Anomalous Attachment of Mitral Valve Causing Subaortic Atresia
Observations in a Case with Other Cardiac Anomalies and Multiple Spleens

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Summary
In a newborn female infant with persistent common atrioventricular canal, subaortic atresia resulting from attachment of the anterior mitral leaflet to the ventricular septum was observed. The cavity of the left ventricle was hypoplastic, while the wall was hypertrophied. The case illustrates the most severe degree of subaortic obstruction that may result from anomalous attachment of the mitral valve to the ventricular septum. Also present were multiple spleens, and other cardiovascular anomalies. The last included junction of a persistent left superior vena cava with the left atrium, absence of the coronary sinus, absence of the limbi of the fossa ovalis, and an anomalous connection of the right pulmonary veins to the right atrium.

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Congenital heart disease

As the Horizon on the causes of subaortic stenosis widens, anomalies of the mitral valve gain increasing emphasis. In the case to be reported, an anomalous mitral valve was responsible for atresia of the left ventricular outlet. Atresia of the aortic valve is frequently observed, being the most common congenital cardiac malformation which causes death in the first week of life. Atresia of the subaortic portion of the left ventricle, however, is rare among all causes of obstruction to the egress of blood from the left ventricle. We are not aware that a similar case has previously been reported. It is hoped that this report will add to the understanding of the potential basis for subaortic obstruction by an anomalous mitral valve.

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Report of Case
The specimen was obtained from a full-term 2-day-old female infant who shortly after birth exhibited respiratory distress associated with cyanosis and dyspnea. Physical examination revealed a gallop rhythm and cardiomegaly. No cardiac murmur was heard.

A frontal roentgenogram of the thorax revealed a massively enlarged globular heart in the left hemithorax and increased pulmonary vascular markings (fig. 1). The electrocardiogram (fig. 2) exhibited an ectopic right atrial pacemaker with a calculated mean electrical QRS axis in the frontal plane of +110°. The upright T wave and small q wave in lead V1 were considered normal for age. Despite intensive medical measures, the infant expired on the second day of life.

Pathologically, in addition to the cardiac findings, there were features consistent with the presence of multiple spleens. These included the fact that each lung was formed by two lobes, the middle lobe of the right lung being absent. Also, the abdominal viscera exhibited partial situs inversus, and the major lobe of the liver and the gallbladder were on the left side of the body. The spleen was represented by two masses of equal size and, in addition, two small accessory spleens were in the same area. The
SUBAORTIC ATRESIA

Figure 1
Frontal roentgenogram of the thorax.

Figure 2
Electrocardiogram.

duodenal loop was reversed from its normal direction. The distal portion of the small bowel and the large bowel were in normal positions. The mesenteric attachment to the posterior abdominal wall was incomplete, yielding considerable mobility of the bowel.

The heart lay in the left side of the thorax. The great vessels were normally related and the pulmonary trunk was considerably dilated and distinctly larger than the ascending aorta, the latter vessel being hypoplastic. The ductus arteriosus was patent. The inferior vena cava and right superior vena cava joined the right atrium normally. This chamber was dilated and hypertrophied. A persistent left superior vena cava connected with the left atrium directly. The coronary sinus was absent, but at the expected location of the ostium of the coronary sinus, a narrow opening of a cardiac vein was present.

In the atrial septum the upper and lower limbs of the fossa ovalis could not be identified (fig. 3 a). Two atrial septal defects were present. The first, measuring 8 by 5 mm, lay at the anticipated area of the fossa ovalis. The second defect was in the lowermost portion of the atrial septum and showed the characteristics of the atrial septal defect in classical persistent common atrioventricular canal. The posteroinferior portion of the atrial septum was intact.

The septal leaflet of the tricuspid valve and the anterior leaflet of the mitral valve each exhibited a cleft, but the two clefts were not continuous, as they were separated by a bridge of valvular tissue. Beneath the posterior components of the septal tricuspid and anterior mitral leaflets, narrow interventricular communications between chordae were present. The free edge of the anterior component of the cleft anterior mitral leaflet was directly adherent to the ventricular septum (fig. 3 b) at a point superior to a large defect in that septum. The adhesion caused the subaortic zone of the left ventricle to be divided into two separate regions (fig. 3 c). The lower component was continuous with the inflow portion of the left ventricular cavity, which was markedly hypoplastic. The upper component was represented by a segment of the left ventricle which was only a few millimeters in length and width. The latter segment of the left ventricle communicated with the aortic valve above (figs. 3 d and 4). The wall of the proximal compartment of the left ventricle was markedly hypertrophied.

Three hypoplastic aortic valvular cusps could be identified as guarding a hypoplastic orifice. The coronary arteries arose in normal fashion from the aorta (fig. 5).

