Case Report Section

Congenital Aortic Stenosis, Coarctation of the Aorta and Patent Ductus Arteriosus: Report of Two Cases*

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Aortic stenosis combined with coarctation of the aorta has been reported by a small number of writers. In 1955 Smith and Matthews1 were able to collect 24 autopsied cases from the literature. Marquis and Logan2 have listed three additional patients. The clinical diagnosis has been made in a few others.1, 3, 4 In the majority of instances the aortic stenosis was believed to be an acquired lesion.1 That the combination is more common than these figures would indicate and that the aortic stenosis may frequently be of congenital origin is attested by its presence in eight of 37 individuals reported by one of us5 in whom the stenosis could not be considered to be acquired.

Two patients have been seen who, in addition to congenital aortic stenosis and coarctation of the aorta, had a patent ductus arteriosus. Because the ductus introduces certain further physiologic and therapeutic problems, these cases are being reported.

Case Reports

Case 1: E. M., boy age 14. A diagnosis of congenital heart disease had been made at age three months. When he entered elementary school it was noted that on moderate exertion he would tire and become short of breath more quickly than his contemporaries. His activities had always been restricted by the parents. Occasional headaches and frequent epistaxis had been present. There had been no illness suggestive of the rheumatic state.

On physical examination, he was found to be well developed and nourished. There was no cyanosis. Carotid pulsations were visible at rest and the jugular veins were slightly distended. There was a prominent venous pattern over the anterior thorax. The heart was enlarged to the left and downward. A strong systolic thrill was palpable in the second and third right interspaces and over the neck vessels. A feeble systolic thrill was felt posteriorly to the right of the second and third thoracic vertebrae. The cardiac rate was 92/minute and there was a normal sinus rhythm. The sounds were normal. A loud harsh systolic murmur was best heard in the second and third interspaces to the right of the sternum and was transmitted over the entire thorax and to the neck. The right radial pulse was stronger than the left. No pulsation was felt over the arteries of the legs. Blood pressure in the right arm was 112/98; in the left, 102/90. No reading could be obtained in the lower extremities.

On the basis of these findings, diagnoses of aortic stenosis and coarctation of the aorta were made. He was admitted for study.

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Laboratory Data—The electrocardiogram showed left bundle branch block and left ventricular hypertrophy.

On films and fluoroscopy the pulmonary vascular markings were normal. The main pulmonary artery and its right and left branches were normal. In the postero-anterior view the ascending aorta could not be defined. The heart was enlarged 2 plus in mass with rounding of the left border. In the left anterior oblique view the ascending aorta was dilated 2 plus. The data obtained by right heart catheterization are presented in the Table.

Hospital Course—From the catheterization there was evidence of a left to right shunt at the level of the great vessels. This was believed to indicate the presence of a patent ductus arteriosus. The pressure in the right ventricle, pulmonary artery and pulmonary venous capillary bed was elevated. The brachial artery tracing was abnormal, there being a delay to the peak of systole and a notch on the aortic limb near the summit. The dicrotic incisura was definite.

Thoracic aortography was performed to delineate the coarctation believed to be present. The catheter could not be guided into the ascending aorta. Fifteen cc's of 70 per cent diodrast were injected in the region of the origin of the left subclavian artery. A short area of coarctation was demonstrated.

He was operated upon by Dr. Charles P. Bailey. When the isthmus of the aorta was dissected free, it was found that the patent ductus measured 0.5 cm. in diameter. It joined the aorta in the area of coarctation. There was a diffuse systolic thrill over the pulmonary artery and ascending aorta. The ductus was divided in the usual fashion. The coarcted segment of the aorta was then resected and an end-to-end anastomosis performed. The thrill over the pulmonary artery could no longer be felt. Through an incision in the myocardium of the left ventricle the aortic dilator was then introduced and the head guided to the region of the valve. The expanding arms were cautiously opened and the stenosed valve dilated. The thrill over the ascending aorta was now definitely less prominent.

Postoperatively the systolic murmur in the second and third interspaces was decreased in intensity and the thrill less marked. There was a faint early diastolic murmur in the same area. No other sign of aortic insufficiency was found.

Case 2: M. B., girl age 6. A diagnosis of aortic stenosis was made at the age of two years. Her exercise tolerance had always been limited and she would become short of breath on moderate exertion. Profuse perspiration had been noted. Headache was a frequent complaint and she occasionally became dizzy after exertion. Left chest pain had been present on two occasions. The circumstances were not recalled. During the two months prior to admission she frequently said that her legs had "gone to sleep."

On physical examination she was well developed and nourished. The bony thorax was asymmetrical, the left anterior thorax being more prominent than the right. The heart was perhaps slightly enlarged to the left on percussion. There was a systolic thrill in the second and third right interspaces which could be felt over the neck vessels. The cardiac rate was 84/minute and there was a normal sinus rhythm. The second sound at the base to the right of the sternum was faint. A loud harsh systolic murmur was best heard in the second and third right interspaces and was transmitted over the anterior chest and to the neck. A systolic murmur of different character, softer and blowing, was heard over the posterior thorax. Femoral pulsations were weak. Blood pressure was 122/60 in the right arm, 106/80 in the left arm. It could not be determined in the legs.

