cated in the deterioration of right ventricular function and the development of arrhythmias after the repair of TOF. In the present case with a dysplastic PV and a hypoplastic pulmonary annulus associated with absent PV syndrome, we performed a minimum ventriculotomy using a well-designed polytetrafluoroethylene monocusp patch with a bulging sinus [4] through a transatrial-transpulmonary approach to reduce the risk of pulmonary regurgitation and maintain right ventricular function for the long term.

In conclusion, we have presented a case of TOF with an absent PV and a single coronary artery with a major coronary fistula. We performed staged surgical repair and the postoperative course was excellent with no evidence of myocardial ischemia.

References


Duplicate Mitral Valve in an Infant With Shone’s Anomaly

Rıza Turkoz, MD, Canan Ayabakan, MD, Can Vuran, MD, Oğuz Omay, MD, Selman Vefa Yıldırım, MD, and N. Kursad Tokel, MD
Departments of Cardiovascular Surgery and Pediatrics, Baskent University, İstanbul Teaching and Medical Research Center, İstanbul, Turkey

Duplication of mitral valve is a very rare anomaly. It is characterized by two independent mitral valve apparatuses (leaflets and annulus) and subvalvular apparatuses (chordae and papillary muscles) that function well by themselves. In this report, we present duplicate mitral valve with parachute chordal attachment and mitral stenosis in an infant. The patient was successfully treated with the reconstruction of the larger valve without any intervention to the smaller one.


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A 10-month-old boy was referred to our hospital with dyspnea and retarded growth. She had already undergone balloon angioplasty twice for aortic coarctation at another institution (at 40 days, 7 months of age). Physical examination revealed a grade 2/6 holosystolic murmur and a diastolic rumble at the left fourth intercostal space. The liver was palpable 4 cm below the costal margin.

On transthoracic echocardiography, double-orifice mitral valve was observed, and the mitral chordae were short and underdeveloped; hence the leaflets attached directly to a single papillary muscle. Although a second papillary muscle was observed, no chordal attachment to this muscle was seen (Fig 1). Therefore a “double-orifice” and “parachute” mitral valve was suspected, and a duplicate mitral valve orifice was undetectable preoperatively. The peak and mean transmural gradients were 27 mm Hg and 11 mm Hg, respectively. There was trivial mitral regurgitation. The left atrium was measured 52 mm × 40 mm. The associated echocardiographic findings were bicuspid aortic valve, mild tricuspid valve regurgitation, and pulmonary hypertension. There was no coarctation subsequent to the balloon angioplasties per-
formed earlier. Cardiac catheterization was performed at another institution and revealed a mitral gradient of 11 mm Hg and an aortic coarctation gradient of 20 mm Hg. The mitral gradient was calculated by the difference between the pulmonary wedge pressure (14 mm Hg) and the left ventricular end-diastolic pressure (3 mm Hg). The pulmonary artery pressure was 29/17 (mean, 23 mm Hg). The second balloon angioplasty was performed during this procedure.

At the operation, after standard aortic cannulation and venous cannulation through both superior and inferior vena cavae, cardiopulmonary bypass with mild hypothermia (34°C) was instituted. The aorta was cross clamped and the myocardium was arrested by antegrade blood cardioplegia. The large left atrium was entered through a standard longitudinal incision. Two completely separate and independent mitral valves were observed. The diameter of the larger mitral valve was 15 mm, and the diameter of the smaller valve was 8 mm, which was located approximately 1 cm to the left side of the larger one (Fig 2). Both mitral valves had anterior and posterior leaflets, but the chordae tendinea were not developed. The leaflets of each mitral valve were directly attached to separate papillary muscles. So the papillary muscle of the larger mitral valve was divided 1 cm longitudinally, providing a wide secondary mitral orifice. A supravalvular ring was attached approximately 1.5 to 2 mm above both mitral valves. This ring-forming tissue was resected from both mitral valves. No intervention was done to the smaller valve. There was minimal mitral regurgitation from the larger orifice after the operation. The patient was easily weaned from the cardiopulmonary bypass with moderate doses of inotropic support.

The postoperative course was uneventful. Postoperative transthoracic echocardiogram showed mild mitral regurgitation. The peak gradient decreased from 27 mm Hg to 9 mm Hg, and the mean gradient decreased from 11 mm Hg to 5 mm Hg. The patient was discharged on postoperative day 5. Serial clinical and echocardiographic evaluations during 8 months of follow-up confirmed good clinical condition, and the patient remained stable. In the last echocardiographic examination at 8 months postoperatively, there was moderate mitral regurgitation, the mean transmitial gradient was 8 mm Hg, and the velocity of the tricuspid regurgitant jet was 3.5 m/sec, indicating a systolic pulmonary artery pressure of approximately 50 mm Hg. There was no coarctation.

