CASE REPORTS

Aberrant Course of the Left Anterior Descending Coronary Artery Associated with Anomalous Left Circumflex Origin from the Pulmonary Artery

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SUMMARY
We have presented a two year nonoperative follow-up of an asymptomatic 14-year-old girl with an unusual anomaly of the coronary arteries. Systolic and diastolic murmurs heard preoperatively did not change following severance of a patent ductus arteriosus. Postoperative selective coronary arteriography revealed (1) anomalous origin of the left anterior descending artery from the right aortic sinus of Valsalva crossing between the aorta and pulmonary artery, (2) aberrant circumflex artery origin from the pulmonary artery and (3) collaterals from the left anterior descending and right coronary arteries to the circumflex artery. This combination of coronary anomalies has never been described previously.

Anomalous origin of the coronary arteries from the aorta or pulmonary artery is infrequent.1, 2 The presence of these anomalies is of some clinical interest since certain types are associated with an increased risk of sudden death.3-6 This paper describes a 14-year-old girl with a patent ductus arteriosus in whom (1) the left anterior descending coronary artery arises aberrantly from the right anterior aortic sinus of Valsalva and courses between the aorta and pulmonary artery and (2) the left circumflex artery arises from the pulmonary trunk and communicates with the left anterior descending and right coronary arteries via collaterals. This particular combination of anomalies has never been previously described.

Case Report
A 14-year-old Spanish girl was the product of a normal pregnancy and delivery. Childhood development was normal except for frequent respiratory tract infections one of which led to the discovery of a heart murmur at the age of 13. She was referred to hospital in May 1973 for further evaluation. Her exercise tolerance was normal and she had never experienced syncope, chest pain or symptoms of congestive heart failure. The remainder of the functional inquiry was negative. On physical examination the blood pressure was 105/70 mm Hg and the pulse rate 80/min. The point of maximal intensity was in the 5th interspace in the left mid-clavicular line and was normal. There were no thrills palpable. Arterial pulsations were within normal limits. Auscultation at the left sternal border revealed a grade II/VI holosystolic murmur and a grade III/VI late crescendo diastolic murmur maximal in intensity at the second left interspace. The remainder of the physical examination was within normal limits. Electrocardiogram was normal. Chest X-rays showed a slight increase in pulmonary vascularity, normal chamber sizes and a normal cardiothoracic ratio. A phonocardiogram was done which documented the auscultatory findings. Right heart catheterization was performed and the diagnosis of a patent ductus arteriosus was made when the catheter passed from the pulmonary artery into the descending aorta. Pressures in the right heart, pulmonary artery and descending aorta were normal. Oximetry revealed a left-to-right shunt at the pulmonary artery level which was 23% of pulmonary flow. She underwent surgery on June 7, 1973 and a small patent ductus arteriosus was found, ligated, and divided. A pulmonary artery thrill was not felt during surgery. The postoperative course was uneventful. However, the systolic and diastolic murmurs remained unchanged. A postoperative phonocardiogram showed no significant change from the one taken preoperatively. Because a coronary arteriovenous (AV) fistula was suspected, she underwent coronary arteriography and aortography. Selective...
right coronary artery injection (fig. 1) reveals this artery to be moderately dilated with a normal origin and distribution. The left anterior descending artery arises normally in the right coronary sinus (fig. 2) and courses between the pulmonary artery and aorta (fig. 3). The left circumflex artery arises from the pulmonary artery and receives collaterals directly from the posterior descending artery, the distal right coronary artery, the proximal right coronary artery via atrial collaterals (figs. 1-3) and the conus branch of the right coronary artery which arises separately in the right coronary sinus (fig. 4). The obtuse marginal branch of the left circumflex receives collaterals from the left anterior descending (fig. 3) and from the posterior descending coronary arteries (fig. 1). The patient was subsequently discharged from hospital and has remained asymptomatic. A submaximal treadmill exercise test performed in April 1975 was negative for ischemia at a heart rate of 170/minute.

