THE ROLE OF THE PULMONARY VASCULAR BED IN CONGENITAL HEART DISEASE

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Life expectancy and the degree of invalidism have been considered a function of the type of cardiac malformation in patients with congenital heart disease. When the abnormality involves a large communication between the ventricles or great vessels, however, the pulmonary vascular bed plays an important role in governing prognosis. The following group of congenital malformations are involved in this category: 1. single ventricle without pulmonary stenosis, 2. large ventricular septal defect, 3. Eisenmenger complex, 4. atrio-ventricularis communis, 5. true truncus arteriosus, 6. aortic septal defect, 7. large patent ductus arteriosus and 8. patent ductus arteriosus associated with coarctation of the aorta.

THEORETIC CONSIDERATIONS

Peripheral resistance in the systemic circulation is maintained chiefly by the arterioles. The caliber of these small muscular vessels is controlled by the autonomic nervous system. Changes in the caliber of the arterioles alter peripheral resistance and, hence, blood pressure. Normal systemic peripheral resistance is maintained at a high level, thus maintaining a relatively high systemic blood pressure. White has stated that the range of normal adult systemic blood pressure is 145/95 to 90/60 mm.Hg. By using the simplified formula set forth by Friedlich, (Resistance (mm.Hg/l./min./M² = Mean pressure (mm.Hg)) / Blood flow (l./min./M²)) mm.Hg/l./min./M².

In contrast to the systemic circulation, the normal pulmonary vascular bed is characterized by great distensibility of the arterial walls and a large capacity of the arterial and capillary beds. A low peripheral resistance and consequent low pulmonary blood pressure is the essential attribute of the pulmonary circulation. The great distensibility of the pulmonary vascular bed in the normal human is illustrated by the effect of increased pulmonary blood flow on pulmonary pressure. Cournand showed that the pulmonary artery pressure does not rise above normal levels until the pulmonary blood flow exceeds three times the basal level. Normal values for pulmonary blood pressure are 19/6 mm.Hg to 26/12 mm.Hg; pulmonary resistance is 2 to 5 mm.Hg/l./min./M². Definite proof of a vasomotor control of the small pulmonary arteries and arterioles is lacking. However, Zimmerman and his co-workers demonstrated the vasoconstrictive action of adrenalin and the vasodilatation effect of aminophyllin on the pulmonary vascular bed. Whether this effect is humeral or nervous could not be stated. Hickam
demonstrated constriction of the pulmonary arteries during excitement and emotion. Motley and his associates demonstrated a distinct rise in pulmonary artery pressure following short periods of induced acute anoxia. Normally, however, changes in pulmonary artery pressure are small as compared to changes in systemic pressure.

In the normal individual, then, the vascular bed offers a considerable resistance to blood flow in the systemic circulation and very little resistance to blood flow in the pulmonary circulation. If this difference were present when the two circulations were joined, as in the case of a single ventricle, it is readily apparent that the major portion of the cardiac output would be directed to the lungs and little to the body. Such a course would hardly be compatible with life.

Civin and Edwards traced the development of the pulmonary vascular bed from a fetal to an adult state. They demonstrated progressive changes in the elastic arteries, the muscular arteries and the pulmonary arterioles. At birth elastic arteries are open but the walls are relatively thick as compared to the size of the lumen. With growth the wall becomes relatively and absolutely thinner and the lumen relatively and absolutely larger. At birth the lumens of the muscular arteries and arterioles are small and their walls are thick. With growth the media thins out and the lumen widens. These changes take place during the first few months of life and are completed for the most part by the age of 6 months.

It seems reasonable to relate these morphologic changes to changes in pulmonary physiology and to suggest that pulmonary vascular resistance falls as the pulmonary vessels thin out and widen. Although pulmonary arterial resistance and blood pressure have not been measured in normal humans at birth, there is evidence to suggest that they are high. Hamilton and his co-workers found that the pressure within the right and left ventricles of the newborn rabbit and dog were essentially similar prior to and immediately after the first few breaths had expanded the lung. Barclay demonstrated by means of angiocardiograms that the right ventricle supplied blood to the descending aorta through the ductus in the living sheep fetus. It is probable that the high pulmonary pressure immediately following birth is responsible for the absence of a murmur in infants with patent ductus arteriosus.

