Origin of the Right Pulmonary Artery from the Ascending Aorta

Unusual Cineangiographic and Pathologic Findings

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The following is a case report with autopsy findings of an infant who had anomalous origin of the right pulmonary artery from the ascending aorta. This case demonstrated two unusual findings: a venous cineangiogram showed sequential filling of the right atrium, left atrium, left ventricle, aorta, and the right pulmonary artery without initial opacification of the right ventricle and left pulmonary artery; at necropsy there was a fibrous cord connecting the main pulmonary artery with the anomalous right pulmonary artery.

Case Report

A.B., F54 423. This 20-day-old male infant was admitted to the hospital because of dyspnea and intermittent cyanosis of five days' duration.

The patient was the fifth child of a 34-year-old mother. The pregnancy was marked by slight uterine bleeding during the seventh month. The delivery was normal, birth weight was 2.9 Kg., and physical examination was normal at the time of discharge from the hospital on the third day of life. The infant did well until the fifteenth day, when the mother noted he had a weak cry associated with a rapid respiratory rate.

On admission the weight was 3.5 Kg., length 54 cm., temperature 37.9 C., pulse 180, respiration 68, blood pressure 60 mm. Hg by the flush method in both arms. The child was in acute respiratory distress and markedly cyanotic. No differential cyanosis was seen. Fine rales were present over the left lung posteriorly. There was generalized increased precordial activity, the heart was enlarged to the left anterior axillary line by palpation, and a grade-III, high-pitched, nonholosystolic murmur was present in the third and fourth left interspaces adjacent to the sternum. A grade-II diastolic rumble was present at the apex. The pulmonic sound was loud and single. The liver was enlarged to 4 cm. below the right costal margin. The brachial and femoral pulses felt normal.

The hematocrit value was 43 per cent, the white blood-cell count was 5,800 cells per mm.³ with a differential of three stabs, 29 polymorphonuclears, and 68 lymphocytes. Urinalysis revealed a trace of protein without other abnormalities.

The chest x-rays (fig. 1) revealed marked cardiomegaly enlargement. The left lung field was almost completely obliterated by the cardiac shadow. The pulmonary vasculature was slightly increased in the right lung field. The electrocardiogram was interpreted as showing right ventricular hypertrophy (fig. 2).

The child was treated with digoxin and mercurial diuretics and antibiotic agents. Despite vigorous therapy for the congestive heart failure, there was only slight clinical improvement in the child's condition.

On the fifth hospital day a venous cineangiogram (60 frames per second) was performed with 6 ml. of 90 per cent diatrizoic acid (Hyopaque—Winthrop Laboratories) injected into the right saphenous vein. The child's condition was unchanged immediately after the procedure. Ten hours later, he developed apneic spells and died.

Cineangiogram showed prompt filling of a large right atrium from the inferior vena cava (fig. 3). The contrast material shunted directly through to the left atrium and into a large left ventricle. During this time there was no filling of the right ventricle or main pulmonary artery. The aorta then filled and had a peculiar appearance with apparent dilatation of the proximal aorta from which a vessel arose going to the right lung. The right ventricle and a normally positioned main and left pulmonary artery filled late.

At necropsy the heart showed marked enlarge-
Cardiac x-rays revealed marked cardiomegaly with the cardiac shadow filling most of the left lung field. The pulmonary vascular markings were increased in the right lung, and were inapparent in the left lung due to the large cardiac shadow.

Anomalous artery (fig. 4) arose 0.5 cm. above the aortic valve from the right side of the ascending aorta and entered the hilus of the right lung, where it divided into three branches. The circumference of this vessel was 1.3 cm., the aorta proximal to it was 1.8 cm., and the distal aorta was 1.1 cm. in circumference. A small patent ductus, 0.5 cm. in length and 0.4 cm. in external diameter, connected the descending aortic arch with the main pulmonary artery. Additionally, there was a fibrous cord 0.2 cm. long and 0.1 cm. in diameter that connected the dorsal surface of the main pulmonary artery to the dorsal surface of the anomalous right pulmonary artery (fig. 5). On the internal surface of the anomalous artery was a small depression at the point of attachment of the cord. The main pulmonary artery arose from the right ventricle in a normal position and supplied the left lung.

The fibrous cord between the pulmonary trunk and anomalous right pulmonary artery was composed of collagen and elastic fibers, which joined with those of the anomalous artery and pulmonary trunk (fig. 6). No lumen was present in the cord. The lungs showed extensive intra-alveolar hemorrhage, chronic passive hyperemia, and focal areas of emphysema and atelectasis. These changes were slightly more prominent in the right lung.

No significant differences were revealed between the pulmonary vascular beds of the right and left lungs. The small pulmonary arteries showed apparently normal thickness of the wall when compared with similar vessels in autopsy specimens of a similar age group.

