Aberrant Left Coronary Artery

Report of a Case and Review of the Literature

By Harvey Cohen, M.B., B.Ch., and Shirley Siew, M.B., B.Ch.

Recent advances in the successful surgical treatment of a number of congenital heart diseases have stressed the importance of diagnosis as a first prerequisite to proper management. Rosenbaum,1 in a recent article deprecated the continued use of "idiopathic hypertrophy of the heart" as a nondescript term for heart conditions in infancy and early childhood, characterized by congestive cardiac failure, normal blood pressure, absence of cyanosis and murmurs, and without evidence of a shunt.

The anomalous coronary artery was included in this group in the past. In 1911, Abrikosoff2 first described the existence of this anomaly; and in 1933, Bland, White, and Garland3 made the first antemortem diagnosis of this condition. Up to now 45 cases have been reported.2-37

Case Report

An African male infant, aged 9 months, was admitted to the hospital because of coughing and restlessness for 2 weeks. He had been born normally after an uneventful pregnancy and appeared healthy at birth.

On physical examination he was in obvious distress, restless, and dyspneic. He was underweight and small.

The left side of the chest bulged prominently, and moved less than the right side, which was resonant and in which the breath sounds were normal. The area of cardiac dullness was increased and there was dullness at the left base. Breath sounds were diminished at the left base and fine crepitations were audible over the lower half of the chest posteriorly.

The heart was enlarged. The maximum cardiac impulse was in the fifth intercostal space in the anterior axillary line. No thrills or murmurs were observed. The heart sounds were clearly audible and apart from a tachycardia the rhythm was normal.

The abdomen was slightly distended, and the liver edge was palpable about 3 cm. below the costal margin.

X-ray of the chest revealed a grossly enlarged heart with "fairly" good pulsation. The enlargement appeared mainly left ventricular, and the left border of the heart gave the impression of an aneurysmal dilatation of the left ventricle. No abnormal calcifications were noted.

Examination of the blood revealed a hemoglobin of 9.7 Gm. per cent, 33,200 leucocytes per mm.3, with 92 per cent neutrophils.

Electrocardiogram showed regular sinus tachycardia of 200 per minute, deep Q waves in leads I, aVL and V5 and V6, and inverted T waves in leads I, II, and aVL (fig. 1).

The patient had several episodes of crying and restlessness, accompanied by dyspnea and sweating, particularly after feeding. Between attacks he was happy and interested, playful, and amenable to examination and handling. He died suddenly 14 days after admission.

Morbid Anatomy of the Heart. The heart weighed 125 Gm. or 3 times the normal weight of 39 Gm. at 9 months. The left ventricle occupied at least three quarters of the anterior surface of the heart and showed a globular enlargement (fig. 2, table 1).

The pericardium was smooth and glistening. The aorta and pulmonary artery were normal. The right coronary artery arose normally from an ostium 2 mm. in diameter from the anterior aortic sinus (fig. 3 Left) and was normally distributed to the right ventricle and posterior half of the left ventricle. No other ostial openings were seen in the root of the aorta.

The left coronary artery originated from the pulmonary artery from a small ostium 1 mm. in diameter, situated in an extremely low position in the sinus of the right anterior cusp (fig. 3 Right). From its origin in the pulmonary artery, the left coronary trunk ran inferiorly along the anterior surface of the right ventricle and down the anterior interventricular sulcus to the incisura apicis cordis, where it bifurcated, one branch running antero-

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inferiorly toward the apex, while the other swept posteriorly to supply the apical portion of the right ventricle. This main trunk thus followed the distribution of the anterior descending branch of the left coronary artery. A large branch followed the course of the left circumflex artery and arose 4 mm. from the root of the pulmonary branch. Several other smaller branches also supplied the anterior surfaces of the left and right ventricles.

