Left Pulmonary Artery from Ascending Aorta in Tetralogy of Fallot

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SUMMARY

Reported is the first surgically corrected case of tetralogy of Fallot with anomalous origin of the left pulmonary artery from the ascending aorta. The diagnosis of origin of the left pulmonary artery from the aorta depends on an aortogram, but clinical hints are increased vascularity in the left lung compared with the right lung and a continuous murmur. In tetralogy of Fallot, when there is late filling of the left pulmonary artery after a right ventricular cineangiogram, anomalous origin of the left pulmonary artery should be suspected and confirmed by an aortogram. Differentiation from complete absence of the left pulmonary artery, which is also often associated with tetralogy of Fallot, is possible by chest roentgenogram.

Additional Indexing Words:
Selective cineangiography
Anomalous pulmonary artery
Surgical correction of tetralogy of Fallot

THE DIAGNOSIS of anomalous origin of the left pulmonary artery from the ascending aorta with tetralogy of Fallot (no left pulmonary artery arising from the main pulmonary artery) can be suspected clinically and after a right ventricular cineangiogram, but an aortogram is essential to confirm the diagnosis. If anomalous origin of the left pulmonary artery from the aorta is not recognized, attempted complete correction of tetralogy of Fallot can be disastrous. This case shows that if anomalous pulmonary artery is diagnosed, it is possible to do a total correction of tetralogy of Fallot, including anastomosis of the anomalous pulmonary artery to the main pulmonary artery.

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Case Report

History

A 20-month-old girl, K.H., had a heart murmur at birth. Cyanosis was first noted at 2 months of age. Intense cyanosis and loss of consciousness for a few seconds occurred after crying on five occasions between 8 and 12 months of age. She had a transient right hemiparesis at 11 months of age.

Physical Examination

She had moderate cyanosis on crying. There was a loud, single second heart sound, a grade III/VI harsh, systolic ejection murmur at the left upper sternal border, and also a grade III/VI continuous murmur at the left upper sternal border.

ECG and X-ray

Electrocardiogram showed right axis deviation and right ventricular hypertrophy. Chest roentgenograms (fig. 1) showed right ventricular enlargement with prominent vascular markings in the left hilum, especially when compared with the decreased vascular markings in the right lung.

Catheterization

Catheterization and cineangiography were typical for tetralogy of Fallot except that on right ventricular cineangiography there was almost simultaneous filling of the ascending aorta and
Figure 1
Chest X-ray. Note the increased vascular markings in the left lung as compared with the right lung.

The right pulmonary artery and late opacification of the left pulmonary artery. Injection of contrast material in the main pulmonary artery (fig. 2) showed that the only continuation of the main pulmonary artery was a right pulmonary artery. An aortogram (fig. 3) revealed a left pulmonary artery originating from the ascending aorta. The left main pulmonary artery appeared to be kinked, but the pulmonary vessels to the left lung were normal after their origin from the ascending aorta.

Surgery
Complete correction was accomplished at 20 months of age, using cardiopulmonary bypass with the heart suspended in an iced saline bath during aorta cross-clamping. The left pulmonary artery came off the left side of the ascending aorta approximately 2 cm from the origin of the aorta (fig. 4). There was a slight kink in the left main pulmonary artery. The right pulmonary artery was normal but came off a stenotic main pulmonary artery. Severe infundibular hypertrophy was corrected, the ventricular septal defect closed with a patch, and an incision was carried out through the stenotic pulmonary valve into the main pulmonary artery. The left main pulmonary artery was disconnected from the ascending aorta and anastomosed to the main pulmonary artery. A pericardial patch was used to widen the main pulmonary artery. The patient tolerated the procedure well.

On follow-up 9 months after surgery, she appeared to be a normal, very active little girl.

Discussion
Although anomalous origin of the left pulmonary artery from the aorta with tetralogy of Fallot is uncommon, it is important that the condition be recognized, especially prior to surgery. In the case reported here, a continuous murmur and slightly increased vascularity in the left lung as compared with the right lung were hints to the diagnosis. On
Figure 4

Schematic drawing of findings at surgery.

A right ventricular cineangiogram the left pulmonary artery filled late, which is suggestive of anomalous origin of the left pulmonary artery from the aorta with tetralogy of Fallot. The ultimate diagnosis of origin of the left pulmonary artery from the aorta depends on an aortogram, as in this case.

