Anomalous origin of the left coronary artery from the pulmonary artery with coronary artery steal in adults
Report of two cases and review of the literature

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Ihekwaba, F. N., Davidson, K. G., Ogilvie, B., and Caves, P. K. (1976). Thorax, 31, 337-345. Anomalous origin of the left coronary artery from the pulmonary artery with coronary artery steal in adults. Patients with anomalous left coronary artery arising from the pulmonary artery rarely survive to adult life. Those who attain adulthood may present with angina indistinguishable from coronary artery disease and are liable to sudden death. Myocardial infarction, though rare in young adults, may occur and may be due to coronary artery steal. Accurate diagnosis requires coronary arteriography.

Two further cases of coronary artery steal in adults with anomalous origin of the left coronary artery from the pulmonary artery are presented. In both patients aorto-coronary bypass grafting using a reversed autogenous saphenous vein with closure of the origin of the anomalous left coronary artery was successfully performed. This operation provided complete symptomatic relief and may protect patients against the risk of sudden death.

Origin of the left coronary artery from the pulmonary artery is a rare anomaly usually first identified at necropsy. The majority of patients are infants and children (Bland, White, and Garland, 1933; Keith, 1959; Agustsson et al., 1962; Wesselhoeft, Fawcett, and Johnson, 1968) in whom ischaemia of the left ventricle is the cause of death (Somerville and Ross, 1970). Approximately 15% of cases survive beyond infancy (Harthorne, Scannell, and Dinsmore, 1966) and, if intercoronary anastomotic channels develop, survival to middle age is possible (Kaunitz, 1947). In adults the mean life expectancy in reported cases has been 33 years. In almost 30% of these patients death occurred suddenly after severe exertion.

Cine-angiocardiography has recently permitted this condition to be recognized during life and, with surgical correction, a normal life span may be a reasonable expectation for these patients. The world literature reveals only 33 adult cases (18 years and over) of anomalous left coronary artery arising from the pulmonary artery (Table). In 18 of these, the diagnosis was made during life and 17 were treated surgically. Among this group five were treated by ligation of the origin of the anomalous left coronary artery and an aorto-coronary vein bypass graft. This paper reports another two such cases of successful surgical correction for this rare anomaly.

CASE REPORTS

CASE 1 Mrs. E. C. (born 18 April 1928) was first seen in October 1969 when she gave a 12-year history of attacks of palpitations associated with breathlessness. In October 1969 she had a severe attack of paroxysmal tachycardia and after this had to reduce her activities because of tightness across the lower chest associated with breathlessness. The chest discomfort was substernal and was relieved by glyceryl trinitrate. She had had two normal pregnancies, the first 23 years previously.

Her electrocardiogram revealed evidence of an established anterior infarct (Fig. 1). A chest radiograph showed no significant cardiac enlargement.
### TABLE