The cavity of the left ventricle was greatly dilated with bulging of the anterior wall and displacement of the interventricular septum to the right. There was hypertrophy with thinning in the apical portion. The endocardium showed marked fibrosis, particularly in the anterior wall and interventricular septum. The myocardium was pale with atheromatous degeneration of the apices of the posterior papillary muscles. The endocardium of the left atrium was also grossly thickened and opaque.

The cavity of the right ventricle was diminished by the displacement of the interventricular septum, but the endocardium and the myocardium were normal. All valves were normal, and there was no other congenital abnormality.

Injection of Barium Sulfate into the Coronary Arteries. Although the heart had already been incised moderately adequate injection of the aberrant left coronary artery was achieved. The right coronary artery had been cut just beyond its origin and only the proximal segment could be filled properly. Radiographic photographs of the injected heart showed a very rich vascular network arising from the left coronary artery, supplying the anterior wall of the left ventricle and the apex of the heart. The trunk of the right coronary artery could be seen and anastomoses between its proximal branches and those of the anomalous left coronary artery were demonstrated.

In the case of the normal control, the heart was injected and photographed before it was cut. The vascular branches were smaller and formed a fine network, in which relatively few anastomoses could be discerned.

Histopathology. Sections of the anterior wall of the left ventricle showed marked para-arterial fibrosis of the myocardium (fig. 4). In addition there were large areas of diffuse fibrosis, identical in appearance to an organized myocardial infarct. Calcification was seen in some of these areas (fig. 5).
The myocardial fibers showed evidence of degeneration—some were vacuolated and others had progressed to recent coagulative necrosis. Irregular cleft-like spaces lined by endothelial cells were seen especially in the more viable parts of the myocardium (fig. 6). They conformed to the description of embryonic sinusoids.

The endocardium of the left ventricle showed an increase of fibrous tissue and a parallel increase of elastic tissue. The larger branches of the abnormal left coronary artery were well formed (fig. 7). The walls of the smaller branches were thinner and had less muscle and elastic tissue than the branches of the right coronary artery. Some of the small arterioles, particularly those in the more fibrosed areas, showed endothelial hyperplasia and a few were almost completely occluded by endarteritis.

The right ventricle and the right coronary artery were normal.

**DISCUSSION**

**Pathogenesis.** The anlagen of the coronary arteries are seen first as outpouching endothelial buds in the wall of the aortic bulb, before the spiral septum of the truncus arteriosus develops and separates the aorta and the pulmonary artery.

Abrikossoff,2 reported the first case of this abnormality and suggested that the anomaly could arise if the primitive bud of the left coronary artery were displaced anteriorly or if there were posterior displacement of the septum. Displacement of the septum would disturb the relative position and size of the aorta and the pulmonary artery. No such evidence was seen in any of the infant cases of the anomalous left coronary artery. The only instance of malposition of the septum together with an aberrant left coronary artery was reported by Konstantinowitsch38 in a 2-day-old girl who had a grossly malformed cor biloculare with a rudimentary stenosed aorta. One must postulate therefore that this lesion is due to displacement of the primitive bud of the left coronary artery.

In the reported cases2–37 the aberrant coronary artery was the sole congenital abnormality except for a patent ductus arteriosus in 2 cases25, 31 patent foramen ovale in 1 case36 and narrowing of the aorta in 2 cases18, 37 The course, distribution, and branching followed that of the normal left coronary artery, but the extent of the vascular network varied from less than half of the heart7 to the entire left ventricle plus a portion of the right ventricle.26

The right coronary artery in its origin, course, and distribution was normal throughout.

