Left-Sided Patent Ductus Arteriosus and Right-Sided Aortic Arch

Angi cardiographic Findings in Three Cases

By Israel Steinberg, M.D.

When there is a right-sided aortic arch, persistence of a left-sided ductus arteriosus can readily be demonstrated by angi cardiography. This is important because in some types of cyanotic congenital heart disease, such as in the tetralogy of Fallot, an artificial aorti co-pulmonary (Blalock-Taussig or Pott's shunt) is often created in order to alleviate the distress produced by anoxia.

Two patients with congenital cyanotic heart disease herein reported had right-sided aortic arches, atretic main pulmonary arteries, right-to-left shunts at the ventricular level, and left-to-right shunts from a patent (left-sided) ductus arteriosus to the left pulmonary artery (pseudotruncus arteriosus). Another patient with tricuspid and main pulmonary atresia had a hypoplastic right ventricle, a right-to-left shunt at the atrial level, a large left ventricle, a dilated ascending aorta with right-sided aortic arch, and a left-sided ductus arteriosus forming a left-to-right shunt with the left pulmonary artery. Demonstration of the patent ductus arteriosus in these cases precluded surgery.

Case Reports

Case 1. Cyanotic Congenital Heart Disease with Left-Sided Patent Ductus Arteriosus, Right-Sided Aortic Arch, and Tetralogy of Fallot (Pseudotruncus Arteriosus).

A 6-year-old girl (N.Y.H. no. 827735) was referred for angi cardiography on June 30, 1959, because cardiac catheterization data at another hospital had suggested the diagnosis of an aorti co-pulmonary window shunt. The patient did not squat but was mildly cyanotic, dyspneic, below average weight (14.9 Kg.), and had mild clubbing of the fingers and toes. The pulmonic second sound was not split, and there were no cardiac murmurs. A continuous murmur was heard over the right lung, posteriorly more than anteriorly. The systolic blood pressure was 120 mm. Hg, and the femoral arteries were palpable. The hemoglobin was 15.4 Gm./100 ml. and the erythrocyte count 5.0 million/mm.3. The electrocardiogram showed right atrial and ventricular enlargement.

Roentgen studies of the chest showed a boot-shaped heart, with concavity of the cardiac silhouette in the region of the pulmonary artery. The pulmonary vasculature of both lungs was decreased (fig. 1A). The esophagus was indented along the right outer border. Serial intravenous angi cardiography disclosed immediate filling of a large ascending aorta, right aortic arch, and descending aorta. A patent ductus arose from the left side of the aortic arch and opacified small branches of both pulmonary arteries (fig. 1B). The main pulmonary artery was not visualized.

Case 2. Cyanotic Congenital Heart Disease with Left-Sided Patent Ductus Arteriosus, Right-Sided Aortic Arch, and Tetralogy of Fallot (Pseudotruncus Arteriosus).

An 8-month-old female infant (N.Y.H. no. 875944) was referred for angi cardiography on September 7, 1960, because of recurrent cyanotic and apneic attacks. These began at the age of 7 weeks and recently had been increasing. Examination revealed mild cyanosis but without clubbing of the extremities. A holosystolic murmur (grade III to IV) was heard over the precordium and throughout the lung. Both femoral arteries were palpable. The hemoglobin was 19 Gm./100 ml. and the hematocrit value was 55 per cent. The electrocardiogram showed incomplete right bundle-branch block and right atrial and ventricular enlargement.

Roentgenography of the chest showed slight enlargement of the heart with concavity of the cardiac silhouette in the region of the pulmonary artery. The pulmonary vasculature was diminished, and a right-sided indentation of the esophagus was characteristic of a right aortic arch (fig. 2A). Simultaneous, serial, biplane, intravenous angi-
cardiography revealed enlargement of the right atrium and ventricle. A right-sided aorta was immediately opacified from the right ventricle. A large 4-mm. left-sided ductus arteriosus emerged from the undersurface of the left subclavian artery and opacified the right and left pulmonary arteries. The main pulmonary artery was not visualized (fig. 2B).


