Surgical Correction of Coarctation of the Main Pulmonary Artery

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Advances in heart surgery, catheterization, and angiography have called attention to constrictions of the pulmonary artery and its branches. Recently Schumacker and Lurie described a calcified stenotic zone at the bifurcation of the pulmonary artery in a 14-year-old girl. This was dilated with a Kelly clamp. No postoperative physiologic studies were reported. Sondergaard reported 3 cases with similar stenosis at the bifurcation. Two of these were associated with the tetralogy of Fallot, while the third had an atrial septal defect and valvular pulmonic stenosis. Although Sondergaard discovered the vascular obstructions at operation, none was corrected.

Several unoperated cases have been reported. Arvidsson and associates demonstrated by angiography what appeared to be multiple congenital stenoses of the peripheral pulmonary arteries in 4 patients. Coles and Walker reported a case of bilateral constriction of the main branches of the pulmonary artery, which was diagnosed by catheterization and angiography. Eldridge, Selzer, and Hultgren reported 5 patients with stenosis of a branch of the pulmonary artery demonstrated by catheterization.

We have recently encountered 2 cases of stenosis of the main pulmonary artery. Although Williams, Lange, and Hecht point out that stenosis is probably not unusual, our first case may represent a rare form, as the constriction was in the middle third of the main pulmonary trunk. The constriction was successfully treated by open operation and as such appears to be the first case treated definitively and studied with preoperative and postoperative cardiac catheterization. Fortunately there was also cinematographic documentation of the operation.

Case Reports

Case 1

E.T., an 18-year-old white college sophomore was first seen by us in April 1957. When 4 years old, he had been hospitalized for pneumonia. At the age of 8 he was thought to have rheumatic fever and was in another hospital for approximately 1 month. His family was then told for the first time that he had a murmur. The record indicates that it was heard as a systolic murmur along the left sternal border and was accompanied by a thrill.

He did well except for episodes of epistaxis until the age of 15, when he began to have occasional "dizzy spells" on exercise. During the 3 years prior to admission he noted slight dyspnea on vigorous exercise. On examination in April 1957 he was well developed, weighed 166 pounds, and was 66 inches tall. The heart was not enlarged to percussion and was in normal sinus rhythm. A systolic thrill was present anteriorly in the second left intercostal space. The pulmonic second sound was heard poorly and was split on inspiration. A grade-III harsh systolic murmur was heard widely at the base but loudest in the second left intercostal space along the sternal border; it was transmitted toward the apex and into the neck.

Electrocardiogram (fig. 1) showed delayed right ventricular conduction, and cardiac fluoroscopy and x-ray films (fig. 2) showed right ventricular enlargement. The pulmonary artery segment was not enlarged on fluoroscopy. Cardiac catheterization in September 1957 showed a mean systolic gradient of 32 mm Hg across what was presumed...
to be the pulmonic valve (fig. 3A). The mean systolic pressure was 45 mm. Hg in the right ventricle and 13 mm. Hg in the pulmonary artery. The oxygen content of all right-sided cardiac chambers revealed no evidence of a left-to-right shunt. The arterial oxygen saturation was 94.5 per cent. A dilution curve with Evans blue dye from the right atrium to the brachial artery was normal. The cardiac index was normal at 3.3 L./min./M². A diagnosis of isolated moderate valvular pulmonic stenosis was made, and the pulmonic valve area was estimated as 1.0 cm².

Since the symptoms were minimal and the electrocardiogram and roentgenograms did not indicate severe right ventricular hypertrophy, surgery was deferred. Meanwhile, the boy worried about an impending operation, was unable to concentrate on his studies, did poorly in college, and surgery was requested sooner than had been elected initially.

On March 15, 1958, under hypothermia, through an anterior transverse bilateral incision in the fourth intercostal spaces, a stenosis or zone coarctation was found in the middle third of the main pulmonary artery (fig. 4). The diameter at this point measured 0.5 cm., whereas the proximal and distal portions of the main pulmonary trunk measured 2 cm. in diameter. Ideally one might elect excision of the zone of coarctation with end-to-end anastomosis. However, hypothermia would allow only a few minutes of circulatory arrest; therefore the stenotic area was incised longitudinally. This incision was extended proximally and distally. The valve was palpated and found to be normal. The lumen at the stenosis was less than 1 cm² in area due to a thickened fibrous wall remarkably like that seen commonly in coarctation of the aorta. The longitudinal incision was then closed transversely (fig. 5).

