Stenosis of a Branch of the Pulmonary Artery
An Additional Cause of Continuous Murmurs over the Chest

By Frederic Eldridge, M.D., Arthur Selzer, M.D., and Herbert Hultgren, M.D.

Three patients have been demonstrated by cardiac catheterization to have stenosis of a main branch of the pulmonary artery. Two exhibited continuous ductus-like murmurs at the chest wall over the site of stenosis. Demonstration of a pressure gradient throughout the cardiac cycle at the site of stenosis in these 2 patients and the experimental production in the dog of a continuous murmur at the site of partial constriction of a pulmonary artery prove that the murmur is due to the stenosis. This anomaly should be added to the differential diagnosis of continuous murmurs over the chest.

STENOSIS of a main branch of the pulmonary artery has been previously considered very rare. Bredt and Mönckeberg believed that it never occurred independently of stenosis of the pulmonary orifice. Schwalbe in his comprehensive survey of malformations referred to 4 reported instances, but a review of the original reports reveals only 2 in which narrowing of a branch of an otherwise normal pulmonary artery was present, the other 2 being patients with absence of either the right or left pulmonary artery. In 1938 Oppenheimer reported a stricture of the right pulmonary artery and of the lower branch of the left pulmonary artery in a 17-month-old infant. Four patients with stenosis at the area of the bifurcation of the pulmonary artery and insertion of the ligamentum arteriosum have been discovered at operation by Schumaker and Lurie and Søndergaard. Kjellberg and his associates mention having seen a case of supravalvular stricture of the main pulmonary artery. Arvidsson and his associates recently reported 4 patients with pulmonary hypertension in whom multiple stenoses of the main branches of the pulmonary artery were demonstrated by selective angiography. In 1 of these patients a continuous murmur was present over the upper chest.

In this communication 3 additional patients with stenotic lesions of the main branches of the pulmonary artery are described including 2 patients manifesting continuous murmurs. Experimental evidence is also presented that a continuous murmur can be produced by constriction of the main branches of the pulmonary artery.

CASE REPORTS

Case 1. G.T., a 7-year-old Negro girl, entered the Stanford University Hospital in February 1956 with the history of a heart murmur and congenital absence of the right radius having been noted at the age of 1 year. She had been asymptomatic except for several episodes of pneumonitis.

She was an alert, active girl. Her brachial blood pressure was 94/66 mm. Hg. There was shortening and radial deviation of the right forearm. The lungs were clear. The heart was regular. At the pulmonic area the second sound was loud and slightly split. There was a grade III murmur filling systole over the left precordium, and a loud continuous murmur of grade IV intensity was heard at the right second intercostal space just medial to the midediavicular line. This murmur was well transmitted to the right axilla and back.

The urinalysis and blood count were not remarkable. The electrocardiogram showed high voltage R waves over the right precordium, compatible with right ventricular hypertrophy. X-ray pictures showed marked pulmonary plethora, right ventricular enlargement, and widening of the superior mediastinum. A phonocardiogram (fig. 1) demonstrated a continuous murmur over the second intercostal space, just medial to the right midclavicular line and a loud, slightly split second sound in the pulmonic area. The continuous murmur was essentially similar to the machinery murmur produced by the usual uncomplicated patent ductus. The murmur appeared about 0.06 second after the peak of the apical first sound, increased to maximal intensity just before or at the time of the second sound, and faded away in diastole. The diastolic component

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Fig. 1. Case 1. The top tracing shows pressures obtained during withdrawal of the cardiac catheter from right lung. The pressure in the distal right pulmonary artery shows a systolic dip due to the Venturi effect. Next is a sharp rise in pressure as the catheter tip is withdrawn to the proximal right pulmonary artery, and then a further sharp rise of pressure in the main pulmonary artery. Both sharp rises of pressure indicate sites of stenosis. The lower 2 tracings are phonocardiograms showing the continuous murmur heard in the right second intercostal space and the heart sounds as heard in the pulmonic area. The first and second heart sounds are shown on both tracings by the numbers 1 and 2.

appeared to diminish in intensity readily during expiration.

