Stenosis of the Right Main Pulmonary Artery

Clinical, Angiocardiographic, and Catheterization Findings in Ten Patients

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Isolated stenosis of a branch of the pulmonary artery is not a rare anomaly in our experience; however, there have been few published reports of this condition. Multiple stenosis in one or both pulmonary arteries has been more frequently reported.

Søndergaard¹ reported 3 patients with stenosis of the main pulmonary artery at the point where it was joined by the ligamentum arteriosum. He called this anomaly "coarctation of the pulmonary artery." Schumacher and Lurie² cited a similar case in a 14-year-old girl. Recently Hodges,³ Powell,⁴ and Coles and associates⁵ reported cases of narrowing of both right and left pulmonary arteries at the bifurcation of the main vessel diagnosed by angiocardiographic and cardiac catheterization studies.

Gyllenswörd and co-workers⁶ and Arvidsson's group⁷ reported 12 cases of multiple stenosis in both branches of the pulmonary artery that were demonstrated by dextro-angiocardiographic studies. Eldridge and collaborators⁸ described 5 patients with stenosis of a branch of the pulmonary artery, 3 of which occurred on the right, 1 on the left, and 1 was bilateral.

The purpose of this paper is to report 10 patients with stenosis of the right pulmonary artery, in all of whom the diagnosis was confirmed by cardiac catheterization and dextro-angiocardiographic studies. They ranged in age from 4 months to 13 years. Four were males and 6 females. Of these, 7 cases had associated cardiac anomalies as follows: 1 with pulmonary valvular stenosis, 1 with pulmonary valvular stenosis and patent ductus arteriosus, 2 with patent ductus arteriosus only, 2 with ventricular septal defects, and 1 with tetralogy of Fallot.

The physical findings depended on the other cardiac anomalies. The effect of the stenotic pulmonary artery alone was assessed by clinical, angiocardiographic, electrocardiographic, roentgenologic, and hemodynamic data obtained in all of the 10 patients; in 2 patients with patent ductus arteriosus these data were also obtained postoperatively.

Case Reports

Case 1

An 8-year-old girl was admitted to the cardiac clinic because of a heart murmur. The blood pressures in the right arm and left leg were 120/80 and 134/80 respectively. The heart was not enlarged to percussion. S₁ was normally split and maximum at the apex; S₂ was normally split and maximum at the second left intercostal space; S₃ was constant at the apex. There was a soft grade-III systolic murmur heard best in the second and third intercostal spaces near the sternum as well as over the second right intercostal space and transmitted to the back. Roentgenograms of the chest revealed no cardiac enlargement. An electrocardiogram showed left axis deviation and an incomplete right bundle-branch block. Angiograms revealed slight dilatation of the main pulmonary artery. Right heart catheterization demonstrated stenosis of the right main pulmonary artery and no shunt between the pulmonary and systemic circulations (tables 1 and 2).
STENOSIS OF PULMONARY ARTERY

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*In retrospect, reviewing the spot films taken during the 2 catheterizations, it was found that the pressure of 52/23 mm of mercury recorded in the first catheterization was proximal to the stenosis as correlated with the angiograms. The pressures taken during the second catheterization show a definite systolic pressure gradient between the right pulmonary artery and the main pulmonary artery. The spot films taken at the time of the second catheterization indicate that the tip of the catheter was beyond the stenotic area visualized on the angiogram.

R.P.A., right pulmonary artery; M.P.A., main pulmonary artery; S, systolic pressure; D, diastolic pressure; M, mean pressure.

Case 2

An asymptomatic 8-year-old girl was admitted for cardiac evaluation. Blood pressures were 110/70 and 114/70 in the right arm and leg respectively. The heart was not enlarged to percussion. S1 and S2 were normal in intensity. A grade-III harsh systolic murmur was heard over the second left and right intercostal spaces. It was transmitted well to the back in the interscapular region. Roentgen rays of the chest and the electrocardiogram were within normal limits. A diagnosis of stenosis of the right pulmonary artery was made. Right heart catheterization and selective angiocardiogram confirmed the clinical diagnosis of isolated stenosis of the right main pulmonary artery (tables 1 and 2). A selective angiocardiogram showed definite stenosis of the right pulmonary artery with poststenotic dilatation.

