Case Reports

Accessory Mitral Valve Leaflet

Causing Severe Left Ventricular Outflow Tract Obstruction in a Preterm Neonate with a Partial Atrioventricular Septal Defect

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Atrioventricular septal defects represent a class of congenital cardiac malformations that vary in presentation and management strategy depending upon the severity of the particular lesions present. We present the case of a premature neonate who had a partial atrioventricular septal defect and an accessory mitral (or left atrioventricular) valve leaflet. The latter caused severe left ventricular outflow tract obstruction and severely depressed left ventricular function. We found only one other report of this atrioventricular valve abnormality in association with atrioventricular septal defect. To our knowledge, our patient (at a body weight of 1,800 g) is the smallest to survive corrective surgery of an accessory mitral valve leaflet with severe left ventricular outflow tract obstruction. In addition to our patient's case, we discuss the relevant medical literature. ***(Tex Heart Inst J 2016;43(6):543-5)***

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Atrioventricular septal defects (AVSDs) are characterized by deficiency of the atrioventricular (AV) septum, abnormalities of the AV valves, and a predisposition to progressive left ventricular outflow tract (LVOT) obstruction. This progressive obstruction is caused by an elongated "gooseneck" LVOT deformity that displaces the aortic valve anteriorly. Left ventricular outflow tract obstruction occurs most often when 2 distinct AV valve orifices are present, as in partial AVSD; and also in complete AVSD when the superior bridging leaflet attaches to the crest of the interventricular septum (creating 2 orifices and further elongating the LVOT). Abnormal left-AV-valve chordal attachments, septal hypertrophy, and abnormally oriented papillary muscles can further exacerbate subaortic narrowing.[1] Accessory mitral valve tissue (AMVT), a rare congenital malformation, can result in LVOT obstruction and is associated with other congenital cardiac anomalies. Accessory mitral valve (MV) leaflet, the typical form of AMVT, has been reported only once before in association with AVSD.[2]

We describe the case of a very small, premature neonate who underwent partial-AVSD repair and accessory MV (or left-AV-valve) leaflet resection to correct severe LVOT obstruction.

Case Report

In May 2014, a 6-day-old preterm male neonate was transferred to our institution from another hospital. The diagnosis was severe aortic stenosis. His mother's pregnancy was notable for dizygotic twins, well-controlled gestational diabetes mellitus, and delivery by cesarean section because of placenta previa and premature membrane rupture. The infant's birth weight of 1,790 g was appropriate for his estimated gestational age of 31 weeks. At birth, he needed continuous positive airway pressure support but overall appeared clinically well; however, a murmur prompted echocardiographic testing, the results of which raised concerns about severe aortic stenosis. His twin brother was concurrently diagnosed with a membranous ventricular septal defect. There was no other known family history of congenital heart disease.

Prostaglandin E_1 therapy had been initiated before the infant's transport to our hospital. Physical examination revealed a nondysmorphic premature neonate. Auscultation yielded a gallop, a harsh grade 4/6 systolic murmur best heard along the mid right sternal border, and a palpable thrill across the sternum. A chest radiograph showed

moderate cardiomegaly with central venous congestion, and an electrocardiogram revealed sinus rhythm with left-axis deviation, biventricular hypertrophy, and T-wave inversion in the anterolateral leads. Echocardiograms revealed a partial AVSD with a small primum atrial septal defect, patent foramen ovale, no appreciable ventricular-level shunting, 2 good-sized papillary muscles within the left ventricle (LV), and 2 distinct right and left AV-valve annuli at the same septal insertion level (Fig. 1). The left-AV-valve tissue appeared to be redundant and crowded the LVOT (Fig. 2A), resulting in severe LVOT obstruction. Continuous-wave Doppler mode revealed a peak gradient of 55 mmHg (mean, 31 mmHg) in the presence of severely depressed LV systolic function. Moderate biventricular hypertrophy was present. A cleft in the left-AV-valve anterior leaflet enabled moderate regurgitation, as can be seen in AVSD.[1] The ventricles and aortic arch were of normal size, and there was a moderate-sized patent ductus arteriosus with exclusive right-to-left shunting.

