CASE REPORTS

Aortic Dysplasia in Infancy Simulating Anomalous Origin of the Left Coronary Artery

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SUMMARY
A case is reported of an infant believed to have had anomalous origin of the left coronary artery. Clinical evaluation and angiography appeared to support the diagnosis. At necropsy, however, the left coronary was seen to arise from the aorta but was totally occluded by dysplastic changes which resulted in myocardial infarction and death of the patient. There was minimal aortic deformity produced by these changes, and no aortic gradient was present. Thus, this very rare case of minimal supravalvular aortic stenosis with left coronary artery occlusion was not recognizable clinically nor angiographically.

Additional Indexing Words:
Supravalvular aortic stenosis  Myocardial infarction

Report of a Case
This white female infant was born of an uncomplicated pregnancy. She weighed 7 lb. 5 oz. at birth. The newborn period was uncomplicated and a well-baby examination at 6 weeks revealed an apparently healthy infant with satisfactory weight gain. At 3½ months of age she was admitted to the Bethesda Naval Hospital because of congestive heart failure and suspected congenital heart disease. Approximately one week prior to admission she began to experience marked diaphoresis, dyspnea and exhaustion with feedings, and during the day preceding admission she manifested persistent respiratory distress and wheezing.

Physical examination at the time of admission was of an acutely ill, tachypneic and restless infant. The blood pressure was 90/60 mm Hg, pulse rate 160 beats/min, and respiratory rate 70/min. Fine crepitant rales were audible over the lower posterior lung fields. Precordial activity was increased and the apical impulse was palpable at the anterior axillary line. A grade III/VI pansystolic blowing murmur was loudest in the apical region and radiated to the left axilla. A loud apical summation gallop was also present. The liver margin was palpable 5 cm below the right costal margin. The remainder of the examination findings were normal.

A chest X-ray (fig. 1) revealed marked cardiomegaly and passive pulmonary congestion. The electrocardiogram (fig. 2) demonstrated left axis deviation and abnormal Q waves in leads I, AVL, and V₄₋₅. The P wave suggested left atrial enlargement and the ST-T waves were abnormal.

The clinical impression was anterolateral myocardial infarction secondary to anomalous origin of the left coronary artery. The murmur was attributed to mitral regurgitation.

Digitalis and diuretic therapy resulted in significant improvement. Conservative management was elected and the infant was subsequently discharged. However, she was readmitted at 4½ months of age because of pneumonia of the right middle lobe. She responded satisfactorily to treatment.

Cardiac catheterization performed during this hospital stay revealed a cardiac index of 4.5 liter/min/M². No left-to-right shunt was evident by oximetry. Left ventricular end diastolic pressure was increased to 16 mm Hg, resulting in modest elevation of the mean pulmonary arterial wedge pressure to 14 mm Hg. The pulmonary arterial and right ventricular pressures were 40/20 and 42/6 mm Hg, respectively. The left ventricular and aortic pressures were 120/16 and 120/60 mm Hg, respectively. There was no pressure
AORTIC DYSPLASIA AND CORONARY OCCLUSION

Figure 1
Roentgenogram of chest on admission of patient, showing enlarged heart and passive congestion.

gradient across the aortic valve or in the aorta. An aortogram (fig. 3) demonstrated a large right coronary artery which filled the distal left coronary arterial system by collateral circulation; however, no communication with the pulmonary artery was observed. There appeared to be some bronchial collateral formation as well. There was slight dilatation of the ascending aorta which appeared otherwise normal.

It was again elected to pursue medical management and she did relatively well until 15 months of age when she again was brought into the hospital with severe left-heart failure. On this occasion there was no response to an increase in digitalis and diuretics, and the patient died of intractable congestive heart failure.