Case 3

A 16-month-old Negro girl with no cardiac symptoms was admitted for diagnosis of a heart murmur discovered at the age of 2 months. Blood pressure in the left arm was 84/60. The apex of the heart was in the fourth left intercostal space at the midclavicular line. A systolic thrill was felt over the third and fourth left intercostal spaces, maximum over the fourth. S1 was normal in intensity and maximal at the apex; S2 was split and normal in intensity. A harsh grade-IV systolic murmur was heard all over the precordium maximal at the third and fourth left intercostal spaces and was well transmitted to the back. Electrocardiogram showed right bundle-branch block with right ventricular hypertrophy. Cardiac catheterization and angiocardiography demonstrated stenosis of the right main pulmonary artery (tables 1 and 2).

Case 4

A 4-month-old girl was referred to the cardiac clinic because of cyanosis and dyspnea at 3 weeks of age. Blood pressures (flush method) of the left arm and left leg were 58 and 68 mm., respectively. The apex of the heart was in the fifth left intercostal space just outside the midclavicular line. S1 was loudest in the second left intercostal space; S2 was split and maximal over the second right intercostal space. A grade-II to -III systolic murmur was heard over the aortic and pulmonary areas, radiating down the left sternal border and transmitted to the interscapular region on the right. An electrocardiogram showed right ventricular hypertrophy. Angiocardiograms revealed stenosis of the pulmonary valve and the right main pulmonary artery with decreased vascularity of the right lung field. Findings on right heart catheterization were consistent with a diagnosis.
of isolated pulmonary valvular stenosis. A gradient could not be demonstrated between the main pulmonary artery and the stenotic right pulmonary artery, since the catheter could not be advanced into the right pulmonary artery (tables 1 and 2). A diagnosis of pulmonary valvular stenosis and isolated stenosis of the right main pulmonary artery was made on the basis of the catheterization and angiocardiographic findings.

Case 5

A 4-year-old Negro boy was admitted to the clinic with a history of frequent episodes of upper respiratory infection. The blood pressures in the right and left arms were 112/50 and 108/50, respectively. The heart was not enlarged. A systolic thrill was felt with maximum intensity in the second and third left intercostal spaces and suprasternal notch. S1 was split, of low frequency, normal intensity, and duration; S2 was well heard at the second left intercostal space where it was split with a dominant aortic component. A grade-IV systolic murmur was best heard in the second left intercostal space and was widely conducted to the left, both in front and back. A grade-IV systolic and diastolic murmur of continuous type was heard best over the second left intercostal space close to the sternum. Roentgenograms showed a mild prominence of the pulmonary artery and enlargement of the left ventricle and atrium. Electrocardiograms showed right and left ventricular hypertrophy. Angiocardiograms revealed poor opacification of the right lung field and blanching at the level of the main pulmonary artery. Right heart catheterization confirmed the diagnosis of stenosis of the right pulmonary artery and patent ductus arteriosus.

After ligation of the ductus examination revealed a systolic murmur over the second and third left intercostal spaces that was transmitted to the back. Repeat right heart catheterization revealed a stenosis of the right pulmonary artery and valvular stenosis (tables 1 and 2).

Case 6

A 4½-year-old Negro girl was admitted to the clinic because of occasional dyspnea and easy fatigability. Examination revealed increased suprasternal and carotid pulsations and pistol-shot femoral pulses; the blood pressure was 116/75. The aortic impulse was in the fifth left intercostal space at the anterior axillary line. A systolic and diastolic thrill and murmur typical of a patent ductus were noted over the upper left intercostal spaces. The murmur was transmitted to both sides of the chest. Roentgenograms showed moderately increased pulmonary blood flow with left ventricular enlargement. An electrocardiogram revealed diastolic over-filling type of left ventricular hypertrophy. Right heart catheterization showed mild pulmonary hypertension with a marked drop in pressure as the catheter was advanced from the main pulmonary artery into the right pulmonary artery, indicating stenosis of the right pulmonary artery and evidence of a patent ductus arteriosus. Angiocardiograms were confirmatory (tables 1 and 2).

Case 7

A 13-year-old Negro boy was referred to the cardiac clinic for a heart murmur. Neck veins were distended and there was a definite increase in arterial pulsation in the neck. The blood pressure was 168/85 and the femoral pulses were bounding. The fingers of both hands were tapered.
The apex of the heart was at the fifth left intercostal space just outside the midclavicular line. Besides the typical findings of patent ductus arteriosus, there was a systolic murmur over the aortic area radiating to the neck with a soft diastolic component. Roentgenograms and fluoroscopy revealed increased hilar pulsations and vascular markings in the left lung field. Electrocardiograms showed left ventricular hypertrophy. Angiocardiograms revealed the left pulmonary vasculature well outlined compared to the right and evidences of a patent ductus arteriosus. Right heart catheterization (tables 1 and 2) revealed stenosis of the right pulmonary artery and patent ductus arteriosus with marked pulmonary hypertension. There was a marked decrease in pressure as the catheter was advanced into the right pulmonary artery close to its origin from the main branch; the pressure tracing had only a vibratory pattern without the distinguishable systolic and diastolic form. Analysis of the blood samples revealed a left-to-right shunt at the aortic-pulmonary level. A preoperative diagnosis of patent ductus arteriosus, stenosis of the right main pulmonary artery, and Marfan's syndrome was made. The ductus was ligated, and the postoperative recovery was uneventful. Examination after ligation of the ductus revealed a grade-IV systolic murmur heard over the aortic area, transmitted to the back and to the right side. Repeat right heart catheterization demonstrated stenosis of the right pulmonary artery and a decrease in the pulmonary hypertension.

