congenital ventricular diverticulum (CVD) may be more common than generally believed and may present asymptotically in adult life. We present a case of congenital left ventricular diverticulum diagnosed in a patient presenting with myocardial infarction (MI). The case highlights the importance of the differential diagnosis of CVD from post infarct left ventricular aneurysms (PILVA) and suggests that adult studies using modern imaging techniques are needed to define the prognosis for asymptomatic CVD in order to guide management.

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Introduction

Congenital left ventricular diverticula and aneurysms (together referred to as ventricular outpouches) are typically considered to be a rare medical condition, with Ohlow [1] citing 411 reported cases since their first description in 1816 [2]. Presentation in adults is particularly uncommon, and a prevalence of 0.04% in adult

patients undergoing echocardiography are reported [3]. However, improvements in cardiac imaging suggest ventricular outpouches (CVO) may be more common than previously described with a prevalence as high as 2.2% using cardiac multidetector computed tomography (MDCT) angiography [4]. This recognition of a significant number of 'incidental' diagnoses in asymptomatic individuals suggests that the prognosis for congenital outpouches may be better than commonly

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believed. This may be particularly relevant when CVO need to be considered as part of the differential diagnosis of PILVA as described here. Differentiation of congenital diverticula from congenital aneurysms may be important, as discussed.

Case report

A 45-year-old male patient presented to the emergency room with acute onset of chest pain. He was a smoker and had a history of type 2 diabetes mellitus, hypertension, and dyslipidemia. He had no previous history of cardiac disease but had a positive family history of coronary artery disease (CAD). Physical examination revealed a blood pressure of 135/65 mmHg, pulse of 60 bpm and normal heart sounds with no added sounds and no audible murmurs. Following admission he developed severe central chest pain with electrocardiography (ECG) showing ST elevation in the inferior leads with posterior extension. He subsequently developed complete heart block for which a temporary pacemaker was inserted. Routine laboratory investigations were unremarkable and chest X-ray was normal.

The patient underwent coronary angiography which demonstrated widespread CAD: Left anterior descending (LAD) coronary artery (CA) 50–60% mid stenosis, first diagonal CA 70% stenosis, second diagonal CA 70% ostial stenosis, left circumflex artery (LCX) CA 60% mid bifurcating lesion, first obtuse marginal CA 70% proximal stenosis, right coronary artery (RCA) total occlusion. Angioplasty and stenting of the RCA was performed with drug eluting stent deployment followed by abciximab (Fig. 1).

Echocardiography demonstrated a mildly dilated left ventricle with severe hypokinesia of

the inferior wall and posterior septum. A narrow necked pulsatile cavity, contracting synchronously with ventricular contraction, was seen posterolaterally to the left ventricular outflow tract (Fig. 2a). Communication with the left ventricular outflow tract was demonstrated, proximal to the aortic valve. A bicuspid aortic valve, severe aortic stenosis (gradient of 79/50 mm/Hg) and aortic regurgitation were also demonstrated (Fig. 2b). Trans-esophageal echocardiogram (TEE) delineated the outpouch and aortic valve, excluding dissection of the aorta (Fig. 2c, d).

Magnetic Resonance Angiogram (MRA) confirmed the anatomy of the left ventricular outpouch showing its position as posterior to the posterior aortic wall and demonstrating delayed hyper-enhancement of the outpouch (Fig. 2e).

After dental clearance, the patient proceeded to operation. Resection and repair of the outpouch was performed using an intracavitary Dacron patch. Aortic valve replacement and coronary artery bypass grafting (CABG) to the LAD and LCX arteries were also performed (Fig. 3). Aortic cross-clamp time was 159 min with a bypass time of 233 min. The patient made an uneventful post-operative recovery with no chest pain or clinical evidence of heart failure, ambulating freely on the cardiology ward. Transthoracic echocardiography demonstrated a well-functioning size 21 mechanical aortic prosthesis, with an acceptable gradient of 40/20 mmHg due to a small aortic root.

