Repair of an isolated huge congenital left ventricular diverticulum

Yoav Paz, MD,^a Eduard Fridman, MD,^b Fayez M. Shakalia, MD,^c Joseph Danieli, MD,^c and David Mishaly, MD,^a
Tel-Aviv, Israel

Congenital left ventricular diverticulum is a very rare condition usually associated with other congenital anomalies, including those of the sternum, diaphragm, pericardium, and abdominal wall (Cantrell syndrome). We describe a 3½-year-old boy with a huge, isolated, congenital left ventricular diverticulum and no other congenital anomaly, the third reported case of its kind and the first of such large dimensions.

Clinical Summary

A 3½-year-old boy was referred to us with the diagnosis of “double-chamber left ventricle.” He had no previous history of chest trauma, tuberculosis, cardiovascular disease, Chagas disease, or Kawasaki disease, but recurrent left-lung pneumonia events had appeared several months before we examined him, and he had experienced severe respiratory failure prior to his referral. At that time, chest radiography revealed an abnormal cardiac silhouette. Two-dimensional echocardiography demonstrated a huge diverticulum located at the posterolateral wall, penetrating just below the mitral valve annulus. Left ventricular contractility was normal, but a Doppler flow study demonstrated mild-to-moderate mitral regurgitation, as well as bidirectional blood flow, to and from the diverticulum. Angiohelical computed tomography showed a large pouch connected to the left ventricle through a narrow opening (Figure 1).

Through a median sternotomy, the pericardium was found to be adherent to the entire heart and to a huge (8 × 9 cm) left ventricular diverticulum. Although the diverticulum was situated within the pericardial sac, the entire mass bulged into the left chest, causing compression of the left lung. After the patient was connected to cardiopulmonary bypass and during heart fibrillation, the diverticulum was opened. Blood was evacuated, and mitral valve components could be seen through the opening to the left ventricle. No thrombotic remnants were found. The diverticulum was resected, and the entry site was obliterated with a polytetrafluoroethylene patch (Gore-Tex patch; W. L Gore & Associates, Inc, Flagstaff, Ariz; Figure 2). Further inspection revealed a normal-sized left ventricle and normal-sized coronary arteries, with no coronary aneurysms. The operation was completed uneventfully, contractility was good, and the previous mitral regurgitation disappeared as well. The postoperative period was uneventful, and the child was discharged from the hospital on the sixth postoperative day.

Pathologic examination of the excised specimen revealed a 0.4-cm-wide connective tissue with marked reticulin fibers (Masson staining). The internal surface was covered by simple epithelia (CD31 and CD34 staining). A few unorganized muscular fibers were found among the reticulin fibers (desmin staining).

Discussion

The terms “congenital aneurysm of the heart” and “congenital diverticulum of the heart” have been used interchangeably, but they have different definitions and should be distinguished from one another. Papagiannis and colleagues¹ suggested that congenital left ventricular aneurysm should be distinguished from congenital left ventricular diverticulum on the basis of the communication morphology of the pouch with the ventricle. An aneurysm has a wide communication with the ventricle, whereas the diverticulum communicates with the ventricle through a narrow sleeve.¹

From the Departments of Pediatric Cardiac Surgery,^a Tel-Aviv, Israel, Pathology,^b and Pediatric Cardiology,^c Sheba Medical Center, Sackler Faculty of Medicine, Tel-Aviv University, Tel-Aviv, Israel.

Received for publication Dec 1, 2003; accepted for publication Dec 15, 2003.

Address for reprints: David Mishaly MD, Department of Congenital and Pediatric Cardiac Surgery, Haim Sheba Medical Center, Tel Hashomer 52621, Israel (E-mail: dmishaly@sheba.health.gov.il).

J Thorac Cardiovasc Surg 2004;128:313-4
0022-5223/\$30.00

Copyright © 2003 by The American Association for Thoracic Surgery

doi:10.1016/j.jtcvs.2003.12.029

Figure 1. Angiohelical computed tomogram demonstrating a huge diverticulum with a narrow opening to the left ventricle (LV).

Figure 2. The repaired diverticulum shown in Figure 1. A polytetrafluoroethylene patch was used to close the diverticular opening to the left ventricle, and most of the diverticular sac has been resected out.

There are 2 types of congenital ventricular diverticula, fibrous and muscular. In 1984, Mardini² reported 2 cases of infants with congenital diverticulum of the left ventricle, one of which was in a submitral position in an infant who had moderate mitral incom-

petence and a poorly functioning left heart. Our patient also had mitral incompetence but without contractility impairment. Between 1992 and 1996, Cavalle-Garrido and associates³ diagnosed 7 cases of cardiac diverticula and aneurysms during fetal life, only one of which was a submitral diverticulum. Ours is the third reported child with isolated congenital submitral diverticulum, and the first report of such huge dimensions.

Importantly, our patient was asymptomatic until the age of approximately 3 years, suggesting an evolutionary process of an isolated cardiac diverticula that was not diagnosed during fetal life or infancy. Although it is well accepted that symptomatic left ventricular diverticula should be treated by means of surgical intervention, resection of asymptomatic diverticula is a matter of controversy.⁴ This kind of diverticulum can potentially cause mitral insufficiency, as we have described here, and can rupture spontaneously, as described by Westaby and coworkers.⁵ Moreover, Skapinker⁶ reported 2 cases of sudden death related to unresected ventricular diverticulum, both of the muscular type. Other potential complications of untreated ventricular diverticula are arrhythmias, heart failure, and thromboembolic events.

The surgical repair of a left ventricular diverticulum is a simple procedure with nominal risk, and we recommend resection and patch closure of the opening in asymptomatic, as well as symptomatic, cases.

We thank Esther Eshkol for editorial assistance.

References

1. Papagiannis J, Van Praagh R, Schwint O, D'Orsogna L, Qureshi F, Reynolds J, et al. Congenital left ventricular aneurysm: clinical, imaging, pathologic, and surgical findings in seven new cases. *Am Heart J*. 2001;141:491-9.
2. Mardini MK. Congenital diverticulum of the left ventricle. Report of two unusual cases. *Br Heart J*. 1984;51:321-6.
3. Cavalle-Garrido T, Cloutier A, Harder J, Boutin C, Smallhorn JF. Evolution of fetal ventricular aneurysms and diverticula of the heart: an echocardiographic study. *Am J Perinatol*. 1977;14:392-400.
4. Gueron M, Hirsch M, Opschitzer I, Mogel P. Left ventricular diverticulum and mitral incompetence in asymptomatic children. *Circulation*. 1976;53:181-6.
5. Westaby S, Katsumata T, Runciman M, Burch M. Ruptured left ventricular diverticulum in infancy. *Ann Thorac Surg*. 1997;64:1181-2.
6. Skapinker S. Diverticulum of left ventricle of heart: review of literature and report of successful removal of the diverticulum. *Arch Surg*. 1951; 63:629-30.