The Heart Surgery Forum #2004-1011
7 (3), 2004 [Epub May 2004]
doi:10.1532/HSF98.20041011

Online address: www.hsforum.com/vol7/issue3/2004-1011.html

Successful Surgical Management of a Double-Chambered Left Ventricle in a 13-Year-Old Girl: A Report of a Rare Case

Omer Faruk Dogan, MD,[1] Dursun Alehan, MD,[2] Umit Duman, MD[1]

Departments of [1]Cardiovascular Surgery and [2]Pediatric Cardiology, Hacettepe University Medical Faculty, Hacettepe University, Ankara, Turkey

ABSTRACT

Background: Double-chambered left ventricle (DCLV) is a rare congenital anomaly, and only a few cases in which a 2-chambered LV is separated by the interventricular septum or an abnormal muscle bundle have been reported in the literature. Frequently, such cases are diagnosed when a patient is admitted to hospital for the evaluation of a cardiac murmur, and most of these patients have isolated DCLV.

Materials and Methods: We describe the case of a 13-year-old girl with DCLV who had twice undergone operation, including mitral valve replacement, in our institution.

Result: No gradient was found after surgical resection of the hypertrophic floating mass, and the patient's symptoms disappeared.

Discussion: Surgical resection can be carried out in patients with DCLV when done sufficiently early, and careful echocardiographic examination is important in a continuing follow-up. Surgeons should keep in mind the possibility of a recurrence of DCLV.

INTRODUCTION

Double-chambered left ventricle (DCLV) is a very rare congenital cardiac anomaly within the broad category of subdivision of the LV cavity that includes the relatively more common congenital aneurysm and diverticula. Here, we report a patient with DCLV who underwent successful surgical treatment. This case is the first report of a patient with a second diagnosis of DCLV after an operation was performed to correct the original DCLV.

CASE REPORT

A 13-year-old girl was referred to our clinic with complaints of diminished exercise tolerance, palpitation, and perioral cyanosis. The patient had originally been admitted for mitral valve replacement, which was performed in our hospital when she was 11 years old. Chordal adhesion and elongation and mitral valve destruction had been seen in the first operation. We also performed the mitral valve replacement because there had been an elongated and thinning chorda, and the hypertrophic anterior mitral papillary muscle was causing LV outflow tract (LVOT) obstruction.

In the physical examination after the patient's first operation, a 3/6-degree systolic sufl was heard on the left sternal border. She underwent evaluation with echocardiography, cardiac catheterization, and angiocardiography. Echocardiography results showed right atrial dilatation and LV hypertrophy, normal prosthetic mitral valve function, and subaortic obstruction together with interventricular septal hypertrophy and thickening. In addition, a 2-dimensional echocardiographic examination revealed 2 chambers divided by the muscular hypertrophic mass. The walls of the apical chamber and the septum were not hypokinetic. A systolic gradient of 65 mm Hg was found across the LVOT. An LV angiographic examination carried out with multiple views confirmed the presence of septum thickening, and the LVOT was obstructed because of septal hypertrophy. The patient underwent operation with cardiopulmonary bypass (CPB), and septal hypertrophic bands were resected via a transverse aortotomy. She was weaned from CPB, and a systolic gradient of 14 mm Hg was obtained in the postoperative period. She was discharged from the hospital in good health 7 days after the operation.

Six months after the operation, the patient was admitted to pediatric cardiology with symptoms of chest pain and early fatigue. Echocardiographic and angiocardiographic examinations were planned for the evaluation. First-degree prosthetic mitral valve insufficiency, second-degree tricuspid valve insufficiency, and a gradient of 13 mm Hg through the prosthetic mitral valve were found in the echocardiographic study (Figure 1). The echocardiogram showed the presence of a floating mass that divided the LV into 2 true chambers (a small basal chamber and a large apical chamber). The echocardiographic examination revealed thinning of the LV wall and the interventricular septum and a tunnel-type subaortic obstruction (Figure 2) but presented no evidence of LV dysfunction. An angiocardiographic examination revealed right ventricular and LV hypertrophy, and a gradient of 110 mm Hg was found between the apical and basal chambers (Figure 3). The patient had moderate pulmonary hypertension (mean pul-

Received February 10, 2004; accepted March 22, 2004.

Address correspondence and reprint requests to: Omer Faruk Dogan, MD, Birlik Mahallesi 59. Sokak, 9/1, 06670, Çankaya, Ankara, Turkey; 90-312-495-68-98; fax: 90-312-311-04-94 (e-mail: ofdogan@hacettepe.edu.tr).

