The Diagnosis of Bilateral Stenosis of the Primary Pulmonary Artery Branches Based on Characteristic Pulmonary Trunk Pressure Curves

A Hemodynamic and Angiocardiographic Study

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In 1938 Oppenheimer\(^1\) reported the first case of bilateral stenosis of the pulmonary main arteries diagnosed at autopsy. Since then several papers have been published describing clinical, cardiac catheterization, and angiocardiographic findings in both unilateral and bilateral stenosis of the pulmonary arteries.\(^2\)-\(^{12}\)

We are reporting cardiac catheterization and angiocardiographic findings in 12 patients with bilateral stenosis of the primary pulmonary artery branches. In all 12 patients characteristic pressure curves were consistently obtained from the pulmonary trunk that were reproduced experimentally in dogs. The hemodynamic explanation and the diagnostic value of these typical pressure curves are discussed.

Material and Method

Of 1,650 cardiac catheterizations performed in the Department of Pediatric Cardiophysicsology of the Cook County Children's Hospital during the last 11 years, 60 patients were found to have stenosis of the primary pulmonary artery branches, 32 of whom had unilateral stenosis and 28 bilateral stenosis. The hemodynamic and angiocardiographic findings in 12 patients with bilateral stenosis, in whom both main pulmonary artery branches were entered, are discussed. The age of these patients ranged from 3 months to 13 years.

All patients underwent routine physical and laboratory examinations, including phonocardiography. A systolic ejection murmur maximal at the pulmonic area with wide transmission to the right and left axillae and the back was a characteristic finding in all. In the absence of an associated lesion the pulmonic component of the second sound was either normal or somewhat diminished and was widely separated from the aortic closure, the degree of splitting depending on the severity of the stenosis. The aortic sound in the pulmonic area was obscured by the murmur in severe cases. The chest roentgenograms of patients with mild bilateral stenosis usually revealed normal pulmonary vascular markings, whereas those with severe stenosis showed diminution of pulmonary vascular markings in proportion to the degree of severity. The electrocardiograms were normal in the milder cases, and showed right ventricular hypertrophy in the more severe types. Right heart catheterization was performed with the patients under intravenous Pentothal anesthesia or meperidine-chlorpromazine-promethazine sedation. The pressures were obtained by use of Sanborn transducers and recorded on a Sanborn 150 direct-writing and photographic recorder. Simultaneous tracings from the right ventricle and from the main pulmonary artery, using 2 catheters, were obtained in four cases. Following the cardiac catheterization, selective angiocardiography was performed with injection of 90 per cent Hypaque solution into either the main pulmonary artery or into the right ventricular outflow tract. An Elema biplane roll film changer with eight film exposures per second was used.

Results

Table 1 shows the pressures obtained at cardiac catheterization in the 12 patients reported in this paper. Six of the patients had associated anomalies, three with pulmonary anomalies.
Valvular stenosis, one with supravalvular pulmonary stenosis, one with atrial septal defect, and one with ventricular septal defect. The angiocardiograms revealed varying degrees of poststenotic dilatation of the primary pulmonary artery branches in 10 patients (fig. 1). In two patients the pulmonary artery branches were uniformly hypoplastic (fig. 2). Stenosis of the right branch was easily noted in the anteroposterior projection of the selective angiocardiograms in all 12 patients. The stenotic segment appeared wider during ventricular systole than during diastole (fig. 5). The stenosis of the left main pulmonary artery was more difficult to determine in this view because of superimposition on the pulmonary trunk. The lateral angiocardiogram...
BILATERAL STENOSIS OF PULMONARY BRANCHES

Figure 3A
Pressure tracings in bilateral stenosis. MPA, main pulmonary artery; RV, right ventricle.

Figure 3B
Pressure tracings in normal subjects. MPA, main pulmonary artery; RV, right ventricle.

ograms often revealed the stenosis of this artery better than the frontal angiocardio-
grams. In a few instances the existing stenosis

of the left pulmonary artery could not be ascertained from the angiocardigrams.

The systolic pressure in the pulmonary trunk varied from 27 to 95 mm. Hg and the diastolic pressures from 5 to 20 mm. Hg. A systolic gradient was demonstrated in all 12 cases between the main pulmonary trunk and both main pulmonary artery branches. In case 12, with associated coarctation of the pulmonary trunk, proved at surgery, systolic pressure proximal to the stenosis was 130 mm. Hg and 95 mm. Hg distally.

The pulmonary trunk pressure tracings in all patients showed a characteristic pattern entirely different from normal curves (fig. 3A and B). During ventricular systole the main
Selective angiocardiogram with injection of contrast substance into the right ventricle (RV) and schematic drawings. During ventricular systole the pulmonary artery (PA) dilates and the area of stenosis (arrow b) widens. In diastole the pulmonary valves (arrow a) bulge into the right ventricular outflow tract (I = infundibulum) and the stenotic area (arrow b) constricts.