A residual cavity was demonstrated posterior to the aorta with no flow demonstrated into it and no pulsatility. No significant pericardial effusion nor thrombus were seen (Fig. 4). Histology of the resected outpouch demonstrated that the wall of this cavity contained the endocardium, myocardium and epicardium, confirming it to be a ventricular diverticulum.

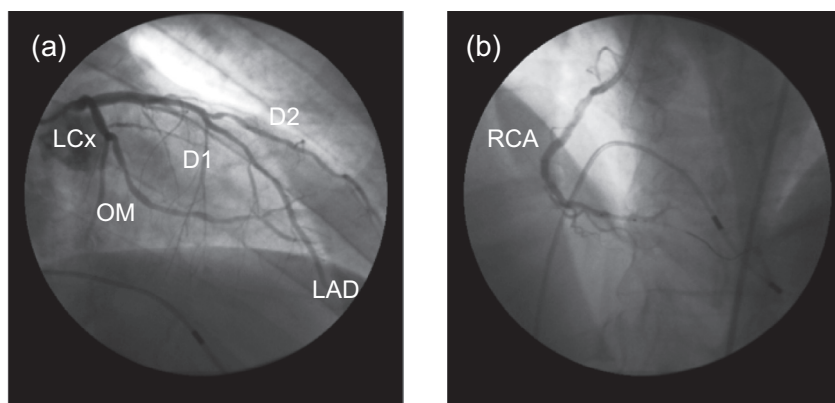


Figure 1. Coronary angiography and PCI to RCA. Left circumflex coronary artery (LCx), obtuse marginal coronary artery (OM), left anterior descending coronary artery (LAD), right coronary artery (RCA).

