Cor Triloculare Biatriatum

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Cor triloculare biatriatum is a rare form of congenital heart disease that needs to be considered in the differential diagnosis of the important group of patients with cyanotic congenital heart disease with pulmonary hypertension. The clinical and pathologic features of an example of this disease are reported and discussed in a review of previously reported cases. The small pulmonary blood vessels showed some features of hypertensive pulmonary vascular disease and evidence of an increased bronchial arterial collateral circulation to the lung. An interpretation of these histologic changes relating them to the abnormal pulmonary physiology in cor triloculare biatriatum is suggested.

Cor triloculare biatriatum is a rare form of heart disease that accounted for 13 of the 1,000 cases of congenital heart disease collected by Abbott and 7 of Clawson's 141 cases. In 1954 van Buchem and his co-workers described a case with data gained from angiocardiography and cardiac catheterization, and estimated that 60 cases had been reported, only 3 not having been associated with transposition. The purpose of this paper is to present the clinical and pathologic features of another patient who survived to the age of 23 years, together with the results of special investigations, including a histologic study of the pulmonary vasculature. There have been very few descriptions of the lung in this disease although pathologic changes in the small pulmonary blood vessels exert great influence on both the clinical picture and the prognosis.

Case Report

A married woman, aged 23 years, who had been deeply cyanosed and dyspneic on exertion since birth, was admitted to hospital for investigation. Congenital heart disease was not diagnosed until the age of 3 years. In childhood she was unable to join in games or physical training on account of dyspnea but later she led a normal life and worked in an office, suffering from only occasional ankle swelling. She was able to lie flat in bed but walking up hills rapidly produced breathlessness. In the year preceding admission her dyspnea worsened and became associated with an unproductive cough and persistent ankle edema. She had 2 epistaxes but no hemoptysis. On examination there was severe central cyanosis and clubbing of the fingers and toes. There was no cardiac enlargement, and the apex beat was tapping in character. There was a heave over the lower sternum, suggesting right ventricular hypertrophy. On auscultation the second sound in the pulmonary area was loud and split and in the third left intercostal space there was a late systolic murmur. The systemic blood pressure was 135/80 mm. Hg. The hemoglobin level was 130 per cent (19.3 Gm. per cent) and the hematocrit 69 per cent. The electrocardiogram showed sinus rhythm at 85 per minute and right axis deviation with right ventricular preponderance. A teleradiogram showed a global cardiac silhouette without cardiac enlargement or prominence of the pulmonary conus. In the left anterior oblique view, right ventricular enlargement was considered to be present, although the other chambers were of normal size. There was an increased density of the hilar shadows, which were stippled in character suggesting a collateral circulation to the lung; hilar dance was absent. The angiogram showed a considerable shunt of blood passing from the right side of the heart into the aorta. These angiocardiographic appearances, coupled with the subsequent inability to demonstrate the pulmonary artery blood pressure at cardiac catheterization, led to an erroneous diagnosis of Fallot's tetralogy. In the right atrium the mean blood pressure was -2 mm. Hg and the blood oxygen saturation was 68 per cent. Attempts to catheterize the right ventricle and the pulmonary artery were abandoned as they provoked frequent runs of extrasystoles. The oxygen saturation of the brachial artery blood was 71 per cent. The patient was considered suitable for a Blalock-Taussig operation. At thoracotomy the main pulmonary artery was found to be very large and tense, causing great technical difficulty. The patient died a few hours later.

Autopsy Report

The heart weighed 250 Gm. (fig. 1). The right atrium was of normal volume and its walls of normal thickness; it contained no antemortem thrombus. There was no atrial septal defect. The right atrium opened into a single ventricle through a right atrioventricular orifice, which was 8 cm. in circumference (fig. 2). The right atrioventricular valve was