Edwards and Civin and Edwards suggested that the pulmonary muscular arteries retain their fetal characteristics and high resistance in patients with certain types of congenital malformations of the heart. Furthermore, they have argued that, as a result of the high pulmonary resistance and consequent high pulmonary arterial pressure, secondary changes take place within the muscular pulmonary arteries causing a further reduction in lumen size and a further increase in resistance. These changes consist of the deposition of hyaline material in the intima and the fragmentation and necrosis of the elastic tissue and smooth muscle tissue within the media. They suggest that the maintenance of the fetal state is compensatory but that the secondary changes that occur in the intima and media, causing further increases in pulmonary resistance, act to the detriment of the patient. In a series of articles, Edwards and his co-workers have cited numerous examples including coarctation of the aorta proximal to the patent ductus, the Eisenmenger complex and single ventricle without pulmonary stenosis; Adams suggested that examples of simple septal defects be included as well. Goldberg and his associates gave additional weight to the argument by presenting catheterization data on a case of Eisenmenger complex with an anomalous pulmonary vein entering the right auricle. The pulmonary venous pressure was found to be normal but the pulmonary arterial pressure
high and the pulmonary venous oxygen saturation was normal although saturation within the aorta was low. The present investigators have recently produced luminal narrowing, medial hypertrophy and intimal thickening in the small pulmonary arteries of dogs subjected to a large pulmonary blood flow and high pulmonary artery pressure. This will be reported in detail in a separate communication.

Theoretically, in patients with the type of congenital heart disease under discussion, there are three possible courses that the pulmonary vascular bed may follow after birth. 1. The thick-walled, small-lumened pulmonary vessels may retain their fetal characteristics for a period of months and then may gradually thin out and approach the adult type of pulmonary vessel. As the walls thin out and the lumens widen, pulmonary resistance falls. More blood will be directed out the pulmonary artery and relatively less blood to the aorta. To maintain systemic blood flow at a level compatible with life, either systemic resistance must fall or cardiac output must rise. Evidence obtained by catheterization indicates that cardiac output is high and systemic resistance is unchanged. If this sequence of events continues, high output cardiac failure ensues. The signs and symptoms in this group of patients are those due to pulmonary congestion and cardiac decompensation. Because of the high volume of pulmonary blood flow, cyanosis will be absent until cardiac failure becomes marked. (See Fig. 1A.)
2. The thick-walled, small-lumened pulmonary vessels and high pulmonary vascular resistance present at birth may continue without appreciable change. A balance between the two circulations thus may be maintained which permits sufficient oxygenation of systemic blood for normal activity, yet prevents a high enough pulmonary blood flow to cause cardiac decompensation. Such a balance is theoretically possible but is rarely seen. Signs and symptoms are minimal or absent. (See Fig. 1B.)

3. The thick-walled, small-lumened pulmonary arteries and consequent high pulmonary vascular resistance may continue after birth. Perhaps as a result of this high resistance and consequent pulmonary hypertension, secondary intimal changes may take place, causing a further narrowing of the pulmonary arteries and a further rise in pulmonary vascular resistance. More blood will be directed out the aorta and less through the pulmonary artery. The patient will suffer from increasing anoxia, cyanosis and polycythemia. The patient’s symptoms will resemble those of pulmonary stenosis. In actuality, a pulmonary stenosis does exist. In patients with tetralogy of Fallot, the stenosis lies in the outflow tract of the right ventricle. In the conditions under discussion, the stenosis lies in the distal small muscular pulmonary arteries and arterioles. The end result is the same. This sequence of events is classically seen in patients with Eisenmenger complex, in whom cyanosis is absent until about the age of puberty and then becomes progressive. (See Fig. 1C.)

The foregoing considerations have caused the authors to alter their approach to the therapy of this group of congenital malformations of the heart. Wherever possible a direct approach to the malformations should be made, such as closure of a large patent ductus or aortic septal defect. When the direct approach is not feasible, as in instances

![Fig. 2.](http://pediatrics.aappublications.org/)

**Fig. 2.** Drawing demonstrating method of producing pulmonary stenosis. Section of wall of main pulmonary artery has been removed and pulmonary artery is then wrapped with polyethylene film and cotton tape.
of single ventricle without pulmonary stenosis, the surgical creation of pulmonary stenosis may effectively reduce the strain on the heart and lungs and return the circulation to a more normal state. The procedure which the authors used to create pulmonary stenosis consists of excision of a segment of wall of the main pulmonary artery and the placement of a band of polyethylene film and cotton tape around the artery. The lumen is reduced in cross sectional area to about 25% of its previous size. (See Fig. 2.)