**CASES OF ANOMALOUS LEFT CORONARY ARTERY (LCA) IN ADULTS (OVER 18 YEARS OF AGE)**

<table>
<thead>
<tr>
<th>Case</th>
<th>Data Source and Date</th>
<th>Age</th>
<th>Sex</th>
<th>Clinical Course</th>
<th>LCA Anatomy</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Abbott 1908</td>
<td>60</td>
<td>F</td>
<td>Accidental death</td>
<td>LCA veinlike large, thin walled</td>
</tr>
<tr>
<td>2</td>
<td>Kockel 1934</td>
<td>38</td>
<td>M</td>
<td>10-year chest pain. Sudden death</td>
<td>Grossly dilated</td>
</tr>
<tr>
<td>3</td>
<td>Rüberdt 1937</td>
<td>27</td>
<td>M</td>
<td>Sudden death while operating</td>
<td>Thin, long</td>
</tr>
<tr>
<td>4</td>
<td>Dietrich 1939</td>
<td>53</td>
<td>M</td>
<td>Heart murmur at 17, progressive dyspnoea and chest pain. Congestive heart failure</td>
<td>Large, tortuous</td>
</tr>
<tr>
<td>5</td>
<td>Ponsold 1939</td>
<td>24</td>
<td>M</td>
<td>Soldier—died after long distance race</td>
<td>Hypoplastic circumflex branch</td>
</tr>
<tr>
<td>6</td>
<td>Ruddock and Stehly 1943</td>
<td>30</td>
<td>M</td>
<td>Labourer—sudden death</td>
<td>Enlarged collaterals. Large heart</td>
</tr>
<tr>
<td>7</td>
<td>Kaunitz 1947</td>
<td>32</td>
<td>M</td>
<td>Sudden death</td>
<td>Dilated and tortuous</td>
</tr>
<tr>
<td>8</td>
<td>Gouley 1950</td>
<td>34</td>
<td>M</td>
<td>Labourer—sudden death</td>
<td>Veinlike, large, tortuous</td>
</tr>
<tr>
<td>9</td>
<td>Wüthrich 1951</td>
<td>27</td>
<td>M</td>
<td>Sudden death</td>
<td>Grossly dilated, scarred myocardium</td>
</tr>
<tr>
<td>10</td>
<td>Rotter 1952</td>
<td>58</td>
<td>M</td>
<td>Engaged in argument—sudden death</td>
<td>Thin, veinlike</td>
</tr>
<tr>
<td>11</td>
<td>Jurishica 1957</td>
<td>18</td>
<td>M</td>
<td>Associated patent ductus. Operative death. Aortic damage</td>
<td>Thin, veinlike</td>
</tr>
<tr>
<td>12</td>
<td>George and Knowlan 1959</td>
<td>26</td>
<td>M</td>
<td>Congestive heart failure, rheumatic heart disease—sudden death</td>
<td>Thickened. Mitral insufficiency</td>
</tr>
<tr>
<td>21</td>
<td>Summer and Hendrix 1968</td>
<td>22</td>
<td>M</td>
<td>Continuous murmur noted at 18.</td>
<td></td>
</tr>
<tr>
<td>22</td>
<td>Sabiston et al. 1968</td>
<td>29</td>
<td>F</td>
<td>LCA. Ligated. Alive</td>
<td>Dilated</td>
</tr>
<tr>
<td>23</td>
<td>Sabiston et al. 1968</td>
<td>31</td>
<td>F</td>
<td>Exertional chest pain. Systolic dyspnoea. LCA. Ligated. Alive</td>
<td>Large, thin walled</td>
</tr>
<tr>
<td>24</td>
<td>Yamane et al. 1969</td>
<td>41</td>
<td>M</td>
<td>Exertional dyspnoea, chest pain. Continuous murmur noted at age 30</td>
<td>Large, thin walled</td>
</tr>
<tr>
<td>25</td>
<td>Dalton et al. 1969</td>
<td>39</td>
<td>F</td>
<td>LCA. Ligated. Alive</td>
<td>Large, thin walled</td>
</tr>
<tr>
<td>26</td>
<td>Reis et al. 1969</td>
<td>20</td>
<td>M</td>
<td>Heart murmur at 15. Palpitations, syncope while playing football. ACBG + ligation of LCA. Alive</td>
<td>Thin walled</td>
</tr>
<tr>
<td>27</td>
<td>Bergnes 1970</td>
<td>23</td>
<td>M</td>
<td>‘Asthmatic’ from age 4. Athlete. Sudden death while jogging</td>
<td>Thin walled</td>
</tr>
<tr>
<td>29</td>
<td>Ogden 1970</td>
<td>20</td>
<td>F</td>
<td>Exertional angina. LCA. Ligated</td>
<td>Thin walled</td>
</tr>
<tr>
<td>33</td>
<td>Thomas et al. 1973</td>
<td>40</td>
<td>F</td>
<td>Heart murmur at age 5 years. ‘Bursting’ substernal pain on exertion. Mitral insufficiency. ACBG with LCA occlusion. Alive</td>
<td>Thin walled</td>
</tr>
<tr>
<td>35</td>
<td>Present case 1974</td>
<td>20</td>
<td>F</td>
<td>Cyanotic attacks in infancy. ACBG + ostial-occlusion. Alive</td>
<td>Thin walled</td>
</tr>
</tbody>
</table>

ACBG = aortocoronary vein bypass graft
Origin of left coronary artery from pulmonary artery with coronary artery steal in adults

A diagnosis of coronary artery disease was made and she was followed up until October 1973 when she stated that she had become increasingly breathless with more frequent episodes of tachycardia. Ordinary housework produced upper chest tightness partially relieved by glyceryl trinitrate.

On physical examination the pulse now felt jerky. The first sound was accentuated, and a mid-systolic murmur grade 2–3/6 and a third heart sound were present in the mitral area. The pulmonary second sound was not accentuated and there was no evidence of cardiac failure. The ECG and plain chest radiograph were unchanged. An echocardiogram showed no abnormality. Serum cholesterol and lipids were normal.