Cardiac catheterization revealed a high oxygen saturation in the superior vena cava (86 per cent) and the right atrial oxygen saturation was higher than that in the inferior vena cava. In addition, there were arterial anoxemia, moderate pulmonary hypertension, and at least 2 sites of stenosis in the right main pulmonary artery (table 1 and fig. 1). The pressure tracing recorded during the withdrawal of the catheter from the wedged position revealed first a typical, normal, wedged-pressure tracing (fig. 1, far left), then an abrupt change in the pressure curve with a distinct dip in pressure during systole and then an abrupt increase in pressure with a positive pressure pulse during systole, and finally a second abrupt rise in pressure. The systolic dip in the pressure trace is probably due to the blood flow through the constricted vessel producing a Venturi effect, such as is occasionally seen when the catheter tip is at the stenotic orifice in valvular pulmonic stenosis. This portion of the tracing and the 2 abrupt rises in pressure in the pulmonary artery conclusively establish the presence of 2 areas of stenosis in the right branch of the pulmonary artery.

Venous angiography subsequently confirmed the presence of a total anomalous pulmonary venous return to the right atrium by means of a persistent left superior vena cava, and demonstrated a moderate dilatation of the right pulmonary artery. The first portion of the right pulmonary artery was poorly seen on the film because it was located over the spine. No distinct area of stenosis could be seen.

Comment. This patient had multiple congenital anomalies. Two sites of stenosis of the pulmonary artery were demonstrated by catheterization. The continuous murmur was located directly over the point where the stenosis of the right main pulmonary artery was demonstrated. The quality of the murmur was quite unlike that of a venous hum, which has been occasionally described over the area of insertion of anomalous pulmonary veins into the right atrium or venae cavae. The rise in the intensity of the murmur a short interval after the first heart sound and the decrease in intensity during diastole correspond to the phasic
pressure changes recorded from the pulmonary artery at the point of the demonstrated stenosis.

**Case 2.** R.W., a 7-year-old boy, was first seen at the Stanford University Hospital at the age of 6 months, when he was noted to have a thrill at the fourth intercostal space along the left sternal border and a loud systolic murmur in the same area. At the pulmonic area a systolic murmur of grade III intensity was followed by a short early diastolic murmur. While not a truly continuous murmur, several observers believed it so on the basis of auscultation alone, and a diagnosis of a probable patent ductus arteriosus was made. At the age of 2 years a thoracotomy was performed at this hospital. The surgeon discovered a small ligamentum arteriosum and ligated it. There were no changes in the murmurs following this surgery.

The patient remained asymptomatic and again entered the hospital at the age of 7½ years for further studies. He appeared well. His brachial blood pressure was 98/60 mm. Hg. There was a systolic thrill and a harsh grade IV systolic murmur loudest at the left sternal border in the fourth intercostal space. In the pulmonic area a grade III systolic murmur was immediately followed by a grade III diastolic murmur of diminishing intensity during diastole.

Routine laboratory studies were normal. The electrocardiogram revealed somewhat high QRS voltage over the precordium but was otherwise normal for a child of 7 years. X-rays showed somewhat prominent right and left ventricles and a large main pulmonary artery. The pulmonary vasculature was prominent. A phonocardiogram (fig. 2) revealed a loud crescendo systolic murmur compatible with a shunt through an interventricular septal defect. At the pulmonic area a systolic ejection murmur with its maximum intensity in midsystole was recorded. It was followed by a murmur of lesser intensity continuing into diastole.

