Severe Congenital Stenosis of the Left Coronary Artery Ostium and Its Possible Pathogenesis According to Current Knowledge on Coronary Artery Development

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We report a 72-year-old woman with severe congenital stenosis of the left coronary artery orifice and clinically significant atherosclerotic changes in both the right and left coronary arteries. The stenotic ostium was located at the point at which the left and posterior aortic valve leaflets joined to form the left commissure, just at the distal vertex of the left interleaflet triangle, between the left and posterior aortic sinuses. The right coronary artery was more developed in size than usual, whereas the left coronary artery consisted of a short left main coronary trunk that bifurcated into left anterior descending and left circumflex arteries. The left coronary artery system was filled retrogradely through two vessels proceeding from the right coronary artery, namely, the conal artery and a well-developed branch that ran across the interventricular septum. This abnormal arrangement of the coronary arteries showed striking functional similarities with atresia of the left main coronary artery.

Current knowledge on the morphogenesis of the coronary arteries suggests that the present anomalous coronary artery pattern resulted from the penetration of the anticipated left coronary artery system into the aorta at a totally erroneous site. This hindered the normal development of the ostium, which subsisted as a punctiform, practically nonfunctional opening.


Congenital obstructive anomalies of the left coronary artery ostium are uncommon events: left main coronary artery (LMCA) atresia [see Byrum et al. (1), Debich et al. (2), and Musiani et al. (3) for extensive reviews of the literature], ostial occlusion resulting from supravalvular aortic stenosis (4–9), and ostial stenosis caused by a congenital ostial membrane (10), congenital syphilis (11), or defective development of the proximal segment of the LMCA (12).

The aim here is to describe the case of a 72-year-old woman with severe congenital stenosis of the left coronary artery ostium due to its erroneous location at the left commissure of the aortic valve. To the best of our knowledge, this report is the first to describe this kind of congenital obstruction of a coronary artery orifice.

Case Report

Clinical History

The patient was admitted to the University Hospital “Carlos Haya” of Málaga on October 29, 1995, because of angina pectoris of 2 hours’ duration which was relieved by continuous nitroglycerin perfusion. Relevant past medical history included diabetes mellitus type II and anginal chest pain for the past 6 months. Clinical examination revealed no...
cardiac murmurs; blood pressure was 110/65 mm Hg. Venous pressure, measured in the external jugular vein, as well as carotid pulse and precordial palpation were normal. Fine crepitant reals were audible over the basal lung fields. Electrocardiographic (ECG) tracings showed sinus rhythm and an acute “non-Q wave” anterolateral myocardial infarction. Chest X-ray findings revealed no other complications than aortic calcification.

Cardiac catheterization on October 31, 1995, showed a morphologically normal left ventricle, with a ventricular ejection fraction of 0.60. On selective coronary angiography, cannulation of the left coronary artery was not possible due to complete obstruction of the coronary ostium. Injection of the right coronary artery (RCA) demonstrated the existence of a well-developed collateral branch arising from the RCA stem, just before the origin of the posterior descending artery. The collateral vessel crossed the interventricular septum to connect with the left anterior descending artery (LAD) and filled the entire left coronary system in a retrograde fashion (Figure 1). After filling the LAD and left circumflex artery (LCx), which were placed normally, contrast material flowed retrogradely through a short left main coronary artery trunk to opacify a blind sac. The RCA, which originated normally from the right aortic sinus, was enlarged and tortuous. Clinically significant atherosclerotic irregularities included 50% stenosis just before the distal bifurcation of the RCA, 90% narrowing at the origin of the posterolateral branch of the RCA, and 70% obstruction at the beginning of the distal portion of the LAD.

The diagnosis was complete occlusion of the left coronary ostium, with collateral heterocoronary circulation from the RCA, and ischemic cardiomyopathy. Therefore, the patient was referred for surgery.

Operation was performed on November 8, 1995. The left internal mammary artery was anastomosed to the distal LAD, and an aortocoronary saphenous vein bypass was constructed to the posterior descending artery. The patient was not weaned from cardiopulmonary bypass because of poor contractility of the left ventricle after 2 hours’ assistance with bypass and intraaortic balloon pumping IABP.

Pathologic Findings

Postmortem examination disclosed a normal heart, with no significant alterations in chamber size or wall thickness. All cardiac valves were normal. There were numerous calcific deposits in the aortic wall. Fibrotic foci were observed at the apical portion of the left ventricle free wall.