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tricuspid. The cusp analogous to the septal cusp of the normal tricuspid valve was large and free and not tethered as in the normal heart (fig. 2). The bases of the cusps analogous to the anterior, medial, and inferior cusps of the normal tricuspid valve measured 3, 3.5, and 1.5 cm. respectively, while the corresponding maximum lengths were 1.2, 1.5, and 1.0 cm. There were 2 main papillary muscles attached to the valve; one, 14 mm. in width at its base, provided chordae tendineae for attachment to the free margins of the anterior cusp while the other, 7 mm. in width at its base, provided chordae for attachment to the contiguous margins of the septal and anterior cusps (fig. 2). One of the columns carneae running from the base of the smaller papillary muscle insinuated between the right and left atrioventricular valves but it did not form a partial ventricular septum. The right atrioventricular orifice led into a single dilated ventricle, which was 11 mm. thick at its base, 8 mm. half way between base and apex, and 2 mm. in the region of the apex (fig. 2). The right ventricular cavity was subdivided into a main chamber, which gave rise to the dilated pulmonary artery, and a rudimentary chamber 33 mm. in length from which the aorta arose (fig. 2). The left atrium was of normal volume and thickness and contained no antemortem thrombus. The mitral valve was of normal structure.

There was transposition of the great vessels, the aorta lying in front and to the left of the pulmonary artery (fig. 3). The aorta was smaller than the pulmonary artery with a caliber of 16 mm. and a wall thickness of 1.5 mm., the caliber of the pulmonary artery being 20 mm. and the wall thickness 3 mm. Unlike the ascending aorta, the pulmonary artery was dilated and atheromatous, the areas of atheroma being continuous with others on the tricuspid and mitral valves. The pulmonary valve consisted of 3 semilunar cusps: 1 was anterior, 1 posterolateral, and 1 posteromedial. The aortic valve also consisted of 3 semilunar valves: 1 was anterior, 1 posteromedial, and 1 posterolateral. There were 2 coronary arteries (fig. 1). The left arose normally from the left posterior sinus but the right coronary artery arose abnormally from the right posterior sinus. This latter artery, which normally arises from the anterior sinus, divided into 3 main branches that ramified over the anterior,
Fig. 3. View of the base of the heart showing the right and left atria (RA and LA) and the transposed aorta (A) and pulmonary artery (PA). The pulmonary artery was dilated and thick walled compared to the aorta, and showed a patch of atheroma, indicated by a white arrow.

HEATH

lateral, and posterior aspects of the right ventricle. The left coronary artery divided into a descending branch that supplied the anterior and marginal portions of the left ventricle and a circumflex branch that passed to supply the posterior surface of the left ventricle. The coronary arteries were not atheromatous.

**Pulmonary Vasculature**

The lung was very vascular owing to numerous thin-walled blood vessels lined by a single elastic membrane, which appeared to arise from both bronchial and pulmonary arteries (fig. 4C).

Those from the bronchial circulation were derived from true bronchial arteries (fig. 4B), the vasa vasorum of elastic pulmonary arteries (fig. 4A), and the visceral pleural arteries (fig. 4D). The bronchial walls and the adventitia of the elastic pulmonary arteries contained numerous dilated, thin-walled blood vessels, lined by a single elastic lamina, which distributed many branches to the lung parenchyma (fig. 4A and B). Pleural bronchial arteries were prominent as vessels about 250 μ in diameter, which consisted of an outer layer of circular smooth muscle and an inner layer of longitudinal smooth muscle separated by a thick internal elastic membrane, and a faintly staining external elastic membrane that was external to the circular muscle (fig. 4D). The internal elastic membrane was split to form fibrillae that surrounded the longitudinal muscle bundles in the subendothelial layer.

The muscular pulmonary arteries (100 to 1000 μ in diameter) had a thick muscular media, intimal proliferation of fibrous tissue leading to almost total occlusion of the vessels and a thick adventitia; they were surrounded by clusters of their dilated thin-walled branches, which were lined by a single elastic lamina (fig. 5). One muscular pulmonary artery, 80 μ x 200 μ, had a media 32 μ thick and was surrounded by dilated branches about 150 μ in diameter that were indistinguishable from those of the bronchial arteries. These thin-walled branches of pulmonary and bronchial arteries anastomosed to form a network of vessels in the lung parenchyma, the average diameter being 150 μ, although some were as large as 600 μ; in some there were eccentric intimal thickenings that appeared to be organizing blood clot.

None of the pulmonary arterioles (less than 100 μ in diameter) had a muscular media with 2 elastic laminae, a configuration usually found in association with pulmonary arterial hypertension and the type of muscular pulmonary artery described above. Instead they consisted of a single elastic lamina between the endothelium and a thin adventitia and very few showed the normal age change of intimal sclerosis. The alveolar walls were distended by small pulmonary arterioles and capillaries, 10 to 20 μ in diameter (fig. 4C).

