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Clinico-Pathological Correlations

Congenital atresia of the orifice of the left coronary artery

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Abstract A 3-month-old infant developed signs of cardiac failure, which was initially attributed to cardiomyopathy. At 8 months, further investigations showed evidence of myocardial ischaemia with reversal of the flow of blood in the left coronary artery, which received no demonstrable inflow from the aorta. An anomalous connection of this artery with the pulmonary trunk was diagnosed but, at surgery, it was found that the arterial orifice was completely atretic, although the main stem was of normal size. A left internal thoracic arterial graft to the anterior descending coronary artery was performed, but he died on the third day after the operation. Postmortem examination showed a small dimple within the aorta at the site of the orificial atresia, extensive myocardial infarction, and two zones of myocardial bridging of the anterior descending coronary artery. We discuss the relationship of coronary orificial atresia with single coronary artery. Although they are related, they typically have different and contrasting clinical presentations. The possible role of the myocardial bridging is also considered.

Keywords: Coronary arteries; congenital anomalies; coronary ostial atresia; single coronary artery; myocardial infarction; myocardial bridging

IT HAS BEEN ESTIMATED THAT CONGENITAL anomalies of the coronary arteries occur in from 0.2 to 1.2 per cent of the population, but most of these are minor variations in the position and numbers of the aortic orifices, possibly demonstrated during angiography, and are of no clinical significance.[1,2] Isolated coronary arterial atresia, and isolated atresia of the orifice of the left coronary artery, are amongst the rarest of reported cardiac anomalies.[3] The latter is of particular importance, as it usually generates critical myocardial ischaemia. In this respect, it is almost identical clinically with anomalous origin of the left coronary artery from the pulmonary trunk, with which it may be confused.

Clinical history

This infant was well until the age of 3 months, but then suffered frequent episodes of respiratory infection. At 8 months he had a suspected viral infection, causing diarrhoea and vomiting, which led to dehydration for which he was hospitalized for 4 days. A chest X-ray showed cardiomegaly, while echocardiography revealed a dilated and hypocontractile left ventricle, with enlargement of the left atrium. A provisional diagnosis of an underlying cardiomyopathy was supported by electrocardiography, which showed biventricular hypertrophy with inverted T waves (Fig. 1). After referral, further echocardiography demonstrated a reversal of flow in the left coronary artery. This was considered to be indicative of an anomalous origin of the left coronary artery from the pulmonary trunk, although this was not demonstrated. At cardiac catheterisation, the left ventricular function was found to be extremely depressed. The right coronary artery was dilated, with retrograde filling of the bed of the left coronary artery (Fig. 2). Although no dye was seen to enter the pulmonary arteries, an anomalous connection of the left coronary artery was still considered to be the underlying condition. At the age of eight and a half months, therefore, he was submitted for surgical correction. At operation, the pulmonary trunk was opened, but there was no evidence of an anomalous

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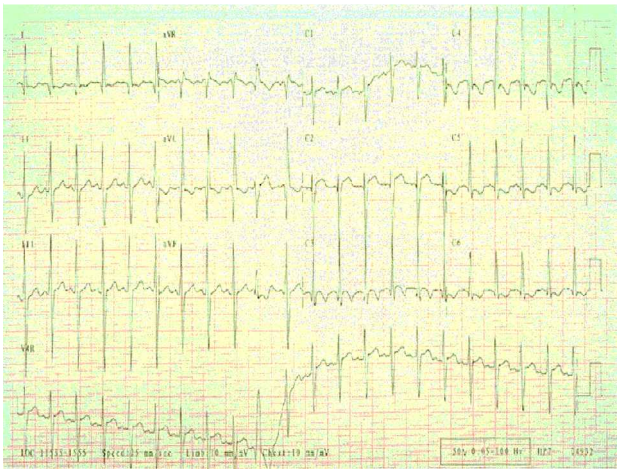

Figure 1.
The electrocardiogram prior to surgery showing left ventricular hypertrophy, changes in the lateral T waves, and deep lateral Q waves.