Discussion

We have presented an asymptomatic 14-year-old girl with a patent ductus arteriosus and very unusual coronary artery anatomy. The anatomical variations are particularly interesting because (1) the left circumflex artery arises from the pulmonary artery and receives collaterals from both the left anterior descending and the right coronary arteries and (2) the left anterior descending coronary artery originates in the right coronary sinus and passes between the aorta and pulmonary artery.

Clinical findings resulting from retrograde flow into the pulmonary artery from a solitary coronary AV fistula can lead to the erroneous diagnosis of a ductus.9, 10 Similarly, anomalous origin of a coronary artery from the pulmonary trunk can mimic the auscultatory findings of a ductus if a significant amount of blood is flowing through the anomalous vessel. Aberrant origin of a coronary artery from the pulmonary trunk was not suspected in this patient because the clinical findings were compatible with a ductus and a ductus was found at the time of catheterization and surgery. Postoperatively, when the murmurs persisted, the correct diagnosis was postulated and subsequently documented by coronary arteriography.

Anomalous origin of the left main coronary artery from the pulmonary trunk is poorly tolerated with the majority of patients dying in infancy.6, 7 Detection of this lesion in adult life is an indication for prompt corrective surgery because of the high risk of sudden death.8 Aberrant origin of the right coronary artery from the pulmonary trunk is better tolerated. This lesion has not been associated with myocardial infarction or sudden death and in several cases it has been discovered as an incidental postmortem finding.11-13 There are no data on the longevity of patients with anomalous left circumflex origin from the pulmonary artery because of the rarity of this anomaly.

There have been several postmortem reports of sudden death during or just after exercise in males in
Selective left anterior descending artery injection in the 30° right anterior oblique position (left) and in the 45° left anterior oblique position (right). These photographs show a left anterior descending artery (LAD) arising aberrantly in the right coronary sinus, coursing between the aortic root and the pulmonary artery (PA) and giving off small septal branches (S) and numerous collaterals to the marginal branch (Marg) of the circumflex artery which fills the proximal circumflex artery (Prox Cx) by retrograde flow. This marginal branch also receives collaterals from the posterior descending artery (fig. 1).

Figure 3

whom the left coronary artery arose from the right coronary sinus of Valsalva or right coronary artery and subsequently coursed between the aortic root and pulmonary artery. In most cases, sudden death appears to have been the result of an inability to increase coronary blood flow acutely. Several hypotheses postulated to explain this phenomenon include 1) spasm or compression of the aberrant artery between the aorta and pulmonary trunk, 2) an anatomical derangement at the takeoff of the anomalous vessel, 3) congenital underdevelopment of the left coronary artery system and 4) a combination of the above. For these reasons, it is difficult to know the role or type of cardiac surgery that should be done in the asymptomatic patient with this anomaly who does not exhibit arrhythmias or have signs and symptoms of myocardial ischemia.

The patient we have presented has an anomalous left anterior descending artery arising from the right coronary sinus and passing between the aorta and pulmonary artery, and an anomalous left circumflex artery arising from the pulmonary artery and receiving collaterals from the left anterior descending and right coronary arteries. We have decided not to reoperate on this patient at the present time for three reasons. 1) The patient is asymptomatic. 2) Anomalous origin of the left circumflex artery from the pulmonary trunk has not been associated with myocardial infarction, congestive heart failure or sudden death. 3) We do not know if there is an indication for prophylactic cardiac surgery in a young asymptomatic girl with these anomalies. Should this

Figure 4

Selective injection of the conus branch in the frontal view. The conus branch (CB) arises separately from the right coronary sinus and gives collaterals to the proximal circumflex artery (Prox Cx) which drains into the pulmonary artery (PA).
patient develop significant cardiac symptoms she will be re-evaluated. Surgical intervention would involve severance of the aberrant circumflex from the pulmonary trunk and aortocoronary bypass grafts to the proximal circumflex and left anterior descending arteries.

References
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