The postoperative course was uneventful except for right bundle-branch block that was first noted on the day after the operation. He was discharged on the fourteenth day after surgery.

Cardiac catheterization 6 weeks after surgery showed that the pressure gradient had been reduced from 32 to 9 mm. Hg (fig. 3B), at an identical cardiac index of 3.3 L./min./M². Repeated passage of the catheter across the obstruction under fluoroscopic observation demonstrated that the site of residual obstruction was about half way between the pulmonic valve and the bifurcation of the pulmonary artery. The estimated area of constriction had been increased to 2.1 cm², twice its preoperative size. Again blood samples

**Figure 1**

Case 1. Preoperative electrocardiogram showing delay in right ventricular conduction with pattern of incomplete right bundle-branch block.
from the right-sided cardiac chambers showed no variation in oxygen content, and there was no evidence of other congenital lesions.

The patient is now asymptomatic. The second pulmonic sound is louder than preoperatively and the pulmonic systolic murmur less prominent. The electrocardiogram shows persistent right bundle-branch block.

**Case 2**

B.N., a 12-year-old white girl, was first seen in December 1957. At birth she appeared to be a normal baby. The family reported that since infancy, on exposure to cold her hands and feet had tended to become blue. Although she tired easily, she denied other symptoms. Because of peripheral cyanosis she had been seen at another clinic in 1954, when a heart murmur was noted and the family was told that the child possibly had a "small hole in the heart." During the year preceding our examination she had frequent episodes of epistaxis. When first seen by us, physical examination revealed a well developed, healthy appearing girl with strikingly red cheeks. The hands and feet were slightly moist and cool but there was no cyanosis, clubbing, or venous distention. The heart was not enlarged. The second pulmonic sound was louder than the second aortic sound, but not unusually loud or split. A grade-II basal systolic murmur was present, which radiated along the left sternal border and out toward the apex.

Electrocardiogram, cardiac fluoroscopy, and x-ray films were essentially normal. Right-sided heart catheterization in April 1958 showed a consistent fall in systolic pressure from the proximal to distal pulmonary artery, amounting to a mean systolic gradient of 6 mm. Hg. The site of transition appeared to be just proximal to the bifurcation of the pulmonary artery. There was no pressure gradient across the pulmonic valve. There was no evidence of coexistent cardiac lesions. Oxygen contents throughout the right side of the heart were identical. Arterial oxygen saturation was normal at 96.3 per cent. Indicator-dilution curves from the superior vena cava and the pulmonary artery to the brachial artery were normal. The cardiac index was rather high at 6.0 L/min./M.² Thus a mild constriction of the pulmonary artery proximal to its bifurcation seemed to be defined. The hemodynamic disturbance did not require surgical correction.

**Discussion**

The pathogenesis of constrictions of the pulmonary artery has been debated.² ⁵ ⁷ ⁸ Most authors favor a congenital origin according to the Skodaic theory. This postulates a stricture produced by an extension of tissue from the ductus arteriosus into the pulmonary artery or aorta which contracts as the ductus undergoes obliteration. This explanation seems untenable in our first case since there was no

*Figure 2*

*Case 1. Preoperative posteroanterior and right lateral chest roentgenograms suggesting right ventricular enlargement.*
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Figure 3
Case 1. Right heart pressures during preoperative (A) and postoperative (B) cardiac catheterization. Preoperatively (A) one pressure pulse was recorded from the pulmonary artery proximal to the region of obstruction. Although this is compatible with a diagnosis of coarctation of the pulmonary artery, this diagnosis was not made inasmuch as artifactual pressure tracings obtained from a catheter tip in a stenotic pulmonary valve may have the same appearance. The preoperative mean systolic pressure gradient of 32 mm Hg (A) was reduced to 9 mm Hg postoperatively (B). The cardiac output was the same during both studies: cardiac index = 3.3 L/min/M². The estimated area of constriction was increased from 1.0 cm² preoperatively to 2.1 cm² postoperatively.

apparent connection between the ligamentum arteriosus and the stricture.

Attention has been directed to thrombosis of the pulmonary artery,⁹⁻¹¹ which at times has simulated pulmonary stenosis. We have recently operated on a patient for anomalous venous drainage associated with long-standing silent massive thrombosis of the pulmonary arteries. He succumbed when the anomaly was corrected at open-heart operation. It seems
likely that some of the cases diagnosed by cardiac catheterization and angiocardiography but without anatomic clarification might represent partially resolved occluding thrombi or emboli with recanalization. This is unlikely in our first case as the actual outside diameter of the pulmonary trunk at the coarctation was markedly narrowed and there was no evidence of thrombosis.