Case 8

A 4-month-old Negro girl was admitted to the hospital for cardiac evaluation. The blood pressures in the right arm and right leg were 100/60 and 110/60, respectively. Cardiac examination revealed a forceful apical beat at the fifth intercostal space in the anterior axillary line. A thrill was felt at both the base and the apex. $S_1$ was increased in intensity, split, and best heard at the apex. $S_2$ was split, normal in intensity, and best heard in the second left intercostal space. A

*Phonocardiogram, in a case of isolated stenosis of the right pulmonary artery at its bifurcation from the main pulmonary artery, shows the systolic murmur in the second left and right intercostal spaces and transmitted to the interscapular region and left axilla.*
Figure 2

Preoperative phonocardiogram, in a case of isolated stenosis of the right pulmonary artery distal to the bifurcation of the main pulmonary artery with patent ductus arteriosus, does not show the typical murmur of patent ductus. Postoperatively only the systolic murmur persisted, still heard best in the second right intercostal space.

A 27-month-old Negro boy was admitted for cardiac evaluation. A clinical diagnosis of a ventricular septal defect was made at the age of 6 weeks and was confirmed by cardiac catheterization. The branches of the pulmonary artery were not entered so that their condition was not known (tables 1 and 2). The blood pressures were 90/60 and 100/60 mm. over the right arm and leg respectively. A diffuse apical impulse was felt in the fifth left intercostal space at the anterior axillary line. There was a thrill at the lower left intercostal space and a grade-V harsh pansystolic murmur of a ventricular septal defect was heard along the left sternal border with maximum intensity over the fourth left interspace; it was transmitted to the apex, axilla, and back. Roentgenograms showed increased pulmonary vascular markings and left atrial and ventricular enlargement. An electrocardiogram revealed right ventricular hypertrophy. Angiocardiograms showed stenosis of the right pulmonary artery and evidence of a ventricular septal defect. Right heart catheterization confirmed the diagnosis (tables 1 and 2).
stenal border. A typical murmur of ventricular septal defect was heard, but it was transmitted to the axilla and the back. A grade-II diastolic rumble was heard at the apex. Roentgenograms of the chest showed evidence of increased pulmonary blood flow and left atrial and ventricular enlargement. Electrocardiograms showed left ventricular hypertrophy. Right heart catheterization demonstrated a ventricular septal defect with slightly decreased pulmonary artery pressure, as well as stenosis of the right pulmonary artery (tables 1 and 2). Angiocardiograms revealed the existence of both the ventricular septal defect and a dilated main pulmonary artery which was larger than the aorta, with stenosis of the right pulmonary branch.

Case 10
A 3-year-old Negro boy with a clinical diagnosis of ventricular septal defect, confirmed by angiocardiograms and right heart catheterization (tables 1 and 2), was admitted for re-evaluation of cyanosis of recent onset. The blood pressures were 90/50 and 100/50 in the right arm and left leg, respectively. A grade-III pansystolic murmur and thrill were noted at the second left intercostal space. S2 was loud and split and best heard over the second left intercostal space, close to the sternal border. Right heart catheterization revealed a bidirectional shunt at the ventricular level, pulmonary stenosis of the infundibular type, right ventricular hypertension, and stenosis of the right pulmonary artery (tables 1 and 2). Angiocardiograms showed early opacification of the aorta and diminished vascularity of the right lung. A diagnosis of tetralogy of Fallot was made and the patient was operated on with use of a pump oxygenator. He had an uneventful postoperative recovery.

