Hemodynamically Significant Primary Anomalies of the Coronary Arteries

Angiographic Aspects

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SUMMARY Hemodynamically significant primary anomalies of the coronary arteries are those which alter myocardial perfusion. There are four major types: coronary artery fistulae, origin of the left coronary artery from the pulmonary artery, congenital coronary stenosis or atresia, and origin of the left coronary artery from the right sinus of Valsalva, with subsequent passage of the vessel between the aorta and right ventricular infundibulum. The angiographic features of these lesions are discussed.

PRIMARY CONGENITAL ANOMALIES of the coronary arteries (that is, those which can occur as isolated anomalies and are not necessarily associated with other types of congenital heart disease) can be divided into hemodynamically significant and insignificant lesions.

The hemodynamically insignificant anomalies are principally those of abnormal aortic origin or distribution of the coronary arteries. They occur in slightly less than 1% of adult patients undergoing coronary arteriography and in children with certain forms of congenital heart disease, such as d- and l-transposition, tetralogy of Fallot, double outlet right ventricle and single ventricle. Myocardial perfusion is not altered in these individuals.

The hemodynamically significant anomalies of the coronary arteries are characterized by abnormalities of myocardial perfusion. There are four major types: 1) coronary fistulae, 2) origin of the left coronary artery (LCA) from the pulmonary artery (PA), 3) congenital coronary stenosis or atresia and 4) origin of the LCA from the right sinus of Valsalva, with subsequent passage of the vessel between the aorta and right ventricular infundibulum. Although angiography provides the key to diagnosis, there have been no previous comprehensive discussions of the radiologic aspects of these lesions. In this paper we review their angiographic appearance, based upon a survey of previously published reports and our own experience with 34 cases. Twenty-one of our patients had coronary artery fistulae, 12 had anomalous origin of the LCA from the PA and one had atresia of the origin of the LCA.

Coronary Artery Fistulae

A pre-capillary fistula connecting a major coronary artery directly with a cardiac chamber, coronary sinus, superior vena cava or PA, is the most common form of hemodynamically significant coronary anomaly. Physiologically, a left-to-right shunt exists in such cases, except in the rare instances when the termination is in a left heart chamber. Approximately half of these patients are asymptomatic and undergo cardiac catheterization because of a continuous precordial murmur. The rest may develop congestive heart failure, subacute bacterial endocarditis, myocardial ischemia or infarction resulting from a "steal" phenomenon, or rupture of an aneurysmal fistula.

The symptoms, murmur, plain radiographic findings and electrocardiographic changes of coronary fistulae may closely resemble those of other lesions, such as patent ductus arteriosus, aortopulmonary window, ventricular septal defect with aortic regurgitation, rupture of a sinus of Valsalva aneurysm and some cases of origin of the LCA from the PA. In some cases, an atypical location of the murmur will suggest that it is not caused by a patent ductus arteriosus.

Twenty-one of our 34 cases had coronary artery fistulae. Seven arose from the right coronary artery (RCA) or one of its branches, while 14 arose from the LCA or one of its branches. Fifteen drained into the right ventricle (RV), two into the right atrium (RA), two into the main PA and two into the coronary sinus. To thoroughly ascertain the patterns of origin and drainage of these lesions, we reviewed an additional 342 previously reported cases. Among the entire group of 363 cases with these lesions, 181 (50%) arose from the right coronary tree, 151 (42%) from the left coronary tree and 19 (5%) from both vessels. The vessel of origin was not specified in 12 cases (3%). Drainage occurred into the RV in 150 cases (41%), into the RA in 94 cases (26%), into the PA in 63 cases (17%), into the coronary sinus in 23 cases (7%), into the left atrium (LA) in 18 cases (5%), into the left ventricle (LV) in 11 cases (3%) and into the superior vena cava in 4 cases (1%). Thus, approximately half of these
fistulae arise from each coronary artery, but over 90% drain into the systemic venous side of the circulation.

