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Partial A-V Canal with Subaortic Stenosis, A Case Report

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Abstract: **A case report of partial atrioventricular canal with subaortic stenosis in a 3 years old boy emphasized the importance of surgical approach for left ventricular outflow tract through the interventricular septum. Not only for reasons of gaining good exposure of all important structures in the left ventricular cavity, this approach also facilitated effective removal of all fibrous tissue and abnormal muscular tissue from the left ventricular outflow tract. In addition, the left ventricular outflow tract which may be hypoplastic can be simultaneously enlarged by a pericardial patch with this approach. The conventional approach through a small aortic incision for the release of outflow tract obstruction may be dangerous in this situation.**

Patient with atrioventricular canal may be associated with left-sided obstruction, either left-sided inlet obstruction or outlet obstruction. De Biase, et al provided prevalence of left-sided obstructive lesions in patients with atrioventricular canal in ninety consecutive patients[1]. They divided the left-sided obstructive lesions into inlet and outlet obstructions. Conotruncal malformations can greatly influence the natural history of this disease.[4,5] Potential factors predisposing left ventricular outflow tract obstruction included abnormal position of papillary muscle and intrinsic hypoplasia of the outflow tract.[2] The functional obstruction most commonly encountered in atrioventricular canal defects must be clarified from any anatomical obstruction. The left ventricular outflow tract has been usually described as being narrowed during diastole[6-14] in left ventricular septal defect seen during left ventriculography examination. This is usually considered a consequence of a functional anomaly of the anterior component of left atrioventricular valve. Other authors had shown that discrete anatomic malformations can complicate atrioventricular defects, producing stenosis of the left ventricular outflow tract.[15-21]

Case Report

A 3 year-old boy had heart murmur detected at birth without apparent cardiopulmonary problem. He was delivered by Cesarean section for breech primigravida with birth weight of 3200 gms. At the age

of 3 years-old, a chest roentgenogram showed mild cardiomegaly with normal vascularity of both lungs. EKG showed axis-60° and incomplete right bundle branch block. Echocardiogram revealed dilatation of the right atrium as well as right ventricle. The left ventricle was hypertrophied. The tricuspid valve showed mild regurgitation with peak gradient of 29 mmHg. There was subvalvular aortic stenosis and mild aortic regurgitation. The tricuspid and mitral valves were attached at the same level. An ASD primum of 1.4 cms in diameter was demonstrated with left to right shunt. Right and left heart catheterization was done to confirm the diagnosis and to gain more information for further management. The data of cardiac catheterization was shown in Table1.

Left ventriculography showed no left ventricle to right atrium shunt and no ventricular septal defect. The aortic arch was normal. There was moderate degree of narrowing at left ventricular outflow tract, compatible with subaortic stenosis. There was no significant mitral regurgitation. The pressure gradient between left ventricle and aorta was 38 mmHg.

Table 1 Data from right and left heart catheterization.

Site	O2sat	Pressure
HSVC	61.1	-
LSVC	59.0	-
HRA	60.6	m = 7
MRA	76.1	-
LRA	76.7	-
IVC	62.8	-
RV	81.9	26/7
MPA	79.7	21/8, m = 14
LPA	83.1	22/8, m = 14
PCW	-	m = 8
LA	92.9	m = 10
LV	93.1	128/0-14
A.Ao	92	90/56, m = 75

$$Qp/Qs = \frac{92-61.1}{92.9-79.7} = 2.3$$

Surgical technique

The operation was performed through a median sternotomy incision. Cardiopulmonary bypass was instituted with bi-caval and aortic cannulation with systemic hypothermia to 25° C. Intermittent cold crystalloid cardioplegic solution was given in an antegrade fashion for myocardial protection.

The right atrium was opened to expose the ASD primum and the mitral valve directly. There was no cleft of the mitral valve and no VSD was found. The ASD primum was 1.4 cms in diameter. No other abnormalities could be detected from the right atrial exposure. Thus, the ASD primum was closed by pericardial patch. Then the aorta was opened to explore the aortic valve and the subvalvular area. The aortic valve appeared normal with the three components of leaflets and commissures but the subvalvular area could not be clearly visualized. Hegar dilator was used to detect the diameter of subvalvular orifice which measured about 10 mms. The aortic incision was closed and the rewarming process was started. Hemodynamic could be maintained after cessation of cardiopulmonary bypass for only 10 minutes when hypotension rapidly developed simutaneously with the rising of left atrial pressure. Therefore, cardiopulmonary bypass was instituted. The right ventricle was opened transversely to expose the interventricular septum. Cardioplegic solution was perfused via the aortic root for myocardial protection. The interventricular septum was opened about 1 cm below the aortic valve annulus (Fig. 1). Thickened interventricular septum was meticulously resected with care