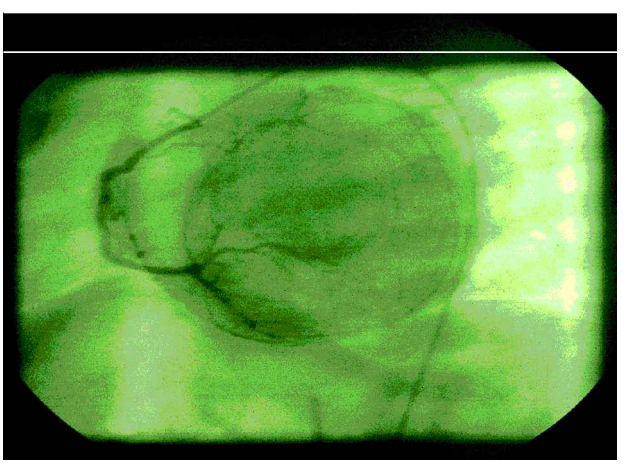

Figure 2.
Selective angiogram of the right coronary artery, showing opacification of the right coronary artery and its branches with retrograde filling of the left system.

coronary arterial connection to the main trunk, the bifurcation, or the right and left pulmonary arteries. Nor was there any evidence of return of cardioplegic solution to the pulmonary circulation after infusion into the root of the aorta. The diagnosis was therefore revised to that of atresia of the orifice of the left coronary artery, and the left internal thoracic artery was anastomosed to the left anterior descending coronary artery. Discontinuation of cardiopulmonary bypass was delayed for about an hour because of a rise in left atrial pressure, after which normal circulation was established with inotropic support. He remained very sick, with poor urinary output, but the arterial pressure gradually improved. On the next day, he developed erythematous bullas in both axillas, which were thought to be due to infective hidradenitis and were drained. Two days later, he developed a narrow-complex tachycardia, which did not respond to adenosine or direct current defibrillation. Intravenous flecainide was commenced, but shortly after he had a sudden increase in heart rate, followed by recurrent episodes of ventricular fibrillation. His condition remained unstable, and he died 3 hours after the original collapse, on the third day after the operation.

Postmortem findings

There were no significant abnormalities apart from the cardiovascular system and ulcerated areas in both axillas. The heart was dilated, and the external appearance was consistent with a dilated cardiomyopathy. The atriums were of usual morphology, with normal systemic and pulmonary venous connections. The atrioventricular and ventriculo-arterial connections were concordant, and the cardiac valves were normal. There were two main coronary arteries, each of good size and apparently normal distribution. The right coronary artery had a wide orifice from the right coronary sinus of the aorta. It divided immediately into a small infundibular branch, and a large main vessel which ran normally to the right in the atrioventricular groove, giving off the usual branches to the right ventricle and the posterior walls of both ventricles. The main stem of the left coronary artery was of normal size, approximately two millimetres wide, and appeared to originate in a normal manner from the aortic root. Examination of the interior of the aorta, however, showed no connecting orifice, although there was a small dimple just above the posterior commissure of the left coronary sinus in a position corresponding to the site of the external connection of the artery (Fig. 3). The main stem was 9 mm long, and, apart from complete atresia of the origin at the level of the external aspect of the aorta, the lumen was fully patent, being free from thrombus, with the wall otherwise being normal. The artery divided normally into circumflex, obtuse marginal, and anterior interventricular branches. The circumflex branch extended to the posterior aspect of the left ventricle, but there was no visible major anastomosis with the more dominant right coronary artery. The anterior interventricular artery divided into two branches of equal calibre which extended to the apex. The right branch passed down the interventricular groove. In its central part, it had a patent end-to-side surgical anastomosis with the left internal thoracic artery. On either side of the graft, the artery had segments of marked myocardial bridging, with a proximal zone 5 mm wide, and an apical zone of 15 mm width. Small

