Case Report

Congenital Left Ventricular Diverticulum Associated with ASD, VSD, and Epigastric Hernia

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Abstract

Congenital left ventricular diverticulum is a rare cardiac malformation. Two categories of congenital ventricular diverticulum have been identified with regard to their localization: apical and non-apical. Apical diverticula are always associated with midline thoraco-abdominal defects and other heart malformations. Non-apical diverticula are always isolated defects.

Diagnosis is established by imaging studies such as echocardiography, magnetic resonance imaging, or left ventricular angiography. Mode of treatment has to be individually tailored and depends on clinical presentation, accompanying abnormalities, and possible complications.

We report a 10-month-old girl with left ventricular apical diverticulum, large atrial septal defect, two small muscular ventricular septal defects, and pulmonary hypertension, associated with epigastric hernia. This patient underwent total surgical repair for intra-cardiac defects as well as diverticular resection.

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Introduction

Congenital left ventricular apical diverticulum is a very uncommon cardiac malformation.[1-5] It is always associated with midline thoraco-abdominal defects and other heart malformations.[1,2,6] All patients with a midline thoraco-abdominal defect and a pulsatile mass should be evaluated carefully to rule out this anomaly. In symptomatic patients, medical and surgical treatment can relieve symptoms and prevent further complications.[1,7]

Case report

A 10-month-old girl referred to us for cardiac evaluation before surgical repair of epigastric hernia. The cause of referral was heart murmur and cardiomegaly, which were found in routine physical examination and chest roentgenogram, respectively.

The patient, weighing 6.5 kg, had normal facies and a normal developmental history.

A relatively large hernia was clearly apparent in the epigastric region with a pulsatile mass upon it (Figure 1).

A heavy cardiac impulse was felt at the left side of the anterior chest wall at the normal site. There was an approximately 3-cm distance between the point of the maximal cardiac impulse and the pulsatile mass. There were no other pulsations or thrills.

Cardiac auscultation revealed a widely split second heart sound with an accentuated pulmonary component. A holosystolic murmur, grade 3/6, was heard along the left sternal border, as well as an ejectional murmur, grade 2/6, at the upper left sternal border.

Electrocardiography showed a sinus rhythm with a heart rate of 120, right axis deviation, and right ventricular hypertrophy.

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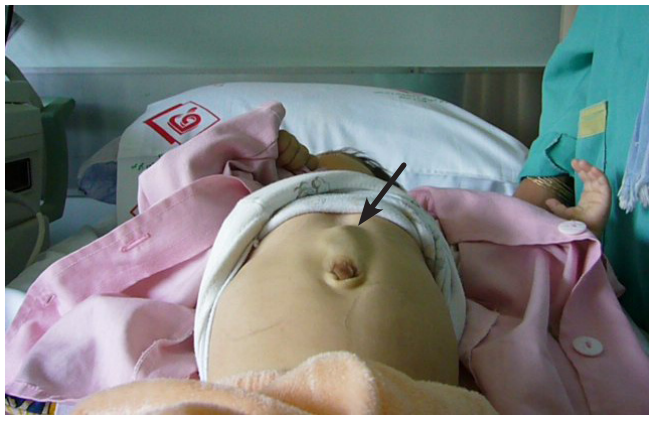
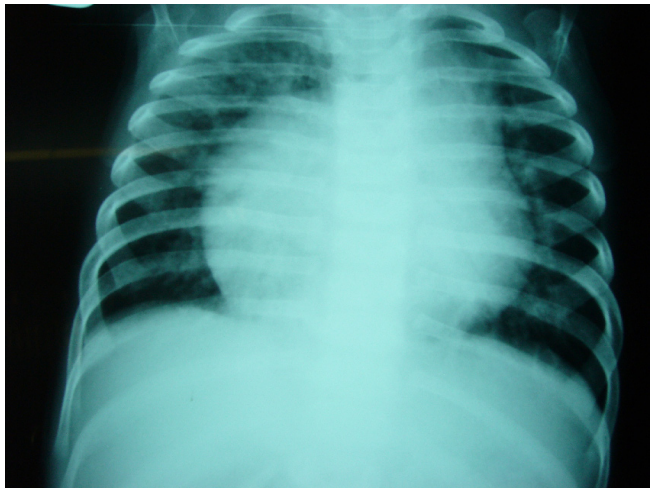


Figure 1. Simple photograph of the patient in supine position. Note the eminency on the epigastric region (arrow)

Chest roentgenogram revealed moderate cardiomegaly, prominent pulmonary artery shadow, and increased pulmonary vascular markings. There was no other structural finding in the anteroposterior and lateral films (Figure 2).



