Pulmonary Artery Stenosis

By Margaret B. Vermillion, M.D., Leonard Leight, M.D., and Lawrence A. Davis, M.D.

Although the number of cases of stenosis of a branch or branches of the pulmonary artery reported in the literature is small, this congenital lesion is being recognized with increasing frequency. This report is a description of 2 cases seen within 1 year. Both cases were studied by means of cardiac catheterization and angiocardiography. In both there were complicating congenital heart lesions.

Since the description by Möller in 1953 of stenosis of peripheral pulmonary arteries, this congenital lesion has been recognized with increasing frequency. The lesion is of interest both from a clinical and physiologic standpoint. From the latter standpoint, there is sufficient documentation to warrant the inclusion of this congenital anomaly as one of the causes of right ventricular hypertension. From a clinical standpoint, pulmonary artery stenosis appears in some instances to be an additional cause of a continuous murmur heard over the thorax.

We have recently seen 2 cases studied both by cardiac catheterization and by selective angiocardiography that are examples of stenosis of a peripheral pulmonary artery. The first case is of interest in that a continuous murmur had been heard over this patient's thorax for some years. The second case is an example of stenosis of a peripheral pulmonary artery demonstrated both by cardiac catheterization and angiocardiography.

Case Reports

Case 1. A 10-year-old white girl was admitted to the hospital on July 9, 1956. She and her identical twin were the product of a 7 months' pregnancy. Each twin weighed 3 pounds at birth. The patient was not known to be cyanotic during her 2-month stay in the premature nursery, but on discharge from the hospital the mother was informed that the patient had a heart lesion. The patient gained weight slowly but was able to play, run, and ride a bicycle without difficulty. No cyanosis had ever been noted. At the time of admission, the patient weighed 10 pounds less and was 2 inches shorter than her twin, who had no known congenital anomaly. The remainder of the history was noncontributory.

Physical examination revealed a frail child appearing younger than the stated age of 10 years. There was a suggestion of cyanosis of the lips and nailbeds. Weight was 45 pounds, height 52 inches, respiratory 16 per minute, cardiac rate 108 per minute, and blood pressure 110/80 mm. Hg in both arms. The positive physical findings were limited to the thorax. The point of maximum impulse was located 1.5 cm. to the left of the midclavicular line. No thrills were palpable. Regular sinus rhythm was present. A continuous murmur of grade II to III intensity was audible at the second right intercostal space, 4 cm. from the sternal border. The continuous murmur changed little in character throughout the cardiac cycle and was not modified by respiration or change in body position. A grade III, harsh, blowing, systolic murmur was heard along the left sternal border. The second sound at the pulmonic area was accentuated.

Red blood cell count was 4.6 million and the hemoglobin was 12.5 Gm. per 100 ml. Electrocardiogram revealed right ventricular hypertrophy. X-ray of the chest and cardiac fluoroscopy revealed a heart normal in size in its transverse diameter. There was marked prominence of the main pulmonary artery segment. The hilar shadows appeared accentuated and showed brisk pulsation. The peripheral pulmonary vascularity was interpreted as being within normal limits. The aorta descended on the left. The left atrium was not enlarged.

Cardiac catheterization was performed in the usual fashion on July 10, 1956, under rectal Avertin anesthesia. The findings are summarized in table 1. During catheterization, the catheter entered the aorta from the right ventricle. In addition, the catheter appeared to enter an anomalous pulmonary vein draining into the right atrium.

The data were interpreted as indicating the presence of a high interventricular septal defect with considerable pulmonary hypertension and possibly an anomalous pulmonary vein draining into the right atrium; however, no left-to-right shunt at the level of the atrium was demonstrated.

Immediately after pressure recordings and blood sampling were completed, the catheter was reinserted into the right ventricle. Twenty-two milliliters of 70 per cent sodium acetrizoate (Urokon)
TABLE 1.—Case 1, Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Pressure Location</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary artery</td>
<td>102/59 (80)</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>115/4/7</td>
</tr>
<tr>
<td>Right atrium, mean</td>
<td>2</td>
</tr>
<tr>
<td>Aorta</td>
<td>100/61 (84)</td>
</tr>
</tbody>
</table>

Blood oxygen content, volume per cent
- Inferior vena cava: 13.91%
- Superior vena cava: 12.76%
- Right atrium, near superior vena cava: 13.21%
- Right atrium, mid: 13.10%
- Right ventricle, mid: 13.88%
- Pulmonary artery, left: 14.87%
- Aorta: 17.64%
- Blood oxygen capacity: 19.22%
- Arterial oxygen saturation: 91.78%

**Fig. 1.** Case 1. Angiocardiogram demonstrating stenosis of a branch of the pulmonary artery with poststenotic dilatation. Catheter in right ventricle.

were injected under pressure through the catheter with film exposure beginning automatically at the time of injection. By means of an Elema bi-plane angiocardiogram x-ray apparatus, 6 films per second were obtained simultaneously in the anteroposterior and lateral positions for a total of 4 seconds. The opaque material was seen entering the right ventricle and passing through a high septal defect into the left ventricle and then into the aorta. The aorta appeared to be normally located with no overriding of the interventricular septum. The main pulmonary arteries appeared enlarged. An unusual stenosis of one of the superior branches of the right pulmonary artery with a poststenotic dilatation was seen (fig. 1). No other areas of narrowing of the pulmonary vasculature were demonstrated. The dye returned normally to a moderately enlarged left atrium.

