CASE REPORT

Anomalous Origin of Left Coronary Artery from the Pulmonary Artery with Ventricular Septal Defect

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SUMMARY Only two cases have been reported previously of the association of ventricular septal defect (VSD) with anomalous origin of the left coronary artery (ALCA) arising from the pulmonary artery. The purpose of this paper is to present two additional cases, to describe the pathophysiology, and to emphasize how the clinical course of this combination of defects differs from that of isolated ALCA.

Patients with both of these anomalies present in infancy with manifestations only of a large left–right ventricular shunt and pulmonary hypertension. Initially the ALCA is well perfused from the high pressure in the pulmonary artery. In these instances in which the pulmonary artery pressure subsequently decreased because of spontaneous reduction in size of the VSD, the left coronary arterial system became less well perfused. Because of this decreased perfusion in association with the left ventricular myocardial stress initially caused by volume overload, myocardial ischemia and ultimately infarction occurred. Early identification and repair of the anatomic abnormality could prevent irreversible myocardial damage.

SINCE ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY (ALCA) from the pulmonary artery was described by Abbot in 1908 and the natural course further characterized by Bland, White and Garland in 1933, numerous reports have been published describing symptoms and modes of therapy. Associated congenital cardiac anomalies are extremely rare. Since many cardiac centers are repairing ventricular septal defects in infants when medical management is not satisfactory, then it becomes important to properly identify associated defects. To the best of our knowledge, there are only two published cases concerning the association of ALCA with ventricular septal defect (VSD).

It is the purpose of this paper to report two additional cases of ALCA associated with VSD, to describe the hemodynamics, and to emphasize the difference in the clinical course of ALCA, both with, and without additional congenital cardiac defects.

Patient Material

Case 1

T.S. was brought to the emergency room at 11 weeks of age because of poor feeding, tachycardia, and tachypnea. On examination she was underdeveloped, irritable, and acyanotic. The respiratory rate was 50 per minute and the heart rate was 140 beats per minute. The first heart sound was normal but the second sound was single and of increased intensity. There was a plateau, III/VI holosystolic murmur at the left lower sternal border with radiation to the base and to the posterior chest wall bilaterally. There was a short, low-frequency mid-diastolic murmur at the apex. The liver was enlarged 3 cm below the right costal margin.

The chest radiograph demonstrated cardiomegaly with increased pulmonary arterial markings. The electrocardiogram demonstrated biventricular hypertrophy (fig. 1a). There were no electrocardiographic changes suggestive of ALCA. Digitalis and diuretics were administered with prompt improvement. The clinical impression was VSD. Cardiac catheterization six days later revealed a VSD. There was a significant increase in oxygen saturation in the right ventricle, with no further increase in the pulmonary artery. Other hemodynamic data are listed in table 1.

During the next six months, the patient had numerous episodes of unexplained irritability, crying and tachycardia. However, the systolic murmur decreased in intensity and the diastolic murmur disappeared. Thus, despite auscultatory evidence of decrease in size of the VSD, the patient continued to have symptoms of distress and cardiomegaly by chest radiograph. There was diminution of the R wave in the anterior and mid precordial chest leads and there were ST-T wave abnormalities (fig. 1b) The vectorcardiogram was interpreted as anterolateral myocardial infarction (fig. 2a). Repeat cardiac catheterization demonstrated that the VSD had closed spontaneously. There was no increase in oxygen saturation at either the right ventricular or pulmonary artery level. The left coronary artery arose from the pulmonary artery. The LV ejection fraction was less than 20%.

Failure to improve with medical treatment prompted referral for surgical repair of the ALCA. At operation, the anterior lateral surface of the left ventricle was pale,
ANOMALOUS LEFT CORONARY ARTERY AND VSD/Pinsky et al.

ANOMALOUS LEFT CORONARY ARTERY AND VSD/Pinsky et al.

a) 

**Figure 1.** Electrocardiograms from TS at a) 11 weeks of age, demonstrating increased midprecordial voltage indicative of biventricular hypertrophy and b) at 10 months of age demonstrating marked generalized ST-T wave abnormalities and decreased R wave over anterior precordium.

b) 

aneurysmal and akinetic. The anomalous coronary artery was reimplanted directly into the aorta. Despite the appearance of adequate left coronary artery perfusion after reimplantation, systemic blood pressure could not be maintained and the patient died. At autopsy, the anterior-lateral surface was fibrotic and thin. There was evidence of an old myocardial infarction. The anastomotic site was patent.

Case 2

M.U. was admitted to the hospital at six weeks of age because of tachypnea and tachycardia. On examination, he was underdeveloped, acyanotic, and irritable. The first heart sound was normal and the second single and increased in intensity. There was a grade III/VI plateau holosystolic murmur at the left lower sternal border followed by a diastolic

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**Table 1.** Catheterization Data

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Abbreviations: PA = pulmonary artery; press = pressure; AO = aorta; Qp/Qs = ratio pulmonary to systemic flow.
flow murmur at the apex. The pulses were normal. The electrocardiogram revealed biventricular hypertrophy (fig. 3a). As with T.S., there were no classical electrocardiographic findings of ALCA, although there was a Q wave in aV_{L} and some ST depression in the precordium. Chest radiograph demonstrated cardiomegaly with increased pulmonary arterial markings. The patient was treated with digitalis and diuretics with improvement in symptoms. Catheterization revealed a type II VSD. There was an increase in oxygen saturation in the right ventricle and no further increase in the pulmonary artery. Other hemodynamic measurements are recorded in table 1.