The elastic pulmonary arteries (more than 1000 μ in diameter) were normal apart from the adventitia, which contained many branched vasa vasorum as described above (fig. 4A). The media was about 10 per cent of the external diameter of the vessel.

In serial sections of the only block of lung available for examination large branches of a pulmonary and a bronchial artery came into close apposition (fig. 6) and their radicles appeared to be forming a bronchopulmonary anastomosis distinct from the numerous anastomoses between the much smaller thin-walled branches of bronchial and pulmonary circulations in the lung parenchyma already described. Unfortunately the actual point of communi-
Fig. 4. Sections of lung stained by Verhoeff’s and van Gieson’s stains to demonstrate elastic tissue, illustrating the increase in blood supply to the lung by the various branches of the bronchial arteries. A. Dilated vasa vasorum in the adventitia of an elastic pulmonary artery (X 95). B. Dilated branches of true bronchial arteries in the wall of a small bronchus (X 30). C. Dilated, thin-walled blood vessels in the lung parenchyma derived from the bronchial and pulmonary arteries (X 38). D. Prominent pleural bronchial artery (X 95).

cation was not reached in the block and, as no further lung tissue was available for study, proof of the existence of the anastomosis was not obtained.

Discussion

Symptomatology

Most of the reported adolescent or adult cases of cor triloculare biatriatum in common with the present patient have had few symptoms and have led fairly normal lives with dyspnea, cyanosis, and tiredness only on marked exertion, but these cases are artificially selected in that they have probably had adaptive mechanisms that have enabled them to survive to adult life. Most have been either acyanotic or only slightly cyanosed, with autopsy revealing a large pulmonary artery arising from the main part of the common
ventricle, allowing a great volume of blood to be oxygenated in the lung. The commoner type of case with severe cyanosis from birth is characterized by activity being considerably curtailed by dyspnea and palpitation. In such cases the pulmonary artery is small and arises from the rudimentary chamber of the common ventricle.

**Signs**

A systolic murmur such as that heard in the present patient has been noted in most of the reported cases. This murmur is usually maximal to the left of the sternum in the second to fourth intercostal spaces, as in the present case and those reported by Edwards and Chamberlain, Rogers and Edwards, and van Buchem and his co-workers, but it may be best heard at the apex. When there is associated dextrocardia, the systolic bruit will naturally be maximal over the right side of the chest. The murmur may be accompanied by a thrill. The typical murmur may be absent in infancy as in the case of a 3-day-old girl described by Richman. A diastolic element may occur to produce distinct systolic and diastolic murmurs, a "to and fro" murmur, or a continuous murmur. It is probable that, as in pulmonary atresia and tricuspid atresia, the "to and fro" and continuous murmurs are due to a collateral bronchial circulation. Tausig believed that the murmurs lack the rasping quality so characteristic of most forms of congenital heart disease as there is no shunting of blood from one chamber to another. The second sound in the pulmonary area is loud, and indicative of pulmonary arterial hypertension, in those cases with a large pulmonary artery from the main chamber of the common ventricle.

In most cases of cor triloculare biaatriatum with transposition, there is an increased pulmonary blood flow and a diminished blood supply to the rest of the body. This reduced
systemic blood flow is expressed by a frail infantile build.\textsuperscript{5, 7, 18} The patient of van Buchem and his co-workers\textsuperscript{5} had "always been very thin" and weighed 37 Kg. with poorly developed musculature and slight development of mammary glands.