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**Table 1.—Catheterization Findings in Case 1**

<table>
<thead>
<tr>
<th>Artery</th>
<th>Oxygen content, ml./100 ml.</th>
<th>Pressure, mm. Hg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left axillary vein</td>
<td>16.0</td>
<td></td>
</tr>
<tr>
<td>Superior vena cava*</td>
<td>15.7</td>
<td></td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>11.5</td>
<td></td>
</tr>
<tr>
<td>Right atrium*</td>
<td>13.5</td>
<td>9 (mean)</td>
</tr>
<tr>
<td>Right ventricle*</td>
<td>13.6</td>
<td>82/11</td>
</tr>
<tr>
<td>Main pulmonary artery*</td>
<td>13.6</td>
<td>82/20</td>
</tr>
<tr>
<td>Left pulmonary artery</td>
<td>13.9</td>
<td>80/20</td>
</tr>
<tr>
<td>Proximal right pulmonary artery</td>
<td>13.8</td>
<td>39/13</td>
</tr>
<tr>
<td>Distal right pulmonary artery</td>
<td>14.0</td>
<td>19/8</td>
</tr>
<tr>
<td>Pulmonary artery wedge</td>
<td>—</td>
<td>14 (mean)</td>
</tr>
<tr>
<td>Femoral artery</td>
<td>13.9</td>
<td></td>
</tr>
<tr>
<td>Arterial oxygen saturation</td>
<td></td>
<td>77%</td>
</tr>
</tbody>
</table>

* These figures represent the average value of 2 or more samples.

**Table 2.—Catheterization Findings in Case 2**

<table>
<thead>
<tr>
<th>Artery</th>
<th>Oxygen content, ml./100 ml.</th>
<th>Pressure, mm. Hg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>13.2</td>
<td></td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>12.5</td>
<td></td>
</tr>
<tr>
<td>Right atrium*</td>
<td>12.0</td>
<td>2 (mean)</td>
</tr>
<tr>
<td>Right ventricle*</td>
<td>13.5</td>
<td>20/3</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>13.9</td>
<td>21/6</td>
</tr>
<tr>
<td>Right pulmonary artery</td>
<td>13.3</td>
<td>21/6</td>
</tr>
<tr>
<td>Left pulmonary artery</td>
<td>—</td>
<td>15/8</td>
</tr>
<tr>
<td>Femoral artery</td>
<td>15.0</td>
<td>115/60</td>
</tr>
<tr>
<td>Arterial oxygen saturation</td>
<td></td>
<td>91%</td>
</tr>
</tbody>
</table>

* These figures represent the average of 2 or more samples.

**Table 3.—Catheterization Findings in Case 3**

<table>
<thead>
<tr>
<th>Artery</th>
<th>Oxygen content, ml./100 ml.</th>
<th>Pressure, mm. Hg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>10.6</td>
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</tr>
<tr>
<td>Inferior vena cava</td>
<td>11.0</td>
<td></td>
</tr>
<tr>
<td>Right atrium*</td>
<td>12.6</td>
<td>7 (mean)</td>
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<tr>
<td>Right ventricle*</td>
<td>12.3</td>
<td>42/8</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>12.6</td>
<td>38/10</td>
</tr>
<tr>
<td>Left pulmonary artery</td>
<td>—</td>
<td>41/9</td>
</tr>
<tr>
<td>Right pulmonary artery</td>
<td>12.2</td>
<td>20/8</td>
</tr>
<tr>
<td>Femoral artery</td>
<td>14.0</td>
<td>94/50</td>
</tr>
<tr>
<td>Arterial oxygen saturation</td>
<td></td>
<td>87%</td>
</tr>
</tbody>
</table>

* These figures represent the average of 2 or more samples.

**Fig. 2.** Case 2. Phonocardiogram taken in the pulmonic area showing the systolic ejection murmur and the diastolic murmur. The location of the heart sounds is shown by the numbers 1 and 2.
Cardiac catheterization was performed and indicated a ventricular septal defect with a small left-to-right shunt (table 2). During the procedure it was noted that the catheter passed in an unusual fashion posteriorly and laterally into the left pulmonary artery and that there was a definite sudden pressure drop in this portion of the left main pulmonary artery (table 2).

