Postvalvular Stenosis of the Pulmonary Artery

By C. Basil Williams, M.D., Ramon L. Lange, M.D., and Hans H. Hecht, M.D.

Constrictive lesions of the pulmonary artery or its branches occur distal to the pulmonic valve. Four patients whose surgical or cardiac catheterization findings demonstrated such postvalvular stenosis are reported. The significance of such findings is discussed and diagnostic misinterpretations are mentioned.

RECENT reports have focused attention on the occasional occurrence of localized constrictions in the pulmonary vascular system distal to the valve of the pulmonary artery. Such lesions have been called "coarctation of the pulmonary artery."1-3 We believe the less specific term "postvalvular stenosis" to be somewhat more inclusive and more descriptive of these defects.

Schumacher and Lurie4 have described a calcified stenotic lesion at the bifurcation of the pulmonary artery. This constriction was subsequently successfully dilated by spreading a Kelly clamp within the lumen of the artery. Sondergaard1 has reported 3 cases of similar constrictions of the bifurcation encountered at surgery during a 3-year period. In each case, a fibrotic band, believed to be the ligamentum arteriosum, was attached to the constriction. Arvidsson, Karnell, and Möller5 have demonstrated multiple stenotic lesions of the peripheral pulmonary vasculature in 4 patients by means of selective angiography. Coles and Walker6 have presented a case of "coarctation of the pulmonary artery" that was diagnosed by cardiac catheterization and angiography. They also mentioned a case of Hodges that presented a similar picture. Recently, Eldridge, Selzer, and Hultgren7 have reported 5 patients with stenosis of a branch of the pulmonary artery. In Bailey's text8 is mentioned the existence of localized strictures of the main pulmonary artery termed "coarctation."

Since 1951 we have encountered 4 patients who apparently presented instances of this previously rarely reported anomaly.

Case Reports

Case 1. G. E., a 15-year-old white boy, was first seen here in August 1951. At 2½ months gestation, his mother had rubella. At 20 months of age, deafness was discovered. His childhood was marked by slow development, easy fatigue, frequent upper respiratory infections, and exertional dyspnea. His nails were always grayish. Orthopnea, edema, and squatting were not noted. Physical examination in 1951 revealed a small deaf mute with long digits and a prominent mandible. His heart was not enlarged to percussion. A brief systolic thrill and a harsh, loud systolic murmur with wide transmission were observed at the second and third left intercostal spaces along the left sternal border. The pulmonic second sound was louder than the aortic second sound. The remainder of the physical examination was unremarkable.

Cardiac fluoroscopy, x-ray films, and repeated electrocardiograms demonstrated right ventricular hypertrophy. The hemoglobin measured 16.8 Gm. per cent and the arterial oxygen content was normal at 20.4 volumes per cent (93 per cent saturated).

At cardiac catheterization, spot pressure readings revealed a right ventricular pressure of 64/0 mm. Hg. No "pull through" was obtained and no abnormal shunts were demonstrated. A diagnosis of pulmonary valvular stenosis was made but at surgery on September 8, 1952, a normal valve was discovered. The operative report stated: "The pulmonic valve was found to be approximately 25 per cent of normal size for the age and general size of the patient prior to dilatation and approximately 40 per cent postoperatively. The valve appeared normal in all respects. A diffuse low grade thrill was palpable over the pulmonary artery but not over the ventricle. A small urethral sound was inserted through the small incision and through the valve with no difficulty. A curved, flexible Bailey-Brock knife was then inserted through the incision and also passed through the valve with ease. The surgeon's finger was then likewise inserted and the valve was found to be essentially normal, but the artery was found to be markedly hypoplastic in its first part. A no. 34 uterine sound was then inserted into this hypoplastic artery segment with the resulting distinct increase in diameter of the artery."

When seen again in June 1953, the patient noted slight increased exercise tolerance, and less lethargy.

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His murmur, thrill, and roentgenograms were unchanged.

**Impression.** A 15-year-old boy whose history and preoperative findings suggested valvular pulmonary stenosis. At surgery, a normal valve was present but a hypoplastic pulmonary artery segment was discovered and partially dilated.

**Case 2.** A. G., a white male infant, was 6 months old when first seen in April 1952 with a history of poor growth. A heart murmur had been present since birth. No cyanosis and no dyspnea had been observed. He had not been ill. Physical examination revealed a small infant with a loud blowing systolic murmur heard best in the third intercostal space along the left sternal border. The pulmonic second sound was decreased. The examination was otherwise unremarkable. At age 1½, he returned with the same findings. In addition, his parents noted that his lips became blue following exertion and that he seemed to tire easily. On August 27, 1953, a benign lymphangioma was removed from the right inguinal area. At that time an electrocardiogram suggested right ventricular hypertrophy. Cardiac fluoroscopy and x-ray films demonstrated the heart to be within normal limits.