The right ventricle was dilated and hypertrophied. The left pulmonary veins joined the left atrium, while the two right pulmonary veins joined the posterior aspect of the right atrium directly.

Comment
It is evident from the foregoing that anomalous attachment of the anterior leaflet of the mitral valve to the ventricular septum was responsible for separation of the left ventricular outlet into two compartments. When obstruction with preservation of a channel is present in this region, the term subaortic stenosis is applied. Consequently, we considered it appropriate to refer to the condition found in our case as subaortic atresia.

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Figure 3

The heart in the case reported. (a) Right atrium and right ventricle. Two atrial septal defects are present. One is located at the fossa ovalis (F. O.), the other (D.) is in the lowermost portion of the atrial septum and has the characteristics of the atrial septal defect in persistent common atroventricular canal. Upper and lower limbs of the fossa ovalis absent. Right pulmonary veins (R. P. V.) connect anomalously with the right atrium. At the expected location of the ostium of the coronary sinus a cardiac vein joins the right atrium. Right ventricle (R. V.) hypertrophied.

(b) Left atrium and left ventricle. Left atrium (L. A.) dilated and hypertrophied. Defect (D) of the atrial septum in position of defect in persistent atroventricular canal. Cleft of the
Figure 4

Apresia of subaortic region caused by adherence of the anterior leaflet of the mitral valve to the ventricular septum which was observed in our case.

The two subaortic compartments in the present case represent a phenomenon different from the absolute closure of the left ventricular outflow tract which may be associated

Figure 5

Aortic orifice viewed from above. The hypoplastic aortic cusps (A. C.) are identified. Through the orifice of the aortic valve, that part of the mitral valve (A. M.) which crosses the outflow tract of the left ventricle is visible.

with aortic valvular atresia. In aortic valvular atresia with intact ventricular septum a column of myocardial tissue may be found lying beneath the valvular tissue. As this is immediately subjacent to the atretic valve, the left ventricular outlet is not subdivided into two compartments, and it is appropriate to continue to use the usual designation of aortic atresia or aortic valvular atresia in such a situation.

Stenosis of the subaortic region of the left ventricle resulting from an anomalous mitral valve is not unique, having been reported by several others and from our laboratory

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*Anterior leaflet of the mitral valve. Anterior portion of the anterior leaflet (A. A. M.) adherent to the outflow tract of the left ventricle (L. V.). P. A. M. = posterior portion of anterior mitral leaflet.*

(c) Section through the heart showing left ventricle (L. V.) and its outflow tract, ventricular septum (V. S.) and right ventricle (R. V.). Anterior portion of the anterior leaflet of the mitral valve adherent (Adh.) to wall of outflow tract of the left ventricle, causing subaortic atresia. A. = ascending aorta; L. A. = left atrium.

(d) Low-power photomicrograph of section made in the same plane as that made to expose the heart shown in c. Segment of the aortic valve is visible at the origin of the ascending aorta (A.). The anterior half of the left anterior leaflet of the mitral valve (A. A. M.) is adherent to the ventricular septum (V. S.) immediately beneath the aortic valve. This causes an absolute obstruction between the lower portion (L. V.) and the subaortic region of the left ventricle. A defect exists in the ventricular septum through which the left ventricle communicates with the right ventricle (R. V.). The posterior half of the left anterior mitral leaflet (P. A. M.) is also apparent. Elastic tissue stain; reduced from original magnification of 5.
by Sellers and associates. A variety of mitral valvular lesions may be responsible for subaortic stenosis. There is, however, a consistent association between the mitral valvular lesion of persistent common atrioventricular canal (so-called endocardial cushion defect) and actual or potential subaortic stenosis. In a current study on obstruction of the left ventricular outflow tract in persistent common atrioventricular canal, we noted a variation in degree of subaortic obstruction which was dependent upon (1) the degree of deficiency in the ventricular tissue, (2) the matter of focal hypertrophy in septal muscle bulging into the tract and (3) encroachment by mitral valvular tissue. The case herein reported represented the most severe degree of subaortic obstruction in the form of atresia. The basis for the obstruction was fusion of the free edge of the anterior mitral leaflet to the septal wall of the left ventricular outflow tract.

An additional feature of interest in our case was the presence of many features of the syndrome of multiple spleens. These included bilateral superior venae cavae in which the left joined the left atrium, anomalous pulmonary venous connection to the right atrium, and an abnormality of the atrial septum characterized by absence of the upper and lower limbi of the fossa ovalis. Extracardiac findings included the bilateral presence of two-lobed lungs and partial situs inversus of the abdominal viscera. The frequent association of multiple spleens and continuity of the inferior vena cava with the azygos venous system was, however, not present.

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References

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