Pulmonary artery curves were identical in contour and timing to those of the corresponding right ventricular curves. The level of the systolic pressure depended on the severity of the stenosis, but the diastolic pressure remained within normal limits with resulting increase in the pulse pressure. The descending limb of the pulmonary trunk tracing was
BILATERAL STENOSIS OF PULMONARY BRANCHES

Figure 6
Pressure tracings in mild bilateral stenosis.

Figure 7
Pressure tracings in main pulmonary artery and right ventricle in moderate bilateral stenosis.

Figure 8
Pressure tracings in severe bilateral stenosis. MPA, main pulmonary artery; RV, right ventricle; RFA, right femoral artery.

Figure 9
Pressure tracings in unilateral and bilateral stenosis.

revealed a gradual descent instead of a plateau, suggesting runoff during diastole (fig. 4). The characteristics of these tracings were not altered in the presence of left-to-right shunts or of associated pulmonary valvular stenosis.

Discussion
The characteristics of the pulmonary trunk pressure curves obtained in patients with bilateral stenosis of the primary pulmonary artery branches has, to our knowledge, not been previously described. The pressure curves in the pulmonary trunk and in the right ventricle, in the absence of pulmonary valvular stenosis, are identical in contour and timing. The pulmonary trunk functions as an outlet chamber during ventricular ejection.
and the valves remain open so long as the right ventricular pressure is greater than that of the pulmonary arteries. The pulmonary trunk dilates during ventricular systole and the pulmonary valves are forced toward the wall of the artery (fig. 5). This explains why the descending limb of the pulmonary trunk pressure curve is steeper than normal. During early isometric relaxation the pulmonary valves close and bulge toward the right ventricular cavity, thus increasing suddenly the volume capacity of the pulmonary trunk (fig. 5), with a corresponding fall in the systolic pressure and formation of the dicrotic notch on the descending limb of the pressure tracing. Since the stenosis of both main pulmonary arteries, especially when severe, restricts the normal transient backward flow of blood from the distal pulmonary arteries toward the pulmonary trunk in diastole (fig. 5), the dicrotic notch becomes deeper than normal. Normally the elastic recoil of the large pulmonary arteries during diastole produces reversal of the transient backward flow of blood, and instead results in runoff to the peripheral branches. In the presence of bilateral stenosis the diastolic runoff is obstructed and consequently the diastolic tracing shows less descent. In fact, in severe stenosis this descent may be absent and the diastole then assumes a plateau (figs. 6, 7, and 8).

The presence of associated left-to-right shunts or pulmonary valvular or supravalvular stenosis did not alter the characteristics of these pressure tracings. Similar tracings were not seen in cases with unilateral pulmonary artery stenosis (fig. 9). The pressure tracings in mild pulmonary valvular incompetence and mild bilateral pulmonary artery stenosis may simulate each other. When pulmonary insufficiency is pronounced, the systolic pressure also becomes elevated, as in bilateral pulmonary artery stenosis, but the dicrotic notch usually appears at a high level and is followed by further rapid descent in the diastolic pressure curve until it reaches a pressure level, nearly identical to that of the right ventricle. The presence of a regurgitant murmur at the pulmonic area on clinical examination, or of a diastolic murmur in the right ventricular outflow tract, demonstrated by intracardiac phonocardiography, is most helpful in differentiating these lesions. The pulmonary trunk pressure curves in pulmonary hypertension, associated with increased pulmonary blood flow or increased peripheral resistance, have a normal contour and show an elevated diastolic pressure.

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BILATERAL STENOSIS OF PULMONARY BRANCHES

Cole and Walker showed a similar characteristic pressure tracing from the pulmonary trunk in a proved case of bilateral pulmonary stenosis, but the authors did not discuss the significance of the curve. Similar pressure curve phenomena have also been shown in tracings obtained from the proximal aorta in cases of congenital and experimental supravalvular aortic stenosis. Proximal to the supravalvular stenosis the systolic pressure is elevated, whereas the diastolic pressure and the dicrotic notch are abnormally low. It seems likely that the same explanation applies to the pressure changes in supravalvular aortic stenosis as to the bilateral pulmonary artery stenosis. Experimentally, we were able to reproduce these typical tracings in three dogs. Figure 10 shows the effects of sudden complete occlusion of one pulmonary artery, with some elevation of systolic pressure, but otherwise a normal arterial tracing. Complete occlusion of one branch and partial (about 50 per cent) occlusion of the other main branch resulted in the characteristic configuration of the pulmonary trunk tracing previously described.

Summary

The cardiac catheterization and angiocardiographic findings in 12 cases with bilateral stenosis of the primary pulmonary artery branches are described. Characteristic pressure tracings were consistently obtained from the main pulmonary trunk in all cases. These typical curves were reproduced in animal experiments. Associated lesions with left-to-right shunts and mild pulmonary valvular stenosis did not alter the characteristics of the curves. An attempt is made to explain these hemodynamic phenomena.

These tracings obtained from the main pulmonary trunk appear to us to be diagnostic of bilateral stenosis of the main pulmonary arteries even if, as frequently happens, one is unable to enter both pulmonary artery branches during cardiac catheterization or to demonstrate definitely the stenosis of both pulmonary arteries by angiocardiography.

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