Although oximetry and other catheterization data are helpful in evaluating these lesions, angiography provides the definitive diagnosis. If the shunt is large, the involved coronary artery will virtually always be grossly enlarged, and the fistula visualized by supravalvular aortography. If the shunt is small, the involved coronary artery is only slightly enlarged and selective coronary arteriography may be required to define the abnormality. In either event, contrast material passes directly from the coronary artery through the single or multiple communications which comprise the shunt, and then into the recipient cardiac chamber or blood vessel. Beyond the origin of the fistulous communication, the coronary artery itself reverts to normal caliber, but this distal segment may not be visualized because of the "steal" phenomenon.

Figure 1 shows a left circumflex-coronary sinus fistula with a 1.9 to 1 pulmonary-to-systemic flow ratio. The left circumflex artery is massively enlarged and the lesion is easily seen on a supravalvular ascending aortogram. Figure 2 shows a RCA-RV fistula arising from the distal portion of the vessel near the crux of the heart. Supravalvular aortography had demonstrated enlargement of the RCA, but selective arteriography was required to precisely locate the lesion. Figure 3 is a selective left coronary arteriogram of an adult patient with a left anterior descending (LAD)-PA fistula. These particular fistulae are frequently composed of multiple small and tortuous channels and may not require repair because of the small amount of blood shunted.

In approximately 3% of cases of congenital coronary artery fistulae, there is associated absence of the contralateral coronary artery. An example of such a case is shown in figure 4. This patient had a RCA-
After locating the fistula, the surgeon usually occludes it transiently to assure that the thrill is eliminated and that no ischemic electrocardiographic changes develop. It is then permanently ligated, using an exterior approach, if possible. If the lesion is inaccessible, or if multiple channels are present, cardiopulmonary bypass may be required. Successful repair depends to a large extent upon precise angiographic demonstration of the points of origin and drainage and the number of channels present.

**Origin of the Left Coronary Artery from the Pulmonary Artery**

The second most common hemodynamically significant anomaly is origin of a coronary artery from the main PA. In approximately 90% of such cases, it is the LCA which originates from the PA. In most of the others, the RCA originates from the PA. In addition, there are isolated reports of origin of the LAD artery from the PA, and origin of both coronary arteries from the PA. The latter is incompatible with life. The discussion below focuses primarily upon anomalous origin of the LCA from the PA.

When this lesion is present, myocardium in the distribution of the LCA is perfused both at low pressure and low oxygen tension after birth. The low perfusion pressure stimulates development of collateral circulation from the RCA to branches of the LCA. However, the beneficial effect of collateralization is usually offset by a "steal" mechanism, whereby blood passing from the RCA through collaterals to LCA branches...
fails to effectively perfuse myocardium, but instead flows retrogradely through the LCA back into the PA. A left-to-right shunt is thereby created.

Seventy-five percent of all patients with this anomaly develop symptoms of congestive heart failure and myocardial ischemia in the first four months of life. Chest radiographs show cardiac enlargement and failure, and electrocardiograms suggest anterolateral ischemia or infarction. Most patients who develop early symptoms die during the first two years of life. The remaining 25% with this anomaly survive infancy, but later in life usually develop mitral regurgitation, angina, a continuous murmur or sudden death.

The clinical course of the patient seems to be determined largely by the adequacy of collateral circulation from RCA to LCA. The greater the degree of collateralization, the more favorable is the prognosis. Perry and Scott found that patients with a left-to-right shunt through collaterals, which was demonstrable angiographically and was associated with an increase in PA oxygen saturation, all survived infancy, regardless of type of treatment employed. Where the left-to-right shunt was only sufficient to be detected angiographically or by dye dilution curves but did not produce an increase in PA oxygen saturation, survival was only approximately 50%.

The original surgical approach was ligation of the proximal LCA at its origin from the PA. This eliminated the "steal" and thereby enhanced myocardial perfusion by the collaterals emanating from the RCA. Obviously, this was only a partial solution to the problem, since perfusion was still via low pressure collateral circulation rather than at systemic arterial pressure. An additional problem was that ligation sometimes was incomplete, with recurrence of retrograde flow from the LCA into the PA. Recently, saphenous vein bypasses have been used in addition to proximal ligation to provide direct recanalization of the LCA system. Another approach is that of Neches et al., who have detached the LCA and the surrounding cuff of tissue from the PA and anastomosed it to the aorta. This obviates the need to suture bypasses to coronary arteries in young infants.