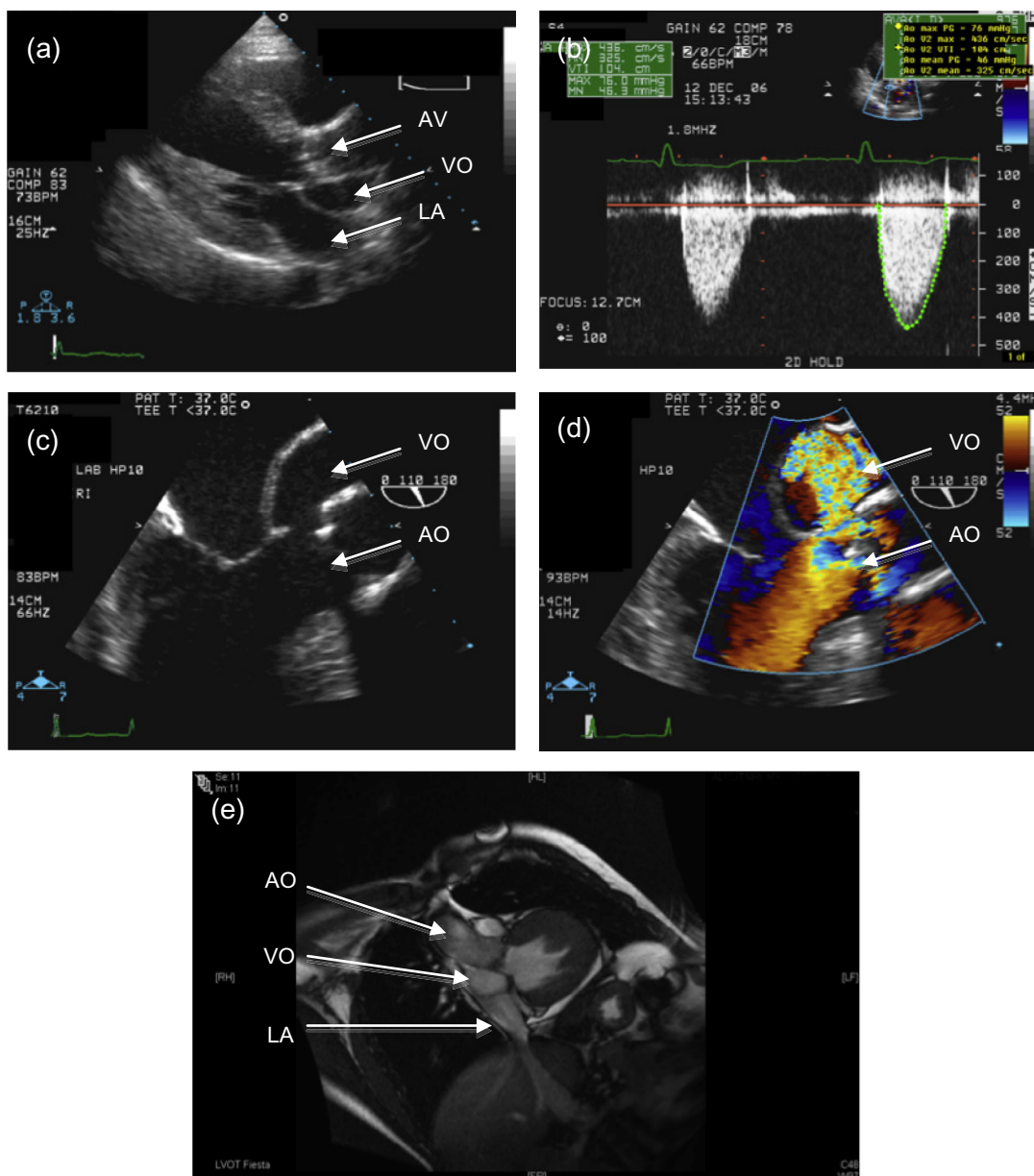


Figure 2. Preoperative Echocardiography and Magnetic Resonance Angiography. Pre-operative transthoracic echocardiography demonstrated: (a) A narrow necked pulsatile cavity postero-lateral to the left ventricular outflow tract. The aortic valve (AV), ventricular outpouch (VO), and left atrium (LA) are indicated. (b) A bicuspid aortic valve with severe aortic stenosis (gradient of 79/50 mm/Hg) and mild aortic regurgitation. Pre-operative transesophageal echocardiography demonstrated. (c) The ventricular outpouch (VO) lying posterior to the aorta (AO) and excluded aortic dissection. (d) Doppler ultrasound confirmed flow in the outpouch. Pre-operative magnetic resonance angiography. (e) Further delineated anatomy with the ventricular outpouch (VO) being visualized lying between the aorta (AO) and left atrium (LA).

Discussion

Congenital ventricular diverticula were first described in 1816 [2] and then by O'Bryan in 1837 [5], this latter case being associated with a diaphragmatic defect, as later described by Cantrell [6]. It has been suggested that given the frequent lack of clarity with regard to differential diagnosis, the term 'congenital ventricular outpouch' (CVO) is useful to describe all congenital

diverticula and aneurysms. Congenital ventricular outpouches are uncommon, being identified in less than 0.1% of congenital heart operations [7]. They are most commonly found arising from the left ventricle and are most often associated with other congenital abnormalities as described by Cantrell [6], and reviewed by Ohlow [1]. However, the occurrence of CVO in the absence of thoracic, diaphragmatic or abdominal wall abnormalities are also described [1,8,9].