Laboratory Data—The electrocardiogram showed left ventricular hypertrophy. On films and fluoroscopy the pulmonary vascular markings were accentuated 2 plus. The

TABLE I
CATHETERIZATION DATA

<table>
<thead>
<tr>
<th>O₂ Content (Vol. %) (Average of Three Samples)</th>
<th>Arterial Saturation</th>
<th>Pressure (Mm. Hg.)</th>
</tr>
</thead>
<tbody>
<tr>
<td>VI</td>
<td>RA</td>
<td>RV</td>
</tr>
<tr>
<td>---</td>
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<td>----</td>
</tr>
<tr>
<td>E. M.</td>
<td>13.2</td>
<td>12.7</td>
</tr>
<tr>
<td>M. B.</td>
<td>10.4</td>
<td>9.9</td>
</tr>
</tbody>
</table>

Key: VI—venous inflow. RA—right atrium. RV—right ventricle. PA—pulmonary artery. PVC—pulmonary venous capillary. BA—brachial artery.

Figures in parentheses indicate mean pressure.
main pulmonary artery was dilated 2 plus as was its right branch. The left branch could not be defined. The ascending aorta was dilated and the knob prominent. The heart was enlarged 2 plus to the left with lengthening of the left border. In the left anterior oblique view there was 2 plus posterior prominence of the cardiac shadow. The data obtained by right heart catheterization are presented in the Table.

Hospital Course—From the catheterization there was evidence of a left to right shunt at the level of the great vessels. The pressure in the right ventricle, pulmonary artery and pulmonary venous capillary bed was elevated. The brachial artery tracing showed a slow rise to the peak of systole.

Thoracic aortography was performed to define the extent of aortic coarctation. It was found to be confined to a short segment just distal to the left subclavian artery. There was a suggestion of simultaneous opacification of aorta and pulmonary artery.

The child was operated upon by Dr. Charles P. Bailey. Over the ascending aorta and pulmonary artery a systolic thrill was felt. The ductus was found to be 1 cm. by 1 cm. and to join the aorta in the area of coarctation. It was divided and the narrowed area of the aorta resected. Aortic valve dilatation was then carried out via the transventricular route. No thrill could now be felt over the pulmonary artery and that over the ascending aorta was softer and more diffuse.

The postoperative course was uneventful. The murmur and thrill in the second and third right interspaces were now much less prominent. The posterior systolic murmur could no longer be heard.

Discussion

This complex of anomalies is unusual in that for each malformation a satisfactory surgical procedure exists. The recognition of the presence of all of the components and of their relative severity is necessary.

The murmur and thrill of aortic stenosis were unmistakable in both of these patients. Nothing was heard which would suggest a patent ductus arteriosus. Knowledge of its existence required catheterization. The character of the peripheral pulses was of importance in the diagnosis of coarctation. Femoral pulsations were weak. Although the blood pressure in the arms was not elevated, that in the legs could not be measured. The aortic stenosis, interfering with left ventricular output at a proximal level, discouraged the development of the hypertension in the aortic arch and its vessels which would be present in uncomplicated coarctation.

Assessment of the relative importance of the defects perforatively depended upon consideration of the electrocardiogram and the catheterization data. In both patients, there was a pattern of left ventricular hypertrophy. Each of the lesions may cause this. It was obvious that the coarctation did not contribute in these patients because of the lack of hypertension in the arch. In other words, under the circumstances, the coarctation demanded no extra work on the part of the left ventricle. The specific effect of the ductus could not be determined. Flow could have been greater in the past and might have required ventricular hypertrophy. Pulmonary vascular changes taking place over a period of time would then have decreased the shunt. However, the observations in regard to the relative importance of the stenosis and the coarctation indicated that the stenosis must be more important than the ductus.

It followed that relief of the aortic stenosis must be the prime consideration in therapy. If the coarctation were to be resected and the ductus interrupted without relieving the stenosis, the burden on the left ventricle would remain. It was clear, too, that were the stenosis alone attacked there would still be a major and a minor cause of left ventricular over-
Atrioventricular Nodal Rhythm with Antegrade Block

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Atrioventricular nodal rhythm is an infrequent, electrocardiographic finding in which the pace-maker has shifted from the sinus node to the atrioventricular node. The characteristic electrocardiographic findings of this rhythm are as follows:¹-⁶

1. In the limb leads the P waves are inverted in II, III, and aVF, and are upright in aVR, aVL and I (small amplitude).
2. It is usually associated with bradycardia.
3. The PR interval may vary from 0.12 second to negative value, i.e., an RP interval.

Nodal rhythms are classified as upper, middle, and lower nodal rhythm according to the positions of the retrograde P wave to the QRS wave. This classification is based on the assumption that the impulse originates from a nodal focus and spreads at normal speed in upper and lower directions. If there is some conduction disturbance of the nodal impulse on its way to the auricle (retrograde block) or ventricle (antegrade block), this classification can not be used.¹ A PR interval of more than 0.12 second can be observed in cases of atrioventricular nodal rhythm associated with antegrade block.¹ ³ ⁶ Because of its rarity, we are reporting an atrioventricular nodal rhythm with antegrade block observed during cardiac surgery and later spontaneously in the post-operative period, in which the auricular conduction was delayed as long as 0.25 second.

Case Report: J. B., a 56 year old white man, was admitted to the City of Hope Medical Center on October 22, 1956. He had developed shortness of breath on exertion

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REFERENCES