Idiopathic Dilatation of the Pulmonary Artery

By Madhukar Deshmukh, B.S.M.B.B.S., Sulahattin Guvenc, M.D., Lamberto Bentivoglio, M.D., and Harry Goldberg, M.D.

IDIOPATHIC dilatation of the pulmonary artery has been recognized as a clinical entity since 1923, when Wessler and Jaches described the first case. Since dilatation of the pulmonary artery is frequent in many congenital as well as acquired heart lesions, differentiation of this benign anomaly from the latter group is of prime importance. With advances in knowledge of congenital heart disease and improvement in the diagnostic methods, antemortem recognition of this condition has become possible. This report of 13 patients, in whom heart catheterization was performed, aims at drawing attention to this anomaly and elaborating its clinical and physiologic aspects.

Material

The 13 patients of this series were referred to the Bailey Thoracic Clinic for evaluation of their cardiac status. Eight were male and 5 were female, and their ages ranged from 3 to 35 years. All these patients had complete clinical examinations including history, physical examination, complete blood count, Kolmer's serologic reaction, and other pertinent laboratory studies. Roentgen examination of the heart was performed in the 4 conventional positions. Every patient had a 12-lead electrocardiogram and, in some, leads V3R and V4R were recorded. Right-sided heart catheterization was performed in the normal way. A systolic pressure of 30 mm. Hg was regarded as the limit of normal in the right ventricle.

Results

All but one of the patients were referred for cardiac evaluation because of the incidental discovery of a heart murmur during routine physical examinations. In 1 patient the abnormality was noted for the first time in a routine roentgenogram of the chest. The heart murmur in this patient was so faint that it escaped attention. Absence or mildness of symptoms was the prominent finding in this series. Five of the patients complained of mild fatigue and 2 of dyspnea of mild degree on exertion; 1 had pain in the chest and 3 had palpitation.

The abnormal physical findings were limited to cardiac palpation and auscultation. A systolic murmur was heard in the left second and third interspaces, and the pulmonic second sound was accentuated. The character and the intensity of this systolic murmur varied from patient to patient. In 3 patients a systolic thrill was felt where the murmur was loudest, i.e., the second and third left intercostal space. In addition, in 5 patients splitting of the first heart sound was heard at the apex and was interpreted as a systolic ejection click (fig. 1). In 1 patient an early diastolic murmur, blowing in character, was heard in the pulmonary area.

The laboratory studies were within normal limits. Kolmer's serologic reaction was negative in all.

The roentgen examination showed a normal cardiac silhouette and, in striking contrast, dilatation and prominence of the pulmonary artery segment in posteroanterior and right anterior oblique positions (figs. 2 to 4). In one case the dilatation extended to the left and right main branches (fig. 3). The peripheral lung fields were of normal vascularity in each instance, and there was no chamber enlargement. The electrocardiogram was normal in all instances.

Cardiac catheterization showed no evidence of left-to-right shunt. The right ventricular and pulmonary artery pressures were within normal limits. A small systolic pressure gradient was noted across the pulmonic orifice in some cases. The cardiac output was normal. The peripheral oxygen saturation was normal except in patients 4, 5, and 8. In these patients the catheterization was performed under
general anesthesia, and the slight peripheral unsaturation was attributed to diminished pulmonary ventilation (table 1).

Discussion

In the patient described by Wessler and Jaches, the diagnosis of idiopathic dilatation of the pulmonary artery was made on the basis of roentgenographic findings.1 Oppenheimer in 1933 described a series of 8 patients having this condition.2 Two of these had postmortem examination and the main finding was dilatation of the pulmonary artery without any other congenital defect or malformation. The pulmonary vasculature revealed intimal thickening. In the other 6 patients the diagnosis was made on the basis of clinical and roentgenologic similarity with these 2. All the patients in the group had severe symptoms. All but 1 of them had shortness of breath; 5 had cyanosis, and 4 had evidence of congestive failure. In this respect the above group of patients differs markedly from subsequent reports4-6 in which the patients had few or no symptoms. It is difficult, therefore, to maintain the diagnosis of idiopathic dilatation of the pulmonary artery in his patients. It is probable that they had some other disease, such as primary pulmonary hypertension with secondary dilatation of the pulmonary artery.