Comment. The ventricular septal defect was the principal cardiac lesion in this case. The stenosis of the left pulmonary artery was an incidental finding, and was probably the cause of the systolic and diastolic murmur closely resembling a continuous murmur that had previously led to a surgical exploration for a patent ductus arteriosus. Since the murmur was present before the thoracotomy, it is unlikely that the operation produced the narrowing of the pulmonary artery. It is of interest that pulmonary hypertension was not present.

Case 3. J.C., a 5-year-old boy, was examined in April 1956. A heart murmur had been heard at birth but he had developed normally and had experienced no symptoms except questionable easy fatigability.

He looked well. His brachial blood pressure was 100/60 mm. Hg. There was a slight precordial bulge of the left chest wall parasternally. A thrill and a grade IV systolic murmur were present along the left sternal border in the third and fourth intercostal spaces. The pulmonic second sound was loud.

Routine laboratory studies were not unusual. The electrocardiogram revealed a pattern consistent with incomplete right bundle-branch block, right ventricular hypertrophy, and abnormal P waves. X-ray studies revealed a prominent main pulmonary artery, increased pulmonary vascularity, and enlarged right and left ventricles.

Cardiac catheterization demonstrated an atrial septal defect with a left-to-right shunt (table 3) and mild pulmonary hypertension. In addition, there was a definite drop of systolic pressure in the right pulmonary artery 4 to 5 cm. beyond the bifurcation of the main pulmonary artery (fig. 3) indicating the presence of a stenosis of the right pulmonary artery.

Mild arterial unsaturation was thought to be due to hyperventilation accompanying the tribromoethanol (Avertin) anesthesia.

Comment. The atrial septal defect was the principal lesion in this patient. Since a continuous murmur was not present, there appears to be nothing in the clinical findings that would enable one to make the diagnosis of stenosis of the right pulmonary artery.

Discussion

The exact nature of such stenotic lesions involving the pulmonary artery distal to the valve is obscure because of the rarity of such lesions and the paucity of descriptions of their pathologic anatomy. Partial obstruction by thrombosis or embolus or a congenital developmental defect are the 2 most likely possibilities.

Partial occlusion by thrombosis, secondary to trauma of the chest wall, of the main pulmonary artery in an adult has been described by Dimond and Jones. The resulting clinical picture resembled that of pulmonary stenosis with an intact septum. Pulmonary thrombosis or embolism with partial recanalization could result in single or multiple areas of narrowing of the branches of the pulmonary artery; since resorption of an occluding thrombus or embolus may be variable, complete restitution of the lumen may occur or a residual area of stenosis may remain. For example, Saphir, has concluded that small bands or valvelike structures found occasionally in the pulmonary artery represent vestiges of resorbed mural thrombosis. Partially resorbed multiple thrombi or emboli similar to those illustrated in Möller's paper could easily give the curious rounded or scalloped appearance demonstrated so clearly in the angiograms of one of Arvidsson's pa-
tients\textsuperscript{2} (case 1). However, it seems unlikely that this was the etiology of the stenotic lesions present in our patients, for there was no clinical history compatible with pulmonary thrombosis or embolism. Although such a process could have occurred during intra-uterine development, there are no descriptions in the literature of any such lesions in the fetus or newborn.

A developmental, congenital defect of the pulmonary artery seems more likely. The occurrence in childhood, the association with other congenital cardiac lesions, and the absence of any evidence of an acquired etiologic process all suggest this possibility. Søndergaard\textsuperscript{8} suggested that the cause of stenosis of the pulmonary artery at its bifurcation was due to the incorporation of the pulmonary artery by the tissue of the closing ductus arteriosus (the Skodaic theory). This theory was first suggested by Craigie\textsuperscript{20} and later supported by Skoda to account for the origin of the adult type of coarctation of the aorta. Although one of the patients in the present report (case 2) probably had a single area of stenosis at the bifurcation, such a theory would not explain the stenosis of the more peripheral branch of the right pulmonary artery (case 3) and the multiple stenoses observed in case 1.