**Pathology.** The morbid anatomy of the heart in our case was quite typical with its
characteristic left ventricular hypertrophy. This reached aneurysmal proportions in 12 of the 45 cases.2, 5, 6, 8, 13, 15, 17, 18, 20, 23, 29, 30 Vivas-Salas39 considered this abnormality to be a most important cause of cardiac aneurysm in early life. Marked fibrosis particularly in the anterior wall and the interventricular septum may lead to extreme thinning and rupture.28, 31 Thrombi were present in this dilated, fibrose cavity in 4 cases.5, 7, 12, 33 The papillary muscles, being farthest from the blood supply, may show evidence of severe ischemia: yellow-gray patches of degeneration,25 as were present in our case; atrophy,36 gray-white scars,35 fibrosis,20 replacement by a fibrous band.23

The valves were usually normal but the mitral valve was thickened in 1 case,16 thickened and stretched in another,30 and showed verrucous projections due to an increase in the myxomatous ground substance in a third.36

Histologically, well marked para-arterial fibrosis, organized and recent myocardial necrosis, foci of calcification (54 per cent), persistence of embryonic sinusoids (33 per cent), and sclerotic and endarteritic vascular changes in the smaller branches of the left coronary (33 per cent) were frequently observed. A marked increase of subendocardial fibrous and elastic tissue may be present. As a result of the anomalous origin of the left coronary artery from the pulmonary artery, the left ventricle receives blood of low oxygen saturation and at the low pressure of the pulmonary circuit. It suffers from anoxemia which causes parenchymatous degeneration of the muscle fibers and condensation of the fibrous tissue, particularly in the para-arterial zone. This is followed by necrosis and fibrosis. Heitzmann5 first observed the similarity between this lesion and that of coronary occlusive disease in the adult. Fisher and Lloyd29 noted fibrous tissue surrounding individual muscle fibers and stated that this appearance was quite distinct from that of adult ischemic fibrosis, in which one sees islands of surviving muscle in a sea of fibrous tissue. But most workers agree that these lesions are essentially similar, being only more widespread and of greater magnitude in infants. Myocardial calcification, which is rare in adult occlusive coronary lesions, was seen.

**TABLE 1.—Cardiac Measurements after Fixation (cm.)**

<table>
<thead>
<tr>
<th></th>
<th>Heart of case A.M.—9 months</th>
<th>Heart of normal control—10 months</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weight in Gm.</td>
<td>125</td>
<td>40</td>
</tr>
<tr>
<td>Left ventricle:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Transverse diameter</td>
<td>7.2</td>
<td>5</td>
</tr>
<tr>
<td>Longitudinal diameter</td>
<td>5</td>
<td>3.2</td>
</tr>
<tr>
<td>Length of posterior</td>
<td></td>
<td></td>
</tr>
<tr>
<td>surface</td>
<td>5.7</td>
<td>4</td>
</tr>
<tr>
<td>Thickness of myocardium</td>
<td></td>
<td></td>
</tr>
<tr>
<td>base</td>
<td>0.8</td>
<td>0.6</td>
</tr>
<tr>
<td>Mid ventricle</td>
<td>0.8</td>
<td>0.6</td>
</tr>
<tr>
<td>Apex</td>
<td>0.4</td>
<td>0.4</td>
</tr>
<tr>
<td>Left Atrium:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Length of posterior</td>
<td>2</td>
<td>1.2</td>
</tr>
<tr>
<td>surface</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Thickness of myocardium</td>
<td></td>
<td></td>
</tr>
<tr>
<td>base</td>
<td>0.35</td>
<td>0.2</td>
</tr>
<tr>
<td>Right ventricle:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Length of posterior</td>
<td>5.5</td>
<td>4.1</td>
</tr>
<tr>
<td>surface</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Thickness of myocardium</td>
<td></td>
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<tr>
<td>base</td>
<td>0.3</td>
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<tr>
<td>Mid ventricle</td>
<td>0.2</td>
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<tr>
<td>Apex</td>
<td>0.15</td>
<td>0.1</td>
</tr>
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**Fig. 4.** A section of the wall of the left ventricle showing well-marked para-arterial fibrosis.
in 54 per cent of cases of the aberrant left coronary artery and was believed by Soloff\textsuperscript{10} to be pathognomonic in the radiologic diagnosis of this abnormality. Abrikosoff\textsuperscript{2} suggested that it followed the necrosis of the muscle fibers, and its prevalence in infants has been ascribed to the relatively higher content in infant myocardium of inorganic calcium, phosphorus, and phosphatase.