Kourilsky and his associates7 presented 10 cases of this condition, 9 with postmortem examination. Only 6 cases can be accepted as having idiopathic dilatation of the pulmonary artery because 2 had atrial septal defect and the third had mitral stenosis.

Gold8 stressed the presence of a hypoplastic aorta in the diagnosis of idiopathic dilatation of the pulmonary artery. In the absence of the hypoplastic aorta, he considered the dilatation of the pulmonary artery to be due to Ayerza's disease or primary pulmonary sclerosis. The deformity was thought to result from unequal division of the truneus arteriosus communis.7,8 Hypoplasia of the aorta, however, is an inconstant finding,7 and its estimation in clinical practice by routine roentgenologic examination is far from satisfactory.

Differentiation of idiopathic dilatation of the pulmonary artery from other congenital and acquired lesions with dilated pulmonary

Figures 1 and 2
Figure 3
Case 7. Roentgenograms of the heart showing dilatation of the main pulmonary artery segment and its extension to the main branches.

Figure 4
Case 10. Roentgenograms of the heart showing more severe dilatation of the pulmonary artery.

artery is of primary importance because most of the latter abnormalities can be corrected by surgical procedures (table 2). Idiopathic dilatation of the pulmonary artery is on the other hand a benign anomaly and does not require surgical intervention. Clinically, an important diagnostic lead is absence or mildness of symptoms.4-6 Careful history taking in some of our symptomatic patients suggested anxiety of iatrogenic origin as the cause of their symptoms rather than the presence of cardiac disease. Severe restrictions were imposed because of the murmurs. In fact, 1 man was rejected from the military service because of the murmur, and 1 woman was advised not to have children.

The systolic murmur, which is the most prominent physical finding in this condition, is quite inconstant in character and intensity.5,6 In some cases the murmur is quite rough and resembles that of pulmonary stenosis, whereas in others it is blowing in character, giving the auscultatory impression of an atrial septal defect. The systolic ejection click, which we heard in 5 patients, is of no diagnostic aid because it may be present in other conditions in which there is dilatation of the great vessels (fig. 1).9

Roentgen examination of the heart is important, particularly for comparison of the
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Table 1

<table>
<thead>
<tr>
<th>Case</th>
<th>PCV</th>
<th>Pressure (mm. Hg)</th>
<th>BA</th>
<th>Oxygen content of blood (vol. %)</th>
<th>Oxygen saturation (%)</th>
<th>Cardiac output (L/min)</th>
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<td></td>
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<td>RV</td>
<td>RA</td>
<td>SVC</td>
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<td>(64)</td>
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* Catheterization performed under general anesthesia.
† Figures in parentheses represent mean pressures.
BA, brachial artery; IVC, inferior vena cava; PA, pulmonary artery; PCV, pulmonary capillary venous; RA, right atrium; RV, right ventricle; SVC, superior vena cava.

size of the pulmonary artery with the total heart size and with the peripheral pulmonary arteries. The paradox of enlarged main pulmonary artery and diminished vascularity of the periphery of the lung fields, so characteristic of pulmonary stenosis, is absent in this condition. The hyperemic lung fields and the hilar dance, which are common in large left-to-right shunts, are also lacking. The heart size is normal; care should be taken, however, because the prominent pulmonary artery segment may give a false impression of cardiac enlargement. The cardiothoracic ratio is within normal limits.