It might be expected that central cyanosis would be severe in this disease since arterial and venous blood both enter the common ventricle, and indeed in some cases, as in the present patient, persistent cyanosis from birth has been reported.\textsuperscript{5, 9, 12, 13} However, cyanosis was absent at the age of 9 years in a patient described by Glendy and co-workers\textsuperscript{7} and in 2 cases reported by Young\textsuperscript{19} and Mann\textsuperscript{20} and was never prominent in a man of 49 years reported by Herndon and his colleagues.\textsuperscript{11} Central cyanosis was present only at birth and death in Kornblum's\textsuperscript{10} patient, while the patients of Greenberg\textsuperscript{8} and van Buchem and his associates\textsuperscript{8} were only slightly cyanosed. Brown\textsuperscript{21} thought that the explanation for absent or only slight cyanosis lay in the usually associated transposition, which brought the aorta close to the mitral valve and the pulmonary artery near the tricuspid valve, with little chance for any great admixture of venous and arterial blood. Rogers and Edwards\textsuperscript{6} believed that the moderate degree of central cyanosis in 6 of their 9 patients with cor triloculare biaatriatum was related to the absence of pulmonary stenosis or atresia, which allowed an adequate pulmonary blood flow. Taussig\textsuperscript{18} believed the absence or presence of cyanosis depended on the volume of blood reaching the lungs for aeration, for when the pulmonary artery arises from the rudimentary chamber (see discussion on pathology) it is of small caliber and allows only a small volume of blood to reach the lung for aeration. Thus a small volume of oxygenated blood is mixed with a large volume of unoxgenated venous blood in the common ventricle, leading to intense cyanosis. Conversely, if there is transposition so that the pulmonary artery arises from the main ventricle, it is large, allowing a large volume of oxygenated blood to return to the ventricle to mix with a small volume of unoxgenated venous blood. Hence cyanosis is minimal or absent as in the case of Greenberg.\textsuperscript{8}

As would be anticipated, the hemoglobin level is high in those cases where cyanosis is both early and persistent. Thus in the present case the hemoglobin level was 130 per cent while levels of 170,\textsuperscript{6} 122,\textsuperscript{12} and 135 per cent\textsuperscript{13} have also been recorded. In acyanotic cases the hemoglobin level is normal; it was 80 per cent in the patient of Glendy and co-workers.\textsuperscript{7}

Although cardiac enlargement may be marked,\textsuperscript{5, 7, 12, 13} the heart may be of normal size, as in the case under discussion.

Electrocardiography

Taussig\textsuperscript{18} pointed out that the electrocardiogram is of little value in diagnosing the anomaly under discussion for it may or may not show right axis deviation; it did occur in the present case and was reported by Glendy and associates,\textsuperscript{7} Rogers and Edwards,\textsuperscript{6} and Edwards and Chamberlain.\textsuperscript{18} The course of the bundle of His in cor triloculare biaatriatum must be grossly abnormal but, although evidence of a disturbance of the conduction of impulses through the ventricle has occasionally been reported, intracardiac conduction time is usually normal and the QRS complex normal in appearance.\textsuperscript{18} The electrocardiogram of the patient of van Buchem and his colleagues\textsuperscript{8} showed incomplete right bundle-branch block but these authors also remarked that, despite the absence of a ventricular septum, the QRS complexes had been repeatedly found to show no abnormality in cor triloculare biaatriatum. Rogers and Edwards\textsuperscript{6} observed left axis deviation in 2 of their 4 patients with cor triloculare biaatriatum and believed this to be of some clinical importance, as left axis deviation in the presence of central cyanosis is usually suggestive of tricuspid atresia.\textsuperscript{17}

Radiology

In cor triloculare biaatriatum the cardiac silhouette may be rounded or globular,\textsuperscript{8} as in the present case. In infants the rudimentary chamber occupies the position of the normal pulmonary conus and produces a bulge in this region so that, although the heart is not really large, the marked dilatation in the region of the conus erroneously suggests that the heart is huge.\textsuperscript{18} Marked cardiac enlargement has been reported by Glendy and co-workers,\textsuperscript{7} Edwards
and Chamberlain, and Rogers and Edwards. However, in the left anterior oblique position their is no projection of the right ventricular shadow to the chest wall, proving the absence of right ventricular hypertrophy. The combination of the posteroanterior and left anterior oblique views forms a very characteristic radiologic picture and makes it possible to diagnose cor triloculare biaatriatum radiologically in young children. When the pulmonary trunk arises from the common ventricle, there is an excessive blood flow to the lungs leading to pulmonary congestion.