The right coronary ostium, located in the right aortic sinus, was 8.5 mm in diameter. The RCA, which was dilated and strikingly tortuous, branched normally. There were numerous atherosclerotic changes along its proximal portion.

The left coronary artery consisted of a short LMCA that bifurcated into LAD and LCx (Figure 2). However, no left coronary ostium could be recognized; the LMCA seemed to end as a blind sac inserted into the wall of the aortic root, approximately at the level of the left commissure of the aortic valve (Figure 3).

Histologic examination revealed the existence of an extremely stenotic, practically nonfunctional opening that connected the lumen of the LMCA with the most distal part

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**Figure 1.** Preoperative right coronary angiogram. A well-developed collateral branch (arrows) arising from the right coronary artery (RCA), crosses the interventricular septum to connect with the left anterior descending artery (LAD).

**Figure 2.** Left lateral view of the left ventricle and aortic root (asterisk). The short left main coronary artery (arrowhead) bifurcates into left anterior descending and left circumflex arteries. Note that the left main coronary artery is somewhat narrower than usual.
the subaortic outflow tract (Figure 4). This punctiform orifice was located at the point at which the left and posterior aortic valve leaflets joined to form the left commissure, just at the distal vertex of the left interleaflet triangle, between the left and posterior aortic sinuses (Figure 5).

The LAD was normal in size along its proximal middle portion; thereafter, its lumen was partially occluded by an atherosclerotic plaque, and from this point onward the caliber of the vessel became remarkably narrower. The LCx was short and relatively thin; it gave off two small marginal branches. The posterolateral aspect of the left ventricle was supplied by posteroventricular branches of the RCA.

The conal artery coming from the RCA was more developed in size than usual; it contributed to the blood supply of the left coronary system by connecting the RCA with the proximal segment of the LAD. In addition, a well-developed collateral vessel arose from the posterior segment of the RCA stem and ran intramyocardially across the interventricular septum to anastomose with the LAD at its middle portion.

Discussion

Anatomical and Clinical Aspects

It is well known that one of the main causes of congenital left coronary ostial occlusion is LMCA atresia. In this congenital anomaly there is no left coronary orifice or functional LMCA trunk; the LAD and LCx are connected as usual, and their blood supply proceeds from one or more collateral vessels arising from the RCA (2,3). Occasionally, LMCA atresia occurs in association with supravalvular aortic stenosis (2,7,13,14).

Sometimes, ostial occlusion is directly related with supravalvular aortic stenosis. In such cases, the occlusion may be due to (i) fusion of the free edge of the left aortic valve leaflet with the narrowed supravalvular segment (4–8); (ii) medial thickening and premature atherosclerosis of the aorta and left coronary artery (7); (iii) fibrotic changes in the aortic media involving the coronary ostium and the proximal coronary artery (7); or (iv) the thickened ridge of the aortic wall itself (15). In this context, it should be noted that the obstruction of the left coronary orifice may also result from the isolated adherence of the left aortic valve leaflet to the aortic wall in the absence of supravalvular stenosis (16–18).

Congenital stenosis of the left coronary ostium is an extremely rare occurrence. It may be due to the existence of a congenital membranous structure that is continuous with the
aortic intima. This condition was reported in two children aged 2 and 8 years, respectively, the latter with truncus arteriosus type I (10). Congenital syphilitic aortitis has been suggested as a cause of stenosis (11), whereas the possibility that the congenital narrowing of the ostium as isolated cardiac anomaly might be due to a developmental defect was only adduced in the case of an infant, aged 5 months, with a left coronary artery originating from a punctiform ostium located in the left aortic sinus (12).

Anatomically, the obstructive anomaly of the left coronary ostium described herein differs from those mentioned above. Indeed, in the present case, the obstruction of the ostium resulted from its erroneous location at the distal vertex of the left interleaflet triangle of the aortic valve. To a certain extent, our case resembles that of the infant reported by Verney et al. (12), in whom a normal LMCA arose from a punctiform ostium. In the infant’s heart, however, the ostium was located normally in the left aortic sinus.