The patient subsequently developed increasing signs of low cardiac output with an increasing obstruction across the LVOT (mean gradient, 42 mmHg). Despite the substantial operative risk associated with his low weight and tenuous status, we concluded that medical management had been maximized and that surgical repair was the best option.

On day 11 of life and at a body weight of 1,800 g, the patient underwent median sternotomy and was placed under cardioplegic arrest on full-flow cardiopulmonary bypass. After performing an oblique right atriotomy, we were struck by the extreme hypertrophy of the atria and ventricles. The AV valve tissue was almost as delicate as tissue paper. The left AV valve was competent, so we did not close the cleft. We closed the primum atrial septal defect with use of an autologous pericardial patch and interrupted 8-0 Prolene horizontal mattress sutures. Working through the aortic valve, we saw an accessory AV valve leaflet that was causing nearly total

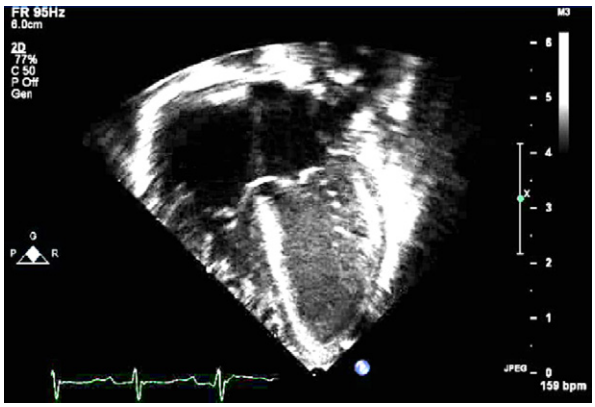

Fig. 1 *Transthoracic echocardiogram shows 2 distinct right and left atrioventricular valve annuli at the same septal insertion level.*

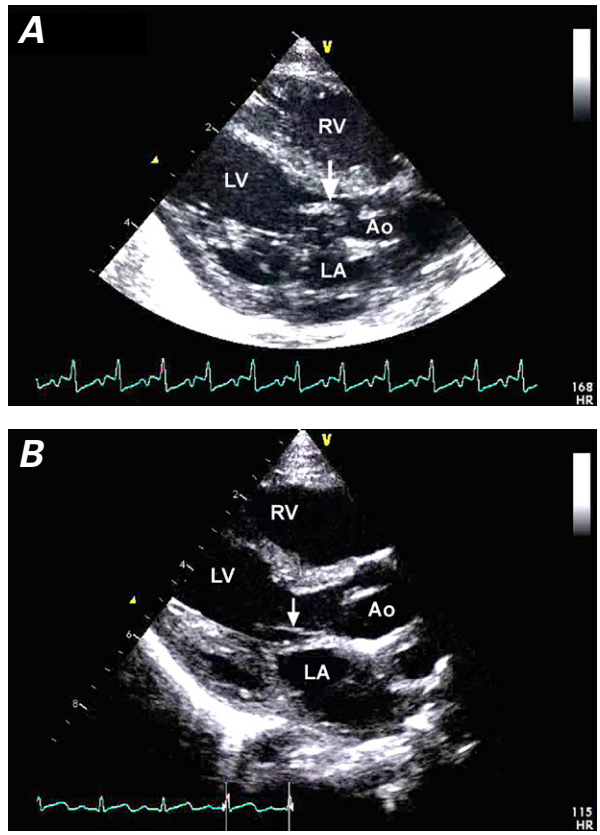

Fig. 2 *Transthoracic echocardiograms.* ***A****) Crowding of the left ventricular outflow tract by a redundant mitral leaflet (arrow) is revealed intraoperatively to be an accessory mitral valve leaflet.* ***B****) Postoperatively, accessory mitral valve leaflet remnants arise from the anterior mitral valve leaflet (arrow) without evidence of left ventricular outflow tract obstruction.*

Ao = aorta; LA = left atrium; LV = left ventricle; RV = right ventricle

obstruction of the LVOT. This leaflet was redundant and dysplastic in appearance. Leaflet resection opened the LVOT satisfactorily. The aortic cross-clamp time was 126 min. Sinus rhythm was restored, and the patient was uneventfully weaned from cardiopulmonary bypass.