Cardiac catheterization and selective coronary angiography were performed on 25 October 1973. Right heart catheterization provoked paroxysms of atrial tachycardia, and samples were not obtained from the right side of the heart.

Repeated attempts to catheterize the left coronary artery were unsuccessful. Selective right coronary angiography together with an aortogram were therefore performed. The origin of the right coronary artery was high, and both the main artery and its branches were greatly dilated and tortuous. The left coronary artery did not arise from the aorta but was filled retrogradely by the large collateral branches from the right artery. The left coronary artery system was also dilated and tortuous, and originated from the main pulmonary artery. Contrast material flowed via the right and left arteries to the main pulmonary artery, clearly outlining both it and the left and right pulmonary arteries. The left ventricular end-diastolic pressure was 20 mmHg, and a left ventricular cine-angiogram showed impaired contractility of the anterior wall with a localized dyskinetic area (Fig. 2). There was no significant mitral incompetence.

Operation was undertaken in November 1973 through a vertical median sternotomy incision. The heart contracted well, except for a scarred area on the anterior wall of the left ventricle.

FIG. 1. Case 1. Electrocardiogram showing an established anterior myocardial infarct.

FIG. 2. Case 1. Systolic frame of left ventricular cine-angiogram showing dyskinetic area on anterior wall.
right coronary artery was very dilated and tortuous with numerous dilated and tortuous anastomotic branches running over the anterior aspect of the right ventricle. The proximal part of the left coronary artery was hidden behind the pulmonary artery but the left anterior descending coronary artery and its branches were also remarkably dilated and tortuous. There was no evidence of atheroma. On cardiopulmonary bypass the pulmonary artery was opened, and a large retrograde flow of blood from the orifice of the left coronary artery into the pulmonary artery was demonstrated. The orifice was in the posterolateral sinus and measured 7 mm in diameter. It was oversewn from inside the pulmonary artery, and an aorto-c coronary bypass graft was then established to the proximal part of the left anterior descending coronary artery using a suitable length of reversed autogenous long saphenous vein.

The patient made a smooth postoperative recovery and was discharged from hospital on the eighth postoperative day. Oral anticoagulation was given for one month. She remained asymptomatic and the third heart sound and systolic murmur were no longer audible. Eight months postoperatively repeat cardiac catheterization revealed normal intracardiac pressures. Graft angiograms revealed vein patency and showed good filling of the left coronary system in a normal antegrade fashion. The previously grossly dilated and tortuous right coronary artery had shrunk to normal size and the large collateral branches were no longer present.

**CASE 2** Miss J. McC. was born five weeks prematurely in July 1954, a twin weighing 2.2 kg. She was cyanosed at birth and remained in hospital for 11 weeks, during which time she had several cyanotic attacks. She was thought to have congenital heart disease, the exact nature of which was not clear. At the age of 7 months she was readmitted to hospital with a chest infection and was found to be of normal appearance and was not cyanosed. Her pulse rate was 130–150 per minute and there was a soft systolic murmur at the left sternal edge.

Her ECG showed Q waves, ST depression, and inverted T waves in leads I and aVL. Chest radiography revealed a minimally enlarged heart (cardiothoracic ratio 130:240) with normal pulmonary vascularity.

A provisional diagnosis of anomalous origin of the left coronary artery from the pulmonary artery was made.

During the first year of life she required anti-biotics for recurrent chest infections and was given digitalis for a short period. No other investigations were performed at that time and no specific treatment was offered. She remained well over the next few years, her activities were entirely unrestricted, and her development progressed normally. At regular review there was no evidence of cardiac failure, but an apical systolic murmur and a blowing early diastolic murmur persisted.

Cardiac catheterization and coronary arteriography were performed in March 1967 and the latter demonstrated a huge tortuous right coronary artery (Fig. 3a).

The left anterior descending and circumflex arteries filled from the right and the flow in them was retrograde, with contrast opacifying the pulmonary artery (Fig. 3b). There was a step-up in oxygen saturation from 67% in the right ventricular outflow to 81% in the main pulmonary artery.

Over the next seven years she remained well, with no clinical, radiological or electrocardiographic evidence of deterioration (Fig. 4), and in 1974, then aged 20, she was referred for surgical treatment.

