Echocardiographic Diagnosis of Double-Chambered Left Ventricle in an Infant

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INTRODUCTION

Double-chambered left ventricle (DCLV) is a very rare entity that occurs when there is division of the left ventricular cavity into two chambers by muscular tissue. The relation of the accessory chamber may be in a left-right or superior-inferior configuration to the main left ventricular cavity. Fewer than one third of cases are associated with congenital heart disease.1

Precise diagnosis of the entity poses a dilemma, with the differential diagnoses including ventricular pseudoaneurysm, true aneurysm, diverticulum, and left ventricular noncompaction. Differentiation among these pathologies remains difficult. The presence of three-layered contractile myocardium distinguishes DCLV from the aforementioned diagnoses. Diagnosis can be made by echocardiography as early as fetal life.2,3 Three-dimensional imaging modalities can further aid the distinction.3,4

We present a case of DCLV in a patient with D-transposition of the great arteries who developed progressively worsening biventricular systolic function after complete repair and was referred for cardiac transplantation. Diagnosis was further corroborated on computed tomography (CT) and examination of the explanted heart after transplantation.

CASE PRESENTATION

A 4-month-old infant with a postnatal diagnosis of D-transposition of the great arteries and multiple ventricular septal defects underwent complete surgical repair (arterial switch operation, LeCompte maneuver, and closure of ventricular septal defects) on day of life 19 at another institution. The postoperative course was complicated by significant bleeding, low cardiac output syndrome, and cardiac arrest requiring venaarterial extracorporeal membrane oxygenation, atrial tachyarrhythmias, mediastinitis, and progressively worsening biventricular systolic dysfunction. Diagnostic cardiac catheterization at 3 months of age revealed combined systolic and diastolic dysfunction with a left ventricular end-diastolic pressure of 12 mm Hg and a right ventricular end-diastolic pressure of 9 mm Hg. There were small residual ventricular septal defects, but no significant shunt (Qp/Qs = 1.1). The patient was transferred to our institution for potential heart transplantation evaluation on milrinone and epinephrine after multiple failed attempts at weaning inotropic support.

On presentation, transthoracic echocardiography demonstrated severely diminished biventricular systolic function with severe tricuspid and mitral valve regurgitation. In addition, the left ventricular cavity appeared to be divided into two chambers by a muscle bar at the level of the papillary muscles, with the smaller chamber being located inferior and posterior to the main cavity (Figure 1, Video 1). There was no obstruction at the point of communication between the two chambers (Figure 2, Videos 2 and 3). Initially, the diagnosis of a left ventricular aneurysm was considered, but subsequent echocardiograms demonstrated similar wall thickness and contractility of the accessory chamber compared with the main ventricular wall.

To better assess the accessory chamber, CT was performed. The patient was too unstable for cardiac magnetic resonance imaging, which would have been our first choice to define the myocardium. CT showed no coronary artery obstruction and uniform wall thickness in the entire left ventricle, including the second chamber, in agreement with the echocardiographic diagnosis of DCLV (Figure 3). However, this chamber appeared to be dyskinetic on CT, more consistent with a true ventricular aneurysm.

Attention was turned to retrospective review of the first postnatal echocardiogram at 4 days of age. This demonstrated the presence of this additional chamber in the subcostal sagittal (Video 4) and para-sternal short-axis views (Videos 5 and 6, Figure 4). In the setting of normal biventricular function, no dyskinesia was observed. This established the presence of this chamber at birth with normal contractility, consistent with DCLV.

The patient was subsequently listed as status 1A on the transplantation list secondary to requirement for intensive care and the need for continuous inotropic support. After various discussions, the left

![Figure 1 Subcostal sagittal view showing the muscle bar at the level of the papillary muscles in the left ventricle. The smaller chamber is located inferior and posterior to the main cavity. LV1, Left ventricular chamber 1; LV2, left ventricular chamber 2; RV, right ventricle.](https://doi.org/10.1016/j.case.2020.12.008)
Ventricular morphology in this patient was deemed to be a contraindication to mechanical circulatory support in the event of acute decompensation while on the waiting list. This added to the urgency of the need for transplantation.

Within 4 weeks a suitable donor was available, and the patient underwent orthotopic heart transplantation. On examination of the explanted heart, a muscular “pseudoseptum” was noted extending from the apex of the left ventricle to the inferior aspect of the posterior mitral valve leaflet. This divided the left ventricle into a larger anterior chamber and a smaller posterior-inferior chamber (Figure 5). The mitral valve papillary muscle was noted to be split at the level of the muscle bar (Figure 6). Gross examination of the wall of the second chamber showed presence of all three layers in the ventricular wall (Figure 7). Microscopic examination revealed endocardial fibroelastosis and prominent myocyte hypertrophy in the walls of both chambers.

Unfortunately, the patient’s postoperative course was complicated by primary graft failure, venoarterial extracorporeal membrane oxygenation, mediastinitis, atrial fibrillation, and cardiac arrest followed by brain death. Care was withdrawn at approximately 6 weeks after transplantation.

