Massive Pulmonary Arteriovenous Fistula
in the Newborn

A Correctable Form of “Cyanotic Heart Disease;”
An Additional Cause of Cyanosis with
Left Axis Deviation

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Persistent cyanosis in the newborn period is most frequently due to serious cardiac malformations or extensive pulmonary pathology. Too often little interest is shown the intensely cyanotic and desperately ill newborn infant because the majority of causative lesions do not lend themselves to correction and infrequently to satisfactory palliation. A noteworthy exception is congenital pulmonary arteriovenous fistula. Although infrequently included in the differential diagnosis of persistent neonatal cyanosis, it is an important cause, since it is a life-threatening, yet potentially curable lesion. To emphasize this condition, a case diagnosed and successfully treated in the first week of life will be presented.

Case Report

The patient, an 8-pound 2-ounce full-term boy, was born on April 14, 1962, the product of an uncomplicated pregnancy of a 19-year-old primipara. Delivery was uneventful, and spontaneous respirations began within 2 minutes. Shortly after birth he was noted to have grunting respirations and generalized cyanosis. Examination revealed a tachycardia of 180 per minute, precordial prominence, and a diffuse precordial thrill. A grade-VI harsh systolic murmur was audible over the anterolateral aspect of the left chest. No other notations of initial auscultatory findings were made. Coarse rales were present throughout both lung fields. Some improvement followed the administration of digitalis. Five hours after birth the cardiac murmur was noted to have changed in character; the systolic murmur was replaced by a harsh continuous murmur audible over the entire precordium and left lateral chest. During the first 2 days of life the clinical condition improved little. He remained desperately ill with persistent cyanosis and tachypnea. He was transferred to Brooke General Hospital on April 16, 1962, at 48 hours of life.

Physical examination revealed a listless, seriously ill, deeply cyanotic infant with a respiratory rate of 60 a minute and a heart rate of 110 a minute. Examination was hampered because it was not possible to remove the baby from his high oxygen environment without immediate profound cyanosis. Peripheral pulse volume was increased, and all pulses were readily palpable. The precordium was diffusely hyperdynamic to palpation, but no thrill could be felt. Splitting of the second heart sound could not be defined. A grade-III pansystolic murmur was audible over the low precordium and far lateral over the left chest. The murmur appeared to carry faintly through the second sound, but no distinctive continuous murmur was heard. The liver was palpable 2 cm. below the right costal margin and was regarded as enlarged. There were small vascular discolorations on the left ear, under the chin, and on the chest and lower back. The remainder of the physical examination was negative.

The hemoglobin was 19 Gm. per cent, hematocrit value was 67 per cent, and the white blood cell count and differential were normal. Portable chest x-ray (fig. 1) showed cardiomegaly, hyperlucent lung fields, paucity of pulmonary vascular markings, and an ill-defined density in the left base, originally regarded as an area of atelectasis. An electrocardiogram (fig. 2) revealed si-
massive pulmonary arteriovenous fistula

Figure 1

Chest x-ray demonstrating cardiomegaly, hyperlucent lung fields, indistinct main pulmonary artery silhouette, paucity of vascular markings, and an ill-defined radiodensity in the left base.

nus rhythm of 90 a minute, P-wave prominence suggesting right atrial enlargement, and a frontal plane QRS axis of zero degrees. Right ventricular "dominance" anticipated at this age was not present; instead, the precordial progression was that of an older child or adult, and suggestive of left ventricular preponderance.

The auscultatory findings, persisting deep cyanosis, ischemic lung fields on x-ray, and the electrocardiographic changes of left ventricular hypertrophy, simulated the clinical picture of tricuspid atresia and closing ductus arteriosus. On the third day of life, while an electrocardiogram was being recorded, the child became intensely cyanotic and apneic. Heart action remained obvious. Because a right precordial recording electrode was in place on the chest, the left hemithorax was forcefully compressed several times as an aid to respiration. The infant immediately became bright pink and resumed breathing. Although the significance of this was not appreciated at the time, we suspect in retrospect that chest compression must have partially restricted flow through the arteriovenous fistula, decreasing the venoarterial shunt and allowing more adequate perfusion of the normal lung. On the fourth day of life, biplane angiocardiography was

Figure 2

Electrocardiogram reveals a frontal plane QRS axis of zero degrees, P-wave prominence and left ventricular preponderance. All precordial leads are recorded at one-half standard.
performed via the saphenous vein with injection of 8 ml. of 85 per cent Cardiografin into the high inferior vena cava. Angiocardiograms (fig. 3) revealed an enlarged right atrium, ventricle, and main pulmonary artery which were displaced to the right. A huge elongated left pulmonary artery entered a large vascular malformation in the left lower lung. The pulmonary venous channels filled almost instantly from this lesion, outlining an enlarged left atrium and ventricle. The aorta and its branches were elongated. Most of the contrast material followed this abnormal pathway with scanty opacification of the remainder of the pulmonary vessels.