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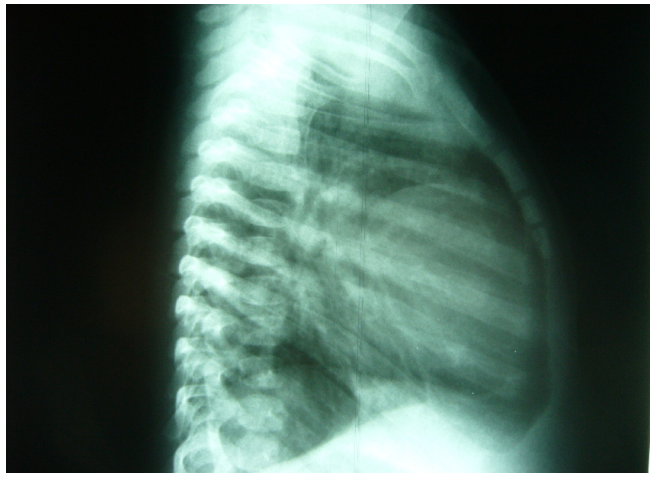

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Figure 2. Chest X-ray of the patient. A, anteroposterior view; B, lateral view. Cardiomegaly and increased pulmonary vascular markings are seen in both images, but the diverticulum is not seen directly

Two-dimensional echocardiography revealed a relatively large secundum type atrial septal defect (ASD), 2 small muscular ventricular septal defects (VSDs), and a dilated pulmonary trunk. A long outgrowth (diverticulum) was also detected in the left ventricular apex with simultaneous contractions (Figure 3). The thoracic and abdominal aorta seemed normal without any abnormal dilatation. A Doppler color examination showed left-to-right flow across the atrial and ventricular septal defects. There was no significant flow gradient through the diverticulum origin.

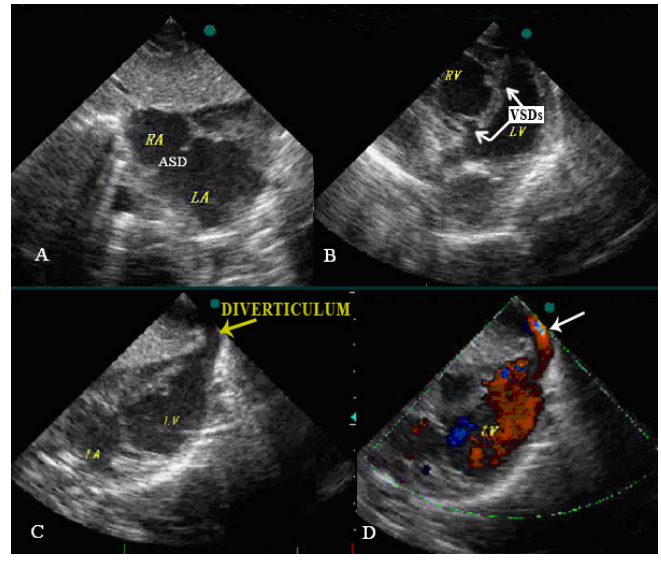


Figure 3. Two dimensional and color Doppler echocardiograms of the patient. A, large large atrial septal defect (ASD) and relatively enlarged left atrium (LA) are seen in subcostal coronal view. B, two small muscular ventricular septal defect (VSDs) (arrows) are seen in apical five- chamber view. C, an apical outgrowth (diverticulum) is seen in modified four-chamber view (arrow). D, non-turbulent antegrade-retrograde flow is seen in the diverticulum by color Doppler imaging (arrow)

Cardiac catheterization and angiography was performed to delineate the exact anatomy and measure the pulmonary artery pressure and pulmonary vascular resistance.