Creation of pulmonary stenosis has two distinctly different applications. First, in patients in whom the pulmonary vessels are becoming progressively thinner-walled and larger-lumened, with a consequent fall in pulmonary vascular resistance and rise in pulmonary blood flow, the creation of pulmonary stenosis acts to reduce excessive blood flow to the lungs, reduce total cardiac output, and relieve the symptoms of high output cardiac failure. Systemic blood flow and coronary blood flow are increased. Second, in patients in whom the fetal pulmonary vascular bed has been retained and secondary intimal changes of an irreversible nature have begun, the creation of pulmonary stenosis acts to remove the stimulus causing pulmonary artery narrowing thus permitting the pulmonary arteries to progress in a normal manner from thick-walled, small-lumened vessels to thin-walled, large-lumened vessels. The nature of the stimulus is not clearly understood but experimental work in animals and study of autopsy or biopsy material suggest that it is related to arterial pressure and possibly to volume of blood flow as well.
The creation of pulmonary stenosis may be contraindicated when pulmonary resistance is high and irreversible due to advanced intimal changes within the small pulmonary arteries.

CASE REPORTS

Case I. S. C. was first seen at the age of 6 mo. because of a heart murmur. At that time she weighed 6.8 kg. Birth and neonatal period were normal. At the age of 3 mo. she developed bronchitis. Rapid respirations became constant. Weight gain was poor as compared to her 2 older siblings. There was no history of cyanosis.

On physical examination her color was good; she appeared moderately well developed and well nourished. Blood pressure was 124/50/0 mm Hg; pulse was 120; respiratory rate was 40. Femoral and brachial pulses were full and bounding. Apex of the heart was felt in the 5th left interspace.

**Fig. 4.** Small muscular pulmonary artery showing hypertrophy of media and relatively small lumen and normal intima. (Verhoeff's elastic tissue stain and van Gieson's connective tissue stain ×900) Case 1: S. C., 7 mo. old infant with patent ductus arteriosus three-fourths size of aorta. Photomicrographs made by Zane A. Price.
FIG. 5. Direct pressures from pulmonary artery and aorta recorded at surgery from A. W. Note increase in pulse pressure from 36 to 25 in aorta following closure of ductus and marked drop in pressure in pulmonary artery.

almost in the anterior axillary line. Over the heart there was a "confusion of sound." At the base, a loud, long, harsh systolic murmur was present which possibly extended into diastole. Low over the sternum and out toward the apex was a second systolic murmur, lower pitched, not as loud, but rough. In this area, too, there was a long, rough, diastolic murmur. The second heart sound was accentuated and split. Lungs were clear. Liver was 2 cm. below the costal margin.

There was right axis deviation in the ECG. The precordial leads gave evidence of right ventricular hypertrophy and suggested left ventricular hypertrophy as well. On fluoroscopy and roentgen examination the base of the heart was obscured by a large thymus gland. Heart appeared overly active. Lung fields were overly vascular and pulsed actively. Right ventricle was definitely enlarged; the left auricle appeared enlarged.

During the following month her respirations increased and her rate of weight gain decreased. She was brought into the hospital for retrograde aortography. Diodrast injected into the left brachial artery appeared within 1 sec. in the pulmonary vascular bed, probably through a patent ductus. Closure of the ductus was decided upon. At the time of surgery, a large patent ductus, about 3/4 the size of the aorta, was found. The pressure in the pulmonary artery (70/50 mm.Hg) was lower than that in the aorta (110/65 mm.Hg). (See Fig. 3.) Pressures in the pulmonary artery were unchanged by closure of the ductus. However, there was a slight fall in aortic systolic pressure and rise in aortic diastolic pressure. Figure 4 represents a typical small pulmonary artery seen on examination of lung biopsy. Patient's postoperative course was uneventful. At the present time, 6 mo. after surgery, she has gained 3.0 kg. in weight, breathes quietly, and is normally active. All murmurs have disappeared. She shows no evidence of cardiac decompensation.

Case 2: A. W., age 13 mo. This little Negro girl appeared perfectly normal at birth but during the first few months of life it was noted that her weight gain was poor, appetite was poor, and breathing at all times was noisy and rapid. She was subject to repeated infections of the upper respiratory tract. She had not had pneumonia. Cyanosis had not been noted.

She appeared small and underdeveloped on physical examination. Respiratory rate was rapid. Blood pressure was 90/0 mm.Hg; pulses were bounding. She weighed 5.3 kg. There was no bulge
of the left side of the chest. Heart was enlarged. A diffuse apical beat was felt over the sternum, suggesting right ventricular enlargement. Maximal in the 2nd left interspace was a loud, long, rough, systolic murmur which seemed to extend slightly into diastole. Chest was clear. Liver was not enlarged.