On April 20, 1962, the sixth day of life, surgery was performed. The basal portion of the left lower lobe was found to be almost replaced by the vascular malformation. Occlusion of the left-lower-lobe pulmonary artery resulted in prompt disappearance of cyanosis and marked reduction in heart size. A lobectomy was performed. Figure 4A of the operative specimen, with the vessels distended with saline, demonstrates the dilated tortuous varices with little normal lung tissue. Subsequent injection of the specimen with latex and digestion by alkali-corrosion revealed the extensiveness of the arteriovenous malformation (fig. 4B).

The postoperative course was uncomplicated and there was no return of cyanosis. The child was discharged on May 6, 1962, on the twenty-third day of life. At the time of last follow-up at 9 months of age, the child weighed 25 pounds and had grown and developed normally. Physical examination disclosed a normal heart without murmurs. Electrocardiograms at that time continued to show a frontal plane QRS axis of zero degrees with a normal precordial QRS progression. Physical examination and chest x-rays of both parents revealed no stigmata to suggest familial telangiectasia or pulmonary arteriovenous fistula.

Discussion

Incidence and Clinical Features

Congenital pulmonary arteriovenous fistula is an infrequent lesion with the majority of the reported examples in adults.1 Symptoms may begin in early life,2 and it would appear that many cases are overlooked in infancy and childhood.3 Kafka et al.4 stressed this point in their review of pediatric cases through 1959. Shumacker and Waldhausen5 operated upon five cases 16 years or younger and reported 26 additional cases from the literature. Of these 31 cases, signs and symptoms of the lesion were present from birth in

Figure 3

Angiocardiogram frames: At 1.5 seconds after injection (A) there is opacification of a vascular mass in the left lower lung. Rightward displacement of the right chambers and elongation of the left pulmonary artery is seen. At 2.0 seconds (B) there is dense opacification of the pulmonary arteriovenous fistula and beginning left atrial filling. Scant filling of the remaining normal pulmonary arteries is noted. At 4.0 seconds (C) the left chambers and the elongated aorta and branches are seen.
seven cases, but only two were diagnosed under 1 year of life. Our patient appears to be the youngest in which the diagnosis was made and successful corrective surgery was accomplished.

The diagnosis of pulmonary arteriovenous fistula depends upon recognition of central cyanosis, polycythemia, a roentgenographic pulmonary density, the overlying continuous bruit and associated mucocutaneous telangiectasia. Associated complications such as epistaxis, hemoptysis, cerebral embolism, and cerebral abscess may be present. While the differential diagnosis of each separate manifestation might be exhaustive, the combination of the signs and symptoms in the adult usually leads to the suspicion of pulmonary arteriovenous fistula, and angiocardiology confirms the diagnosis. In infants, however, polycythemia is “physiologic” and, since other pulmonary or cardiac malformations are more commonly the cause of cyanosis, there may be little to direct the physician to the true problem. In our case the significance of the left-lower-lobar opacity was not immediately appreciated.

**Hemodynamic and Electrocardiographic Sequelae**

It is generally considered that pulmonary arteriovenous fistulas do not increase cardiac output and that right heart hemodynamics are usually normal. The clinical observations of a precordial prominence, a hyperdynamic precordium, and the impressive immediate reduction in heart size at surgery would indicate that the cardiac output in this case must have been considerably increased. From the scanty contrast material distributed to the uninvolved pulmonary vasculature and the large quantity shunting through the fistula, one can infer that the resistance to flow through the anomaly was very low, in spite of contrary statements in the literature. This low-resistance pulmonary bypass must have existed throughout intrauterine development instead of the normal high fetal pulmonary vascular resistance. As a consequence, an unusual volume load was prematurely imposed upon the left ventricle, contrary to the normal prenatal circulatory dynamics, accounting for the unusual electrocardiographic findings in this infant. Although most reports of this malformation in children record normal or right ventricular preponderance, the observed electrocardiogram in this infant is consistent with a large prenatal pulmonary arteriovenous shunt and its serious dynamic consequences: marked total reduction in pulmonary vascular resistance and premature volume overload of the left ventricle.