As the patient survives to childhood, the heart becomes long and narrow and the prominence of the pulmonary conus may disappear, as in the present case and as noted by Taussig and van Buchem and his colleagues. The absence of a prominent pulmonary conus does not imply that the pulmonary trunk is small, as in the present case, where the pulmonary artery was shown at autopsy to be big but transposed. If the pulmonary artery arises from the common ventricle, conspicuous hilar shadows and hilar pulsation may occur, although hilar dance was absent in the patient of Edwards and Chamberlain. In the present case there were conspicuous hilar shadows but no hilar pulsation. The shadow of the aortic knuckle produced by the small aorta arising from the rudimentary ventricle is small. In this way, in the adult, the combination of hilar pulsation and a prominent pulmonary conus may suggest Eisenmenger’s complex or any of the congenital cardiac disorders associated with severe pulmonary hypertension, and this makes it very difficult to diagnose cor triloculare biaatriatum radiologically in older children or adults. On the other hand, if a large aorta arises from the common ventricle, while a small pulmonary artery arises from the rudimentary ventricle, there is no prominence of the pulmonary conus or increased hilar shadows and the cardiac silhouette is suggestive of Fallot’s tetralogy.

Angiocardiography

Angiocardiography demonstrates simultaneous filling of the great vessels from a single ventricle, although the dilated vessel arising from the main chamber of the ventricle will cast a denser shadow than the vessel springing from the rudimentary chamber. Thus in the patient of van Buchem and his associates 2 seconds after the injection of radiopaque dye there was filling of the main and rudimentary ventricle resulting in large ventricular shadow; while the aortic shadow was only faint, the pulmonary artery produced a distinct shadow and dye remained in this vessel even after 5.5 seconds. In the present case early filling of the aorta from the right ventricle led to an erroneous diagnosis of Fallot’s tetralogy.

Cardiac Catheterization

Cardiac catheterization gave little information in the present patient, as the introduction of a cardiac catheter into the right atrium provoked numerous runs of extrasystoles. However, van Buchem and his colleagues found it possible to enter the common ventricle, pulmonary artery, and aorta in their patient; they demonstrated that there was severe pulmonary arterial hypertension (125/70 mm. Hg), the blood pressures in the pulmonary artery being equivalent to those in the aorta and the common ventricle. The blood oxygen saturation in the pulmonary artery was 75 per cent, a value almost identical with those found for the blood in the aorta and the common ventricle. The findings of van Buchem and his associates prove that pulmonary arterial hypertension occurs in cor triloculare biaatriatum and provides a basis for the hypertensive changes in the pulmonary vessels in the present case.

Blood Oxygen Saturation

This is of diagnostic importance in cor triloculare biaatriatum since, even though cyanosis may not be detectable, there is always an increased amount of reduced hemoglobin in the arterial blood. In the case of van Buchem and his co-workers the brachial artery blood oxygen saturation was 77 per cent, while in the present case it was 71 per cent. Oxygen inhalation will not raise the blood oxygen saturation to normal. This finding is of importance in distinguishing cor triloculare biaatriatum from Eisenmenger’s complex, a disease with which it may be confused radiologically. In Eisenmenger’s complex the blood oxygen saturation in the pulmonary
artery and right ventricle is lower than in the aorta\textsuperscript{3, 22} in contrast to the condition under discussion where the blood oxygen saturation is the same in the common ventricle, pulmonary artery and aorta.\textsuperscript{3}

**Pathology**

In cor triloculare biaatriatum the ventricular septum is absent or rudimentary, and both tricuspid and mitral orifices communicate with a common ventricle. In the present case, a dividing muscular band, such as described by Taussig,\textsuperscript{24} was found at the outflow tract running from the base of the dorsal wall to the ventral wall. This band, regarded by some authors as a rudimentary ventricular septum and by others as an aberrant crista supra-ventricularis was considered by Taussig\textsuperscript{18} to develop as follows. When the primitive cardiac tube bulges forward and swings to the right, the anterior portion of it becomes the bulbus cordis and the posterior loop becomes the common ventricle and normally the ridge separating bulbus from ventricle atrophies and the aortic septum develops. The aorta arises from the common ventricle posterior to the bulbus cordis while the anterior portion forms the outflow tract of the right ventricle from which the pulmonary artery arises. In cor triloculare biaatriatum the development of the heart is arrested at this stage, so that the bulbus cordis persists as a rudimentary outlet chamber. Both great vessels may arise from the primitive outflow tract or one may arise from the common ventricle and one from the rudimentary outlet chamber. This conception of the origin of the great vessels explains to some extent the presence or absence of cyanosis. If both great vessels arise from the diminutive chamber, the mixture of arterial and venous blood is approximately equal and the patient is cyanosed but if one of the great vessels arises from the common ventricle, the mixing is disproportionate. When the pulmonary artery arises from the main common ventricle a large volume of blood goes to the lung for aeration and much oxygenated blood returns to the left atrium to mix with the small amount coming from the systemic veins, cyanosis is usually absent or minimal. Conversely, when the aorta arises from the common ventricle, the situation is reversed and cyanosis is frequently intense. However, this is not invariably so, for in the present case in which deep cyanosis occurred, the pulmonary artery was large and arose from the main chamber of the common ventricle, while in other reported cases\textsuperscript{11} cyanosis was never prominent although the aorta arose from the main ventricular chamber. Rogers and Edwards\textsuperscript{6} believed that the band usually divides the ventricular outflow tract into a large dorsal subpulmonary part and a small ventral subaortic part. This is certainly true in the case under discussion in which not only was the subpulmonary region bigger than the subaortic outlet chamber but the pulmonary artery was also larger than the aorta. This added disproportion in the size of the great vessels has been noted in previous reports.\textsuperscript{7, 13} Occasionally the pulmonary artery arises from a persistent truncus arteriosus.\textsuperscript{12}