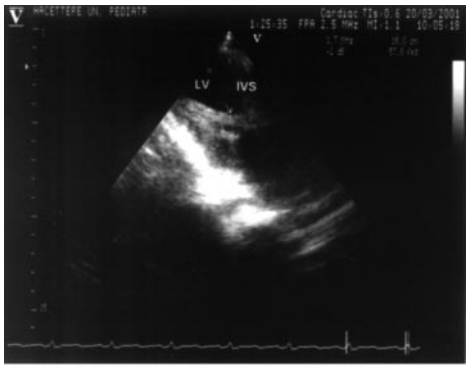

Figure 1. A subcostal short-axis section of an echocardiogram showing a hypertrophic muscle mass originating from the left ventricular (LV) free wall. IVS indicates interventricular septum.

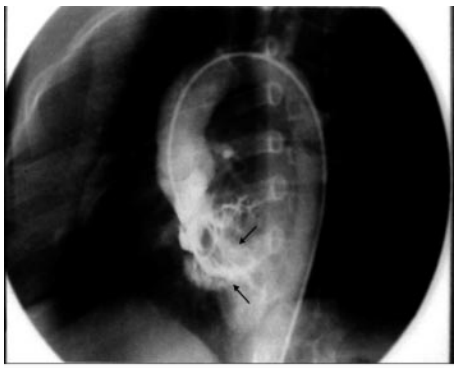

Figure 3. A left ventricular angiogram in left anterior oblique cranial view showing a floating hypertrophic muscle dividing the left ventricle into 2 true chambers. Arrows show the apical and basal chambers.

monary arterial pressure, 35 mm Hg; pulmonary wedge pressure, 25 mm Hg). The systolic and diastolic pressures of the apical LV chamber were 200 mm Hg and 12 mm Hg, respectively, and the systolic and diastolic pressures of the basal chamber were 90 mm Hg and 6 mm Hg, respectively. She underwent operation with CPB with a diagnosis of DCLV and mild aortic and moderate subaortic stenosis. In the operation, a midline sternotomy was performed, and CPB was instituted. Before CPB began, systolic pressures of 170 mm Hg from the basal chamber and 70 mm Hg from the LV apical chamber were measured while the aortic pressure was 90 mm Hg. A wide fibromuscular floating mass divided the LV into 2 chambers. The accessory chamber had a communicating flow with the basal chamber through an orifice with a diameter of 1.5 to 2 cm (Figure 4). No other congenital malformations were found intraoperatively. The fibromuscular mass was excised from the LV free wall, the interventricular septum, and the subaortic area. After the patient was weaned from CPB, the intraventricular gradient disappeared, and the difference between the systemic and the LV pressures decreased to 10 mm Hg. A histopathologic examination of the resected wall tissue showed hypertrophic myocardial fibers and fibrosis in the intermyocardial space. The patient was discharged from the hospital 8 days after the surgery. Her echocardiographic examination in the third postoperative month revealed a gradient of 20 mm Hg on the LVOT, and the patient was free of symptoms (Figure 5). One year after the operation, septal hypertrophy and a band formation were found on the LV free wall through the interventricular septum, and a systolic gradient of 35 mm Hg was observed. The patient was observed to be asymptomatic in the 2 years of follow-up.

DISCUSSION

DCLV is a very rare congenital cardiac anomaly within the broad category of subdivision of the LV cavity, which includes the relatively more common congenital aneurysm and diverticula. This pathology is more rare than double-chambered right ventricle [Kay 1981, Vaidyanathan 2001]. We found a few case reports of subdivision of the LV in the MEDLINE database [Kay 1981, Caron 1991, Rickli 1999, Vaidyanathan 2001]. Gerlis et al reported the largest series (3 cases), which consisted of patients who had endomyocardial fibroelastosis

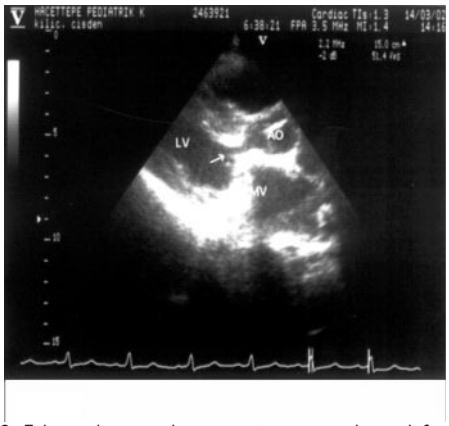

Figure 2. Echocardiogram demonstrating a tunnel-type left ventricular (LV) outflow tract obstruction and a normal prosthetic mitral valve (PMV). AO indicates aorta.