Electrocardiography is very useful in the differential diagnosis. The patterns of so-
Table 2

Conditions Accompanied by Dilatation of the Pulmonary Artery

<table>
<thead>
<tr>
<th>Congenital A.</th>
<th>With intracardiac or extracardiac shunts</th>
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<tbody>
<tr>
<td>Atrial septal defect</td>
<td>Anomalous pulmonary venous drainage (partial or total)</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>Patent ductus arteriosus</td>
</tr>
<tr>
<td>Aortopulmonary septal defect</td>
<td>Aortic-right heart fistula</td>
</tr>
</tbody>
</table>

B. Without shunts
- Pulmonary stenosis (poststenotic dilatation)
- Congestion of the pulmonary artery or its branches
- Pulmonary vein stenosis
- Idiopathic dilatation of pulmonary artery

Acquired A.
- Cardiac diseases
  - Mitral valve disease
  - Left atrial tumors
  - Chronic left ventricular failure
- Pulmonary diseases
  1. Parenchymal
     - Pulmonary agenesis, cysts, extensive pulmonary emphysema, and fibrosis, granulomas (sarcoid, berylliosis, milary tuberculosis), carcinoma of the lung, and Hamman-Rich syndrome.
  2. Vascular
     - Primary pulmonary hypertension
     - Secondary pulmonary hypertension due to repeated pulmonary emboli, collagen diseases, schistosomiasis,
     - Syphilis
- Hyperkinetic circulatory states
  - Anemia
  - Hyperthyroidism
  - Paget’s disease
  - Beriberi
  - Systemic A-V fistulas

called diastolic and systolic overload of the right ventricle found, respectively, in atrial septal defect and pulmonary stenosis are not present. The pattern of diastolic overload of the left ventricle commonly found in interventricular septal defect and patent ductus arteriosus is also absent. The electrocardiogram in this condition is normal; if significant abnormality is present, other types of cardiac disease should be suspected.

In spite of the well-marked clinical, radiologic, and electrocardiographic features, the diagnosis of idiopathic dilatation of the pulmonary artery is one of exclusion. We think that cardiac catheterization is superior to angiography as a diagnostic tool. Absence of left-to-right shunt, normal pressure relationships in the cardiac chambers, and normal oxygen saturation are the important findings. A small pressure gradient between right ventricle and pulmonary artery during the systolic ejection phase may exist (fig. 5). Chisholm advocated the concept of trignonodation of the pulmonary orifice as the mechanism for the basal systolic murmur and the relative stenosis of the pulmonary orifice in simple dilatation of the pulmonary artery. Since the pressure difference in idiopathic dilatation of the pulmonary artery is present without right ventricular hypertension, its probable cause, in our opinion, is the deceleration of blood flow in the dilated pulmonary artery, rather than true anatomic obstruction of the pulmonary valve. This is quite apparent from the hemodynamic expression: resistance = pressure/flow. Flow being constant, pressure varies directly with resistance. Resistance in its turn is related inversely to the radius of the vessel as expressed in the formula $R = \frac{8LV}{\pi r^4}$, in which $L$ and $r$ are the length and the radius of the tube and $V$ is the viscosity of the fluid. In idiopathic dilatation of the pulmonary artery, resistance to blood flow in the pulmonary artery is diminished because of the dilatation of the lumen; consequently there is a proportional drop in pressure, which accounts for the pressure gradient between the right ventricle and the pulmonary artery during systole.

Greene and associates reviewed the literature on idiopathic dilatation of the pulmonary artery and established the following pathologic criteria for its diagnosis: (1) simple dilatation of the pulmonary trunk with or without involvement of the rest of arterial tree; (2) absence of intracardiac or extracardiac shunts; (3) absence of chronic cardiac or pulmonary disease; (4) absence of arterial disease, such as syphilis, or more than minimal atheromatosis or arteriosclerosis of the pulmonary vascular tree. All these criteria, except atheromatosis of the pulmonary vasculature, can be evaluated clinically and by routine laboratory tests, cardiac catheterization, and pulmonary function studies. All the
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patients in our group fulfilled these clinical criteria. In addition all had normal hemodynamic findings. In our opinion, the normal pressure in the right ventricle and pulmonary artery should be introduced as the fifth criterion in the diagnosis. The presence of even a mild degree of right ventricular hypertension associated with a systolic gradient across the pulmonic valve should be considered as sufficient evidence for the diagnosis of pulmonary stenosis.