DISCUSSION

Diagnosis of DCLV is often challenging because of multiple overlapping features among DCLV, pseudoaneurysms, true aneurysms, and diverticula. A ventricular pseudoaneurysm comprises dyskinetic tissue that does not contain all three layers of the ventricular wall and is usually a result of myocardial infarction. A ventricular aneurysm contains all three layers but is dyskinetic in comparison with the main ventricular wall. A diverticulum has all three layers but is dyskinetic in comparison with the main ventricular wall. A diverticulum has all three layers but is dyskinetic in comparison with the main ventricular wall. A diverticulum has all three layers but is dyskinetic in comparison with the main ventricular wall. A diverticulum has all three layers but is dyskinetic in comparison with the main ventricular wall. A diverticulum has all three layers but is dyskinetic in comparison with the main ventricular wall.

View the video content online at www.cvcasejournal.com.
Transthoracic echocardiography is considered sufficient for the diagnosis of DCL V. The original location of the abnormal muscle bundle and communication between the main and second chamber can be delineated in subcostal and parasternal views. Synchrony of the myocardium of both chambers can also be established. CT was performed on our patient to rule out coronary artery obstruction, and the findings confirmed our diagnosis. There are reports of the use of magnetic resonance imaging to complement findings on echocardiography.1,3,4,7 We were fortunate to be able to obtain pathologic examination of the explanted heart to substantiate our diagnosis. Unlike ventricular aneurysms and pseudoaneurysms that are attributed to ischemia and infections, the pathogenesis of DCL V is largely unknown. Recently, Anderson and Gufler8 postulated that it is a result of abnormal coalescence of the trabecular component of the developing ventricular walls during papillary muscle formation. This is consistent with the idea that this entity is congenital and not acquired. Our diagnosis was aided by a prior echocardiogram obtained at birth that showed the presence of this chamber at birth with all the properties of a DCLV. Most cases of DCLV are found incidentally, and the entity by itself is believed to have little to no bearing on clinical outcome.3,4,7,9 Management is varied and generally driven by accompanying diagnoses such as systolic dysfunction and/or arrhythmias.10 In our case, however, the division of the left ventricular cavity added a layer of complexity to our patient because of the critically ill condition. With severely depressed ventricular function requiring inotropic support, our patient was at high risk for waiting-list mortality. Typically, such patients are offered mechanical circulatory support in the event of further hemodynamic compromise. Positioning an inflow cannula to adequately offload the entire left ventricle was not feasible because of the muscle bar originating from the apex in this patient. Therefore, mechanical circulatory support was not considered an option, and the risk for waiting-list mortality was substantially increased.

Early and accurate diagnosis is crucial to differentiate DCLV and similar entities because unlike most cases of DCLV and diverticula, ventricular aneurysms and pseudoaneurysms are associated with ominous causes such as myocardial infarction or myocarditis, and patients are at increased risk for ventricular arrhythmias, thrombi, and sudden cardiac death.

**CONCLUSION**

Distinguishing between DCLV and similar entities is critical because of different treatment modalities and prognosis. Echocardiography is considered sufficient for the diagnosis, but additional modalities may prove useful.

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**SUPPLEMENTARY DATA**

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2020.12.008.

**REFERENCES**

1. Yuan SM. Double-chambered left ventricle: clinical features comparison between children and adults. J Coll Physicians Surg Pak 2019;29:1087-91.
2. Xue C, Zhao Y, Zhang Y, Gu X, Han J, Henein M, et al. Double-chambered left ventricle diagnosis by 2D and 3D echocardiography: from fetus to birth. Echocardiography 2019;36:196-8.

3. Li T, Han J, Li Y, Gao S, Hao X, He Y. Diagnosis of double-chambered left ventricle by echocardiogram and cardiac magnetic resonance imaging: a rare case in an adult. Echocardiography 2020;37:368-9.

4. Masci PG, Pucci A, Fontanive P, Coceani M, Marraccini P, Lombardi M. Double-chambered left ventricle in an asymptomatic adult patient. Eur Heart J Cardiovasc Imaging 2012;13:E1-3.

5. Krasemann T, Gehrmann J, Fenge H, Debus V, Loeser H, Vogt J. Ventricular aneurysm or diverticulum? Clinical differential diagnosis. Pediatr Cardiol 2001;22:409-11.

6. Zhang XY, Cao TS, Yuan LJ. Double-chambered left ventricle in echocardiography. Echocardiography 2012;29:E67-8.

7. Mordi I, Carrick D, Tzemos N. Diagnosis of double-chambered left ventricle using advanced cardiovascular imaging. Echocardiography 2013;30:E206-8.

8. Anderson RH, Guller H. Commentary: what makes the morphologically left ventricle double chambered? J Thorac Cardiovasc Surg 2020;159:e195-6.

9. Gilbert JD, Byard RW. Double-chambered left ventricle—a “heart within a heart”. Forensic Sci Med Pathol 2018;14:545-7.

10. Sharma S, Dinwoodey DL, Chaudhry GM, Labib SB. Congenital double-chambered left ventricle presenting as monomorphic ventricular tachycardia. CASE (Phila) 2019;3:51-5.