A similar electrocardiogram is seen in a case reported by Lucas and associates. This 3-year-old cyanotic child had a large and direct communication between the right pulmonary artery and the left atrium, a variant of
the condition under discussion. We believe the generation of the electrocardiogram in these cases is similar. One may speculate that a markedly reduced intrauterine pulmonary vascular resistance would cause a reversal of flow through the ductus arteriosus from the aorta into the pulmonary arteries, producing even further left ventricular volume overloading. Additional information cannot be brought to bear on this point, since evidence of patency of the ductus arteriosus was not observed at surgery, nor was it heard clinically after surgery or at follow-up. Indeed, premature closure of the ductus is a possibility under these circumstances.

Differential Diagnosis

The combination of central cyanosis, ischemic lung fields, and left axis deviation most often calls to mind the diagnosis of tricuspid atresia or right ventricular hypoplasia, and such was the consideration in our patient prior to study. The hyperdynamic precordium and the density in the left lower lobe should have modified this consideration. Recently, Shaher\(^5\) has reviewed the cases of cyanosis with electrocardiographic findings of left axis deviation and left ventricular preponderance. Massive pulmonary arteriovenous fistula must be added to this differential diagnosis, since these electrocardiographic changes should be expected whenever this lesion is of sufficient magnitude at birth to produce clinical manifestations and difficulty.

Treatment

Treatment of pulmonary arteriovenous fistulas with this degree of physiologic disturbance should be surgical extirpation. In the review by Stringer and associates\(^2\) it is noted that 30 of 140 patients suffered severe complications or died as the result of their fistulas. In Shumacker and Waldhausen's\(^3\) collected series of 31 children, two brain abscesses were recorded and four others had central nervous system symptoms. Brain abscess and associated meningitis was also the cause of death in the patient reported by Lucas.\(^7\) Surgical results in the children reviewed by Shumacker and Waldenhausen\(^3\) were excellent with one death in 29 operated cases.\(^9\) The present authors have seen another child, 21 months old, with multiple bilateral fistulas. These were resected in two stages, with initial removal of two lesions from the left side and subsequently of the remaining lesion on the right with excellent results.

Hemodynamic Parallel to Massive Systemic Arteriovenous Fistula

An interesting parallel to this case of massive pulmonary arteriovenous fistula is seen in the massive systemic arteriovenous fistula recently reported in a newborn by Walker and associates.\(^10\) In both anomalies, a large, low-resistance run-off is present in the involved vascular bed. In massive pulmonary arteriovenous fistula, total pulmonary resistance is markedly decreased, imposing an excessive venous return to the left heart in the face of unaltered peripheral resistance. The electrocardiogram reflects this left ventricular dominance and the peripheral pulses are full. Cyanosis is the result of the large intrapulmonary shunt. In massive systemic arteriovenous fistula the systemic resistance is markedly reduced, the peripheral pulses are weak, and the right heart chambers are overloaded by a large venous return in the face of unaltered high neonatal pulmonary resistance. Cyanosis results from reversal of flow through the foramen ovale and possibly the ductus arteriosus. In both lesions a continuous murmur may serve to direct attention to the correct diagnosis. Awareness of the existence and the dynamics of these congenital malformations is essential for recognition and important because both types are potentially curable by relatively simple surgical procedures.

Summary

A case of massive pulmonary arteriovenous fistula, diagnosed and treated in the first week of life is reported. A concept of severely altered intrauterine and neonatal pulmonary blood flow is proposed, with consequent left ventricular overload manifested electrocardiographically by left axis deviation and left ventricular preponderance. Consideration of this malformation in seriously ill cyanotic infants.
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with a radiographic density and these electrocardiographic changes is essential, since this is a surgically curable lesion.

References


No university is worthy of the name, that does not do everything in its power to promote original research in its laboratories. It is the duty of the university to see that its professors and teachers are not overburdened with routine teaching, but are given time for investigation and provided with research laboratory facilities and the necessary funds for this purpose.—E. RUTHERFORD.
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