Postoperative transthoracic echocardiograms showed accessory left-AV-valve leaflet remnants arising from the anterior leaflet and protruding into the LVOT without evidence of obstruction, no left-AV-valve stenosis, and only mild regurgitation (Fig. 2B). Although the biventricular hypertrophy persisted, ventricular function was normal.

The patient was weaned from inotropic support on postoperative day 6. He was extubated and transferred from the cardiovascular to the neonatal intensive care unit on day 7. Ectopic atrial tachycardia was observed during this period, and propranolol was initiated on day 26 of life. Because of his prematurity, the patient had an additional 56-day hospital stay before he was discharged.

Follow-up monitoring in the cardiology clinic revealed an asymptomatic infant reaching appropriate growth and developmental milestones. Because no other organ systems were involved, no genetic testing was performed. The patient had no further arrhythmias after being weaned from propranolol. An echocardiogram one year after repair showed accessory left-AV-valve leaflet remnants arising from the anterior leaflet without evidence of LVOT obstruction, mild left-AV-valve regurgitation without stenosis, normal biventricular systolic function, and no ventricular hypertrophy. Findings on the most recent echocardiogram, 2 years after repair, were unchanged.

Discussion

To our knowledge, our patient is the smallest to undergo and survive surgery to correct LVOT obstruction caused by an accessory MV (or left AV valve) leaflet. In addition, we think that this condition has been reported in association with AVSD only once before,[2] in a 2-year-old boy who had undergone complete AVSD repair as an infant and then presented with 2 syncopal episodes when the AMVT caused severe LVOT obstruction. Although the AMVT had not been detected during initial surgical repair, it was later noted upon a review of the preoperative echocardiograms.

Accessory MV tissue, which can be found on the ventricular face of the anterior MV leaflet, chordae tendineae, or papillary muscle attachments, is a rare cause of LVOT obstruction.[3] Severe obstruction can present with substantial cardiovascular compromise in infancy, whereas less severe obstruction can go undetected into adulthood and be identified after detection of a murmur or presentation with dyspnea, fatigue, palpitations, chest pain, or syncope.[3-5] Most cases of AMVT are associated with other cardiac abnormalities. Manganaro and colleagues[4] identified 104 patients who were diagnosed with AMVT from 1963 to 2012. Most (86.6%) had signs of LVOT obstruction, and AMVT was most frequently associated with ventricular septal defect (19.2%), followed by subaortic membrane (9.6%), LV hypertrophy (8.6%), and transposition of the great arteries (7.7%).[4]

After reviewing 90 reports that detailed postoperative findings of LVOT obstruction involving AMVT, Prifti and colleagues[6] classified AMVT into 2 main types: fixed and mobile. Type I (fixed) can be nodular (type IA) or membranous (type IB), and type II (mobile) is pedunculated (type IIA) or leaflet-like (type IIB). Accessory MV leaflet, an alternative term for type IIB, corresponds to approximately half of reported cases of congenital LVOT obstruction caused by AMVT.[6]

Abnormalities of the AV valves in the partial and complete forms of AVSD include a cleft in the anterior leaflet and, less often, a double-orifice or parachute left AV valve with a single papillary muscle.[1] Among 155 cases of surgically repaired AVSD, 8 patients had accessory bridging-valve tissue connecting the anterior and posterior leaflets to form a double-orifice valve with resultant mitral stenosis; however, there were no reports of LVOT obstruction caused by AMVT.[7] Although reports of an accessory MV leaflet causing LVOT obstruction in the presence of AVSD appear to be rare, it stands to reason that an accessory leaflet might easily exacerbate the subaortic narrowing that is caused by the LVOT gooseneck deformity seen in AVSD.

Surgical correction can damage the MV, especially in smaller infants. A combined approach to the MV and aorta by means of atriotomy and aortotomy is thought to enable safer complete removal of AMVT.[8,9] In our patient, a right atriotomy enabled inspection of the AV valve tissue through the primum atrial septal defect; however, the absence of a ventricular septal defect and the infant's small size necessitated that the accessory valve leaflet be resected entirely through an aortotomy.

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