The only detailed report of a patient with stenosis of the main branches of the pulmonary artery in which a complete autopsy study was described is by Oppenheimer.\textsuperscript{8} In that 17-month-old child, the right pulmonary artery and the lower branch of the left pulmonary artery were constricted by thickened intimal tissue beneath which were extensive deposits of calcium in the media. The right pulmonary artery at the stenotic area was completely encircled by a ring of calcium. Calcification was also present in the media of the aorta. Five other instances of calcification of the pulmonary artery were described but in none of these patients was there any narrowing of the pulmonary arteries. The author suggested a congenital origin for these lesions. It is possible that a similar process is present in our patients, but a careful review of the x-ray studies revealed no evidence of calcifications either in the pulmonary arteries or aorta.

The question of the proper terminology of such lesions of the pulmonary artery should be considered. Søndergaard\textsuperscript{8} suggested the term "coarctation of the pulmonary artery" because of the resemblance in structure and, as he believed, in origin to the adult variety of aortic coarctation. This term would be desirable if the stenosis were isolated and located near the bifurcation and if the Skodaic theory were the proper explanation for such a lesion. Many of these lesions are multiple, however, and they may be located at various points in the pulmonary artery. Furthermore, the Skodaic theory of the origin of the adult variety of coarctation of the aorta has been attacked for compelling reasons by Edwards and associates,\textsuperscript{21} and the possibility of such a process being the cause of a stenosis of the pulmonary artery is only conjectural. Oppenheimer\textsuperscript{8} referred to the lesions as "partial atresia of the main branches of the pulmonary artery." This is a poor term because atresia means "without a lumen," hence it cannot be partial. Considering the fact that such areas of narrowing may occur at many sites in the pulmonary artery distal to the valve, that they may be multiple, and that their exact origin is still unknown, it might be best to refer to them as "stenoses of the pulmonary artery" specifying the area involved, i.e., "stenosis of the main pulmonary artery, stenosis of the right or left pulmonary artery, etc."

Arvidsson and co-workers\textsuperscript{12} suggested that this anomaly may be the cause of hypertension in the main pulmonary artery. The fact that in 2 of their patients with multiple stenoses no other anomaly was demonstrated suggests that this may be the case; but such a relationship is by no means certain. It should be pointed out that abundant clinical and experimental evidence is available to show that even total occlusion of 1 main pulmonary artery does not increase the resistance enough to elevate pulmonary arterial pressure if the flow is not increased and if the pulmonary vascular bed is normal. Localized narrowing would be even less likely to cause pressure rise per se, unless both branches were involved. Furthermore, in 2 cases of the 3 reported here, in which pulmonary hypertension was present, there existed another congenital defect (anomalous venous return and ven-
tricular septal defect) which in itself may be associated with pulmonary hypertension.\textsuperscript{15}

The physiologic effects of stenosis of a main branch of the pulmonary artery are not readily apparent unless a relationship to pulmonary hypertension can be demonstrated. No measurements of relative blood flow to the lungs have been made, although on theoretic grounds one might expect some changes from the normal.

In one of the reported cases the stenotic area was dilated surgically\textsuperscript{2} and clinical improvement was thought to have taken place. Whether surgical intervention for the sole purpose of dilating a localized stenosis of a pulmonary arterial branch is justified, cannot be answered on the basis of information at hand. One is inclined, however, to lean toward the view that this is a benign, clinically insignificant lesion that would not justify thoracotomy under ordinary circumstances.

Unfortunately, there are no distinctive clinical signs of the presence of stenosis of the main branches of the pulmonary artery, unless a continuous murmur be present. In order to detect such lesions during cardiac catheterization, both branches of the pulmonary artery should be entered and withdrawal tracings recorded from each branch. Since continuous murmurs are produced by this lesion only when the degree of stenosis is severe, one might expect the murmur to be extinguished or suddenly changed in intensity when the catheter is passed through the area of narrowing.