The present anomalous arrangement of the coronary arteries has striking similarities with LMCA atresia from the functional viewpoint. As in LMCA atresia (3,19), in the present specimen, coronary blood flow had to be in part centripetal, since the circulation was from the right coronary artery collaterals to the left coronary system vessels of increasingly larger size. Thereafter, flow had to reverse in the left coronary artery system. In this context, it should be noted that both the present anomalous coronary artery pattern and LMCA atresia functionally differ from the so-called solitary coronary ostium in aorta which comprises the single right coronary artery and the anomalous origin of the left coronary artery from the right coronary artery or from the right aortic sinus. Actually, in solitary coronary ostium in aorta there are no collateral vessels, and the blood flow pattern is centrifugal and not centripetal; blood flows from the center toward the periphery through progressively smaller vessels (3,19) and is not subjected to a reverse flow.

Congenital atresia of the LMCA often becomes symptomatic during childhood and adolescence. In contrast, it usually remains asymptomatic between the late teens and the early forties (3). Survival to adulthood seems to depend mainly on the rate of development of the collateral circulation and the severity of the coexistent congenital cardiac lesions (2,19,20). It is thought, however, that even in the absence of acquired atherosclerosis, vasospasm, or other concomitant cardiac defects, patients with LMCA atresia will inevitably become symptomatic from the fifth decade onward (3,19,21). In the patient reported herein, anginal chest pain did not appear before the beginning of the seventh decade, and was mainly attributable to the atherosclerotic lesions of the coronary arteries.

Surgical management of congenital LMCA atresia has been reported in nine adults, aged 38–68 years (3). Coronary atherosclerosis occurred in two of these nine cases only (19,22). In both patients, surgical revascularization using a saphenous vein bypass graft (19,22) and a mammary artery graft (22) was successful.

**Pathogenetic Aspects**

Recent work, carried out in quail (23), chick (24,25), quail-chick chimera (26), and rat (27) embryos, has contradicted the classic hypothesis that the proximal coronary arteries develop as coronary artery buds that hollow out from the aorta to connect with the peritruncal plexus of capillaries located in the subepicardial layer of the developing heart [see Tomanek (28) for a review]. These studies have demonstrated that the proximal coronary arteries do not grow outward from the aorta, but into the aorta. They develop with the penetration of endothelia into the media of the aorta from the peritruncal ring of coronary arterial vasculature (23,24). The main coronary arteries form by coalescence of discontinuous colonies of microvessels (25,27). As observed in quail-chick chimeras, the quail endothelial cells grow into the aorta at several sites, but only two of these sites undergo coalescence of the capillaries to develop coronary arteries (26). Therefore, from the embryological viewpoint, a left coronary that arises normally from the left aortic sinus has to be regarded as a left coronary artery connected to the left aortic sinus, whereas a solitary coronary ostium in the aorta is the consequence of sole successful
penetration of the peritruncus plexus of capillaries into the aorta.

On this basis, the pathogenesis of the present coronary artery arrangement is easy to explain. The anomaly resulted from the penetration of the anticipated left coronary artery system into the aorta at a completely erroneous site, namely, just at the point at which the developing left and posterior leaflets joined to form the commissure. The wrong emplacement of the ostium hindered its normal development, subsisting as a punctiform, practically nonfunctional opening. As a result, the left coronary system had to be supplied through a collateral circulation coming from the right coronary artery system. Nevertheless, the etiologic factor or factors inducing the penetration of the capillary system into the aortic wall at a wrong site remain unknown.

The anomalous embryologic process described above undoubtedly diverges from that leading to LMCA atresia. According to current knowledge on the morphogenesis of the coronary arteries, the atresia of the coronary artery trunk and its ostium must be the result of a pathogenic process that takes place after the formation of the normal left coronary system, that is, after the normal penetration of the anticipated left coronary system into the aorta. This suggestion is mainly supported by the fact that in most cases of LMCA atresia there is a cordlike structure occupying the position of the LMCA and a dimple instead of the left coronary ostium (29–32). However, the nature of the pathogenic process excluding the lumen of the LMCA and its ostium is uncertain. Interesting hypotheses, such as failure of canalization (arterIALIZATION) of the proximal segment of the LMCA (33,34) and coronary obstruction by infection of thrombosis in the early embryonic stage (32), can be gained from classical papers. Further studies are needed, however, to decide the mechanism by which an anticipated main coronary artery trunk becomes occluded during fetal life.

Supported by grant PB96-0475 from the DGICYT (Ministerio de Educación y Cultura, Spain).

References