ECG revealed right axis deviation and left ventricular hypertrophy. There was a suggestion of right ventricular hypertrophy as well. On fluoroscopy the main pulmonary artery segment was full and the lungs appeared overly vascular and active. Left ventricle appeared enlarged, more than the right ventricle. Left auricle was normal. Patient was admitted to the hospital for retrograde aortography. Aortograms revealed rapid filling of the pulmonary vessels after injection of diodrast into the left brachial artery. At the age of 14 mo. she underwent surgery for closure of a large patent ductus arteriosus. Ductus equalled the aorta in size. Pressures in the aorta and the pulmonary artery were essentially the same prior to closure of the ductus, 104/67 and 98/73 mm.Hg, respectively. Following closure of the ductus there was a significant drop in pulmonary artery pressure.

Fig. 6. Small muscular pulmonary artery and arteriole showing medial hypertrophy, small lumen and normal intima. (Verhoeff's elastic tissue stain and van Gieson's connective tissue stain X900)

Case 2: A. W., 13 mo. old infant with patent ductus arteriosus size of aorta.
Fig. 7. Small muscular pulmonary artery showing thin media, normal intima and normally wide lumen. (Verhoeff's elastic tissue stain and van Gieson's connective tissue stain ×900) Five year old child with typical small patent ductus and no symptoms.

and a drop in aortic pulse pressure of 12 mm Hg as shown in figure 5. Figure 6 represents a typical vessel seen on examination of the lung biopsy. Postoperative course was without difficulty. At the present time, 6 mo. after surgery, she is gaining weight well; respirations are slow and quiet. She is active. The systolic murmur has gradually faded but is still faintly heard over the base of the heart. The diastolic murmur, heard for the 1st 4 mo. after surgery, has completely disappeared.

Comment on Case 1 and Case 2

In each patient there was an early onset of symptoms suggestive of cardiac decompensation. The symptoms increased in severity sufficiently to warrant early study and early surgical intervention. At surgery a ductus of large size was found and closed. Pressures taken prior to and following closure of the ductus showed definite evidence of
pulmonary hypertension. In the first patient, pulmonary pressure was not as high as that found in the aorta. Although the authors have not been able to make a consistent correlation, it is of interest that the ductus in this patient was not as large as the aorta, suggesting that the ductus itself put some limitation on the amount of blood which could pass through it. In the second patient the ductus equalled the aorta in size and pulmonary and aortic pressures were almost identical. Examination of the lung biopsies in both instances demonstrated marked thickening of the media and lumens of decreased caliber in the small muscular arteries and arterioles. No intimal changes were noted. These changes were deemed sufficient to cause the elevation of pulmonary pressure. In both instances the postoperative course was excellent. All signs of cardiac decompensation disappeared; murmurs decreased or disappeared. For comparison with Case 1 and Case 2, see figure 7 which is a representative sample of a small pulmonary artery from a 5 year old child who was free of symptoms, had a classical continuous murmur, minimal cardiac enlargement and no evidence of decompensation. At surgery the patent ductus was found to be small.

Case 3: L. B.* This 3 1/2 mo. old infant was referred because of continuously rapid respirations, orthopnea, a constant dry cough, and the presence of a cardiac murmur.

**TABLE 1**

<table>
<thead>
<tr>
<th>Cardiac Catheterization</th>
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<tr>
<td>L.B.</td>
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<tr>
<td><strong>O₂ Content (Vol. %)</strong></td>
</tr>
<tr>
<td>Right Auricle</td>
</tr>
<tr>
<td>Right Ventricle</td>
</tr>
<tr>
<td>Brachial Artery</td>
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<td>Capacity</td>
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Assumed O₂ consumption = 50 cc./min.

Pulmonary blood flow = 6.2 l./min./M²

Systemic blood flow = 1.06 l./min./M²

On physical examination the baby weighed 3.7 kg. He appeared underdeveloped and undernourished. There was no cyanosis. His breathing was rapid; he was definitely orthopneic. There was a rough, long, loud, systolic murmur maximal in the 3rd and 4th left interspaces, close to the sternum. The second sound at the base was accentuated and split. A diastolic gallop was heard at the apex. Moist rales were heard over both chests. Liver edge was 3 cm. below the right costal margin.

On fluoroscopy and roentgen examination there was definite enlargement of both auricles. The apex was seen extending below the diaphragm in the posterior-anterior projection. Lung fields appeared notably vascular and vigorous pulsations were seen. The main pulmonary artery segment, however, was only slightly convex in the posterior-anterior projection; it became more prominent in the right anterior oblique projection. These findings suggested displacement of the pulmonary artery.