Discussion
No satisfactory etiology has been given to explain the occurrence of stenosis in the right pulmonary artery. A congenital developmental defect has been cited. That absence of a pulmonary artery or its stenosis has been reported more often on the right side, suggests a common embryologic explanation for both these anomalies. The pulmonary arch is formed by the establishment of a connection between the dorsal aorta and a pair of arteries extending backward from the aortic sac. This is joined by the primitive pulmonary artery. Next, on the right side, the segment of the arch lying distal to the primitive pulmonary artery degenerates. The angle between its proximal part and the primary artery is gradually lost and the segment now forms part of the proximal segment of the adult right pulmonary artery. The distal segment of the left pulmonary arch, however, does not degenerate but forms the ductus arteriosus of the full-term fetus. Embryologically, the main pulmonary trunk is anatomically to the left of the aorta in the midsagittal plane and therefore its left branch has a much shorter and more direct route to the dorsal aorta and thus receives a better and more direct blood supply. Madoff and co-workers10 suggested that these factors might explain the occasional absence of the right pulmonary artery occurring more often on the right than on the left. Isolated stenosis of the right pulmonary artery may be an intermediate stage between the normal development or complete absence of the right pulmonary artery.

We are concerned here with isolated stenosis of the right main branch of a pulmonary artery, with or without associated anomalies. The cases of multiple stenosis of the smaller branches of the pulmonary arteries may very well be due to developmental anomalies that occur in the transformation of the primitive pulmonary artery from the plexus of vessels.
to its adult form. Pulmonary embolism with partial recanalization could result in single or multiple sites of narrowing in branches of both pulmonary arteries. However, pulmonary embolism is unlikely in our group of young patients. Williams, Lange, and Hecht have shown that a considerable degree of constriction must exist before a measurable pressure gradient in the pulmonary artery can be obtained for a given flow. They also demonstrated that a 50 per cent reduction in diameter will result in a drop of only 9 mm. of mercury mean pressure across the constriction. It is obvious, therefore, that minimal stenosis may occur without demonstrable pressure change. Conversely, when the flow and pressure are raised, as in the presence of a patent ductus arteriosus (cases 5 and 7) or in the presence of a ventricular septal defect, a high gradient may occur even though the stenosis may be mild. A moderate stenosis with a coexistent pulmonary valvular (case 4) or infundibular stenosis may only be detected after the latter abnormalities are relieved. It is quite possible that the diastolic murmur heard in our case 7, who also had an associated patent ductus arteriosus, was not due to the ductus alone, in that there was a wide diastolic gradient across the stenotic area prior to surgery. The reduction in this diastolic pressure gradient following ligation of the patent ductus is easily explained by the resultant reduction in pulmonary blood flow, with consequent disappearance of the diastolic murmur due to both the ductus and the stenotic pulmonary artery.

In recent years, this anomaly has been diagnosed with increasing frequency by both clinical and laboratory means. We presume that the normal life span of these patients should not be affected in any way by the presence of this isolated anomaly. In the past, before angiocardiography and cardiac catheterization became part of the routine investigative procedure in undiagnosed cardiac lesions, some of the so-called "functional murmurs" could

Figure 5
Dextroangiocardiogram shows overlapping of the right atrial appendage obscuring the stenotic right main pulmonary artery.
very well have been due to this condition. The systolic murmur in this entity is of the mid-systolic ejection type. It does not involve the first heart sound because it always starts after the closure of the atroventricular valves. An early systolic click is not present as the normal anatomy of the pulmonary valve is not involved, and there is no dilatation of the main pulmonary artery. Since significant right ventricular hypertrophy is not present, ventricular systole is not prolonged. The second sound is normally split with normal intensity of its 2 components over the pulmonary area.

The location of the murmur depends upon whether the stenosis is at the point of bifurcation of the main pulmonary artery or distal to it. In the former site, it is maximal in the second left intercostal space and may even be transmitted to the left axilla or interscapular region (fig. 1). In the latter case (fig. 2) it is maximal in the second right intercostal space and may then be transmitted to the back.

The electrocardiogram of patients with stenosis has a normal pattern unless associated lesions are present. The roentgenograms of the chest may show evidence of decreased vascularity of the right lung as compared to the left (fig. 3). This finding, together with the systolic murmur and the normal electrocar-

diagram, should permit clinical diagnosis of this anomaly. Angiocardiograms usually demonstrate the stenosis of the right main pulmonary artery (fig. 4). At times, the stenotic area may not be visualized because of overlapping of the right atrial appendage (fig. 5).

Right heart catheterization in the isolated form shows no alteration in hemodynamics except for a pressure gradient across the stenotic area (fig. 6). This was obtained in all cases except 1 (case 4) in which the catheter could not be advanced beyond the main pulmonary artery. The pressure-pulse distal to the stenosis may have a vibratory curve (fig. 7), depending upon the degree of stenosis; at times this may make its identification as an artery difficult. Right ventricular and pulmonary hypertension are unlikely to occur unless other congenital cardiac defects are present.