In most cases, as in that under discussion (fig. 3), transposition of the great vessels has been associated with cor triloculare biaatriatum, the aorta lying in front and to the right of the pulmonary trunk. In some cases the aorta lies to the left.\textsuperscript{6, 7, 10} It is exceedingly rare to find cor triloculare biaatriatum unassociated with transposition, although it has been reported.\textsuperscript{4, 5, 24}

Many other congenital anomalies as well as transposition occur in association with cor triloculare biaatriatum. Rogers and Edwards\textsuperscript{6} found hypoplasia of the aortic arch and patent ductus arteriosus, bicuspid pulmonary and aortic valves, and mongolism in their 9 patients with cor triloculare biaatriatum. Patent ductus arteriosus has also been reported by Favorite\textsuperscript{9} and Richman,\textsuperscript{14} while associated atrial septal defect has been described by Glendy and co-workers,\textsuperscript{7} Mehta and Hewlett,\textsuperscript{12} and Richman.\textsuperscript{14}

The heart in cor triloculare biaatriatum may be of normal size (fig. 1) but it is frequently large and heavy\textsuperscript{7, 19} with thick ventricular walls.\textsuperscript{5, 7} In particular, the right atrium may be dilated and hypertrophied.\textsuperscript{5, 12}

**Pulmonary Vessels**

Edwards and associates\textsuperscript{13, 22} stated that in cor triloculare biaatriatum the undivided ventricle ejects blood into both the aorta and the pul-
monary trunk and the relative volume of blood flowing into each of these vessels will depend upon the level of resistance in the systemic vascular bed as compared with that in the pulmonary. If the pulmonary artery blood pressure were normal, a great flow of blood to the lungs would occur at the expense of systemic blood flow and this may explain deaths from cor triloculare biaatrium in infancy. Edwards\(^2\) considered that patients who live to adult life, such as the woman reported here, probably have a regulatory mechanism such as a reduced caliber of the pulmonary vessels that diminishes pulmonary blood flow. He concluded that the small pulmonary vessels in the anomaly under discussion would probably show similar changes to those found in association with pulmonary hypertension. In fact, in the present case the muscular pulmonary arteries showed medial hypertrophy and intimal fibrosis with dilated thin-walled branches, appearances found by Brewer\(^2\) and Heath and co-workers\(^6\) to be characteristic of pulmonary arterial hypertension. There were, however, no vessels below 100 \(\mu\) in diameter with a distinct media and internal and external elastic laminae, which have been found in association with pulmonary arterial hypertension in Eisenmenger's complex,\(^7\) patent ductus arteriosus with pulmonary hypertension,\(^8\) and idiopathic pulmonary hypertension.\(^9\) Similar findings in the pulmonary vasculature have been reported by Edwards and Chamberlain\(^10\) and a photomicrograph of small pulmonary blood vessels in a case of cor triloculare biaatrium described by Favorite\(^9\) shows dilated thin-walled vessels. In the present case there was also one probable bronchopulmonary anastomosis together with hypertrophy of the various branches of the bronchial arteries (true bronchial, pleural, and vasa vasorum), which supplied the lung with numerous thin-walled vessels. These changes might be interpreted as follows.