The most apparent significance of stenosis of a main branch of the pulmonary artery lies in its relationship to the origin of continuous murmurs. Continuous, ductus-like murmurs have been observed in 3 cases: in 1 patient of Arvidsson\textsuperscript{23} and in 2 reported here. The location of the continuous murmur over the thorax at the site of the stenosis and the appearance of the phonocardiogram suggest strongly that the murmur originated at the stenosis. It was thought, however, that in order to prove this relationship beyond doubt, an examination of the nature of the pressure gradient across the stenotic areas should be made and an experimental reproduction of such a murmur would have to be accomplished.

Several conditions must be fulfilled for a continuous murmur of this sort to be produced. There must be narrowing of the blood channel and sufficient flow through the constricted area to produce turbulence during both systole and diastole. In order to have flow in diastole of sufficient volume and velocity to produce this turbulence, a reservoir of blood under pressure must exist proximally to the constriction. These conditions are met in the case of patent ductus arteriosus, peripheral and pulmonary arteriovenous fistulas, and in some of the more unusual arteriovenous communications. They are also met in the case of stenosis of a pulmonary artery branch where the proximal pulmonary arterial bed acts as the reservoir under pressure.

The presence of a sufficient volume and velocity of blood flow to produce turbulence can best be demonstrated by showing a pressure gradient between the areas proximal and distal to the narrowed area. Such a gradient can easily be demonstrated, both in systole and diastole, between the aorta and the pulmonary artery in a typical case of patent ductus arteriosus with continuous murmur. Furthermore, it has been shown that when pulmonary hypertension develops, leading to an obliteration of the gradient in diastole or in both systole and diastole, there is disappearance of the diastolic component of the murmur in the first instance and total disappearance of the murmur in the second.\textsuperscript{24} Figure 4 shows the gradients of pressure for 1 cardiac cycle across the areas of stenosis in the 3 patients presented in this report. The pressure gradients were constructed from an analysis of the pressure curves obtained proximally and distally to the area of pressure drop. Significantly, the 2 patients with continuous murmurs showed a gradient throughout the entire cycle, whereas the third patient, in whom only a systolic murmur was observed, showed a pressure gradient only in systole. This is offered as evidence that the continuous murmur heard in 2 of our cases originated at the site and was caused by the stenosis of the pulmonary artery. It also explains why such a murmur is present in some cases only.
Experimental Study

The experimental part of our study was performed in acute experiments on dogs anesthetized with intravenous pentobarbital. A special constricting clamp was placed around a large branch of the pulmonary artery and graded constriction applied. Auscultation of the constricted area revealed only a faint systolic murmur. In the next experiment the constricting clamp was placed around the thoracic aorta and graded constriction applied. One could observe and register phonocardiographically a soft systolic murmur with mild constriction, a loud systolic murmur with more severe constriction, then a continuous murmur, first with a faint diastolic phase, and upon further constriction a loud murmur throughout the cardiac cycle. A similar series of experiments, performed independently, was recently reported by Myers and associates. In the final experiment it was thought that if a branch of the pulmonary artery could be constricted and the flow through it increased, a situation similar to that in the thoracic aorta would arise. In this experiment the clamp was placed around the superior branch of the left pulmonary artery (the main branch in the dog is too short before its bifurcation to place a clamp around it) and graded constriction was applied after completely clamping the right pulmonary artery. In this experiment mild constriction of the left pulmonary artery produced a systolic murmur and more severe constriction produced a continuous murmur similar to that of aortic constriction (fig. 5).

The experimental reproduction of a continuous murmur at the site of constriction of a branch of the pulmonary artery fully confirms the clinical impression that the murmurs observed in 2 of our patients were due to the stenosis of the pulmonary artery.

