Left Ventricular Outflow Tract Obstruction Produced by Redundant Mitral Valve Tissue in a Neonate

Clinical, Angiographic, and Operative Findings

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SUMMARY An unusual case of left ventricular outflow tract obstruction associated with severe left ventricular failure in a neonate is reported. The physical and laboratory data were consistent with the diagnosis of infantile valvular aortic stenosis. At operation, however, redundant gelatinous pedunculated tissue attached to the mitral valve annulus appeared to move through and obstruct the aortic valve during systole. The aortic valve showed only minimal thickening of the right and left coronary cusps. A distinct angiographic pattern was demonstrated during left ventricular cineangiography. In the frontal projection a large ovoid filling defect appeared to protrude through the aortic valve during systole and return to a subvalvular location during diastole. Recognition of this angiographic pattern should facilitate diagnosis and subsequent repair. Complete correction is possible by operative excision of the obstructing tissue without damaging the mitral valve. In contrast to isolated congenital infantile valvular aortic stenosis, a condition in which the valve leaflets are often primitive and deformed, aortic valvotomy and/or subsequent valve replacement are not necessary, resulting in a better long-term prognosis.

LEFT VENTRICULAR OUTFLOW TRACT obstruction is an uncommon cause of congestive heart failure in the newborn. Often obstruction is caused by a primitive and markedly deformed aortic valve with thickened or obliterated cusps. Progressive left ventricular failure and diminishing systemic output frequently result despite intensive medical management. The abnormal structure of the valve presents a difficult challenge to the surgeon even with modern surgical techniques. Operative mortality remains high, and may exceed 50% even in the absence of associated lesions. In addition, survivors often require repeat valvotomy or valve replacement in later life.

The purpose of this report is to document the clinical, angiographic and operative findings in an unusual case of left ventricular outflow tract obstruction in a newborn infant who presented with signs and symptoms suggestive of congenital infantile aortic stenosis. Dysplastic mitral, rather than aortic, valve tissue caused obstruction at the aortic valve level and contributed to severe left ventricular failure. This form of left ventricular outflow tract obstruction appears to have a characteristic angiographic appearance, is potentially correctable surgically and, in contrast to most cases of congenital infantile valvular aortic stenosis, should have a better long-term prognosis. To our knowledge no cases have been reported in newborn infants, although MacLean and associates have documented a single case with similar angiographic and morphologic features in a one-year-old infant.

Case Report

This 3.8 kg, 2-day-old female was the product of a full term uncomplicated pregnancy and labor. One and five minute Apgar scores were 7 and 8, respectively, and intermittent grunting and peripheral cyanosis were apparent. There was no audible murmur and the neurological examination was normal. The cardiac and respiratory rates were within normal limits. During the first 36 hours of life the baby fed poorly and developed progressive cyanosis, tachypnea, and tachycardia.

At 38 hours of age, the patient was transferred from a local Salt Lake City hospital to the Primary Children's Medical Center for further evaluation. There was no evidence of central or peripheral cyanosis while the infant was breathing 100% oxygen by face mask. The skin was clear and free of unusual pigmentation. The heart rate was regular at 160 beats/min but respirations were labored at 80/min. Doppler blood pressure measurements from the left arm and left leg were 70 and 60 mm Hg, respectively. The lungs were clear to auscultation. A grade III/VI systolic ejection murmur was audible at the lower left sternal border associated with a prominent S2 gallop. There was no palpable thrill or audible ejection click. A soft liver edge was felt 6 cm below the right costal margin. Bilateral brachial and femoral pulses were readily palpable. There was no evidence of seizure activity, and suck and Moro reflexes were considered normal.