The pulmonary arterial hypertension produces changes in the small muscular pulmonary arteries with partial occlusion of the vascular lumina. There results an increased resistance to pulmonary blood flow and a greater flow of blood to the systemic circulation. While the patient can survive because of the increased systemic blood flow, there is a diminution of blood flow to the lung and progressive pulmonary arterial hypertension. Compensation for the diminution in pulmonary blood flow is probably made by an increase in the bronchial circulation exemplified in the present case by dilated branching bronchial vessels in the pleura, adventitia of elastic pulmonary arteries, and bronchial walls. However, in addition, part of the increased pulmonary vascularity may be due to the thin-walled branches of the muscular pulmonary arteries.\(^2\) \(^7\) \(^6\) It is conceivable that bronchopulmonary anastomoses occur and relieve the pulmonary arterial hypertension, blood passing from the pulmonary artery to the bronchial artery as described by Verloop.\(^9\)

The eccentric intimal thickenings seen in some of the thin-walled blood vessels in this case were probably due to fibrous organization of blood clot, as similar lesions have been found in Fallot's tetralogy,\(^2\) \(^7\) \(^1\) tricuspid atresia,\(^7\) atrial septal defect with pulmonary stenosis, and transposition with pulmonary stenosis.\(^2\) All these diseases are characterized by high hemoglobin levels that were probably the cause of the intravascular thrombosis in the present case, where the hemoglobin level was 130 per cent.

**Differential Diagnosis**

In adults the differential diagnosis of cor triloculare biaatrium is from other forms of cyanotic congenital heart disease. As the cyanosis and murmurs in the anomaly under discussion are variable and so uncharacteristic, diagnosis must rest on special investigations. Of these, electrocardiography is valueless, as it may show right or left axis deviation and QRS complexes may not indicate that the ventricular septum is absent. Radiology is of help in young children, for while the cardiac silhouette appears to be huge in the posteroanterior view, due to the rudimentary ventricular chamber in the position of the pulmonary conus, the left anterior oblique view confirms that there is no right ventricular hypertrophy. In older children or adults, radiology is of little value for, if a large pulmonary artery arises from the main common ventricle and a small aorta from the rudimentary chamber, the radiologic appear-
ances suggest Eisenmenger’s complex or other cardiac diseases associated with pulmonary arterial hypertension and, if a large aorta arises from the main common ventricle and a small pulmonary artery from the rudimentary chamber, the appearances suggest Fallot’s tetralogy. Diagnosis must be based on the combined use of angiocardiography and cardiac catheterization. Angiocardiography demonstrates simultaneous filling of the aorta and pulmonary artery from a common ventricle while cardiac catheterization demonstrates pulmonary arterial hypertension in association with equal blood oxygen saturations in the pulmonary artery, the aorta, and common ventricle. The demonstration of pulmonary hypertension differentiates the condition from Fallot’s tetralogy, while in Eisenmenger’s complex or other congenital cardiac disease associated with pulmonary arterial hypertension the pulmonary arterial blood oxygen saturation is lower than that in the aorta.

Prognosis

The over-all prognosis in cor triloculare biatriatum is bad, but survival to adolescence or adult life may occur if there is a large pulmonary artery arising from the main chamber of the common ventricle with a consequent good pulmonary blood flow; in Deuchar’s33 patient, aged 13, who had little cyanosis at rest, a hilar dance with pulsation was visible out to the periphery of the lung fields. However, there are exceptions to this general rule. In the patient of Herndon and his colleagues,11 who lived to be 49, the aorta, whose diameter was greater than the pulmonary artery, arose from the main ventricular chamber. Similarly in Holmes’s44 patient, aged 22, it was the pulmonary artery that arose from the rudimentary ventricle. Mehta and Hewlett32 described a patient who survived to the age of 56 with cor triloculare biatriatum associated with persistent truncus arteriosus, and in this case the blood supply to the lungs was not determined. In most cases the anomaly under discussion has the best prognosis when associated with transposition in which the pulmonary artery is larger than the aorta and arises from the main part of the common ventricle.