Operation was undertaken in September 1974 through a vertical median sternotomy incision. The cardiac chambers were of normal size, but there was a small scar near the apex of the left ventricle indicative of previous myocardial infarction. The right coronary artery and its branches were enormously dilated and tortuous. The left coronary system had dilated thin-walled vessels but no tortuosity.

On cardiopulmonary bypass the main pulmonary artery was opened and the anomalous origin of the left coronary artery was confirmed. There was a considerable retrograde flow from the ostium, which was 4 mm in size and lay in the posterior sinus. The orifice was sutured, the pulmonary artery repaired, and a reversed saphenous vein bypass graft inserted between the ascending aorta and the proximal part of the left anterior descending artery. There were no operative or postoperative complications, and the patient was discharged home on the tenth postoperative day on no medication. Six months later she remained well with no cardiac murmurs.

Two weeks postoperatively coronary angiography showed that the right coronary artery was smaller than preoperatively and that no contrast passed from the right to left coronary systems. The vein graft was widely patent and filled the left coronary system completely.
Origin of left coronary artery from pulmonary artery with coronary artery steal in adults

Brooks in 1886 reported his observations on two cadavers in whom an anomalous left coronary artery originated from the pulmonary artery. He proposed that the anomalous vessel acted as a vein allowing blood to flow retrogradely into the pulmonary artery. In 1908 Abbott published the finding of an anomalous left coronary artery in a 60-year-old woman who had died accidentally, and argued that the left coronary blood flow was into the pulmonary artery. A further case was described in 1911 by Abrikosoff, but it was left to Bland et al. (1933), whose names are sometimes applied eponymously to the condition, to give clinical and pathological descriptions of the anomaly.

The clinical presentation in infants is well defined, since the majority present with episodic dyspnoea, irritability, crying and pallor, congestive heart failure with gross cardiomegaly, and anterolateral myocardial infarction. Clinical signs of mitral insufficiency may be present (George and Knowlan, 1959; Usman et al., 1961; Burchell and Brown, 1963). Without operation death occurs in 93% of such children during the first year of life (Nadas, Gamboa, and Hugenholtz, 1964; Wesselhoeft et al., 1968). In adults the clinical presentation is quite different (Gouley, 1950; Agustsson et al., 1962). Symptoms may be absent or minimal. Although exertional dyspnoea and angina pectoris may occur, sudden death in an apparently normal individual is the commonest form of presentation (George and Knowlan, 1959; Burchell and Brown, 1962; Liebman et al., 1963). A few cases have been recognized following investigation of a cardiac murmur (Baylis and Campbell, 1952). Palpitations, which were the earliest cardiac symptom in case 1,
are uncommon. The electrocardiogram may be normal. Usually, however, left axis deviation, deep, wide Q waves in leads I and aVL, S-T segment or T-wave changes suggestive of ischaemia, and voltage criteria for left ventricular hypertrophy are seen. Myocardial infarction is thought to be rare in adults (Nadas et al., 1964; van der Hauwaert et al., 1965) but was present in both of our cases. The chest radiograph is usually normal but may show left ventricular and left atrial enlargement. Mitral regurgitation, possibly due to papillary muscle dysfunction, may be demonstrated on left ventriculography but was not seen in these cases.

Cardiac catheterization with selective coronary angiography, first used by Lampe and Verheugt (1960), is the definitive technique for diagnosis in adults. Oxygen saturation studies may be normal or show a slight increase within the pulmonary artery. Pressure readings in the right and left sides of the heart are usually normal. Selective right coronary angiography demonstrates a dilated tortuous vessel with delayed opacification of the left coronary artery through intercoronary collateral communications and passage of the contrast medium into the pulmonary artery via the anomalous origin of the left coronary artery. Edwards (1961) and Nadas et al. (1964) and their co-workers described three haemodynamic phases in the evolution of this anomaly. In the first phase, during the fetal and newborn period, blood flow is from the pulmonary artery into the anomalous left coronary vessels. This forward flow persists in the infant (Stern et al., 1961; Jameson, Ellis, and Levine, 1963) while the pulmonary artery pressure is high. The newborn is asymptomatic and may remain so for five to eight weeks (Wesselhoeft et al., 1968). The second (transition) phase occurs when the pulmonary artery pressure falls and the flow in the left coronary artery becomes retrograde. This phase is critical and during it survival depends upon the establishment of adequate intercoronary collateral vessels to prevent myocardial ischaemia, infarction or death. The third phase, the ‘adult’ type, is characterized by the establishment of large collateral vessels permitting adequate perfusion of the left ventricle from the right coronary artery. A fourth phase, the ‘coronary steal’ phase first proposed by Baue and associates (1967), produces such a large arteriovenous shunt that left ventricular myocardial perfusion is embarrassed. For descriptive purposes this classification has merit since it correlates well with pathology found both at operation and at necropsy. Both our cases clearly have the characteristics of the coronary steal syndrome, for a large retrograde flow was seen at cardiac catheterization and was confirmed at operation. And more patients with symptoms of myocardial ischaemia are investigated by coronary arteriography it seems likely that the syndrome of coronary artery steal will be recognized more frequently.