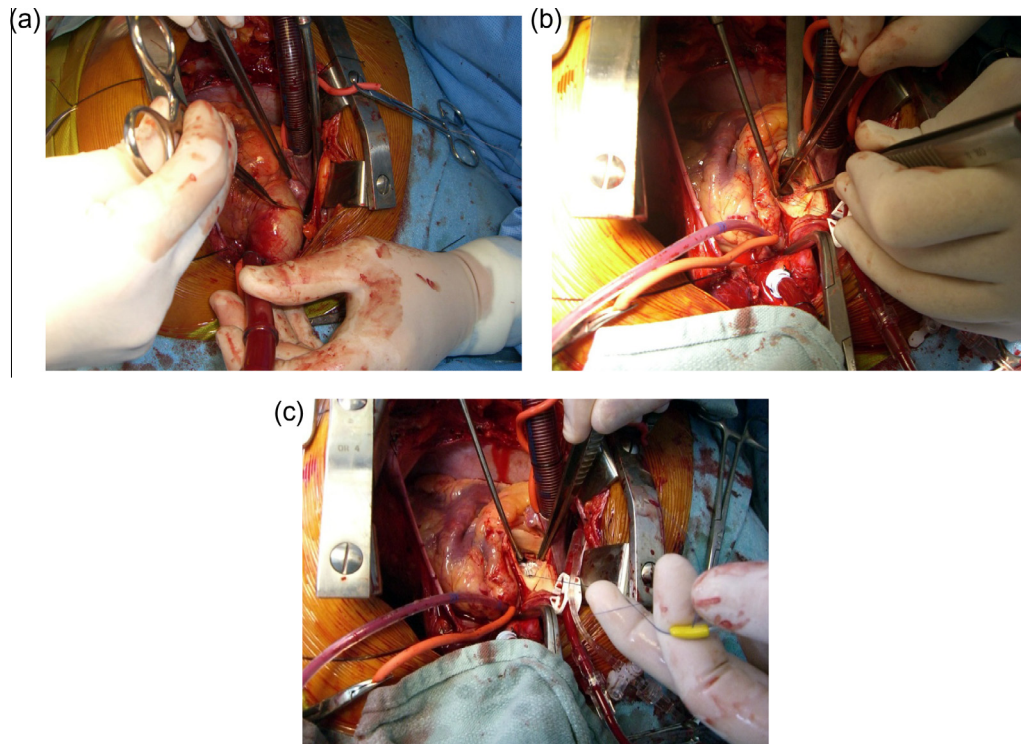


Figure 3. Operative procedure. (a) Native aortic valve (AV) before replacement. (b) Ventricular outpouch. (c) Prosthetic aortic valve (PV) in situ.

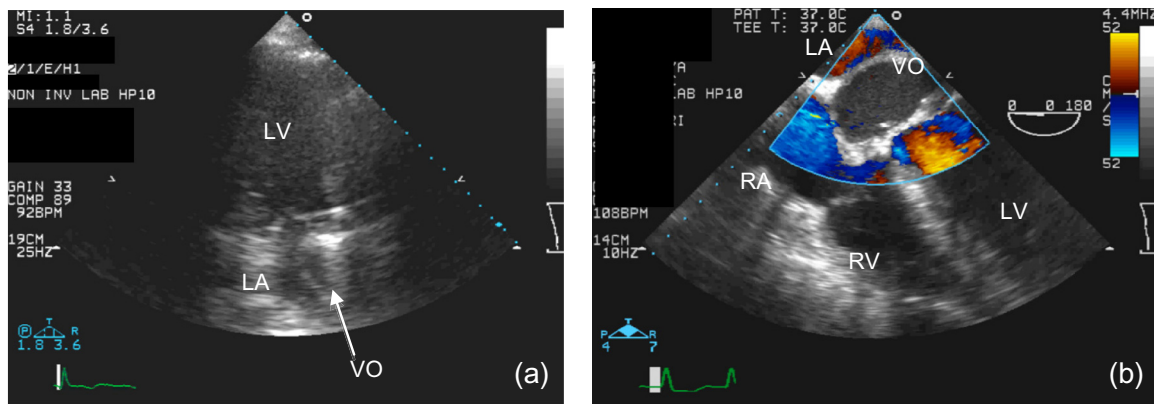


Figure 4. Postoperative Echocardiography. Echocardiographic images acquired post-surgery. (a) The position of the ventricular outpouch (VO) is demonstrated between the left atrium (LA) and left ventricle (LV). (b) Doppler imaging demonstrates complete absence of flow in the ventricular outpouch (VO). The right atrium (RA), left atrium (LA), right ventricle (RV), and left ventricle (LV) are indicated.

Congenital ventricular outpouches are most often present in newborns or children, but have been described in all age groups. Cases presenting in adult life are frequently cases of ‘isolated’ ventricular defect, as distinct from the cases described by Cantrell.