The fact that stenosis of the left branch of the pulmonary artery may produce a murmur indistinguishable in quality and location from that of a typical case of patent ductus arteriosus is of a considerable practical importance. This has been dramatically demonstrated in our case 2, in whom a thoracotomy was performed under the mistaken diagnosis of patent ductus arteriosus. Presently, surgical treatment of patent ductus arteriosus has reached such a degree of safety that in most quarters all cases are being operated on regardless of whether or not the lesions are dynamically significant. Furthermore, the presence of a typical continuous murmur is usually considered pathognomonic for this lesion, so that few cases undergo special procedures to confirm the diagnosis. The possibility of other conditions causing continuous murmurs over the thorax.
should constantly be kept in mind. A review of such lesions has been recently published by Davis and associates. To the list of such conditions should now be added stenosis of a main branch of the pulmonary artery.

SUMMARY

Three patients, all with other congenital cardiac malformations, were shown by cardiac catheterization to have stenosis of a main branch of the pulmonary artery. Two of the 3 patients exhibited continuous ductus-like murmurs heard at the chest wall over the site of the stenosis. The origin of the murmur at the site of the constriction was proved by the demonstration of pressure gradients throughout the cardiac cycle at the site of constriction and by the experimental production of continuous murmurs in dogs in which partial constriction of a pulmonary arterial branch was accomplished. Conditions necessary for the production of such murmurs are discussed. The importance of these observations in the differential diagnosis of congenital heart disease is emphasized and demonstrated by the fact that one of the patients underwent thoracotomy under the mistaken diagnosis of patent ductus arteriosus.

SUMMARIO IN INTERLINGUA

In tres patientes, catheterisation cardiac serviva a monstrare le presentia de un stenosis del branca major del arteria pulmonar. In omnes altere congenite malformationes cardiac esseva presente. Duo del tres patientes exhibiva continue murmures, simile al murmur de patente ducto arterioso, al pariete thoracie supra le sito del stenosis. Le origine del murmur al sito del constriction esseva provate per le demonstration de gradientes de pression durante le integre cyclo cardiac al sito del constriction e per le production experimental de continue murmures in canes in que constriction partial del branca pulmonaria arteriel habeva essite effectuate. Es discutite le conditiones necessari pro le production de tal murmures.

Es sublineate le importantia de iste observationes in le diagnose differential de congenite morbo cardiac. Un illustration multo significative se vide in le facto que un del patientes hic reporte esseva subjicite a thoracotomia in consequentia del diagnose erronee de patente ducto arterioso.

ADDENDUM

Since this paper was submitted for publication, 2 additional patients with stenosis of a branch of the pulmonary artery have been studied in this laboratory.

Case 4. A 4-year-old girl without symptoms in whom a heart murmur had first been heard at the age of 9 months. Positive physical findings included a prominent apex impulse, a loud high-pitched apical systolic murmur, and in the aortic area a systolic murmur of lower frequency, which was well transmitted to the neck.

The electrocardiogram showed high voltage complexes over V1 to V4 and was thought to be suggestive of left ventricular hypertrophy. X-rays were interpreted as showing moderate left ventricular enlargement, slight right ventricular enlargement, and prominent proximal pulmonary arteries.

Cardiac catheterization revealed no intracardiac shunts. The arterial oxygen saturation was normal. Pulmonary artery pressure tracings, however, revealed a sharp pressure drop in the right pulmonary artery distal to the bifurcation of the main pulmonary artery. The following measurements were obtained: right ventricle 38/0 mm. Hg; main pulmonary artery 36/16 mm. Hg; left pulmonary artery 34/15 mm. Hg; right pulmonary artery 24/14 mm. Hg. Analysis of the pressure tracings revealed a gradient only during systole.

The diagnosis was aortic stenosis plus stenosis of the right pulmonary artery.

Case 5. A 5½-year-old girl without symptoms in whom a cardiac murmur was noted at the age of 2 weeks. There was a slight left parasternal lift. A thrill was noted in the left second and third intercostal spaces parasternally. A moderately loud holosystolic murmur was heard in the same area. In addition a louder systolic ejection murmur was heard in the midsternal line at the level of the second intercostal space. This murmur began after the first sound, reached its peak intensity just before mid-systole and decreased in intensity during late systole.