Since the murmurs produced by this anomaly may, in many ways, resemble the group of surgically correctible acyanotic congenital heart disease, it is important to consider the existence of such an anomaly in the differential diagnosis. Foremost among these is pulmonary valvular stenosis because the character, location, and distribution of the systolic murmur usually simulate the entity under discussion. In mild or moderately severe pul-

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monary valvular stenosis, however, with right ventricular pressure below 100 mm. of mercury, an early systolic click is usually present and the second sound over the pulmonary area is widely split (0.06 to 0.12 second) due to the delay in the closure of the pulmonary valves.  
In the severe type with right ventricular pressure over 100 mm. of mercury, the markedly diminished second heart sound over the second left intercostal space, the markedly delayed closure of the pulmonary valve, the presence of a lower parastral lift, and an electrocardiogram showing right ventricular hypertrophy of the systolic overfilling type manifested as a qR or tall R over V4R, V3R, and V1 with or without a tall R in aVR, serve to differentiate it from isolated stenosis of the right pulmonary artery. In atrial septal defects, the systolic murmur that is heard best over the pulmonary area is almost always softer and of less intensity. The wide separation of the 2 components of the second sound together with its relative fixation during the respiratory cycle should help in the diagnosis.

On the basis of our experience with these 10 patients, we believe the auscultatory findings most common to this type of congenital anomaly is a systolic murmur of grade II to IV intensity, heard over the second and third left or right intercostal spaces, depending on the location of the stenosis. This murmur usually radiates to the axillae and the interscapular region. Both the first and second heart sounds in these cases are essentially normal; this makes the differentiation from the other cyanotic congenital cardiac lesions clinically possible, particularly if the roentgenograms demonstrated diminished pulmonary vasculature of 1 lung. A systolic-diastolic murmur reported by others as a finding in this anomaly has not been present in our group of cases, even when there was a gradient of 6 mm. of mercury in diastole across the stenotic area. Such a diastolic murmur may be present in this anomaly if there is a significant diastolic gradient across the stenotic area. In order to produce such a gradient, central pulmonary artery hypertension must be present. Gyllenswärd et al. 6 attributed the diastolic murmur to proximal pulmonary artery dilatation during ventricular systole followed by prolonged emptying into the distal segment during part of the diastolic phase. This was corroborated by experimental studies done by Eldridge et al. 8 However, Gyllenswärd's case was one of multiple peripheral pulmonary artery stenosis with central pulmonary hypertension. This mechanism cannot apply to the isolated stenosis of one branch of the pulmonary artery unless pulmonary hypertension is present due to other causes.

Summary

Ten cases of isolated stenosis of the right main pulmonary artery are presented, 7 with associated anomalies. Although right heart catheterization and angioiardiography are usually necessary to demonstrate this malformation, a knowledge of the existence of this anomaly correlated with auscultatory and roentgenologic findings may often lead to a correct clinical diagnosis.

Addendum

Since submitting this paper for publication, the authors have made a correct clinical diagnosis of this anomaly based on auscultatory and roentgenologic findings in an additional 3 patients. This diagnosis was confirmed by subsequent angiocardiography and cardiac catheterization.

Summario in Interlingua

Es presentate 10 casos de isolate stenosis del artery pulmonar dextero-major, inclus 7 con anormalitates associate. Ben que catheterismo dextero-cardiac e angioiardiographia es usualmente necessari pro demonstrar le presentia de iste malformation, le conosce consideration de su presentia possibile, in correlation con constatationes auscultatori e roentgenologie, resulta frequentemente in le correcte diagnose clinici.

References


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STENOSIS OF PULMONARY ARTERY

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A 20-year-old woman who had experienced repeated attacks of rheumatic fever showed signs of mitral stenosis, atrial fibrillation, and wide Q waves in leads III and aVF. In the previous year deeply inverted T waves of pointed configuration had been present in leads II and III. The patient died of collapse following peripheral embolization. At autopsy mitral, aortic, and tricuspid stenosis with thrombosis in the left and partly in the right atrium was found. The coronary arteries were normal, but the arterioles showed extensive obliterating arteritis. In the posterior wall of the left ventricle there was a large area where the entire myocardium with the exception of a narrow subendocardial layer had been replaced by dense fibrous tissue. Extensive degenerative and fibrotic changes were present throughout the myocardium, and it is probable that the aneurysmal zone of complete fibrosis was due to confluence of islands of rheumatic myocarditis.

LEPESCHKIN
Stenosis of the Right Main Pulmonary Artery: Clinical, Angiocardiographic, and Catheterization Findings in Ten Patients

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