Necropsy
The significant findings were limited to the heart, which was enlarged and weighed 150 g. There was moderate biventricular dilatation with severe left ventricular hypertrophy. The cut surface of the left ventricle (fig. 4) showed old and recent anterior and lateral myocardial infarction involving the anterior papillary muscle. The mitral leaflets were slightly thickened. The ascending aorta was minimally narrowed immediately above the sinuses of Valsalva. Only a small dimple was present in the intimal surface of the aorta marking the origin of the left coronary artery. The right coronary ostium and the dominant right coronary artery appeared normal. The left coronary artery distributed normally over the left ventricle (fig. 5). Microscopic examination of the aorta and the left coronary artery revealed dysplastic medial changes in the left aortic sinus which resulted in fibrous occlusion of the left coronary artery at its origin (fig. 6). This process continued about 1 to 1½ cm out into the left coronary artery, beyond which the vessel appeared normal. The "mosaic-like" dysplastic medial changes were localized to the region of the origin of the left coronary artery; the remainder of the aorta appeared normal.

Discussion
Myocardial infarction is uncommon in infants and in children but may result from one of several conditions. Anomalous origin of the left coronary artery and a variety of rare coronary occlusive diseases including medial calcification, periarteritis nodosa, atherosclerosis, syphilitic arteritis,

Figure 2
Electrocardiogram demonstrating anterolateral myocardial infarction.

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Figure 3
Aortogram in left anterior oblique projection.
Cut surface of the left ventricle showing anterior and lateral myocardial infarction (I) involving the anterior papillary muscle (P).

Normal origin and distribution of the left coronary artery.

(a) Longitudinal section through ascending aorta including left aortic sinus (AS). (The aortic valve (AV) has been prolapsed in preparation of the section.) The ostium of the left coronary artery (LC) is shown filled in with fibrous tissue. (b) A deeper section showing the mosaic appearance of the aortic media and the narrow left coronary artery (LC) proceeding through the aortic wall. Intimal fibrous thickening in the artery is evident. (c) A further section in the same series as (a) and (b) in which the left coronary artery (LC) has left the aortic wall and appears in the epicardium. The artery is narrow and shows intimal fibrous thickening. (d) The left coronary artery after it has emerged from the aorta and as it lies in the epicardium showing an overall narrow state and significant intimal proliferation causing narrowing of the lumen. (e) The anterior descending coronary artery beyond the stenotic zone showing a narrow vessel but with only minimal intimal thickening. (Courtesy of Drs. Jesse E. Edwards and Zeev Vladaver.) (elastic tissue stain, × 13.)

coronary embolism, and fibroelastosis of the coronary arteries have been reported.

Supravalvular aortic stenosis may cause coronary occlusion by one of three mechanisms: 1. fusion of the free margin of the aortic valve to the region of supravalvular aortic narrowing; 2. medial thickening and premature atherosclerosis, presumably secondary to increased coronary perfusion pressure; or 3. fibrotic changes in the aortic media which are a part of the supravalvular obstructive lesion and also involve the coronary ostium and proximal
coronary artery. The case described here is believed to be an example of the latter, wherein the supravalvular aortic narrowing was minimal and resulted in no gradient at catheterization, and only minimal deformity by aortography, but produced total obstruction of the left coronary artery, myocardial infarction and death of the patient.

Myocardial infarction in this patient suggests that the left coronary artery was functional during the early stages of development of the left ventricle. The course was similar to anomalous origin of the left coronary artery and quite unlike single coronary artery wherein the blood supply to the left ventricle remains adequate.

Mullens et al. have recently reported a case of a 14-year-old boy with total fibrous occlusion of the left coronary ostium, who survived to age 14 years without myocardial infarction, but manifested angina and ischemic electrocardiographic changes, and subsequently underwent successful saphenous vein bypass graft. The coronary anatomy and pathology were quite similar to the case reported here. The considerable difference in the clinical courses must have reflected a variation in the rate of development of coronary collateral circulation.

While Peterson et al. have reported 20 of 33 cases of supravalvular aortic stenosis to have some coronary artery abnormality, none of the reported deaths appeared due to acute myocardial infarction. This case is thus reported because of this very rare presentation of the supravalvular aortic stenosis which closely simulated anomalous origin of the left coronary artery. The case also illustrates the necessity of very careful scrutiny of the aortogram for aortic deformity in any instance of suspected anomalous origin of the left coronary artery when no left-to-right shunt is present at right heart catheterization and no communication between the left coronary artery and pulmonary artery is demonstrated by angiography.

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References
2. BLAND EF, WHITE PD, GARLAND J: Congenital anomalies of the coronary arteries: Report of an unusual case associated with cardiac hypertrophy. Amer Heart J 8: 787, 1933
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