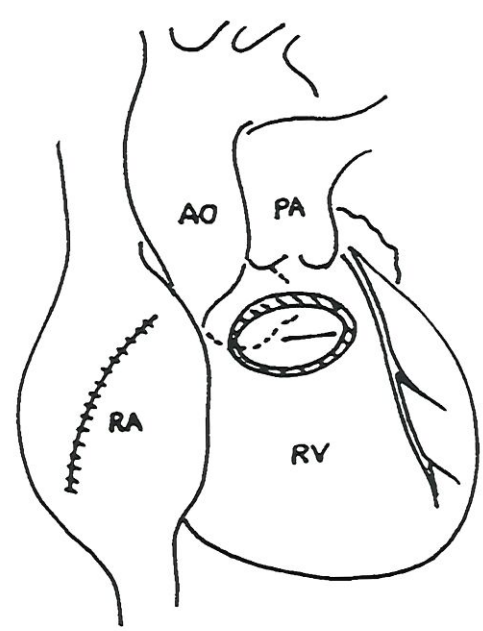

Fig. 1 Diagramatic representation showing the incision at interventricular septum to expose left ventricular outflow tract.

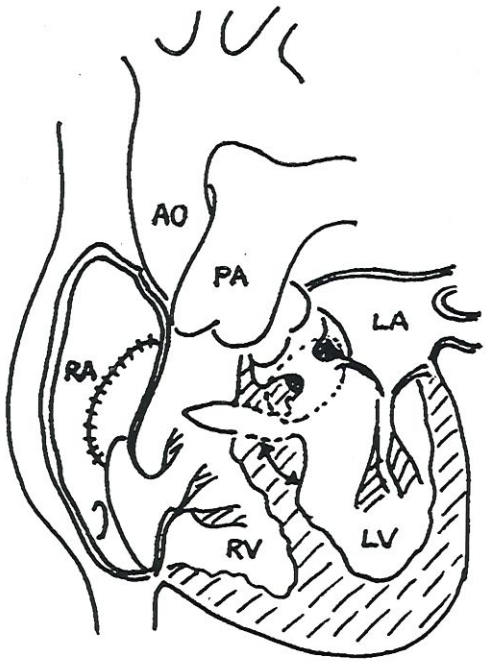

Fig. 2 Diagramatic representation showing thickened interventricular septum and circumferential thickened fibrous band that could be visualized and completely removed by trans interventricular approach.

not to injure the important components of both mitral and aortic valves (Fig. 2). Finally, the circumferential thickened fibrous band below the aortic valve annulus and all abnormal fibrous band responsible for left ventricular outflow tract obstruction were clearly visualized. They were then completely excised to relieve the obstruction. Subsequently, Hegar dilator of 14 mms. in diameter could be introduced from aortic incision through the orifice of subvalvular area. After that the interventricular incision was closed by using a pericardial patch in order to widen the left ventricular outflow tract. The aortic incision and the right ventricular incision were closed respectively. Hemodynamic was well maintained after cardiopulmonary bypass was terminated.

The patient recovered well from surgery and was discharged from the hospital with postoperative echocardiography showing no pressure gradient between left ventricle and the ascending aorta.

Discussion

In making decision for correction of subaortic stenosis, the degree of pressure gradient between the left ventricle and the ascending aorta must be carefully interpreted, especially in patients who has both ASD and subaortic stenosis. Perhaps some degree of minor pressure gradients of subaortic stenosis may become harmful effects after the ASD is closed. There is significant hemodynamic change by the increased blood volume to the left ventricle after ASD is closed. So the degree of subaortic stenosis or aortic stenosis will be increased also. If this obstructive lesion is not corrected promptly, hemodynamic will not be maintained finally. We suggested that surgeons should not over look even a minor degree of pressure gradients in these situations. The increase in pressure gradients is unpredictable since it is related to the size of ASD. Left ventriculography is very useful to confirm the degree of left ventricular tract obstruction by measuring its diameter. If there is definite narrowing of left ventricular outflow tract demonstrable by left ventriculography although with only small pressure gradients, correction of subaortic must be seriously considered.

Correction of subaortic stenosis is very difficult in small children, especially in small aortic root. Exposure of subvalvular structure is limited by a small aortic incision such as in the case here in reported. Inadequate removal of fibrous band and hypertrophied muscular tissue causing obstruction may lead to residual subaortic stenosis in the near future. Furthermore operative injury to important components of both aortic and mitral valves is very dangerous that may later cause severe regurgitation. Therefore, the right ventriculotomy and interventricular incision were more advantageous to obtain good exposure over the aortic incision. Another benefit from this approach was the excellent exposure would also enable us to enlarge the left ventricular outflow tract when necesary. We believed that the incidence of residual and restenosis of subaortic stenosis should be markedly reduced or eliminated by this technical approach.

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