The electrocardiogram (fig. 1) demonstrated a frontal plane axis of +160°, a qR pattern in right precordial leads and ST depression and T wave inversion in left precordial leads. Serum glucose, calcium, hematocrit, and hemoglobin levels were within normal limits. Initial warmed capillary heel-stick blood gases while breathing 100% oxygen by face mask were: pH 7.35, Po2 63 mm Hg, Pco2 30 mm Hg, and HCO3 16.5 mEq/L. The initial plain chest X-ray showed moderate cardiomegaly and severe pulmonary venous congestion. We considered the physical and laboratory findings to be compatible with the diagnosis of left ventricular out-
flow tract obstruction with severe congestive failure.
Therapy consisted of intravenous furosemide, digoxin, and sodium bicarbonate. Over the next 12 hours, cardiac and respiratory rates decreased to 130 and 45/min, respectively, urine output measured 240 cc, and the liver decreased in size by 4 cm. The murmur and gallop rhythm, however, remained unchanged.

Cardiac catheterization was performed at 50 hours of age. We first advanced an umbilical artery catheter in a retrograde fashion into the ascending aorta but could not pass it across the aortic valve. A second catheter was then introduced into the right saphenous vein and advanced across a patent foramen ovale, the mitral valve and into the left ventricle. Simultaneous measurement of left ventricular and ascending aortic pressures demonstrated a peak systolic gradient of 62 mm Hg across the left ventricular outflow tract (table 1). Biplane frontal and lateral cineangiograms performed from the left ventricle disclosed a small, thick-walled chamber, and mildly thickened but freely mobile aortic valve leaflets. An ovoid filling defect was present in the left ventricular outflow tract. This filling defect, which suggested the presence of a mass lesion, appeared to move into the aortic valve during systole (fig. 2B) and return to a subvalvular location during diastole (fig. 2A). Mild narrowing of the transverse aorta between the origin of the left subclavian artery and a small ductus bump suggested the presence of fetal coarctation. However, no significant gradient was present on pull-back from the ascending to the descending aorta.

Aortotomy was performed at 72 hours of age utilizing cardiopulmonary bypass and moderate hypothermia (32°C). The surgeon exposed the aortic valve and found the left and right coronary cusps to be mildly thickened while the noncoronary cusp was normal. When the valve leaflets were retracted, a mass of tissue which protruded into the aortic valve area with each systole could be seen. The pedunculated mass was then excised from its origin at the bicuspid point of the mitral annulus and its anterior leaflet. The mass was smooth, avascular and gelatinous in appearance and measured 2.0 × 0.5 × 0.5 cm. It closely resembled the accessory tricuspid tissue reported by us in two cases of complete d-transposition.5 We are unable to provide photomicrographs of the mass because the specimen was lost in transit to the department of pathology. No other accessory mitral valve tissue was seen, and the aortic annulus was normal in size.

The aorta was closed and cardiopulmonary bypass discontinued. The infant was unable to maintain an adequate systemic pressure, and cardiopulmonary bypass was re instituted. We felt the low output might be due to an additional obstructive lesion in the ventricle, or that excision of the pedunculated mass might have damaged the mitral valve, resulting in mitral insufficiency. A left ventricu lotomy was performed and a small, thick-walled chamber could be seen. The ventricular septum, anterior and posterior mitral valve leaflets, and chordae were normally formed and intact. No other redundant tissue was present in the left ventricular outflow tract and there were no palpable or visible masses in either the ventricular septum or the free walls of the left or right ventricles. There was no visible evidence of endocardial fibroelastosis. The ventriculotomy was closed and cardiopulmonary bypass was again discontinued. Despite vigorous resuscitative efforts, the infant was unable to maintain an adequate systemic blood pressure and died. At the parents' request, necropsy was not performed.

**Discussion**

There are few reports of left ventricular outflow tract obstruction due to accessory or redundant atrioventricular valvular tissue.5-10 MacLean and associates5 described sub-aortic stenosis in a 29-year-old male caused by redundant cup-shaped structures attached to the anterior leaflet of the mitral valve. A ball-valve type of obstruction to left ven-

<table>
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<th>Site</th>
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<th>Pressure</th>
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<tr>
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<td>97</td>
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<td>97</td>
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<td>Pulmonary vein</td>
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*Data collected in 100% O₂.
Abbreviations: a = a wave; m = mean pressure; v = v wave.