A 3½-year-old girl (N.Y.H. no. 900128) was referred for angiocardiography by Dr. Henry P. Goldberg on January 4, 1962. Cyanosis, first noted at the age of 7 weeks, had gradually increased. Examination showed severe cyanosis and

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**Figure 1**

Case 1. A, left. Conventional frontal roentgenogram of the chest showing a boot-shaped heart, concave pulmonary artery segment, and right-sided aortic arch. B, right. Frontal angiocardiography revealing immediate opacification of the ascending aorta from an enlarged right ventricle and a right-sided aortic arch. A left innominate artery arising from the arch bifurcates to give left common carotid and left subclavian arterial branches. The main pulmonary artery is atretic, but the left-sided ductus arteriosus (arrow) arises from the left subclavian artery and opacifies both pulmonary arteries.

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**Figure 2**

Case 2. A, left. Frontal roentgenogram of the chest showing cardiac enlargement, concavity of the pulmonary artery segment, and a right-sided aorta indenting the right outer surface of the esophagus. The pulmonary vasculature is markedly diminished. B, right. Frontal angiocardiogram revealing right ventricular enlargement, an atretic main pulmonary artery, a right-sided aortic arch, and a left-sided patent ductus arteriosus arising at the lowermost and larger branch of the left subclavian artery and opacifying both pulmonary arteries (arrow). Note that the left subclavian and common carotid arteries arise from a left-sided innominate artery.
clubbing of the extremities. A short systolic murmur was heard along the right sternal border and a high-pitched continuous murmur was present at the base of the heart. The hemoglobin was 18.7 Gm./100 ml. and the hematocrit level was 61.5 per cent. The electrocardiogram showed left axis deviation and enlargement of both atria and the left ventricle.

Roentgen studies of the chest showed moderate enlargement of the heart, a concave pulmonary artery segment, a right aortic arch, and diminished pulmonary vasculature (fig. 3A). Simultaneous, serial, biplane, intravenous angiocardiography disclosed tricuspid atresia, atrial septal defect with right-to-left shunt, and a huge ascending aorta with a right aortic arch. A large (8 mm.) ductus filled both pulmonary arteries. The left pulmonary artery was hypoplastic and the main pulmonary artery was atretic (fig. 3B and C).

**Discussion**

Edward's classic paper on the anomalies of the derivatives of the aortic arch system very adequately describes the embryology of a left-sided patent ductus arteriosus and right-sided aortic arch. Since the pattern of the right-sided aortic arch and its branches is a mirror image of the normal aorta, the aortic arch passes over the right major bronchus and blends with the descending aorta (figs. 1B, 2B, and 3B). The upper portion of the aortic arch lies to the right of the esophagus and to the right of the midline of the spine (figs. 2A and 3A). This anomaly is a derivative of the embryonic double aortic arch. Therefore, the left subclavian artery in common with the left common carotid artery arises from a left-sided innominate artery and a patent ductus arteriosus extends from the subclavian branch of the innominate artery to the left pulmonary artery (figs. 1B, 2B, and 3B). The left-sided innominate artery is the first branch of the aortic arch, whereas the right common carotid and right subclavian arteries are the second and third branches, respectively. Because the aortic arch, its branches, and the upper part of the descending aorta are mirror images of the normal aorta, there is no encroachment upon the trachea or esophagus.

Taussig has also pointed out that right-sided aortic arches are frequently associated with congenital cardiac defects, especially tetralogy of Fallot. Indeed, two of the cases

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*Figure 3*

Case 3. A, left. *Frontal esophagram showing cardiac enlargement, a concave pulmonary artery segment, and decreased lung vasculature.* B, center. *Frontal angiocardiogram demonstrating a right-sided aortic arch, the left innominate artery, and the subclavian artery with a large ductus (arrow) opacifying both pulmonary arteries. The main pulmonary artery is atretic.* C, right. *Simultaneous, biplane, lateral angiocardiogram of B, revealing atresia of the main pulmonary artery, a dilated ascending aorta, and the pulmonary arterial branches in the aortic window opacified by a patent ductus arteriosus.*
LEFT-SIDED PATENT DUCTUS

reported above (case 1 and 2) have the latter malformations; the pulmonic stenotic component of the syndrome consists of atresia of the main pulmonary artery, which forms a combination of anomalies known as a pseudotruncus arteriosus. Case 3, in addition, has tricuspid atresia, the main pulmonary artery being also atretic.