ECG was bizarre and suggested right ventricular hypertrophy. The diagnosis of a single ventricle with a large pulmonary artery arising in an abnormal position was made.

At surgery a large pulmonary artery was found arising to the right of its usual position. The pressure within it was high. The aorta, which lay to the right and posterior to the pulmonary artery, was relatively small. A section of the main pulmonary artery was removed and a band of polyethylene tubing was placed around the pulmonary artery. The cross sectional area of the main pulmonary artery was reduced to about one-third of its previous size. At the time the clamp was placed on the pulmonary artery, cardiac action became stronger and color improved. No lung biopsy was taken. Postoperatively for the first few days there was a delicate balance between cardiac decompensation and compensation. After approximately 20 days, however, the patient began to improve.

*This case was reported in Surg., Gynec. & Obst., August, 1952, except for the postoperative cardiac catheterization which had not been carried out at that time.
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Weight gain was steady, breathing became quieter, and evidence of decompensation disappeared. At the end of 3 mo. digitalis was discontinued. Improvement continued for an additional 3 mo. and then the rate of weight gain slowed down. He remained active and alert. A trial of digitalis did not improve him. On fluoroscopy, the lung fields still appeared extremely vascular. It was obvious that, although his condition had been benefited by surgery, the patient was still suffering from excessive blood flow to the lungs. In order to establish this fact, cardiac catheterization was attempted. The small size of the infant made catheterization difficult and it was not possible to get all the data desired because of clot formation within the catheter. From the data 2 conclusions can be drawn. First, pulmonary blood flow was excessively high, being about 6 times the systemic blood flow (6.2 l./min./M² to 1.06 l./min./M'). Second, there was strong evidence of pulmonary stenosis, the pressure within the ventricles being 98 to 60/20 to 12 mm.Hg and that in the pulmonary artery distal to the stenosis being 20 to 10/14 to 8 mm.Hg. (See table 1 and Fig. 8.)

Case 4: S. T. This child was first seen at 4½ mo. of age. Birth weight was 3.4 kg.; at 2½ mo. of age he weighed 4.3 kg.; at 4½ mo., 4.6 kg.; at 5½ mo., 4.8 kg. He appeared normal at birth and for the 1st 6 wk. of life did relatively well. Rapid respirations were present shortly after birth; his respiratory rate seemed to increase steadily after the 1st 3 mo. of life. His color had always been good. During the last few weeks prior to admission he appeared constantly irritable and slept poorly.

He appeared undernourished and chronically ill on physical examination. Pulse was 160, respirations 66. His color was pale and there was a suggestion of cyanosis. There was no bulge of the chest. The apex of the heart was felt in the 5th left interspace in the anterior axillary line. A loud, long, harsh, systolic murmur accompanied by a systolic thrill was heard over the entire precordium, maximal in the 3rd left interspace close to the sternum. A diastolic gallop was present at the apex. The second heart sound at the base was accentuated. Rales were heard throughout the chest. Liver was enlarged 4 cm. below the right costal margin. Femoral pulses were normal.

On fluoroscopy and roentgen examination the heart appeared grossly enlarged with enlargement

![Fig. 8. Pressure recorded from pulmonary artery distal to point of stenosis, ventricle and brachial artery during cardiac catheterization of L. B. Note variations in pressure due to respiration recorded from pulmonary artery. Amplification of pulmonary artery pressure was 5 times that of ventricle and brachial artery pressure.](https://example.com/fig8.png)
Fig. 9. Direct pressures recorded from aorta and pulmonary artery during surgery from S. T. Note increase in aortic and proximal pulmonary artery pressure following creation of pulmonary stenosis. Note drop in distal pulmonary artery pressure and fall in pulse pressure.

of all chambers. The left auricle displaced the esophagus posteriorly in the right anterior oblique view and to the right in the posterior-anterior projection. The pulmonary artery segment was prominent; the right and left pulmonary arteries were enlarged and pulsed actively. The peripheral lung fields appeared congested.

ECG revealed right axis deviation and right ventricular hypertrophy.

The patient was digitalized and for a short period of time appeared improved. Respiratory rate slowed to 40/min. His color improved, appetite improved and he gained a little weight. However, after about 2 wk. of therapy, signs of cardiac decompensation increased. He was admitted to the hospital for further study.

Retrograde aortography revealed a normally situated aortic arch and descending aorta and failed to reveal evidence of a patent ductus arteriosus or aortic septal defect.