*Electrocardiogram demonstrated right axis deviation and a clockwise QRS loop. The prominent Q wave in V1 suggests right ventricular hypertrophy.*

*Figure 2*
Figure 3
Cineangiogram following injection of contrast material into right saphenous vein. RA, right atrium; LA, left atrium; LV, left ventricle; Ao, aorta; Rt. PA, right pulmonary artery. A. Sequential filling of right atrium, left atrium, and left ventricle without opacification of the right ventricle. The "filling defect" in right ventricular area gives picture as is seen in tricuspid atresia. B. Following opacification of the large dilated proximal portion of the aorta, the right pulmonary artery was demonstrated to arise from the ascending aorta. The right ventricle and main and left pulmonary arteries are not opacified.

Discussion
A review of the literature concerning aortic origin of the right pulmonary artery has been made by Griffiths and co-workers. These authors point out that this diagnosis depends on peripheral venous or selective angiographic technics. We have been able to find only two reports of this entity with associated angiographic studies. Caro et al.2 presented the findings in a 23-year-old patient. Their study, with a peripheral venous injection, showed sequential filling of the right heart and the main and left pulmonary arteries, with subsequent opacification of the left heart, ascending aorta, and right pulmonary artery. Selective cineangiography was performed in the 10-month-old patient of Armer and co-workers,8 with injections into the patent ductus arteriosus, main pulmonary artery, right ventricle, left atrium, and ascending aorta.

Following the injection of contrast material into the saphenous vein, the cineangiogram in our patient demonstrated a large right-to-left shunt at the atrial level with subsequent filling of the left heart, aorta, and anomalous right pulmonary artery. Such a shunt at the atrial level in this condition has been suggested by Griffiths et al.1 This may be accounted for primarily on the basis of preferential shunting from the inferior vena cava through the foramen ovale; yet it remains unexplained why the total amount of contrast material was shunted to the left atrium without opacification of the right ventricle and
Figure 5
Posterior view of the heart and right lung showing the fibrous band running from the anomalous right pulmonary artery to the main pulmonary artery. A, right lung; B, anomalous right pulmonary artery; C, fibrous band running from the anomalous right pulmonary artery to the main pulmonary artery; D, main pulmonary artery; E, probe beneath the fibrous band.

Figure 6
Photomicrograph showing anomalous artery on the right side with the fibrous band running to the lower left hand edge. The black-staining elastic fibers in the artery are continuous with those in the fibrous band. (Combined Verhoeff-Masson stain × 84.) A, anomalous right pulmonary artery; B, fibrous band.

main and left pulmonary arteries on its initial passage through the heart.

Schneiderman has reviewed the various possibilities of embryogenesis of aortic origin of the right pulmonary artery. He postulated that the migration of the right sixth aortic arch to the left side of the truncus is retarded so that, with division of the truncus arteriosus, the right sixth arch is caught on the right or aortic side. In the case of Wagenvoort et al. evidence is presented that this malformation results from persistence of the distal (instead of proximal) portion of the primitive sixth right aortic arch and of a portion of the right dorsal aorta. In our case, a fibrous band connected the main pulmonary arterial trunk to the proximal portion of the anomalous right pulmonary artery. The band contained elastic fibers that were continuous with those in the walls of both connected vessels. This is strong evidence that the right sixth aortic arch initially migrated properly; the proximal portion became atretic; and the distal end was retained, with resultant origin of the right pulmonary artery entirely from the aorta. These findings add further support to the embryologic explanation for this malformation suggested by Wagenvoort and co-workers.

Summary
A case of aortic origin of the right pulmonary artery has been presented. A venous cineangiocardiogram demonstrated filling of the left side of the heart from the right atrium with subsequent opacification of the aorta and the anomalous right pulmonary artery without initial opacification of the right ventricle and main plus left pulmonary artery. At necropsy there was a fibrous band connecting the main pulmonary artery with the anomalous vessel. The embryologic implications suggest that this case represents atresia of the proximal portion of the right sixth aortic arch.

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REFERENCES


Effect of Electricity on Muscular Motion

From the outcome and result here expounded, if I do not explain and express myself ill, it is clearly understood that muscular contractions can be produced by natural electricity in the living animal from three different causes: first from a violent overcharge of the muscular cell induced by the powers of the mind, and this seems to occur in voluntary motions; second from a forced overcharge, as when, by some external agent or irritation, the aforesaid electricity is determined to descend forcibly and violently from the brain to the muscles, as in reflex motions, and I call this action an overcharge, assuming that some charge, as seems very likely, is in the muscle constantly and naturally; third and finally from a charge equally violent and forced, as will occur when some external agent, applied to the nerve or to the brain, determines the electricity of the internal surface of the muscle to ascend through the nerve and go back to the external surface of that muscle.

Given these three causes, it seemed to me that I saw open a wide field for the felicitous explanation, not only of voluntary motions, but also of unnatural and violent ones; and of various nervous maladies and their causes, as also of their relations to terrestrial and atmospheric electricity.—Luigi Galvani. Commentary on the Effect of Electricity on Muscular Motion. Translated by Robert Montraville Green, M.D. Cambridge, Massachusetts, Elizabeth Licht, Publisher, 1953, p. 95.
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