On September 25, 1953, cardiac catheterization demonstrated no shunt or arterial desaturation, but did show a right ventricular pressure of 80/0 mm Hg and a pulmonary artery pressure of 5/2 mm Hg. He was observed over the next 2½ years during which time easy fatigue, circumoral cyanosis on exertion, occasional squatting, and frequent upper respiratory infections were noted.

On March 7, 1956, the patient, now 4 years old, was explored using a cardiac bypass technic. At operation the interatrial septum was found intact. A stenotic pulmonary valve was found but, in addition, a marked narrowing of the pulmonary artery was observed 1½ cm distal to the annulus (near the bifurcation). Following dilatation of the valvular stenosis, a right ventricular pressure of 100/0 and a pulmonary artery pressure of 50/18 were recorded while the patient was on the operating room table. Anesthetic difficulties precluded any further attempts to reduce this gradient, and the incision was closed. At the time of discharge on April 4, 1956, the systolic murmur was present as before. The patient moved away and has been lost to follow-up.

**Impression.** A 4-year-old boy with severe valvular pulmonary stenosis. In addition, a constriction of the pulmonary artery near the bifurcation was discovered at surgery.

**Case 3.** J. P., an 11-year-old girl, was seen here in June 1956 with the history that a heart murmur had been noted since age 3. Normal growth and development had occurred and cyanosis had never been observed. She was normally active but her mother noted she needed much sleep (particularly after exertion). She contracted frequent upper respiratory infections, but she had no known history of rheumatic fever or kidney disease. Physical examination revealed a healthy young girl with slight prominence of the left chest. Her heart was not enlarged to percussion. A systolic thrill was present along the left sternal border and a loud systolic murmur was heard over the entire precordium being loudest at the second left intercostal space. The pulmonic second sound was normal or slightly reduced.

Electrocardiograms, cardiac fluoroscopy, and x-ray films suggested right ventricular hypertrophy. The pulmonary artery segment was not prominent and there was no hilar dance. Cardiac catheterization-

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**Fig. 1.** Case 4. This pressure tracing was obtained by pulling the catheter from the distal pulmonary artery back across the pulmonary valve, through the right ventricle to the right atrium. Note the sharp increase in systolic pressure within the pulmonary artery and the additional rise at the pulmonary valve. Repeated "pull-through" tracings showed identical results.
tion and dye oximeter studies demonstrated a moderate left-to-right shunt through an interventricular septal defect. In addition, a stepwise pressure rise was observed in the pulmonary artery, rising abruptly from 26/11 mm. Hg (mean 19) to 33/11 mm. Hg (mean 22). An infundibular pressure was recorded at 33/0 mm. Hg and then rose abruptly again to 55/0 mm. Hg. The arterial oxygen saturation was 94 per cent.

Impression. An 11-year-old girl with an interventricular septal defect and infundibular pulmonary stenosis ("balanced" tetralogy of Fallot) and a postvalvular stenosis of the pulmonary artery.

Case 4. N. A., a 4-year-old white boy, was seen in November 1956. He presented a history of decreased exercise tolerance, rapid heart rate on exertion, and circumoral blueness when cold. A heart murmur had first been noted 1½ years previously. Physical examination revealed a well-developed boy of normal size for his age. Harrison's grooves were present bilaterally. The heart was enlarged to percussion. A systolic thrill was felt over the apex; a harsh holosystolic murmur and a short early diastolic murmur were heard over the same area and along the left sternal border. The pulmonic second sound was split but normal in intensity. Electrocardiograms demonstrated a prolonged P-R interval and an unusual rotation of the electric axis. Cardiac fluoroscopy revealed a globular heart with apparent right and left ventricular hypertrophy. Pulmonary markings and the pulmonary artery segment appeared normal.

Cardiac catheterization and oxygen sampling demonstrated interatrial and interventricular septal defects. The arterial oxygen saturation was 89 per cent. Several "pull-through" pressure tracings revealed a stepwise increase in pulmonary artery pressure from 22/6 mm. Hg (mean 14) to 27/6 mm. Hg (mean 19) with a further rise to 34/0 mm. Hg at the pulmonary valve (fig. 1).

Impression. A 4-year-old boy with interatrial and interventricular septal defects, slight valvular pulmonic stenosis ("pentology"), and a postvalvular stenosis of the pulmonary artery.

Discussion

Although infrequently reported, postvalvular stenosis ("coarctation") of the pulmonary artery is probably not rare. Sondergaard encountered 3 cases at surgery in as many years. We have seen 4 cases in 5 years, 2 of which were discovered at surgery and 2 by cardiac catheterization. A summary of the findings is presented in table 1. During this same period, we have seen a total of 70 patients with true pulmonic stenosis (valvular and infundibular) with or without associated defects.