Angiocardiography demonstrated an enlarged right auricle, right ventricle and pulmonary artery. Early in the series of films, a small amount of diodrast was seen in the left ventricle and aorta. Again, late in the series, after visualization of the pulmonary veins and left auricle, the left ventricle, aorta and pulmonary arteries were reopacified. These findings suggested the presence of a large ventricular septal defect. Because of the continuation of cardiac decompensation, the constant presence of rales over both lungs, the rapid respirations and rapid pulse rate, and the large heart and large liver, it was decided to reduce the pulmonary blood flow by the creation of a pulmonary stenosis.

When the chest and pericardium were opened a large pulmonary artery was found arising in its normal position. To the right and posteriorly, a much smaller aorta was seen. Before the pulmonary artery was narrowed, pressures were taken from both the aorta and the pulmonary artery.
The pressure in the aorta was 82 to 84/58 to 59 mm Hg and that in the main pulmonary artery was 82 to 83/52 to 53 mm Hg. The pulmonary artery was then narrowed between 75% and 80%. A second series of pressures was taken after this was completed. The pressure in the aorta had risen to 115 to 130/100 to 105 mm Hg and in the pulmonary artery proximal to the stenosis 105 to 115/35 to 40 mm Hg. The pressure distal to the stenosis had fallen to 60 to 70/40 to 50 mm Hg (Fig. 9). Pulse rate decreased from 140 to 120 after the pulmonary artery was constricted. Signs of pulmonary edema cleared completely and heart action appeared much improved. His color remained good. Figure 10 represents a typical muscular artery seen on examination of a lung biopsy.

Case 5: M. F. This 20 mo. old girl appeared normal at birth. Birth weight was 3.3 kg. She weighed 8.2 kg, at the time of admission. On the second day of life a murmur was first heard. Respirations were constantly rapid. Her appetite was poor and weight gain was slow. At no time was cyanosis observed. In the few months prior to surgery the parents and the referring physician noted less activity, poorer food intake and slower weight gain.

She appeared underdeveloped and undernourished without evidence of cyanosis or clubbing on physical examination. Her respiratory rate was 72; blood pressure was 100/70 mm Hg. There was a marked bulge of the left anterior chest. The apex of the heart was felt in the 5th left intercostal space in the anterior axillary line. There was a precordial heave and a strong pulmonary shock over the base of the heart. There was a long, loud, moderately high pitched, systolic murmur which did not extend into diastole and which was accompanied by a thrill. This murmur was maximal in intensity in the 3rd left intercostal space near the sternum. A mid-diastolic rumble was audible at the apex. The second heart sound was accentuated and split. The lungs were clear, the liver was not enlarged. Pulses in the extremities were equal and normal in quality.

On fluoroscopy there was marked cardiac enlargement. Both ventricles appeared enlarged, the right more than the left. Right auricle was enlarged. There was suggestive enlargement of the left auricle. The main pulmonary artery was markedly enlarged and the right and left pulmonary arteries were large and abnormally active. Peripheral lung fields appeared overly vascular. Roentgenographic examination confirmed these findings.

ECG revealed right axis deviation and right ventricular hypertrophy. Comparison with an ECG taken when the patient was 7 mo. of age demonstrated an increase in the degree of right ventricular hypertrophy and ST-T changes suggestive of right heart strain.

Patient was admitted to the hospital for retrograde aortography and angiocardiography. The aortogram revealed good filling of the arch of the aorta and descending aorta. No diodrast was visualized in the pulmonary arteries. There was no evidence of a patent ductus arteriosus or an aortic septal defect. The angiocardiogram revealed rapid filling of the right side of the heart and pulmonary arteries. In the second film, diodrast was visualized in the left ventricle. Later in the series the pulmonary veins, left auricle, left ventricle and aorta were clearly outlined. At this time there was reopacification of the pulmonary artery.

A diagnosis of a large ventricular septal defect or functional single ventricle with pulmonary hypertension was made. Digitalization was carried out with improvement in appetite but no change in respiratory rate. A thoracotomy was performed and the pulmonary artery was narrowed 75% to 80%. Pressures taken from the aorta and pulmonary artery before and after constriction of the pulmonary artery demonstrated a definite fall in pressure distal to the stenosis of about 15 mm Hg and a rise in the aortic pressure of about 15 mm Hg (Fig. 11). Preoperative aortic pressure was 85 to 90/55 and the pulmonary pressure was 60 to 65/35 to 40. Following creation of the stenosis, the aortic pressure was 95 to 105/65. Right ventricle pressure was 100 to 105/5 to 10 and distal pulmonary artery pressure was 46 to 52/26 to 28. Figure 12 represents a typical small artery seen on examination of the lung biopsy.