Growth failure, tachypnea, and irritability continued. Although the radiographic heart size remained enlarged, the pulmonary arterial shadows diminished in size. The ECG demonstrated more classical evidence of ALCA with Q waves in I, aV_{L}, V_{6}, and ST depression in lateral precordial leads (fig. 3b). The vectorcardiogram was interpreted as showing an anterolateral myocardial infarction (fig. 2b). Repeat cardiac catheterization at six months of age (table 1) revealed a VSD, an anomalous right ventricular muscle bundle, and ALCA arising from the main pulmonary artery. Again, there was no further increase in oxygen saturation in the pulmonary artery. The LV ejection fraction was reduced to 41%. Because of LV dysfunction and evidence of ischemic changes, surgical closure of the VSD was carried out with resection of the anomalous right ventricular muscle bundle and direct reimplantation of the anomalous coronary artery into the aorta. At the time of operation, the anterior left ventricle was pale, scarred and akinetic. Although post-operatively the left coronary artery perfusion appeared to be adequate, cardiac output could not be maintained and the patient died. At autopsy, the anterolateral surface of the heart was thin. There was diffuse fibrosis in that area and evidence of an old myocardial infarction. The anastomotic site was patent.

**Discussion**

According to Edwards, there is a dynamic coronary flow pattern in ALCA. In the fetal and neonatal period, when there is relatively high pulmonary artery pressure, there is flow from the pulmonary artery to the anomalous coronary artery. Maturation of the pulmonary vascular bed results in a decrease in pulmonary artery pressure and then there is no longer adequate coronary perfusion from the pulmonary artery. The variance in establishment of coronary anastomoses is critical in determining the extent of resultant ischemia or infarction.

In patients with associated VSD due to elevated pulmonary artery pressure, there is presumably a prolonged early phase of antegrade flow from the pulmonary artery to the anomalous coronary artery. Because of this prolonged first phase, adequate collateralization between the right and left coronary anastomoses may not be established. Therefore, as pulmonary vascular resistance and pulmonary pressure decrease, the coronary flow will decrease.

Anomalous LCA with VSD differs in the clinical presentation from isolated ALCA. In the former the initial signs and symptoms are referable to the left-right shunt. The irritability, cardiomegaly, and heart failure often can be improved by proper anticongestive measures. The electrocardiogram is consistent with changes secondary to volume overload rather than ischemia.
FIGURE 3. Electrocardiograms of MU at a) six weeks of age demonstrating increased mid precordial voltage indicative of biventricular hypertrophy and b) six months of age showing abnormal Q waves in left chest with ST-T wave abnormalities.
In the cases presented, we theorize that the subsequent drop in pulmonary artery pressure resulted in decreased coronary flow and increasing myocardial ischemia. In addition to a failure of formation of collaterals vessels, in Case 2, the left ventricle was subjected to increased stress because of the continued volume overload. The patients unexpectedly became worse clinically with increasing myocardial damage. In neither patient during the initial catheterization was a pulmonary artery cineangiogram performed. Because of the pulmonary hypertension, a pulmonary artery cineangiogram would have allowed visualization of the anomalous coronary artery. Neither ventriculogram was adequate to delineate the coronary anatomy. In retrospect, only one (right) coronary artery could be visualized arising from the aorta.

What could be done even if the anomalous coronary artery had been identified at the time of initial evaluation? Several surgical approaches are available that have been successful in infants. These include subclavian-to-coronary artery anastomosis,10,11 direct coronary implantation into the aorta,12,13 and the use of a segment of subclavian artery as a free graft for implantation into the aorta.12 In addition, there have been numerous successes in operation of older children.6 Because of the supposed poor anastomoses between the right and left coronary arteries with associated VSD, ligation of the anomalous coronary artery would be contraindicated.

In each of the two cases presented in this paper, operation was attempted only after extensive myocardial damage had occurred. If the anomalous coronary artery had been identified earlier, before the ECG, radiographic and clinical changes occurred, surgical intervention could have been attempted before the extensive damage resulted. The two previously reported cases of ALCA with VSD both died after the VSD was closed surgically without treatment of the ALCA.7,8 There are reports of patients who had origin of both coronary arteries from the pulmonary artery who died at the time of closure of a ventricular defect.14,15 This re-emphasizes the need for coronary artery identification at the time of catheterization.

On the basis of experience with these patients and currently available diagnostic and therapeutic measures, we therefore recommend: 1) ventricular or aortic cineangiography should be of a quality to be able to identify coronary artery anatomy at the time of initial evaluation; 2) suspect the presence of coronary artery anomalies if myocardial dysfunction occurs, despite evidence of decreasing left-right shunt without pulmonary hypertension; and 3) surgical intervention to create a two coronary artery system before extensive myocardial damage occurs. With increasing success of surgical closure of ventricular septal defects, especially in infants, delineation of anomalies such as the one described is necessary.

References

Anomalous origin of left coronary artery from the pulmonary artery with ventricular septal defect.

W W Pinsky, P C Gillette, D F Duff, N Wanderman, J H Morriss, C E Mullins and D G McNamara

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