Barnard\textsuperscript{17} quoted Barcroft's\textsuperscript{41} work on the fetal circulation and suggested that this abnormality caused anoxemia of the left ventricle during fetal life. The persistence of embryonic structures such as the sinusoids supported this contention.\textsuperscript{20} Taussig\textsuperscript{42} did not accept these conclusions and maintained that there is little or no difference in the oxygen content of the blood in the 2 sides of the heart in utero. Kaunitz\textsuperscript{23} concluded that coronary ischemia does not manifest itself before birth, since advanced degenerative cardiac changes are not present in infants dying in the immediate neonatal period in whom both coronaries originate from the pulmonary artery.\textsuperscript{37, 44, 45} Bassis and Sheinkopf\textsuperscript{36} thought that the high intraventricular pressure in fetal life would assure the myocardium of an adequate supply of oxygenated blood.

\textbf{Clinical Features.} In cases with an aberrant right coronary artery originating from the pulmonary artery there is no evidence of functional disturbance or early death. Four such cases\textsuperscript{46-49} all lived until adulthood; one died at age 74. This longevity indicates that venous blood would appear to be adequate for the nutrition of the right ventricle, but not for that of the left side.\textsuperscript{15} It may still be sufficient for the demands in early infancy, but will fail with the increase of growth and activity of the patient.

The degree of disability and the length of the life span of the individual depend firstly upon the extent of the vascular network from the anomalous vessel—if this is extensive as in Craig's\textsuperscript{26} case, the subject dies in early infancy (2 months); if less than half of the heart is supplied by the left coronary artery,\textsuperscript{7} then there is relative longevity (10 months). This conclusion is also supported by some of the adult cases, e.g., a man aged 27, in whom the abnormal left coronary artery supplied less of the heart than usual.\textsuperscript{50} The second
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factor determining the length of survival, is the extent of the collateral circulation from the normal right coronary artery. This may in turn depend on the congenital inherent pattern of distribution of the coronary arteries, or may be determined by pressure gradients in these hearts. Anastomoses were demonstrable in most of the 8 adult cases reported, but their presence was variable in the infants of this series.

In the 45 previously reported cases there was a preponderance of female children; the sex was stated in 41 instances, 27 of which were female. The age at which the lesion became evident ranged from 7½ weeks to 13 months. In only 1 case was the infant born prematurely.

Maternal health had been normal in all except 3 instances—one had eclampsia, another syphilis, and the third had amenorrhea for 2 years before pregnancy.

The onset of symptoms was usually delayed, except in 1 case in which a persistent cough appeared at 1 week of age.

The usual presenting symptoms were cough and dyspnea; difficulty with feeding (refusing feeding, vomiting, discomfort and restlessness after feeding; difficulty in swallowing; failure to thrive; sweating); and anginal attacks.

Anginal Attacks. Bland, White, and Garland first drew attention to this condition and the similarity between the attacks of restlessness, sweating, and crying and attacks of angina as seen in adults. These attacks may be precipitated by the effort of feeding. The apparent feeling of well-being and amiability between attacks is in marked contrast.

Electrocardiographic Changes. Since the first description by Bland and co-workers most authors have confirmed the findings of inversion of T waves in all 3 standard leads and low voltage. In the present case T-wave

Fig. 6. Persistence of embryonic sinusoids—in such areas the myocardium remains viable.
inversion was noticed in leads I, II, and aV₆, and the voltage was high. Deep, wide Q waves were present in leads I, aV₆, V₅ and V₆.

Delayed Onset of Symptoms. Usually the symptoms were delayed until the third month of life. This delay may be due to the patent ductus arteriosus which allows the pulmonary artery to receive oxygenated blood from the aorta. When the ductus closes off, only venous blood reaches the left coronary and myocardial ischemia results.