Death at 4 years of age occurred in a case of left pulmonary artery from the ascending aorta with tetralogy of Fallot reported by Mudd and colleagues;2 in that case increased vascularity in the left lung was also noted. Attempted total correction in a case of tetralogy of Fallot with left pulmonary artery from the arch of the aorta, described by Czarnecki and colleagues,1 ended in surgical disaster because the anomalous left pulmonary artery was not recognized before surgery. In another reported patient the left pulmonary artery originated from the ductus arteriosus, and complete surgical correction was accomplished.3, 4

Anomalous origin of the left pulmonary artery from the aorta also occurs without tetralogy of Fallot; these cases also usually have increased vascularity in the left lung. The three reported cases of left pulmonary artery from the ascending aorta without tetralogy of Fallot or other heart disease5-6 all had a right aortic arch, whereas the four known cases (including the case presented here) associated with tetralogy of Fallot all had a left aortic arch.

Differentiation from Absence of the Left Pulmonary Artery

Differentiation between complete absence of the left pulmonary artery and anomalous origin of the left pulmonary artery was not attempted in the past.7 The differentiation is now important because complete surgical correction is possible in cases of anomalous origin of the left pulmonary artery but hazardous in tetralogy of Fallot with absence of the left pulmonary artery.

About half of the cases of absence of the left pulmonary artery are associated with tetralogy of Fallot, with either a right or left aortic arch.7, 8 In cases of absence of the left pulmonary artery not associated with tetralogy of Fallot, a right aortic arch appears to be usual.8, 9

The roentgenologic picture is the diagnostic feature in the absence of the pulmonary artery and differs from that of a left pulmonary artery from the ascending aorta. With absence of the left pulmonary artery, there is hypoplasia of the left lung (and possibly increased vascularity in the right lung). In anomalous origin of the left pulmonary artery from the ascending aorta, there is no hypoplasia of the left lung since there is adequate blood flow from the anomalous pulmonary artery.

Right ventricular cineangiocardiology in absence of the left pulmonary artery will reveal no opacification of the left main pulmonary artery, whereas in anomalous origin of the left pulmonary artery from the aorta with tetralogy of Fallot, there will be late filling of the left pulmonary artery.

Absence or Anomalous Origin of Right Pulmonary Artery

Origin of the right pulmonary artery from the aorta has been reported in association with
tetralogy of Fallot in only one case. All of the reported cases appear to have a left aortic arch, and the pulmonary vascularity was usually increased on the right side. \cite{1, 3, 8, 11-13}

Absence of the right pulmonary artery was also never associated with tetralogy of Fallot, except in one case of mirror-image dextrocardia. All of the cases also had a left aortic arch (except the one case of mirror-image dextrocardia) and decreased vascularity and hypoplasia of the right lung.

**Embryology**

The main pulmonary trunk is derived from the truncus arteriosus as a result of its septation. The right and left main pulmonary arteries arise from the proximal sixth aortic arches. The intrapulmonary arteries develop from the primitive vessels in the lung buds.

If there is defective septation of the truncus arteriosus, a pulmonary (sixth) arch will remain connected to the aorta proximally. Involutionary changes occurring in the arch determine the final defect. If the proximal segment remains patent and the distal segment involutes, there will be anomalous origin of a pulmonary artery from the ascending aorta. If the distal segment remains patent and the proximal segment involutes, the anomalous pulmonary artery originates from the arch of the aorta. If both the distal and proximal segments involute, the affected pulmonary artery will be absent.

The three types—absence of a pulmonary artery, anomalous origin of a pulmonary artery from the ascending aorta, and anomalous origin from the arch—should be differentiated.

Unequal division of the truncus arteriosus is unlikely to occur on the left without dislocation of the truncoconal ridge in the conus. Thus, tetralogy of Fallot should be common with the left anomaly, but not with the right.

**Surgery**

There have been reports of successful anastomosis of an anomalous pulmonary artery to the main pulmonary artery in cases without tetralogy of Fallot\cite{5, 6, 11-13} and in one case associated with tetralogy of Fallot where the anomalous pulmonary artery originated from the ductus arteriosus. The first known successful surgery in tetralogy of Fallot with an anomalous pulmonary artery from the ascending aorta was accomplished in the case reported here. If the anomalous pulmonary artery is not diagnosed before surgery, attempted total correction of tetralogy of Fallot may result in exsanguination of the patient. If the anomalous origin of the left pulmonary artery from the aorta is diagnosed by an aortogram, the left pulmonary artery can be anastomosed to the main pulmonary artery at the time of correction of the tetralogy of Fallot. With an anomalous left pulmonary artery from the aorta, there will probably be progressive pulmonary hypertensive vasculature changes in the left lung, and, therefore, complete correction at an early age is probably indicated.

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**References**


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