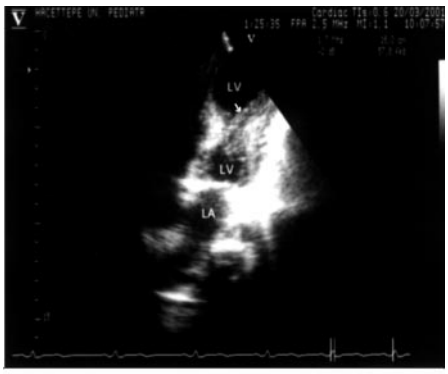

Figure 4. An echocardiographic subcostal long-axis section showing the 2 chambers of the left ventricle (LV) divided by the muscle originating from the LV free wall and the interventricular septum. Arrow shows the floating mass. LA indicates left atrium.

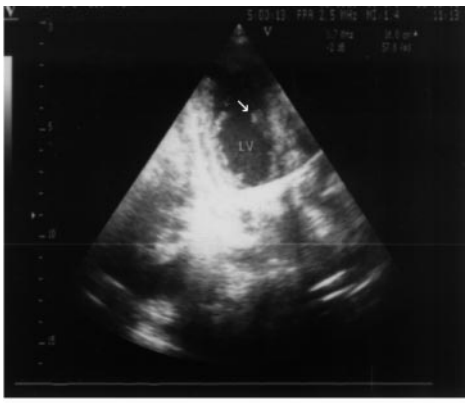

Figure 5. A subcostal echocardiogram demonstrating the left ventricle (LV) after surgical resection. Arrows show the remaining muscular tissue originating from the interventricular septum and the LV free wall.

and cardiomyopathy. All of the patients died in the first year of life from the cardiomyopathy [Gerlis 1981]. The etiologic mechanism is unknown, and there is often an associated cardiomyopathy. However, there was no evidence of LV failure or cardiomyopathy in our case. Gerlis et al suggested that the endocardial fibroelastosis rendered the main chamber noncontractile and that the functional portion of the LV was derived from intratrabecular myocardial sinusoid expansion [Gerlis 1981]. In our patient, the etiology is unlikely to be related to cardiomyopathy because she has a normal ventricular function. One of the reports described a patient with a large accessory chamber lying anterolateral to the main LV, with nonrestrictive communication occurring between the 2 chambers of the LV; however, our DCLV patient had a fibromuscular hypertrophic mass dividing the LV into 2 true chambers with a restrictive communication between the apical and the basal chambers. In addition, the apical chamber was larger than the basal chamber. Joy et al described a case of DCLV [Joy 1980] in which the patient had false tendons of fibrous or fibromuscular bands less than 3 mm in diameter crossing the LV cavity either between the LV free wall and the interventricular septum or between 2 papillary muscles [Joy 1980]. Rickli et al reported a case of DCLV with a prominent fibromuscular ridge distal to the papillary muscles that divided the LV cavity into a small apical portion and a large basal portion [Rickli 1999]. In the present DCLV case, a fibromuscular mass was seen to divide the LV into a small basal portion and a large apical portion.

In our case, the associated septum was derived from the interventricular septum and the LV free wall, and it was in a floating formation that separated the LV cavity into 2 chambers. Furthermore, the patient had previously undergone operation for LVOT obstruction and had presented no evidence of DCLV in the first operation. Regarding our patient, a search of the English-language literature revealed no case with a presentation similar to that of this patient's history following the surgery.

REFERENCES

Caron KH, Hernandez RJ, Goble M, Beekman R. 1991. MR imaging of double chambered left ventricle. J Comput Assist Tomogr 15:140-2.

Gerlis LM, Patridge JB, Fiddler GI, Williams G, Scott O. 1981. Two chambered left ventricle: three new varieties. Br Heart J 46:278-84.

Joy J, Subramanyan R, Sandhyamani A, Balakrishnan KG. 1980. Double chambered right ventricle with anomalous muscle bundles and false tendons in the left ventricle. Indian Heart J 42:58-61.

Kay PH, Rygbi M, Mulholland HC. 1981. Congenital double chambered left ventricle treated by exclusion of accessory chamber. Br Heart J 49:195-8.

Rickli H, Attenhofer Jost CH, Locher JT, Jenni RE. 1999. Double-chambered left ventricle: Unusual presentation with apical defect in a thallium scan in a middle-aged, asymptomatic woman. Echocardiography 16:387-91.

Vaidyanathan D, Prabhakar D, Selvam K, Alegasan R, Thirunavukarasu N, Muthukumar D. 2001. Isolated congenital left ventricular diverticulum in adults. Indian Heart J 53:211-3.