Nothing is known about the etiology of this lesion and relatively little about its pathogenesis and natural history. Assman postulated an unequal division of the truncus arteriosus communis as the possible mechanism, and Kourilsky et al., Laubry and associates, and Gold supported this idea because of the presence of a hypoplastic aorta in some of their cases. Kaplan and others thought the anomaly represented one aspect of the mal-development of the entire pulmonary tree and of congenital weakness of the arterial wall. Our series does not contribute to this issue.

Prognostic evaluation of idiopathic dilatation of the pulmonary artery is limited by the number of clinically studied cases and the lack of adequate follow-up. In the only acceptable autopsied series, however, the 6 proved cases died of unrelated diseases at the ages of 56, 63, 78, 82, 88, and 92. It is therefore believed that idiopathic dilatation of the pulmonary artery is a benign congenital malformation.

Summary

Thirteen patients with idiopathic dilatation of the pulmonary artery are described and their clinical, roentgenologic, electrocardiographic, and hemodynamic features are elaborated. Absence or mildness of symptoms was the most significant finding in this series. A pulmonic systolic murmur was constantly present. The roentgenogram showed various degrees of dilatation of the pulmonary artery in the presence of normal heart size. The electrocardiogram was normal. The hemodynamic studies showed a mild systolic pressure gradient across the pulmonic valve in some cases in the presence of normal right ventricular pressure. A possible explanation of this finding is discussed. Idiopathic dilatation of the pulmonary artery is a benign lesion and does not affect cardiac function to any appreciable degree.

Summario in Interlingua

Es presente 13 casos de idiopathic dilatation pulmonar-arterial, e le observationes clinic, roentgenologic, electrocardiographie, e hemodynamic es describite detaliatemente. Le absentia de symptoms o le presentia de solmente leve symptoms eseva le plus significative constatation in iste serie. Un murmur pulmonar-systolic eseva constantemente presente. Le roentgenogramma monstравa varie grados de dilatation del alteria pulmonar in le presentia de normal dimensiones cardine. Le electrocardiogramma eseva normal. Le studios hemodynamic revelava un leve gradiente de pression systolique a transverso le valvula pulmonar in certe casos in le presentia de normal pressiones dextero-ventricular. Un explication possibile de iste observation es offerite e discutite. Dilatation idiopathic del arteria pulmonar es un lesion benigna e non affic le function del corde a grados notabile.

References

4. GRISHMAN, A., STEINBERG, M. F., AND OPPEN-

A case of congenital complete heart block is described, and the literature reviewed. A brother had also presented bradycardia and had bouts of cyanosis with Stokes-Adams episodes. The incidence, etiology, symptomatology, and prognosis of this condition are reviewed. Other congenital anomalies are usually found, the most common being an intraventricular septal defect. If congenital complete heart block is the only lesion, the prognosis is good and there is usually no limitation on the patient's activities. Even pregnancy and high-altitude flying have not been contraindicated in reported cases. In this patient, the diagnosis was suspected prenatally because of the slow, regular fetal heart rate (64 to 68 per minute). This condition can be confused with fetal distress and be the cause of unnecessary interference with pregnancy or labor. The criteria for the diagnosis of congenital complete heart block are (1) a slow pulse at an early age, (2) proof by graphic methods, (3) other signs of congenital heart disease, and (4) absence of a history of infection, such as diphtheria or rheumatic fever. The pulse rate is usually from 40 to 60 and may increase with exercise or after atropine or epinephrine. The stroke volume is greater than normal but the cardiac output remains normal.

MAXWELL
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