The angiographic findings generally reflect coronary hemodynamics accurately. Supravalvular aortography reveals a large RCA and absence of an aortic LCA ostium. As the filming sequence progresses, collateral vessels can usually be seen arising from the RCA and opacifying the LAD and circumflex branches of the LCA. If collateral flow is large, retrograde filling of the LCA can then be seen with eventual passage of contrast back into the main PA. Angiography may at times be more sensitive in detecting the left-to-right shunt at the PA level than oximetry. Among our 12 patients with this lesion, a broad spectrum of collateral patterns was seen. At one end was a 4-week-old infant with only minimal collateralization and very little opacification of the LCA branches. This patient died of congestive heart failure shortly after admission to the hospital, where she received emergency cardiac catheterization. At the other end was an asymptomatic 10-year-old boy who was studied because of mild mitral regurgitation and a continuous murmur along the left heart border. Cineangiography revealed a dilated RCA with extensive collateralization and dense opacification of all LCA branches and the PA. This patient had a 2 to 1 pulmonary-to-systemic flow ratio. Figure 5 is an angiogram in a 4-month-old girl with congestive heart failure, a systolic murmur, marked cardiomegaly on plain chest radiographs and a 1.2 to 1 pulmonary-to-systemic flow ratio demonstrated at catheterization. The case shown in figure 6 is that of a 13-year-old boy who had had previous attempts at ligation of his anomalous LCA at ages 4 and 6 and an anterior myocardial infarction at age 8. Catheterization did not reveal a recurrent left-to-right shunt by oximetry. However, angiography clearly showed the left circumflex artery, LCA and main PA filling through collaterals from the RCA. Left ventriculography revealed an anterior aneurysm of the LV. At surgery, ligatures were found around the proximal segment of the LAD artery. The orifice of the LCA remained patent and had to be closed from within the PA. This case illustrates a potential complication of surgical therapy in the instances of inadequate ligation and/or recanalization of the anomalous LCA.

**Congenital Coronary Stenosis or Atresia**

Stenosis or atresia of the coronary arteries may occur as an isolated anomaly, but more commonly occurs in conjunction with other congenital lesions, such as calcific coronary sclerosis, supravalvular aortic stenosis, homocystinuria, Friedreich's ataxia, Hurler's syndrome, progeria and rubella syndrome.

Calcific coronary sclerosis is a disease of unknown etiology in which heavy calcification is found in the intima, internal elastic membrane and the adjacent inner portion of the media of coronary arteries in infants. Occlusion of the lumen results from fibroblastic intimal proliferation. In most reported cases, arteries of other organs are involved, such as the lungs, spleen, kidneys and adrenals.

Supravalvular aortic stenosis is occasionally associated with ostial occlusion of a coronary artery. This may be due in some cases to actual atresia of the ostium, as shown in figure 7. In other cases, a fusion of the free edge of an aortic valve cusp with the narrowed supravalvular segment results in occlusion of the underlying coronary ostium.

Arterial lesions in homocystinuria are characterized by extensive intimal proliferation and hyperplasia, and may be complicated by superimposed platelet thrombi. Coronary and cerebral arteries are commonly affected. A clinical study of 38 patients with this disease showed that four developed myocardial infarction or angina before age 45.

Nadas et al. reported on the autopsy findings in one patient with Friedreich's ataxia whose coronary arteries showed marked medial hypertrophy, intimal proliferation and varying degrees of obstruction, and whose LV was diffusely fibrotic. Five other living
FIGURE 5. (A) Simultaneous frontal and lateral views from a supravalvular aortogram in a 4-month-old girl with anomalous origin of the left coronary artery (LCA) and the pulmonary artery (PA). Early films show an enlarged right coronary artery (arrowhead), no evidence of a LCA ostium and early collateral filling of the left anterior descending (LAD) and circumflex branches of the LCA (the white arrow points to the LAD artery). (B) Later films show progressive opacification of the LCA via collateral circulation, with retrograde flow back into the PA (black arrow).

patients with the disease all showed electrocardiographic changes suggesting myocardial ischemia. Based on their experience with these patients and a review of the literature, they felt that coronary lesions probably occur in most patients with Friedreich's ataxia.