Pulmonary systolic pressure was as high as 60 mmHg when aortic pressure was 80 mmHg. Left-to-right shunt was estimated at approximately 2:1.

The left ventricle angiogram in the lateral projection clearly illustrated the diverticulum (Figure 4). ASD and VSDs were detected in their special views. The diverticulum had synchronal contractions with the left ventricle.

Because of the high pulmonary artery pressure and heart failure, total surgical repair (closure of ASD and VSDs and diverticulectomy) was recommended.

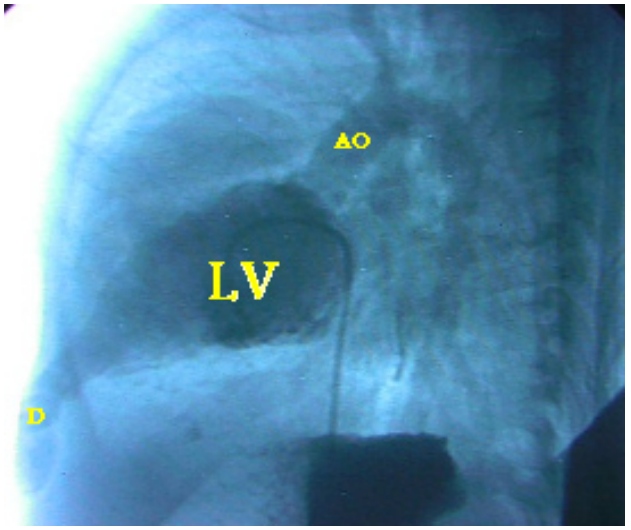
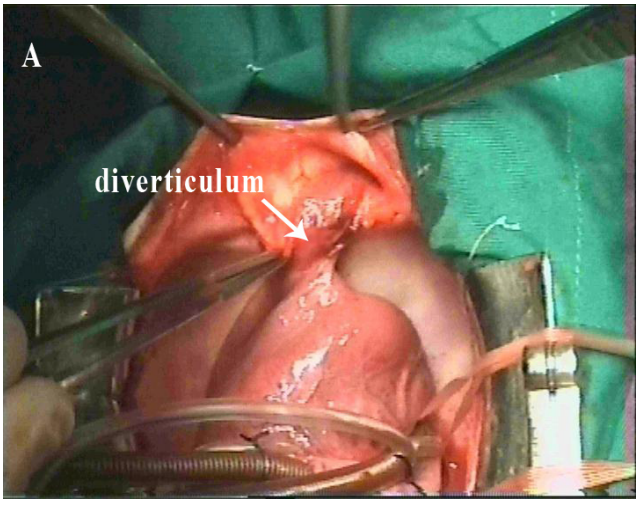
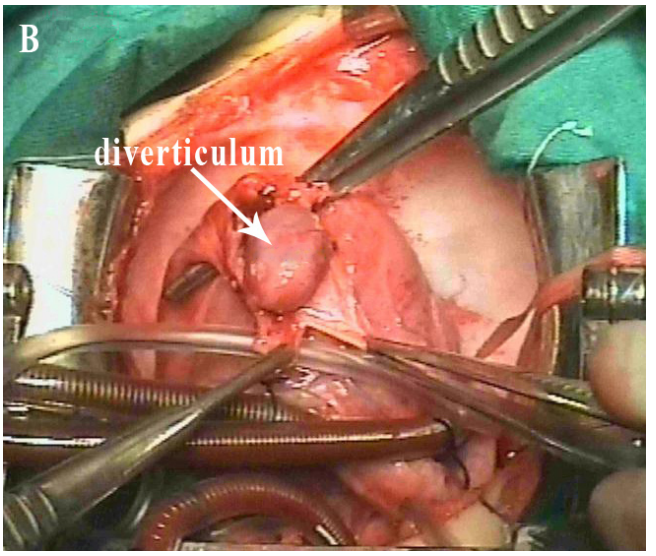


Figure 4. Left ventricle angiogram of the patient
AO, Aorta; LV, Left ventricle; D, Diverticulum

Under general anesthesia and via median sternotomy, cardio-pulmonary bypass (CPB) was established and the heart was arrested with cold crystalloid cardioplagin administration. The diverticulum was herniated into the abdominal wall through a relatively small diaphragmatic defect (Figure 5 A). The diverticulum was released from the surrounding tissues and resected (Figure 5 B, C). The ventricular septal defects were thereafter repaired via right ventriculotomy. The atrial septal defect was closed via right atriotomy without patch usage. The patient was weaned from CPB by using a temporary pacemaker.