Thus none of the prevailing theories of pathogenesis seems to explain the isolated midzone coarctation of the main pulmonary artery of our first case. In the systemic circulation, the aorta may be narrowed at 2 sites and at times in the abdominal region. Perhaps these are comparable vascular anomalies.

Postvalvular stenosis of the pulmonary artery is probably more common than is generally realized. To our knowledge, however, pure stenosis, or what we prefer to call coarctation of the midportion of the main trunk of the pulmonary artery has not been reported.

The corrective technique in the first patient reported here had not been planned preoperatively. It is fortunate that hypothermia was being used. The possibility of unsuspected coarctation of the pulmonary artery may constitute an added indication for the routine use of hypothermia or mechanical bypass in what is thought preoperatively to be pure pulmonic valvular stenosis. In case 1 the stenosis was presumed to be critical on the basis of the preoperative pressure gradient of 32 mm. Hg and an estimated area of constriction of 1 cm. It has been emphasized by Holman that when the diameter of the pulmonary artery is constricted to less than 50 per cent in animals, heart failure results. A critical level undoubtedly existed in our patient with a diameter of 0.5 cm.

The diagnosis of anomalies of the pulmonary artery would be made more frequently if it were routinely considered in the differential diagnosis of pulmonary valvular stenosis. The absence of postvalvular dilatation, as in case 1, might make one consider coarctation of the pulmonary artery.

Catheterization studies should be helpful, but here again special care must be exercised to demonstrate the exact location of the stenosis or stenoses.

At the time of operation, proper exposure is mandatory in dealing with all congenital anomalies so that the unexpected may be detected.

Summary

Two cases of pure congenital coarctation of the main pulmonary artery are reported.

One case may be unique in that it represents the first report of an anatomically proved constriction located in the middle third of the main trunk. In this patient, successful open correction under hypothermia was carried out with cinematographic documentation. The results of preoperative and postoperative cardiac catheterization are described.

A plea is made for the routine use of open operation for correction of "pure pulmonic valvular stenosis." This allows better correction when the diagnosis is confirmed and flexibility for treatment if the unusual coarctation or constriction of the pulmonary artery is encountered.
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Figure 5
Case 1. Illustration of technic. Longitudinal incision of the zone of coarctation and transverse closure.
Summario in Interlingua
Es reportate duo casos de congenite pur coarctation del arteria pulmonar commun.
Un del duo es possiblemente unique in tanto que illo representa le prime reporto de un anatomicamente demonstrate constriction in le secunde terto del trunco principal. In iste patiente, correction aperte esseva effectuate a bon successo sub hypothermia e documentacion cinematographique. Es describite le resultatos de catheterismo cardiae ante e post le operation.
Es recommendate urgentemente le uso de chirurgia aperte in le correction de ‘pur stenosis pulmona-valvular.’ Iste technica permitte un correction superior quando le diagnose es confirmate; illo permette un plus alte grado de flexibilitate in le tractamento in casos in que le phennomeno inusual de coarctation o constriction del arteria pulmonar es inconstante.

References

Vesalius
The influence of Vesalius on the history of science may be regarded on the one hand in its general, on the other in its more special aspect.
Taking the general aspect first we may say that he founded modern anatomy. He insisted upon, and through his early unwearyed labours by his conspicuous example he ensured the success of the new method of inquiry, the method of observation as against interpretation; he overthrew authority and raised up experience, he put the book of nature, the true book, in place of the book of Galen, and thus made free and open the paths of inquiry. . . .
Under a more special aspect he may be regarded as the founder of physiology as well as of anatomy in as much as he was the distinct forerunner of Harvey. For Harvey’s great exposition of the circulation of the blood did, as we shall see, for physiology what Vesalius’ Fabrica did for anatomy; it first rendered true progress possible. And Harvey’s great work was the direct outcome of Vesalius’ teaching, the direct outcome and yet one reached by successive steps, steps taken by men of the Italian school, of which Vesalius was the founder and father.—Sir M. Foster. Lectures on the History of Physiology. London, Cambridge University Press, 1901.
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Circulation. 1960;21:672-678
doi: 10.1161/01.CIR.21.5.672

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/21/5/672

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