CASE DISCUSSION

These three patients had varying malformations of the heart. One had a single ventricle and two had large ventricular defects. In all three there was evidence of pulmonary hypertension. All three were in difficulty because of excessive pulmonary blood flow. The creation of pulmonary stenosis was beneficial because it reduced the pulmonary blood flow, thus relieving cardiac decompensation. In the first patient, postoperative cardiac catheterization done nine months after surgery suggested that, despite the presence...
of pulmonary stenosis, pulmonary blood flow was still too high. The authors have considered the advisability of reoperation with creation of additional pulmonary stenosis but have delayed doing this, hoping that the present degree of stenosis will prove to be more satisfactory when the patient has grown and developed further.

The immediate effect of creating pulmonary stenosis was well demonstrated in the second and third patients. The rise in aortic pressures following the procedure suggests an increase in systemic blood flow. The difference between the pressures obtained from the proximal pulmonary artery or right ventricle and the distal pulmonary artery demonstrates the presence of a definite stenosis. The fact that the distal pulmonary artery pressure did not fall to normal immediately after the creation of pulmonary stenosis is not a matter of concern. A period of time must elapse before the small-lumened, thick-walled pulmonary arteries seen on examination of the lung biopsy can enlarge

Fig. 10. A (above). Small muscular pulmonary artery (×900). B (opposite page). Arteriole (×1091). Note medial hypertrophy and small lumen. There is also slight intimal thickening of small muscular artery. Case 4: S. T., 6 mo. old infant with large ventricular defect.
and thin out. In the third patient, the preoperative pressure difference between the aorta and pulmonary artery of 20 to 25 mmHg suggests that the volume of shunt through the ventricular defect was limited somewhat by the size of the defect. Following constriction of the pulmonary artery, pressures in the aorta and right ventricle were identical.

The long term value of this surgical procedure cannot yet be analyzed since the first patient (L. B.) was operated upon only 1½ years ago. However, the course to date has been most encouraging. Cardiac failure has cleared. Respiratory rates have returned to normal. Weight gain has improved and physical activity has been normal. Postoperative studies including cardiac catheterization and possibly lung biopsies should be done to determine the effectiveness of the procedure and to establish in each instance whether the degree of stenosis created was sufficient and yet not excessive.

Pulmonary stenosis has been attempted but did not significantly lower the distal pulmonary artery pressure in one older patient with a coarctation of the aorta, ventricular
septal defect, a pulmonary artery pressure of 150/60 mm Hg and a pulmonary artery blood flow that was less than the systemic blood flow because of advanced arterial changes. The pulmonary artery was only narrowed 50 to 60% in that patient. A second attempt is planned for the future in an effort to reverse the pulmonary artery changes and prevent irreversible changes.
Fig. 12. Small muscular pulmonary artery showing medial hypertrophy and luminal narrowing.
(Verhoeff's elastic tissue stain and van Gieson's connective tissue stain ×900.) Case 5: 20 mo. old girl with large ventricular defect and excessive pulmonary blood flow.

SUMMARY AND CONCLUSIONS

In the normal human being systemic blood pressure is maintained at a high level by means of the high resistance offered to the flow of blood by the smaller systemic arteries and arterioles. In contrast, pressure in the pulmonary circulation is maintained at a low level because of the relatively large size of the pulmonary vessels. In the fetal and newborn period pulmonary arteries resemble systemic arteries in that they have a small lumen and thick media. In this period pulmonary pressure and resistance are high. As these vessels gradually thin out and enlarge, resistance to pulmonary blood flow falls and consequently pulmonary blood pressure falls.

There is evidence to suggest that the fetal state of thick-walled, thin-lumened pulmonary arteries is retained in instances where the two circulations are joined, such as in patients with a single ventricle, large ventricular defect, aortic septal defect or large
patent ductus arteriosus. As the patient grows, resistance to pulmonary blood flow may: (1) decrease. If the small pulmonary arteries develop normally and become thin-walled and large-lumened, pulmonary resistance will fall. More and more blood will be shunted into the lungs and the patient will develop the signs and symptoms of cardiac decompensation. (2) remain the same. If the small pulmonary arteries retain their fetal characteristics, pulmonary resistance will remain elevated. A balance between systemic and pulmonary blood flow will be maintained compatible with a relatively normal life. (3) increase. If, because of secondary intimal changes, the pulmonary arteries become thicker-walled and smaller-lumened, pulmonary resistance will increase. Progressively less blood will be shunted to the lungs and finally the shunt will reverse and become predominantly venous arterial. The patient will develop the signs and symptoms of pulmonary stenosis with increasing dyspnea and cyanosis.