The electrocardiogram was normal. Cardiac catheterization revealed a rise of 1.0 ml. per 100 ml. in oxygen content in the right ventricle, indicating the presence of an interventricular septal defect. There were sharp pressure drops in both right and left pulmonary arteries at their juncture with the main pulmonary artery. The pressures were: right ventricle 28/1 mm. Hg; main pulmonary artery 28/8 mm. Hg; left pulmonary artery 19/8 mm. Hg; right pulmonary artery 20/7 mm. Hg. Analysis of the pressure tracings again revealed pressure gradients only in systole. The maximum gradient on the
right side occurred 0.18 to 0.22 second after the QRS complex. This corresponded very closely in time to the point of maximum intensity of the murmur recorded phonocardiographically over the midline at the level of the second intercostal space.

This is the only patient of this series who exhibited bilateral stenosis of the pulmonary artery branches.

Comment. Several additional reports of this condition have come to our attention. Powell and Hiller described a 5-year-old child in whom were found pressure gradients between the main pulmonary artery and its branches, as well as angiographic evidence of stenosis of both branches at the bifurcation of the main pulmonary artery. This patient exhibited a continuous murmur in the pulmonic area. Hodges discussed the catheterization findings of a patient who had low pressures in both pulmonary artery branches in addition to a pulmonic valve stenosis. Coles and Walker report a child, 26 months old, in whom angiography demonstrated a narrowing of the initial portion of each pulmonary artery branch, and in whom cardiac catheterization gave evidence of pulmonary branch stenosis as well as pulmonary valve stenosis. Figley reported a patient with stenosis of the pulmonary artery branches at 3 sites. These were shown by angiography and pressure gradients were found at cardiac catheterization.

Nineteen additional cases have been studied but not reported by 4 other workers, T. Schnabel, B. Jonsson, A. Leatham, and M. Figley. In several of these other cardiac malformations were also present. Three of these cases exhibited continuous murmurs.

It becomes apparent from the number of patients discovered by these few investigators over a fairly short period of time that stenosis of the pulmonary branches is not a rare condition. The awareness of its existence will probably lead to many more such cases being discovered.

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ductus arteriosus with pulmonary hypertension. Circulation 8: 15, 1953.


This paper attempts to determine the course and prognosis of coarctation of the aorta, based upon a study of 130 patients. Sixty per cent were first seen when under 20 years and only 14 per cent when over 30. Eighty patients have been followed 5 years or more. Twenty-eight were seen in the first decade. Three patients died, 1 with recurrent bouts of failure, 1 of aortic stenosis and rheumatic heart disease, and 1 of failure after being cured of bacterial endocarditis. The living patients were regarded as normal by their parents. Fifty were seen in the second decade. One died, probably of heart failure. The clinical course was usually uneventful in this decade although the blood pressure rose slowly. Thirty-seven were seen in the third decade. The blood pressure was now stable. Five died, 1 of ruptured aorta, 1 of cerebral hemorrhage, and 3 of congestive failure. Seventeen were seen in the fourth decade. Two died, 1 with aortic regurgitation and 1 with aortic stenosis. Only a few were seen in the fifth and sixth decades.

About 25 per cent had aortic regurgitation and 5 per cent aortic stenosis. Large hearts and electrocardiographic evidence of left ventricular strain were uncommon in the absence of valvular disease. Congestive heart failure was the commonest cause of death. Below the age of 30, death was more commonly due to aortic rupture, bacterial endocarditis (with or without aortitis), and intracranial hemorrhage.

Because of the possibility of sudden and unexpected death, operation was advised for most children. Aortic regurgitation was an added reason for urging operation.
Stenosis of a Branch of the Pulmonary Artery: An Additional Cause of Continuous Murmurs over the Chest
FREDERIC ELDRIDGE, ARTHUR SELZER and HERBERT HULTGREN

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