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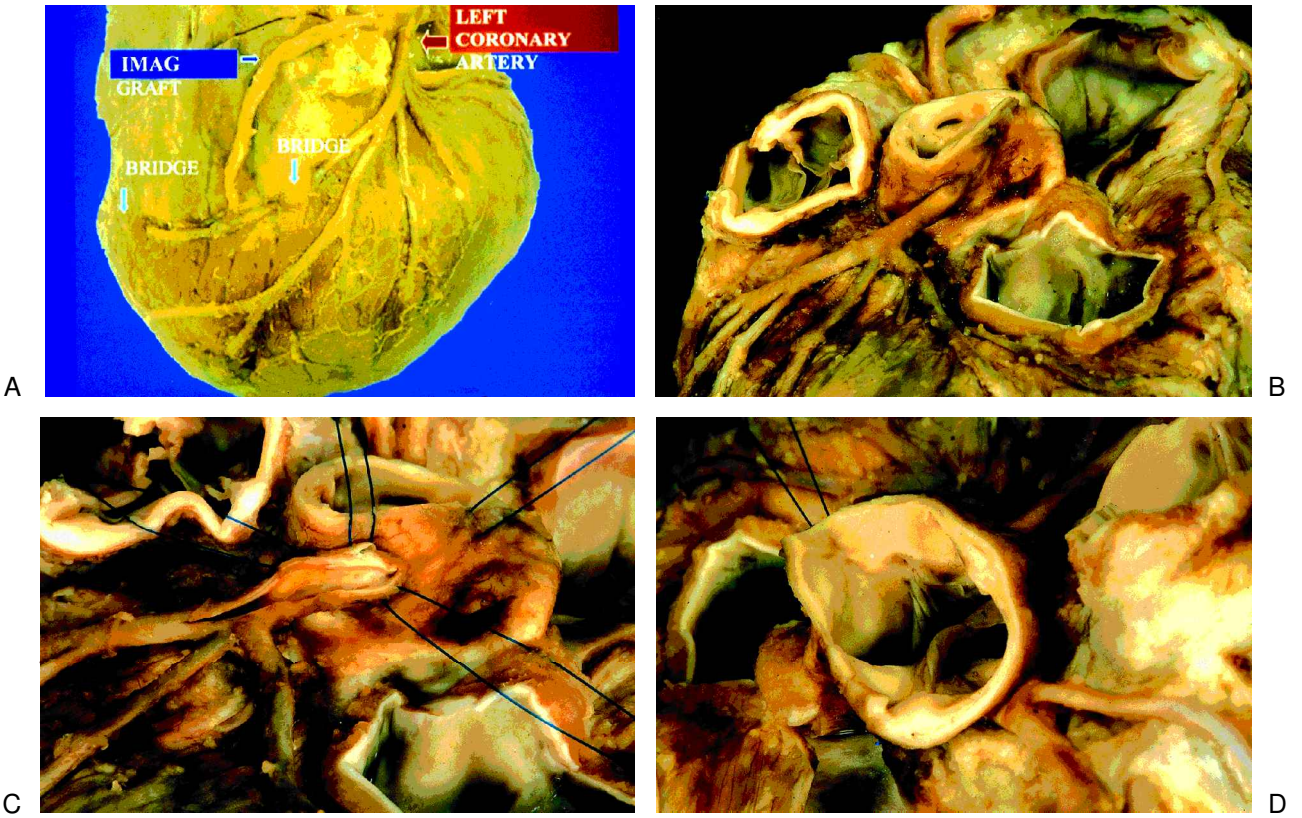

Figure 3.
Gross findings. The left coronary artery of normal size (A) branches in normal fashion. Two large myocardial bridges are seen on the anterior descending branch. Note the internal mammary by-pass graft (IMAG). The artery (B) takes an apparently normal origin. The orifice of the right coronary artery is visible within the aorta. Opening the artery (C) shows the normal lumen extending from the atretic orifice. A view from the interior of the aorta (D), seen from above shows the dimple just above the commissure at the site of the atretic orifice of the left coronary artery.

anastomosic channels were present in the apical region, between the distal portion of the posterior interventricular branches of the right coronary artery and the anterior interventricular branches of the left coronary artery. The territory of the left coronary artery was therefore perfused in a retrograde or centripedal manner from the right artery. The walls of the coronary arteries were normal, and there was no evidence of supravalvar aortic stenosis or of any acquired aortic disease.