In the present case, no history of previous attacks was obtained from the mother who appeared to be a reliable and intelligent informant. It follows, therefore, that if the attacks of dyspnea, restlessness, and sweating, seen in hospital, were the first experienced by the patient, then the onset of symptoms in this case was delayed until 9 months of age.

Diagnosis. The diagnosis of this condition should be considered in all cases of congestive cardiac failure in infancy in which cardiomegaly is not associated with cyanosis or murmurs, with no evidence of a shunt, and with normal blood pressure.

Among the other conditions which may show these features are glycogen storage dis-
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ease, congenital fibroelastosis, medial necrosis of the coronary arteries, and idiopathic myocarditis.

The radiologic features suggestive of the anomalous left coronary artery are gross cardiac enlargement affecting mainly the left ventricle and poor pulsation on fluoroscopy with a difference in the force of contraction of the 2 ventricles. The gross hypertrophy and size of the left ventricle may suggest aneurysmal dilatation.

Electrocardiograms suggest varying degrees of myocardial ischemia. Several patterns have been recorded, particularly inversion of T waves in all 3 standard leads together with low voltage. 

Course. While the anomalous left coronary artery has been reported in 8 adult cases, by far the majority of cases prove fatal in the first year of life. The average age of death in the 45 infant cases reviewed was 4 months.

Treatment. Medical treatment is essentially of a palliative nature and is designed to prevent attacks by the avoidance of those factors which may precipitate these attacks. Sedation is used to relieve the attack.

There has as yet not been any report of surgery to relieve the condition. Procedures have been suggested to increase the supply of oxygen to the left ventricle. Gasul and Loeffler suggested a Potts-Smith operation, in which the left or right pulmonary artery is anastomosed to the descending aorta. Keizer and Rochat thought that the left pectoral artery could be used, and Blalock suggested anastomosis with the internal mammary. Omentopexy might also be considered. McKinley et al. favored the Potts-Smith operation, but objected to Beck's pericardial anastomosis for fear of the development of restrictive pericarditis.

The necessary prerequisite to any proposed treatment is the confident diagnosis of the condition during life. Bland et al. 3 made the first antemortem diagnosis of this condition, and it has been diagnosed clinically 5 times since. 21, 22, 54, 23 (cases 1 and 4)

Summary
A case is reported of the anomalous origin of the left coronary artery from the pulmonary artery in a Bantu male infant aged 9 months. The clinical, radiologic, and electrocardiographic findings, and the cardiac pathology are presented.

The 45 cases previously reported in the literature are reviewed and the clinical syndrome and pathologic physiology of the aberrant left coronary artery are discussed.

Acknowledgment
We are indebted to Drs. M. Tonkin and H. Faleke for permission to publish the case; to Dr. J. Kaye and Miss Dreyer for radiographic assistance; to Miss E. E. A. Gygax for the translation of the German literature; to Mrs. T. E. Lawrence and Mrs. J. E. Bailey for secretarial assistance, and to Miss M. J. H. Barnard for the microscopic sections.

Summario in Interlingua
Es reportate un caso del origine anormal del arteria coronari sinistre in le arteria pulmonar. Le patiente eseva un infante masculino de 9 menses de etate de raccia bantu. Le constataiones clinica, radiologic, e electrocardiographic e le pathologia cardiac es presentate.

Es presentate un revista del 45 casos prevemente reportate in le litteratura. Le syndrome clinica e le physiologia pathologic del aberrante arteria coronari sinistre es discutite.

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I cannot say that I am in the slightest degree impressed by your bigness, or your material resources, as such. Size is not grandeur, and territory does not make a nation. The great issue, about which hangs a true sublimity, and the terror of overhanging fate, is what are you going to do with all these things?—Thomas H. Huxley. American Addresses with a Lecture on the Study of Biology. London, MacMillan and Co., 1877, p. 125.
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