Of the 16 adult patients reported with this anomaly who did not have surgical treatment, only one was submitted to cardiac catheterization (Likar, Criley, and Lewis, 1966). The fate of this woman, 29 years old in 1966, is not known. The other 15 patients were diagnosed at necropsy. In the majority, death occurred suddenly, usually after exertion. Their average age was 33±0 years, which demonstrates the poor prognosis associated with this anomaly in the few patients who achieve adult life. Over the age of 45 years, only three patients with this anomaly, including one of the patients here reported, have been diagnosed during life, the oldest being 49 years.

Because of the poor prognosis in adults, various surgical procedures have been suggested to improve the coronary circulation (Cooley, Hallman, and Bloodwell, 1966). Ligation of the anomalous vessel at its origin (Case et al., 1958; Edwards, 1961) is the procedure which has been performed most frequently. Wide acceptance of this procedure followed observations on instantaneous coronary blood flow made at the time of operation which showed that the pressure in the anomalous left coronary vessel rose to systemic levels after occlusion of the vessel at its origin (Rowe and Young, 1960; Sabiston, Neill, and Taussig, 1960; Baue et al., 1967; Reis, Cohen, and Mason, 1969). Likar et al. (1966) found 16 survivors among 27 infants and adolescents who underwent the operation. Those not surviving were all infants, but several infants demonstrably profited from the procedure. Ten adults have been submitted to this operation (Sabiston, Floyd, and McIntosh, 1968; Summer and Hendrix, 1968; Dalton, Arrington, and King, 1969) without mortality and with symptomatic relief in most cases. Rowe and Young (1960) suggested that if, after occlusion of its origin, the pressure in the anomalous vessel rises to systemic levels, nothing is gained by coronary artery bypass grafting. Although Massih, Lawler, and Vermillion (1963) and later Summer and Hendrix (1968) reported postoperative ECG changes of ischaemia after ligation, it appears that ligation of the anomalous vessel is safe in adults in whom an adequate collateral circulation has developed.
A more desirable procedure and one which would appear to enhance the long-term prognosis would be the creation of a two coronary system. This restores almost normal coronary anatomy and dynamics; for although systemic pressures in the anomalous vessel may be obtained after ligation when cardiac work is low, the intercoronary collateral circulation may not be adequate during exercise. Detachment of the left coronary vessel from its anomalous origin and its direct anastomosis to the aorta is technically difficult and hazardous, especially if the vessel lies partially buried within the myocardium. Cooley et al. (1966) first reported the establishment of a ‘two coronary heart’ in two children in whom they fashioned aortocoronary bypass grafts using in the one case a Dacron prosthesis and in the other a reversed length of autogenous saphenous vein. Thirteen other published cases have had a two coronary system created (Lam et al., 1968; Reis et al., 1969; Somerville and Ross, 1970; Gasior et al., 1971; El-Said et al., 1972; Thomas et al., 1973), of whom five were adults.

The two patients described in this paper are the sixth and seventh reported adults who have undergone successful closure of the origin of an anomalous left coronary with aortocoronary vein bypass grafting. Cine-angiography, eight months and two weeks after surgery, showed unaltered left ventricular contractility and good filling of the left coronary system from patent grafts. The grossly dilated right and left coronary arteries of the first patient had returned to normal size. This patient has achieved complete relief of her palpitations, angina, and breathlessness, and her systolic murmur and third heart sound have disappeared. The second patient remains asymptomatic and has no murmurs.

Since all patients born with this arterial anomaly who survive to adult life are at risk of myocardial infarction and sudden death, we now believe that such patients should be treated by the establishment of a two coronary system before increasing ‘coronary steal’ produces myocardial ischaemia, infarction or death.

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