A transverse macroscopic section through the ventricles showed extensive recent infarction throughout the wall of the left ventricle, with extensive scarring and fibroelastosis throughout the inner zone. This was confirmed on histological examination. Sections of the origin of the main stem of the left coronary artery showed a histologically normal wall with slight endocardial thickening, but the artery ended blindly within the wall of the aorta. There was no evidence of inflammation or fibrosis (Fig. 4).

Death was deemed as due to myocardial failure, due to myocardial infarction associated with isolated atresia of the orifice of the left coronary artery.

Discussion

The first issue to be addressed is whether the orificial atresia was acquired or congenital. There were two points in favour of an acquired aetiology. First, the child showed no apparent evidence of myocardial ischaemia until several months after birth. Second, the size and patency, with absence of thrombosis, in the long segment of the left main stem, proximal to the origin of the circumflex branch, suggested that, at sometime, there had been flow of blood through it. A delayed onset of ischaemia in infancy, nonetheless, is not uncommon with congenital atresias.[4–8] It has been suggested that this may be due to aggravation of an impending ischaemia by growth of the infant and increasing physical activity.[9] Ischaemia may even be delayed until late adult life, when atherosclerosis becomes a complicating factor.[10–12] Koh and colleagues[13] found that only one-twentieth of symptomatic cases presented during childhood, with half seen during infancy and adolescence and one-third in adult life. Acquired stenosis of a coronary artery, or its orifice, is generally associated with more widespread arterial disease.[14–16] Congenital cases

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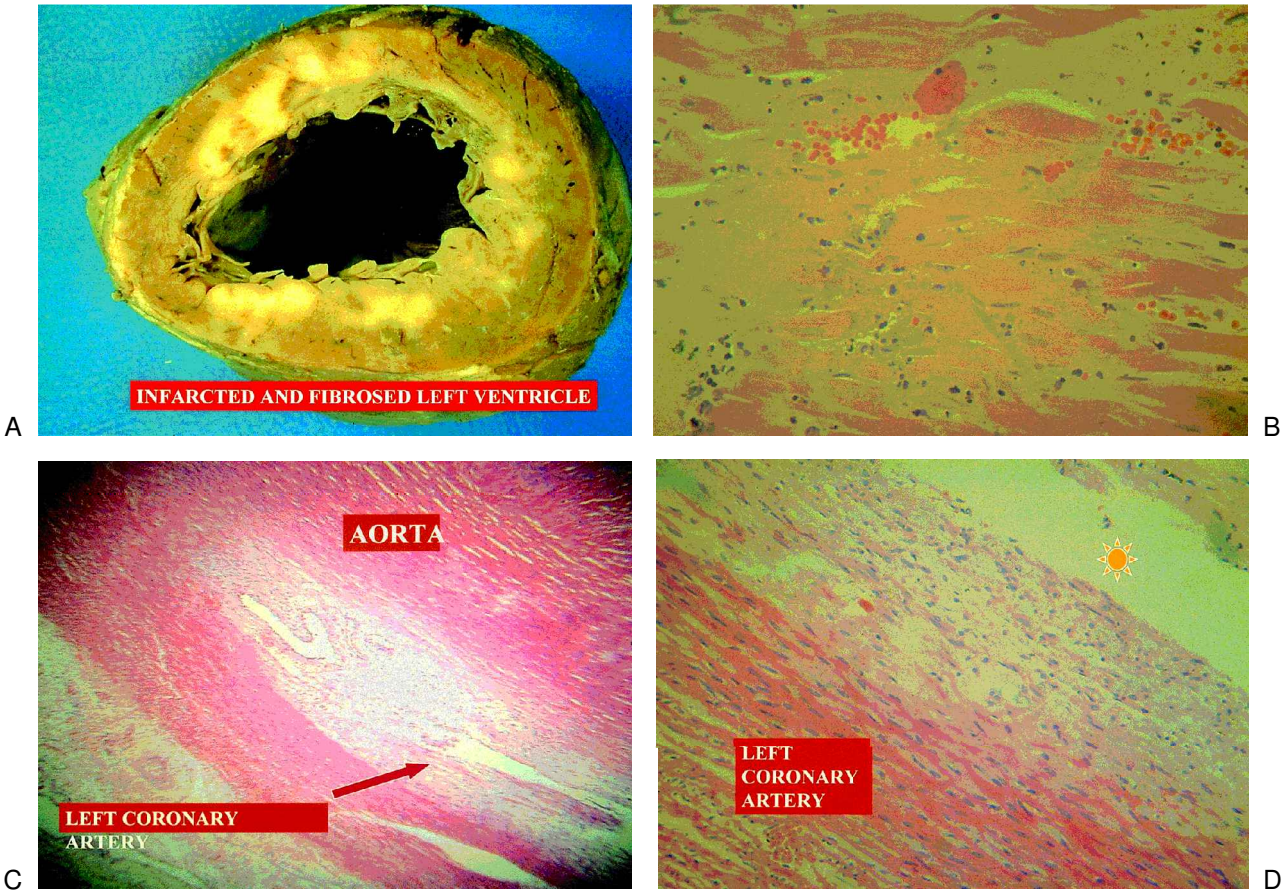