Data from two groups of patients are presented to illustrate the importance of the pulmonary vascular bed. In the first group, early and severe symptoms of cardiac failure necessitated closure of a large patent ductus arteriosus early in life. The ductus was large, comparable to the size of the aorta, pressures from the aorta and pulmonary artery were similar and a study of the lung biopsies demonstrated that the lumens of the small pulmonary arteries were decreased in size and the media were abnormally thick. In each instance closure of the ductus resulted in a cure.

In the second group the two circulations were joined by either a large ventricular defect or a single ventricle. Three patients were in cardiac failure due to excessive pulmonary blood flow. Pulmonary blood flow was decreased by the creation of pulmonary stenosis, the main pulmonary artery being narrowed 60 to 80%. The postoperative course in these three patients illustrates the value of the creation of pulmonary stenosis in instances where the two circulations are joined by a large communication between the ventricles or great vessels.

ADDENDUM

A second operation was performed in Case 3 (L. B.) and the pulmonary artery narrowed still further. Pressures taken before and after from the pulmonary artery and aorta revealed the presence of a pulmonary stenosis which was increased by the additional constriction of the pulmonary artery. The patient’s postoperative course has been excellent suggesting that the degree of pulmonary stenosis present is now adequate.

REFERENCES


SPANISH ABSTRACT

Papel de la Red Pulmonar Vascular en las Enfermedades Congénitas Cardiacas

La presión sanguínea sistémica se mantiene elevada en el sujeto normal gracias a la resistencia ofrecida por las arterias y arteriolas del organismo a la corriente sanguínea. Por el contrario la presión de la circulación pulmonar se mantiene baja debido al tamaño relativamente grande de los vasos pulmonares. En los períodos fetal y del recién nacido las arterias pulmonares se parecen a las del resto del organismo por su pequeña luz y por el gran grosor de sus capas; en estos períodos tanto la presión como la resistencia pulmonar se encuentran elevadas. A medida que se adelgazan y crecen los vasos, disminuye la resistencia a la corriente pulmonar y por lo tanto también la presión sanguínea pulmonar.

Hay hechos que sugieren que se mantiene el estado fetal de paredes gruesas y luz pequeña de las arterias pulmonares en aquellos casos en los que se unen las dos circulaciones, tales como pacientes con ventrículo único, gran defecto interventricular, defecto aórtico y persistencia de un gran conducto arteriovenoso. A medida que crece el paciente, la resistencia a la corriente sanguínea pulmonar puede presentar las siguientes eventualidades:

1. Disminuir.—Si las pequeñas arterias pulmonares se desarrollan normalmente, sus paredes se adelgazan y su luz se amplía, mayor cantidad de sangre se vertirá a los pulmones y el paciente desarrollará los signos y síntomas de la descompensación cardiaca.
2. No ser afectada.—La resistencia pulmonar permanecerá alta cuando las arterias pulmonares conserven sus características fetales. El equilibrio entre la corriente sanguínea sistémica y la pulmonar, mantendrá compatibilidad con una vida relativamente normal.
3. Aumentar.—Así sucederá cuando las arterias pulmonares, debido a cambios íntimos secundarios, engrosan sus paredes y disminuyen su luz; entonces se verá progresivamente menor cantidad de sangre a los pulmones hasta invertirse el ritmo de la corriente, que se vuelve predominantemente venoarterial; el paciente desarrollará signos y síntomas de estenosis pulmonar con creciente disnea y cianosis.

Para ilustrar la importancia de la red vascular pulmonar se presentan datos de dos grupos de pacientes. En el primero, los síntomas intensos de insuficiencia cardíaca necesitaron la sutura de un gran conducto arteriovenoso desde época temprana de la vida; el conducto se encontró grande comparable al tamaño de la aorta, las presiones de la aorta y de la arteria pulmonar fueron semejantes y las biopsias pulmonares demostraron que la luz de las pequeñas arterias pulmonares estaban disminuídas, así como la pared enormemente gruesa. En todos los casos, la sutura del conducto produjo curación.

En el segundo grupo se unían las dos circulaciones por un defecto ventricular o bien por la existencia de un ventrículo único. Se encontraron 3 pacientes con insuficiencia cardíaca debida a flujo sanguíneo pulmonar en exceso. La corriente pulmonar se acentuó al crearse una estenosis pulmonar, estrechándose la principal arteria pulmonar de 60 a 80%. La evolución postoperatoria en estos 3 pacientes mostró el valor de la formación de una estenosis pulmonar en aquellos casos en que las dos circulaciones se unen por comunicaciones amplias entre los ventrículos o los grandes vasos.

University of California Medical Center
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