The disease is compatible with moderate activity until early adult life, as shown by the present patient who was married and employed as a shop assistant until she died at the age of 23. In Abbott’s1 series 1 patient reached the age of 35, while 1 survived to reach the age of 21. Other reported cases have survived to the ages of 18 1/2, 4, 14, 49, 51, and 56.12 However, the author is in full agreement with Edwards,22 who considered that an erroneous impression of the average length of survival has arisen because only special cases surviving to adult life have been described. In the Mayo Clinic series of 9 cases, the longest survival was 8 years, while 6 died below the age of 1 year. While most patients with this disease die from congestive cardiac failure, they may die suddenly from rupture of an aneurysmally dilated pulmonary artery.9 It is probable that the dilatation of the pulmonary artery and its rupture are associated with a severely raised pulmonary artery blood pressure as in patent ductus arteriosus with pulmonary hypertension.22

Treatment

Cases with a large pulmonary artery, a small aorta, and an increased pulmonary blood flow are not amenable to surgery. If a large aorta and a small pulmonary artery arise from a rudimentary ventricular chamber and provide a poor pulmonary blood flow, a Taussig-Blalock operation is of benefit.

Summary

The clinical and pathologic features, including histologic changes in the small pulmonary blood vessels, of a case of cor triloculare biatriatum are recorded together with the results of special investigations.

The patient, a woman of 23 years, presented as a case of cyanotic congenital heart disease with pulmonary hypertension whose main complaints were dyspnea on exertion and ankle edema. The electrocardiogram indicated right ventricular preponderance and the teleradiogram showed a globular cardiac silhouette without prominence of the pulmonary conus. Angiocardiography showed a considerable shunt of blood passing from the right side of the heart into the aorta.
An erroneous diagnosis of Fallot’s tetralogy was made and a Blalock-Taussig operation was performed.

At autopsy the main pulmonary artery was dilated and transposed, arising from the main cavity of the common ventricle, while a small aorta arose from a rudimentary ventricular chamber. The small pulmonary blood vessels showed certain features of hypertensive pulmonary vascular disease together with evidence of increased bronchial artery collateral circulation to the lung. An interpretation of the histologic changes is suggested.

The clinical and pathologic findings of the case are discussed in a review of previously reported cases.

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SUMMARIO IN INTERLINGUA

Es presentate le characteristicas clinic e pathologic, incluse le alterationes in le parve vasos pulmonary de sanguine, de un caso de corle triloculare biatriatum, sequite per le resultatos de investigationes special.

Le patiente eseva un femina de 23 annos de etate. Illa se presentava como un caso de congenite morbo cyanotic del corde con hypertension pulmonar. Su major gravamines eseva dyspnea post effortio e edema tarsal. Le electrocardiogramma indicava preponderantia dextero-ventricular, e le teleradiogramma monstava un contorno cardiac globular sin prominentia del cono pulmonar. Angiocardiographia revelava un derivation considerabile de sanguine currente ab le latere dextere del corde a in le aorta.

Esveva formulate le diagnose erronee de tetralogia de Fallot. Le operation de Blalock-Taussig eseva esecutate.

Al necropsia, le major arteria pulmonar se mostrava dilatate e transpone. Illo prendeva su origine in le cavitate principal del ventriculo commun, durante que un parve aorta proveniva ab le atrophiae secunde camera ventricular. Le parve vasos sanguine del pulmon monstava certe characteristicas de hypertension pulmono-vascular insimul con signos de un augmentate circulation collateral al pulmon via le arteria bronchial. Es presentate un possibile explication del alterationes histologic.

Le constatationes clinic e pathologic in le presente caso es discutite con referentias a previe reportos in le litteratura.

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Eighty-two cases of ventricular aneurysm were collected from various sources in Johannesburg. Sixty-five were Europeans, 15 were Negro (Bantu), 1 was Cape Coloured and 1 was Hottentot. Seventy-four of these patients were studied at necropsy. Coronary atherosclerosis was associated with all 57 aneurysms studied in the Europeans and was considered probably responsible for the cardiac involvement in each instance. Coronary atherosclerosis was not associated with any of the 15 ventricular aneurysms occurring in the Bantu. Six of these 15 cases were due to syphilis, 1 each to tuberculosis of the left coronary artery, Loeffler's parietal endocarditis, mycotic aneurysm, rheumatic disease, and congenital causes, and 4 were considered idiopathic. Among the 74 necropsied cases, rupture of the aneurysm was the cause of death in 16. The authors offer no explanation for the differences in incidence of coronary artery atheroma in the European as contrasted with the Bantu but report that differences in diet and nutrition and histologic differences in coronary arteriolar structure are still to be investigated.

Rosenbaum
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DONALD HEATH

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