Figure 4.
Histological examination. Gross transverse sections of the left ventricle (A) show extensive infarction and ischaemic fibrosis. The section of the wall of the left ventricle (B) shows ischaemic necrosis and fibrosis (haematoxylin and eosin). The wall of the aorta (C) shows the stenosed intramural portion of the left coronary artery adjacent to the atretic orifice (low power, haematoxylin and eosin). A high power view of the intramural portion of the left coronary artery (D) reveals the absence of inflammation or fibrosis (haematoxylin and eosin). The lumen is denoted by an asterisk.

may rarely be associated with localised or widespread supravalvar aortic stenosing arteriopathy.[17–19] As there was no histological evidence of any such abnormality in our case, it was considered, on balance, to be a congenital malformation. The patency of the main arterial segment is rather curious, as a blind-ending vessel would be expected to thrombose and obliterate.[16] We can offer no explanation for this, but we do not consider that this finding invalidates a congenital aetiology. Musiani and his colleagues[20] maintained that, in true congenital atresia of the left coronary artery, there is no coronary orifice and no left main trunk, with blind proximal ends to the anterior descending and the circumflex arteries. Such an arrangement, however, would appear to be rare.[8,13] It represents the extreme condition in a spectrum of changes in which the main segment may be a fibrous band,[11,21] an impervious cord,[10,11,17,22] or a diminutive twig.[3,8] Ours is the first detailed case in which the atresia is limited to the orifice alone, without any stenosis of the main stem of the artery. The arterial orifice may be absent without trace,[13] or may be represented by a dimple in the interior aspect of the aorta,[3,21–23] or by a blind pouch.[24] Although, by definition, outside the range of atretic conditions, severe orificial hypoplasia may well be related.[25–27]

Atresia of the orifice of the left coronary artery is an exceedingly rare condition. Musiani and his colleagues[20] reviewed 26 cases from the literature and added two more. Other reported cases,[3,4,7,8,13,18,28–30] together with the present case, bring the total published, to the best of our knowledge, to 41 cases. Becker[31] considers that the condition fits into the designation of single coronary artery, and from a functional point of view this is logical. On the other hand, some authorities regard the condition to be totally distinct from that of single right coronary artery, even though the entire coronary circulation is supplied solely by the right coronary artery.[20,32] This latter view is based on the difference between the clinical presentations which, in turn, are a consequence

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of different patterns of myocardial perfusion. Generally speaking, with single coronary arteries, there is adequate perfusion due to major anastomoses in the atrioventricular groove. The flow of blood to the bed of the left coronary artery is centrifugal or anterograde. The condition is usually asymptomatic, in the absence of coexisting atherosclerosis or aortic stenosis,[26,33,34] although it has been suggested that it may play a role in some unexpected deaths both in infants,[35,36] and in adults.[37]

In cases with atresia of the left coronary arterial orifice, the collateral supply from the right coronary arterial system to the territory of the left artery usually takes in the region of the apex of the heart. Between the small distal branches of the vessels, the flow is centripedal or retrograde, and is generally inadequate, leading to ischaemia and symptomatic cardiac disease or sudden death.