**Figure 1** Twelve lead electrocardiogram taken at 24 hours of age. The frontal plane axis is +160°. Note the qR pattern in V₃ and V₄, and ST depression and T wave inversion in left precordial leads.
Tricuspid outflow tract obstruction of valvular tissue. Three of these structures attached to the mitral valve during systole. The aortic valve obstruction resulted in the immediate subvalvular region of the left ventricular outflow tract during diastole moving into the area of the aortic valve annulus during systole. The mass occupied approximately three-fourths the diameter of the aortic annulus. The right and left coronary cusps were mildly thickened but freely mobile.

Tricuspid outflow was produced during systole when these structures filled with egressing blood. Sellers and associates\(^8\) described three cases of subaortic stenosis due to accessory atrioventricular valvular tissue. In a postmortem examination of an 11-year-old child, they found five spherical balloon-like masses varying in size from 0.5 to 2.0 cm in diameter. Three of these were attached to the anterior leaflet of the mitral valve causing obstruction to left ventricular outflow during systole. The remaining two masses were attached to the mitral chordae tendineae. At operation these structures were collapsed and were not seen. In the two other patients (ages not reported), there was redundant pedunculated tissue attached to the tricuspid valve, associated with a ventricular septal defect. The authors suggest that under certain circumstances these masses protrude through the ventricular septal defect causing subaortic stenosis. Layman and associates\(^8\) and later Riemenschneider and associates\(^8\) described a similar type of obstruction to left ventricular outflow produced by accessory tricuspid valve tissue in d-transposition of the great arteries.

More recently, Shaher and associates\(^9\) reported successful surgical relief of subaortic stenosis in a one-year-old infant. The obstruction was caused by a large pedunculated spherical mass visualized angiographically as a filling defect immediately below the aortic valve. This mass moved through the aortic valve during systole and returned to a subvalvular position during diastole, simulating the motion of a ball valve. At operation two avascular tumors, measuring 1.5 × 1.5 cm were excised from the anterior mitral valve leaflet. Several other tumors were palpated in the left ventricle. All lesions were identified as rhabdomyomas by microscopic examination. The patient had seizures and skin lesions consistent with the diagnosis of tuberous sclerosis. Keuhl and associates\(^10\) recently described a similar case of severe aortic valve obstruction in a newborn caused by a polypoid rhabdomyoma which moved through the aortic valve during systole. This mass, however, was attached to the anterior wall of the left ventricle inferior to the atrioventricular groove, not to the mitral valve.

In contrast to these cases, the left ventricular outflow tract obstruction in our patient was produced by a gelatinous pedunculated tissue mass which was attached to the mitral valve annulus. To our knowledge there have been no reports describing this type of left ventricular outflow tract obstruction in the newborn period.

The angiographic findings in this case appear to constitute a distinctive pattern. One sees an asymmetric filling defect in the frontal projection during left ventriculography which moves in a ball valve fashion into or through the aortic valve annulus during systole and returns to a subvalvular location during diastole (fig. 2). A similar angiographic pattern was present in the case described by Shaher and associates.\(^9\) We believe that recognition of this cineangiographic pattern will facilitate diagnosis and surgical repair.

In the absence of a deformed stenotic valve, hypoplasia of the left ventricle, and endocardial fibroelastosis, this form of left ventricular outflow obstruction should be surgically correctable. By careful excision of the dysplastic tissue from the mitral valve apparatus, left ventricular outflow obstruction may be completely relieved. The long-term prognosis as compared to isolated infantile valvular aortic stenosis should be much better.
Acknowledgment

We gratefully acknowledge the secretarial assistance of Miss Tina Snow and the art work contributed by Mr. Robert Borstadt.

References

Left ventricular outflow tract obstruction produced by redundant mitral valve tissue in a neonate. Clinical, angiographic, and operative findings.
J W Mathewson, T A Riemenschneider, E C McGough and V R Condon

Circulation. 1976;53:196-199
doi: 10.1161/01.CIR.53.1.196
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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