Some authors differentiate between congenital ventricular aneurysms (CVA) as opposed to congenital ventricular diverticula (CVD). The former often have broad communication with the ventricle unlike CVD, which typically communicate

through a narrow channel [10]. Classically, CVD have thick walls consisting of endocardium, myocardium and epicardium, whereas the wall of CVA consists primarily of fibrous tissue lined by endocardium with few muscle fibers [1,11]. However, these descriptions define the endpoints of a spectrum, all points in between being seen, hence the suggested term CVO is preferable in many cases. Importantly, CVD typically contract during systole, whereas CVA expand. This functional differentiation has clinical significance, as

it may determine prognosis, with progressive thinning of the wall of the outpouch being a possible indication for surgical intervention. Such thinning is suggested to be more likely in CVA than in CVD, although further studies are needed to confirm this.

Identification of CVO in adults presenting with MI has been described. While CVO are rare, left ventricular aneurysms frequently complicate MI [12]. Thus, the possibility of misdiagnosis is real. This is important, as the prognosis of the two may be significantly different, in particular if the CVO is a diverticulum as opposed to an aneurysm. Some authors have suggested criteria identifying CVO that may be managed conservatively, in contrast to post infarct left ventricular aneurysm (PILVA), which has poor prognosis and demands surgical intervention.

The differential diagnosis of CVD and true PILVA may be difficult to make, particularly in patients presenting with MI as reported here. Diagnosis of congenital left ventricular ‘aneurysm’ in a patient presenting with MI has been described [13]. The patient reported here developed complete heart block, which is consistent with a diagnosis of PILVA as a sequel to trans-mural MI [12,14]. In CVO, ECG has been described as mimicking an infarct-like pattern and is therefore of limited use for differential diagnosis [15]. However, this patient’s echocardiography and MRA showed contraction of the outpouch, synchronous with the ventricle, suggesting this outpouch to be a congenital diverticulum. Histology has demonstrated all three normal ventricle wall layers, including the myocardium, confirming this outpouch to be a congenital diverticulum. This case therefore suggests the clinical and radiological criteria for differentiation of CVO from PILVA, as proposed by Jeserich [13], to be useful.

Descriptions of the complications of CVO have led some authors to suggest that repair be universally appropriate. A mortality of up to 50% has been described for complex malformations requiring surgery in the first few days of life [16]. However, CVO that do not present until adult life may have a fundamentally different natural history and prognosis. It could be argued that in contrast to PILVA, surgical correction of CVO – particularly if they are thick walled and show synchronous contraction – is unnecessary. However, progressive thinning of CVO has been described, with development of heart failure and risk of rupture. It has been suggested that surgery is necessary for congenital right ventricular outpouches that are thin walled, but that

conservative care with monitoring is appropriate for thick walled contractile outpouches. Similar considerations may be true for all CVO. However, until definitive guidance is available, many clinicians will prefer to treat CVO surgically.

Conclusion

This case demonstrates that although no conclusive diagnostic methods have been established, TEE and MRA are very helpful in differentiating CVO from PILVA. We suggest that further studies need to be conducted to determine the specificity and sensitivity of echography, to assess TEE and MRA in the differentiation of CVO and PILVA, in particular as the prognosis for these conditions may be markedly different. With the clarification of diagnostic criteria, prognosis can be correctly determined, guiding decisions with regard to intervention for CVO presenting in adult life. Progressive thinning of the wall of the outpouch (suggested as more likely in congenital aneurysms than congenital diverticula) is most probably a criterion for elective repair of these defects [13]. It may be advisable to resect all outpouches if surgery is required for other reasons (such as CABG or valve replacement), as performed here. However, definitive data guiding clinical decision-making in such instances are lacking as the additional morbidity associated with such an approach is not known. Adoption of clear criteria for radiological diagnosis of CVO, in particular, differentiating diverticula from aneurysms in all cases will allow the definition of criteria for surgical versus conservative management of CVO in general, as suggested for right ventricular outpouches.

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