Bifid cardiac apex: A rare morphologic structure

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A 17-year-old patient was diagnosed with a large ventricular septal defect (VSD) (18 mm), a small atrial septal defect (ASD), and a double-chambered right ventricle with a 90 mm Hg gradient. She had been monitored by several pediatric cardiologists since the age of 1 year, and she had no abnormalities except for her cardiac disease. These features were also confirmed with cardiac magnetic resonance imaging (MRI). After that examination, the patient was referred to our clinic for surgical correction. She had no symptoms and her oxygen saturation was 92% to 93% in the operating room before anesthesia.

The operation was begun with a median sternotomy. After the pericardium was opened, the external finding of the heart was not consistent with the preoperative echocardiographic and MRI findings. The heart was not as big as expected, and it had a bifid apex with a 2-cm deep cleft like a crab fork (Figure 1). The pulmonary arteries were of normal size. Right atriotomy revealed a big ASD (2 cm). The right ventricular volume was less than normal and the tricuspid valve was small in diameter. No VSD could be seen through right atriotomy. Normal saline was also flushed through left ventricle but nothing exited via the right ventricle. The pulmonary arteries were controlled and the right ventricular outflow tract was free of obstructive lesions. Because the right ventricular volume was less than normal, the ASD was closed partially.

The patient was weaned from cardiopulmonary bypass without any problem. The postoperative course was uneventful, and the patient was discharged on the seventh postoperative day with an oxygen saturation of 96% to 97%.

When we compared the cardiac MRI after the operation with the intraoperative morphology, we saw that there was a large VSD but that it had no connection with the right ventricle. Another chamber was present between the right and the left ventricles, and the VSD connected only with the left one. A septumlike structure separated this chamber and right ventricle. Also, the heart had a bifid apex as seen in the cardiac MRI (Figure 2).

The right and left ventricles develop as independent chambers on either side of the primitive plate. Subsequently, they merge and muscle fibers form a bridge across the two ventricles.[1] As in our case, the separation of the two ventricles is not complete. Although the bifid cardiac apex is seen in sea mammals, such as the whale, dugong, and manatee, it is a unique congenital morphology for human beings.[1] It is seen rarely in the normal human heart or association with other congenital cardiac anomalies.

With these findings, we believe that this kind of morphology (bifid apex with a small right ventricle, third chamber–like development, and ASD) may be seen rarely and has not been documented before in the literature.

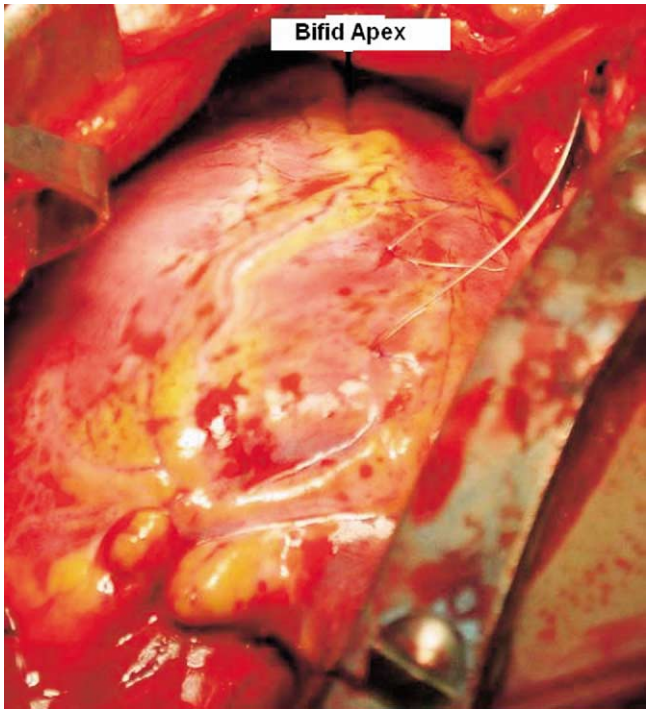

Figure 1. Bifid apex.

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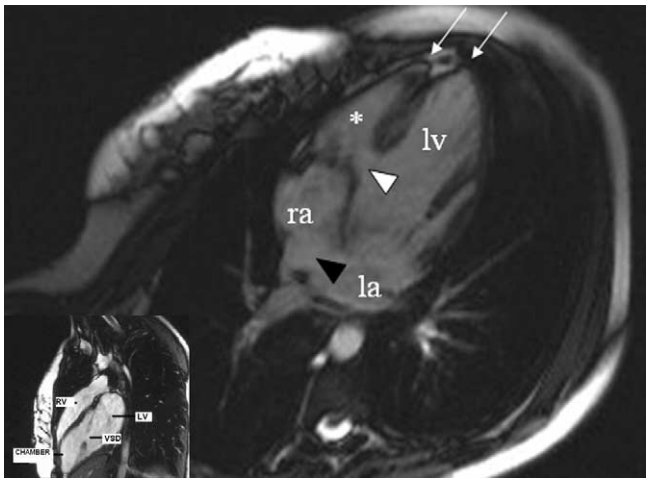

Figure 2. Cardiac MRI of the patient. *ra,* Right atrium, *lv,* left ventricle; *la,* left atrium; *white arrow,* VSD; *black arrow,* ASD; *third chamber.

Reference

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A novel aortic arch reconstruction method for double-inlet left ventricle with interrupted aortic arch and restrictive bulboventricular foramen

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Author photo

Patients with a functional single ventricle, unrestricted pulmonary blood flow, and aortic arch obstruction require staged palliation aimed at the Fontan circulation. Obstruction of the bulboventricular foramen (BVF), the communication between the left ventricle and the rudimentary right ventricle, may complicate the clinical course of patients with double-inlet left ventricle (DILV).[1] In this report we present a new reconstructive approach for such morphology, applying a modification of the Damus-Kaye-Stansel procedure combined with arch reconstruction by suturing the ascending aorta to the descending aorta. This modification is called the swing-back technique.

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Clinical Summary

The patient was a neonate (14-day-old boy weighing 3.0 kg) in whom situs solitus, atrioventricular and ventriculoarterial discordance {S,L,L}, DILV with a rudimentary right ventricle, transposition of the great arteries (TGA), restrictive BVF, patent ductus arteriosus, retroaortic innominate vein, and type A interrupted aortic arch (Seroria-Patton classification) were diagnosed. The diameters of the ascending aorta, main pulmonary artery (PA), and descending aorta were 9.1, 14.5, and 9.8 mm, respectively, and the BVF area index was 1.39 cm^2/m^2. The morphologic diagnosis was based on the evaluation of 2-dimensionalechocardiography and multislice computed tomography.

Surgical Technique

After median sternotomy, cardiopulmonary bypass was instituted with dual arterial cannulations: one cannula was placed into the brachiocephalic artery and the other was placed directly into the