Ordinarily, the diagnosis of the typical patent ductus arteriosus can be made by eliciting the classical continuous (machinery) murmur at the base of the heart. This may be simulated by other conditions, such as a ruptured aortic sinus. Coronary arteriovenous fistulas, adjacent systemic (chest wall), or pulmonary arteriovenous fistulas may also have to be differentiated from a patent ductus. In cyanotic congenital heart disease a continuous murmur may sometimes be produced by bronchial collateral circulation. The murmur of a ventricular septal defect may also be continuous. Furthermore, pulmonary hypertension and reversal of blood flow through a ductus may alter the machinery murmur of a patent ductus arteriosus. Indeed, the murmur may be absent or be replaced by a systolic murmur. Accordingly, contrast visualization of the ductus is therefore indicated.

Countercurrent, retrograde left brachial aortography, and retrograde femoral aortography have become well-established methods for visualizing the ductus. Intravenous right heart catheterization with advancement of the catheter into a patent ductus has also long been recognized as a way of diagnosis of a patent ductus. Because the normally placed aortic arch conceals a patent ductus arteriosus, intravenous angiography has, therefore, depended upon indirect signs, i.e., blanching of the left pulmonary artery following right heart opacification and reopacification of the left pulmonary artery when the left heart structures are opacified. When there is patency of a left-sided ductus arteriosus and the aortic arch is right-sided, especially when there is also pulmonary atresia, direct opacification of the ductus soon after the aorta is visualized is readily achieved (figs. 1B, 2B, and 3B).

Summary and Conclusions

Angiocardiography in three female patients, aged 8 months, 3½ years, and 6 years, respectively, established patency of a left-sided ductus arteriosus and a right-sided aortic arch. All had congenital cyanotic heart disease and atresia of the main pulmonary artery. Two cases fitted into the category of the pseudotruncus arteriosus type of tetralogy of Fallot in that there was atresia of the main pulmonary artery, enlargement of the right ventricle, a right-to-left shunt at the ventricular level, and immediate opacification of a dilated ascending aorta. The other patient had the classic features of tricuspid atresia with a right-to-left shunt at the atrial level, hypoplasia of the right ventricle, and an enlarged left ventricle, which opacified a large ascending aorta. In every instance the ductus arose from an inferior branch of the left subclavian artery and opacified both pulmonary arteries. The left subclavian and left common carotid arteries came off from a left innominate artery which was the first branch of the right-sided aorta.

Since an artificial aortico-pulmonary shunt is often indicated for relief of cyanotic congenital heart disease, demonstration of a persistent ductus arteriosus is important and precludes surgery for the creation of an aortico-pulmonary shunt.

References


ERRATUM

Please substitute the following text for the abstract published in Circulation, October 1963, part two, volume XXVIII, no. 4, page 828, since the study was concerned with myocardial tissue analyses and not serum analyses.

Coronary Stenosis and Myocardial Enzyme Loss

Frederick T. Zugibe, Thomas Conley, and John W. Vester, Pittsburgh, Pennsylvania

Alterations in cardiac function have been frequently observed without complete obstruction of the coronary arteries and without gross or microscopic evidence of infarction. The circumflex arteries of 50 dogs were constricted to varying degrees to determine the effects of partial occlusion on myocardial function. The dogs were sacrificed a week after constriction, and samples of myocardium were taken from an area supplied by a control artery (anterior descending) for glutamic pyruvic transaminase (GPT) and glutamic oxaloacetic transaminase (GOT) analyses. The total content of these components were correlated with objective measurements of degree of constriction by a plastic-mold technique developed in this laboratory.

Our preliminary studies revealed that in the absence of sufficient collateralization, a constriction in excess of 40 per cent resulted in significant loss of GPT and GOT (15-75 per cent of the control). The study was then extended to human autopsy material where similar results were obtained. In these cases, 6 individuals were found to have constriction of the anterior descending in excess of 45 per cent, with 4 individuals showing a significant loss of GPT and GOT (25-75 per cent of the control). The 2 cases that showed no loss in tissue components were induced with well-developed collateral systems, while the remaining 2 showed a poor collateral supply. These findings suggest that partial coronary-artery constriction can result in myocardial alteration without apparent infarction by causing a loss of muscle-cell components.
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ISRAEL STEINBERG

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