Comment

Double-orifice mitral valve is a rare anomaly and is most commonly associated with atrioventricular septal defects [1, 3]. The incidence of double-orifice mitral valve has been reported as high as 9.1% in a large series among atrioventricular septal defects [3]. The clinical presentation of double-orifice mitral valve is variable. Zalzstein and colleagues [4] described echocardiographic findings in 46 patients with double-orifice mitral valve. In this study, mitral insufficiency was the most common functional abnormality, and mitral stenosis, either alone or with insufficiency was rarer. A minority of these patients who underwent repair of associated cardiac lesions required surgical intervention directed at the double-orifice mitral valve. The largest, postmortem study concerning the double-orifice mitral valve was done by Bano-Rodrigo and colleagues [1]. In most of these cases of this study, a double orifice was created by a hole in the anterior or the posterior leaflets (85%), whereas only 15% of the patients had a fibrous tissue separating the two orifices [1]. Completely separate orifices and discrete mitral valve apparatuses, as seen in our patient, were not defined in any of their cases. It is noteworthy that most of the cases (56%) in this study had atrioventricular septal defects [1].

Duplication of the mitral valve is a very rare anomaly [2]. In duplicate mitral valve cases, two independent mitral valve apparatuses (leaflets and annulus) and subvalvular apparatuses (chordae and papillary muscles) function well by themselves [2]. Such an anomaly was described by Ando and colleagues [1] in a patient who also had significant mitral regurgitation. However in our case the predominant lesion was the mitral stenosis. Because the supravalvular mitral ring, mitral stenosis, aortic coarctation, and bicuspid aortic valve were associated with the duplicate mitral valve in our patient, she was considered to be a case of Shone’s anomaly. The most common mitral lesions in Shone’s anomaly consist of parachute deformity, supramitral fibrous rings, and typical congenital mitral stenosis with short chordae tendineae, obliteration of interchordal spaces, and reduction of interpapillary distance [5, 6], which are similarly present in our patient. Our patient had two papillary muscles, and the leaflets of each valve were attached directly to an isolated papillary muscle. Both of the mitral valves were hypoplastic, and there was no chordal tissue. The mitral valve repair directed to the larger mitral valve orifice successfully abolished the transmitral gradient in our patient. Because no chordae tendinea exists, mitral valve dysfunction requiring a mitral valve replacement may be expected in the follow-up; however, delaying it as much as possible is desired, given the high morbidity.
and mortality of the mitral valve replacement at an early age [7].

We believe that the duplication of mitral valve associated with Shone’s anomaly has not been previously published. Our case demonstrates a unique aspect of this very rare anomaly. When the duplicate mitral valve orifice is associated with mitral stenosis, we suggest the reconstruction of the larger valve without any intervention to the smaller one is a feasible option of treatment.

References


Isolated Subclavian Artery: Anatomical and Surgical Considerations

Igor E. Konstantinov, MD, PhD, Pankaj Saxena, MCh, DNB, Yves d’Udekem, MD, PhD, and Christian P. Brizard, MD

Department of Cardiac Surgery, Royal Children’s Hospital, Melbourne, Australia

Isolated subclavian artery is a rare congenital anomaly. Herein we discuss a patient with isolated left subclavian artery and bilateral patent arterial ducts who underwent successful repair. We also describe the detailed anatomy of both recurrent nerves in this condition and embryology of the anomaly.

Comment

The described anomaly derives from an unusual involution of the double fourth aortic arch [2] when the breaks occur between the left common carotid artery and the LSCA, and the LSCA and the descending aorta as well as the bilateral patent arterial ducts and the dorsal remnants of the sixth (pulmonary) embryonic aortic arches (Figs 2A and 2B). Thus, the LSCA remains attached to the pulmonary artery though the left PDA (Figs 2B and 2C). Clear understanding of the course of the recurrent nerves in this rare anomaly is a must for successful repair. The recurrent nerve derives from the vagus nerve that enters the thorax through the carotid sheath adjacent to the common carotid artery, and as such, is always anterior to subclavian artery. The recurrent nerve then turns around the remnants of the sixth aortic arch and travels to the vocal cords in the tracheo-esophageal groove (Fig 2C). Both recurrent nerves were clearly visualized in our patient during surgery (Fig 2D). Because the LSCA is connected to the PA through the left PDA, and to the left vertebral...