It should be recognized that for a given flow a considerable degree of constriction must exist before a measurable pressure gradient in the pulmonary artery can be obtained. For example, a 50 per cent reduction in diameter will result in a 9 mm. Hg pressure drop across the constriction. Obviously minimal constriction may exist without demonstrable pressure changes (fig. 2). In view of these relationships, the case of Coles and Walker4 is not clear-cut. Their pressure tracing is reminiscent of a "pull-out" from the "wedge" position to the pulmonary artery and then across a stenotic pulmonary valve. It seems unlikely that the degree of constriction they delineate with their angiograms could cause the marked pressure changes they record by catheterization. On the other hand, when flow is raised, a pressure

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* Postvalvular stenosis found at surgery.
and solving for $P_2$, the pressure beyond the constriction. 8 We assume low frictional losses and neglect the coefficient of contractibility. $P_1$ is assumed to be a normal mean pulmonary artery pressure of 15 mm. Hg (20 cm. H$_2$O), and $V_1$ (velocity proximal to the constriction) is assumed to be 40 cm. per second. 9 Inasmuch as the velocity through the constricted area ($V_2$) varies inversely as the square of the radius, notice that after a critical amount of constriction is reached the pressure changes effected by constriction become most striking. If shunts are present and pulmonary flow is increased, constriction may produce more profound pressure changes. $W =$ specific weight of blood. $P$ measured in centimeters of water.

gradient may appear even if constriction is mild, a point clearly demonstrable in patients with mild stenosis of the mitral valve.

Certain other pitfalls exist in catheter diagnosis of these lesions. The case of Coles and Walker may point out one such error. When the catheter is pulled from the "wedge" position to the pulmonary artery, it is seen and felt to flip from its peripheral position and an abrupt increase in pressure is recorded (particularly if pulmonary hypertension is present). By sampling the oxygen content of the blood at both positions, the actual position can be determined, since either fully saturated blood or no sample at all will be obtained from the "wedge" position. Inadvertent "wedging" of the cardiac catheter and a spurious pressure rise must be ruled out before pulmonary constrictive lesions can be considered. Severe pulmonary stenosis per se may completely mask peripheral constriction (e.g., case 2), and such defects may be obvious only after the stenosis is relieved. Occasionally recordings taken near a patent ductus arteriosus may show a higher pressure than that recorded distally. 10 This might suggest postvalvular constriction but again judicious oxygen sampling and fluoroscopic examination of the catheter's position will aid in disclosing the true abnormality.

In any event, diagnosis of these lesions is important. Since various degrees of constriction may exist, they may or may not be functionally significant. Certainly marked involvement can mimic pulmonary stenosis very closely (e.g., case 1). The surgeon should be forewarned in such cases and should plan a more extensive procedure than a simple valvulotomy. Conversely, in cases of severe pulmonary stenosis, the surgeon should also search for distal constriction at the time of surgery since the 2 defects may coexist.

The etiology of these lesions is unknown; it is assumed that they are congenital in origin. The Skodaic theory 11, 12 (which emphasizes the obliteration of the ductus arteriosus as a causative factor) has been espoused by Sondergaard. However, it seems equally likely that multiple factors may cause these defects. Defective formation of the aortic septum must account for some cases. At least 5 of the 6 previously reported cases had coexisting pulmonary stenosis as did 3 of our 4 cases. This strongly suggests an association of the 2 defects.

**Summary**

Four cases of postvalvular constrictions of the pulmonary artery are described. Such lesions may be isolated or associated with other defects, and may vary widely in their degree of severity. Certain pitfalls in diagnosis (misinterpretation of normal findings, or artifacts produced by a patent ductus arteriosus) are noted. The etiology is unknown but is assumed to be on a congenital basis. Such lesions are probably not so rare as the paucity of reports would indicate.
SUMMARIO IN INTERLINGUA
Es describite quatro casos de constriction postvalvular del arteria pulmonar. Tal lesiones occurre in isolation o in association con altere defectos. Illos varia grandemente in grado e severitate. Certe riscos diagnostic es signalate que resulta in le misinterpretation de condiciones normal o de artefactos producite per un patente ducto arteriose. Le etiology non es cognoscite sed pare haber un base congenite. Lesiones de iste genere es probablemente minus rar que lo que es indicate per le paucitate de reportos.

REFERENCES


Using rigid criteria for the diagnosis of hypertension the authors examined the records of 509 patients under treatment for malignant neoplasms to determine the incidence of hypertensive disease in such a group. A similar number of control patients without neoplasms, matched for distribution of sex and age were included in the study. It was found that in both sexes and all ages, essential hypertension was less common in the neoplastic than in the control group. The difference was more striking in men than in women. These observations are in agreement with previous studies in which less rigid criteria for hypertension were applied. If the reference standard used is a working population in which the incidences of hypertension are 9 per cent in men and 10 per cent in women, the observed values of 1 per cent and 6 per cent respectively in the neoplastic cases may have further significance. It is suggested that a broad epidemiologic and experimental study is needed to evaluate the dissociation between hypertension and neoplastic disease.

SHUMAN
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