**Case 2.** A 19-month-old white boy was admitted to the hospital on March 23, 1957. He was the product of a 7 months' pregnancy and weighed 3 pounds, 6 ounces at birth. The presence of a heart lesion was noted soon after birth. The patient failed to gain weight normally and tired easily while playing. No cyanosis had been noted by the parents.

Physical examination revealed a small baby, who weighed 173⁄4 pounds and was 32 inches tall. Respirations were 28 per minute, cardiac rate was 88 per minute, and blood pressure was 95/65 mm. Hg. The positive physical findings were limited to the thorax. The point of maximal impulse was in the fifth intercostal space at the midclavicular line. A grade III to IV harsh, blowing, systolic murmur was audible over the entire precordium but was loudest at the second to fourth intercostal space to the left of the sternum, and a thrill was palpable here. The second pulmonary sound was grossly

**Table 2.—Case 2, Cardiac Catheterization Data**

<table>
<thead>
<tr>
<th>Pressure Location</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary artery, right peripheral branch: 20/8</td>
<td></td>
</tr>
<tr>
<td>Pulmonary artery, main: 36/8</td>
<td></td>
</tr>
<tr>
<td>Right ventricle: 59/2/6</td>
<td></td>
</tr>
<tr>
<td>Right atrium, mean: 2</td>
<td></td>
</tr>
</tbody>
</table>

Blood oxygen content, volume per cent
- Inferior vena cava: 7.84%
- Superior vena cava: 8.54%
- Right atrium, near superior vena cava: 9.42%
- Right atrium, mid: 9.46%
- Right ventricle, near tricuspid valve: 9.42%
- Right ventricle, mid: 10.21%
- Right ventricle, near pulmonary valve: 9.46%
- Pulmonary artery, right: 9.63%
- Pulmonary artery, main: 9.59%
- Pulmonary vein, right: 12.48%
- Femoral artery: 11.12%
- Blood oxygen capacity: 12.59%
- Arterial oxygen saturation: 86.68%

**Fig. 2.** Case 2. Pressures obtained on withdrawing catheter from peripheral branch of right pulmonary artery to right ventricle.

diminished. There was no cyanosis and the rhythm was regular. X-ray of the chest revealed an enlarged heart with no unusual or diagnostic configuration. The lung fields showed questionable overvascularization. The fluoroscopist reported enlargement of the left atrium.

The red blood cell count was 4 million, and the hemoglobin was 12.9 Gm. per 100 ml. The electrocardiogram revealed right ventricular hypertrophy.

Cardiac catheterization was performed in the usual fashion on March 25, 1957, under thiopental (Pentothal) and meperidine (Demerol) anesthesia. The findings are summarized in table 2. During catheterization the catheter entered a pulmonary vein on 2 occasions apparently from the right atrium; however, as in the previous case, our failure to demonstrate a left-to-right shunt at the level of the atrium casts doubt as to whether anomalous pulmonary venous drainage was truly present. The catheter was inserted into the right ventricle and then into a pulmonary artery, finally entering a peripheral branch of the right pulmonary artery. The pressures obtained in pulling back from the peripheral branch of the right pulmonary artery to the right ventricle just proximal to the pulmonary valve are illustrated in figure 2.

The location of the catheter at the time pressures were obtained from a peripheral branch of the right pulmonary artery and the main pulmonary artery is illustrated in figures 3 and 4 respectively. The pressure in a peripheral branch of the right pulmonary artery was 20/8 mm. Hg (figs. 2 and 3). As the catheter was pulled out to the main pulmonary artery, there was a sudden rise in pressure to 36/8 mm. Hg (figs. 2 and 4). When the catheter was then pulled back to the right ventricle, there was a further rise in systolic pressure that appeared to begin sharply at the pulmonary valve, the right ventricular pressure just proximal to the valve being 58/2/6 (fig. 2). There was no further rise in right ventricular pressure in other areas of the right ventricle. These pressure relationships were demonstrated several times.

The data were interpreted as indicating both a stenosis of a peripheral branch of the pulmonary artery and a valvular pulmonary stenosis. A mild pulmonary hypertension was present proximal to the peripheral pulmonary stenosis. In addition, the possibility of an anomalous pulmonary vein draining into the right atrium could not be excluded.

Immediately after pressure recordings and blood sampling were completed, the catheter was again placed in the right ventricle. Ten milliliters of 70 per cent sodium acetazolamide (Urokon) were injected under pressure through the catheter with film exposure beginning automatically at the time of injection. Again by means of the Elema bi-plane angiocardiogram apparatus, 6 films per second were obtained simultaneously in the antero-posterior and lateral projections for a total of 4 seconds. Soon after injection of the dye, the catheter was whipped from the right ventricle into the right atrium and most of the dye was injected into the right atrium. A representative film of the angiocardiographic study is illustrated in figure 5. No right-to-left shunt was demonstrated at angiocardiography. A constriction
of the right main pulmonary artery just distal to the bifurcation can be seen.