After surgery, the patient was transferred to the pediatric ICU. However, the patient continued to have complete heart block, which persisted after 10 days, l eading to the insertion of a permanent pacemaker. The patient is healthy otherwise with a good ejection fraction.





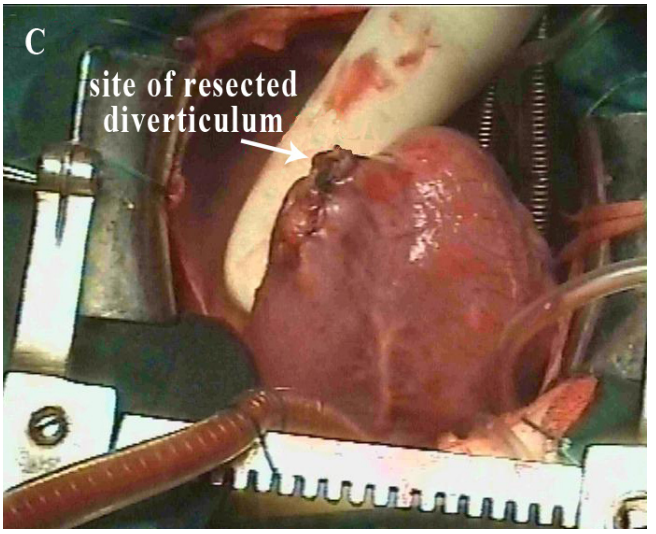


Figure 5. Surgical diverticular resection

Discussion

Congenital left ventricular diverticulum is a rare cardiac malformation.[1-5] It appears to be a developmental anomaly, starting in the 4th embryonic week.[1] Although this anomaly may exist alone, associated cardiac, vascular, or thoraco-abdominal abnormalities are present in some cases.[1,2]

Diagnosis can be made after the exclusion of the ventricular aneurysm, coronary artery disease, local or systemic inflammation, or traumatic causes as well as cardiomyopathies.[1,2,8]

Clinically, most congenital left ventricular diverticula are asymptomatic, but some of them may causesystemic embolization, heart failure, valvular regurgitation, ventricular wall rupture, ventricular tachycardia, or sudden cardiac death.[1] Diagnosis is established by imaging studies such as echocardiography, magnetic resonance imaging, or left ventricular angiography.[1,9,10]

Two categories of congenital ventricular diverticulum could be identified with respect to their localization: apical and non-apical. Apical diverticula are always associated with midline thoraco-abdominal defects and other heart malformations. Non-apical diverticula are always isolated defects.

Ventricular diverticula could be differentiated from ventricular aneurysms by echocardiography and angiography.[1,8] Diverticula are characterized by synchronal contractility, but aneurysms are akinetic with paradoxical systolic motion. The outcome is different in these two types of outpouchings: congenital ventricular aneurysms are associated with adverse outcomes, whereas the prognosis for congenital ventricular diverticula is good.[1]

All patients with a midline thoraco-abdominal defect and a pulsatile mass should be evaluated carefully to rule out ventricular apical diverticulum. Otherwise, it may complicate surgical options for repairing the thoraco-abdominal defects. Patients with cardiac diverticulum should be evaluated for other associated intracardiac and extracardiac lesions.[1,2,6]

Mode of treatment has to be individually tailored and depends on clinical presentation, accompanying abnormalities, and possible complications. Because of the usually benign course of congenital left ventricular diverticula, most of them can be managed conservatively. Treatment options for high-risk cases include surgery, anticoagulant therapy, and management of arrhythmias.

Different surgical approaches have been recommended for the treatment of apical diverticula.[7] Irrespective of the method, however, the surgeon should be aware of surgical complications such as complete heart block as well as other well-known complications.

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