When cardiac symptoms present in infancy, the initial diagnosis is often that of a cardiomyopathy.[4,38] In our case, this was suggested by the preceding presumptive viral infection. Clinical evidence of myocardial ischaemia, and angiographic findings, may lead to a diagnosis of anomalous origin of a coronary artery from the pulmonary trunk.[5–9] As in our case, the true nature of the abnormality may only become evident at surgery. Although there may be different clinical presentations, there does not appear to be a rigid anatomical distinction between single coronary artery, of the types reviewed and classified by Smith[34] and Allen & Snider,[39] and isolated atresia of the left coronary orifice. An infundibular artery, or an infundibular branch of the right coronary artery, is said to be present in approximately half of human hearts. Although usually small, it can provide an important avenue for the development of collateral circulation in occlusive disease.[40] It may sometimes be large enough to present a hazard to surgical incisions into the infundibulum.[41,42] It may also join the anterior descending branch of the left coronary artery to form the anastomatic circle of Vieussens (cited by Vidne et al.[43]). If this occurs in conjunction with left orificial stenosis, the flow of blood to the bed of the left coronary artery would be centrifugal or anterograde. The clinical picture would then be that of a single coronary artery, rather than that of an atretic coronary arterial orifice. This intermediate anatomical state is illustrated by two cases which have been described as being examples of single coronary artery, but which had rudimentary traces of the main stem of the left coronary artery (Fig. 5).[37,44]

Surgery would appear to offer the only chance of survival in cases with myocardial ischaemia. There are several reported instances of successful intervention in infants,[4,5,45] children,[13,17,19,46] adolescents[21,38] and adults,[12,23,43] with ages ranging from two months[6] to 56 years.[22]

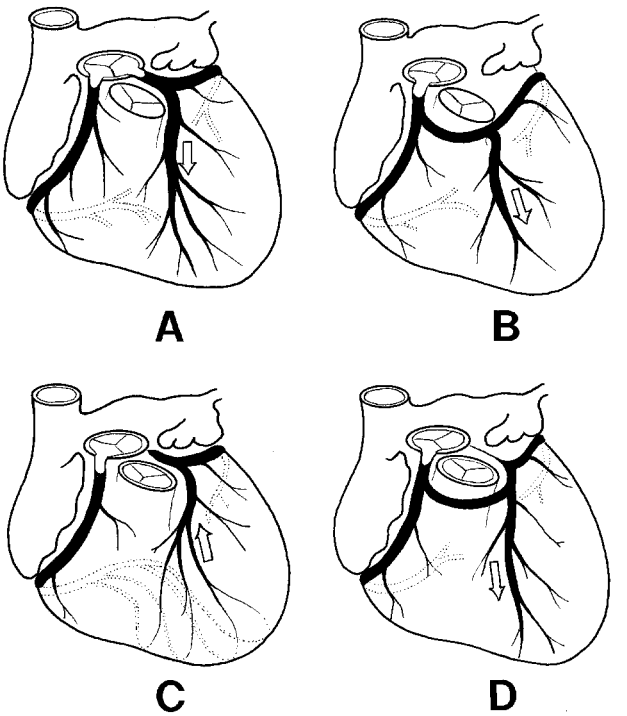

Figure 5.
Diagrams of the coronary arteries. (A) Normal. (B) A usual type of single (right) coronary artery with anterograde flow in the "left" branches through a large canus artery. (C) Ostial stenosis of the left coronary artery as in the present case, with retrograde flow from apical anastomoses with branches of the right coronary artery. (D) Intermediate form; atresia of the left main coronary artery with anterograde flow into its branches through a large conus branch of the right coronary artery.

In our case, the unsuccessful outcome was probably due to the extensive myocardial infarction that had already been sustained, but it is possible that the additional presence of the wide intramural course of segments of the anterior descending coronary artery may have had an adverse effect.[47–49] This combination of anomalies has been reported previously.[46] As myocardial bridging is so very common in normal hearts,[50–52] this was probably a chance association.

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