**DISCUSSION**

Stenosis of peripheral branches of the pulmonary artery was first described by Möller in 1953. This author presented a patient with a continuous murmur over the aortic area, electrocardiographic evidence of right ventricular hypertrophy, and cardiac enlargement with prominence of the pulmonary conus on roentgenographic examination. Cardiac catheterization revealed only hypertension of the pulmonary artery and of the right ventricle. Arterial oxygen saturation was normal. Selective angiocardiography of the pulmonary artery showed multiple constrictions with poststenotic dilatations. In 1955 Powell and Hiller described a 5-year-old patient with a history of tiredness, failure to gain weight, and dyspnea on exertion. A continuous murmur was heard over the pulmonic area. On angiocardiography a narrowing of both the right and left pulmonary arteries immediately distal to the bifurcation of the main pulmonary artery was found. Dilatation of the main pulmonary artery proximal to the narrowing was seen. Of the 4 cases of peripheral pulmonary stenosis and associated pulmonary hypertension reported in 1955 by Arvidsson and associates, 1 patient had a continuous murmur, which was heard over the entire chest. A selective angiocardiogram of the pulmonary artery of this patient revealed a dilated pulmonary artery with marked narrowing of short areas of the lumina of the lobar arteries. The other 3 patients, who had similar angiocardiographic findings, had a coarse murmur widely transmitted over the back and precordium that was audible, however, only during systole.

Thus it would seem that stenosis of a peripheral branch or branches of the pulmonary artery should be included as one of the causes of a continuous murmur heard over the precordium and we believe our case 1 is another example of the association of this defect and the accompanying murmur. In this case, the stenosis of the pulmonary artery played a questionable part in the pulmonary hemodynamics in view of the demonstration of a high interventricular septal defect.

Since the original description of stenosis of branches of the pulmonary artery, several other reports in addition to those enumerated above have appeared. In 1953, Schumacher and Lurie referred to a 14-year-old patient with cyanosis, clubbing, physical incapacity, and lethargy. At thoracotomy, the main pulmonary artery was described as approximately normal in size or a little smaller than normal. About 2 cm. distal to its origin at approximately the region of the bifurcation, the artery became very small and a calcified stenotic lesion was found in this area.

Søndergaard in 1954, reported 3 cases of peripheral pulmonary stenosis. All of these patients had associated lesions, 2 suggestive of tetralogy of Fallot and 1 of an atrial septal defect.

Gyllenswärd and co-workers have recently reported 8 cases of the anomaly under discussion, in 1 of which the presence of a stenosis of a peripheral branch of the pulmonary artery was demonstrated at the time of cardiac catheterization by means of pull-out pressures. Coles and Walker have also described a case in which pull-out curves demonstrated the presence of pulmonary artery stenosis. We believe
PULMONARY ARTERY STENOSIS

our second case is another example of this lesion demonstrated at the time of catheterization. Our case apparently has an associated valvular pulmonic stenosis. There appears to be a mild pulmonary hypertension present proximal to the stenotic pulmonary artery.

Whether or not significant right ventricular hypertension will result from stenosis of the pulmonary artery undoubtedly depends on the degree and location of the stenosis and the number of branches involved. Although we are unaware of the description of this lesion in adults, there appears to be no reason why in some cases this abnormality should not be compatible with a relatively long life span. From the small number of cases thus far reported, it is apparent that many of the cases of stenosis of the peripheral pulmonary artery have other congenital cardiac defects. This was so in both of our cases.

SUMMARY

Two examples of stenosis of a branch of the pulmonary artery are presented. Both patients were studied by means of cardiac catheterization and angiography. One case presented with a continuous precordial murmur and the presence of the stenosis and poststenotic dilatation was demonstrated by means of angiography. The second case is another report of the demonstration at the time of cardiac catheterization of stenosis of the pulmonary artery. Both patients had associated cardiac anomalies, the first case a high interventricular septal defect and the second case a valvular pulmonic stenosis.

SUMMARIO IN INTERLINGUA

Es presentate duo exampli de stenose de un branca del arteria pulmonar. Ambae le patientes esseva studiate per medio de catheterisation cardiac e angiocardiographia. Le prime veniva al consulta con un continue murmure precordial, e le presentia del stenose e de dilatation poststenotic esseva demonstrate per medios angiocardiographic. Le secunde representava un exemplo addicional del demonstration de stenose del arteria pulmonar al tempore de catheterisation cardiac. Ambae le patientes habeva associate anormalitates cardiac: le prime un defecto del septo alto-interventricular, le secunde un stenose del valvula pulmonar.

REFERENCES


Cerebral symptoms, of a remarkable character, are commonly present in this disease. These consist in the occurrence of repeated pseudo-apoplectic attacks, of various degrees of intensity and duration. They are seldom followed by paralysis. Attacks of vertigo, dimness of vision, and syncope, are observed.—William Stokes. The Diseases of the Heart and the Aorta. Dublin, 1854.
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