Hurler's syndrome is a genetic deficiency of lysosomal hydrolases which leads to the accumulation of acid mucopolysaccharides in fibroblasts and other cells of numerous organs. Patients with this syndrome may have multiple severe narrowings of the major coronary arteries. Histologically, these changes result from marked intimal thickening caused by the presence of abnormal storage cells.

Progeria and rubella syndrome are other diseases which may be characterized by coronary artery
FIGURE 6. (A and B) Early simultaneous frontal and lateral views of a supravalvular aortogram in a 13-year-old male who had undergone two earlier attempts at ligation of an anomalous left coronary artery (LCA) arising from the main pulmonary artery (PA). There is a large right coronary artery (RCA) but no LCA can be seen arising from the aorta. (C and D) Simultaneous frontal and lateral films slightly later during the same injection show filling of the left circumflex artery and LCA via collaterals from the RCA. The retrograde flow pattern opacifies the main PA. The white arrow in figure 6C points to the origin of the LCA from the PA, and the black arrowhead in figure 6D points to the left circumflex artery. Reoperation revealed that only the left anterior descending artery had been ligated previously.

FIGURE 7. (right) (A) Simultaneous biplane views of a left ventriculogram in a 17-year-old male with supravalvular aortic stenosis and marked left ventricular hypertrophy. The large white arrow shows the typical narrowing of the aorta just above the sinuses of Valsalva. There were no symptoms or electrocardiographic evidence of coronary disease. A large right coronary artery (RCA) is seen (small white arrow) but there is no definitive visualization of a left coronary artery (LCA). (B and C) Early and late phase biplane views of a selective right coronary arteriogram. The RCA gives rise to extensive collateral circulation which opacifies the left anterior descending artery (arrow). In the late phase, the LCA fills in retrograde fashion as far as its atretic proximal segment (arrowhead).
obstruction, although relatively little is known of their frequency or location. In patients with progeria, advanced atherosclerosis may develop as early as the second decade of life, leading to premature coronary artery disease.

Origin of the Left Coronary Artery From the Right Sinus of Valsalva

The final, and probably least common, hemodynamically significant coronary artery anomaly is origin of the LCA from the right sinus of Valsalva or the RCA itself, with subsequent passage of the vessel between the aorta and right ventricular infundibulum. After passing between them, the LCA bifurcates into normal LAD and circumflex arteries. We have not had experience with this lesion, but it is diagramatically represented in figure 8. Cheitlin et al. reviewed the autopsy findings in 33 such cases and found that sudden unexplained death occurred at an early age in nine (27%). Seven of these nine deaths occurred during vigorous exercise. It had been postulated that death results from acute occlusion of the aberrant LCA, and that the occlusion could occur at two possible sites: the sharp leftward bend made by the LCA immediately upon arising from the right sinus of Valsalva, or the passageway between the aorta and the right ventricular infundibulum. As cardiac output rises with exercise, the LCA could either become compressed between the dilated aorta and right ventricular infundibulum, or stretched between them with resultant occlusion of the sharply angulated proximal segment of the vessel. In any event, this is a dangerous lesion which should be treated by bypass surgery. There have been isolated reported instances of myocardial infarction in cases in which the LCA originated from the right sinus of Valsalva and passed either anterior to the PA or posterior to the aorta. Since the aberrant LCA in these cases did not pass between the two great vessels, and these patients had no angiographic evidence of atherosclerotic disease, the etiology of their infarcts is obscure.

Two cases have recently been reported in which chest pain suggesting angina pectoris occurred in patients whose right coronary artery originated from the left sinus of Valsalva and then passed between the aorta and right ventricular infundibulum. There have been no reported deaths with this anomaly, and its hemodynamic significance has not yet been firmly established.

Discussion

Radiologists and cardiologists dealing with adult patients are used to diagnosing coronary artery lesions by standard selective coronary arteriography. Many patients with the anomalies discussed above are initially studied during childhood, however, and selective coronary arteriography is not a standard part of pediatric angiocardiac diagnostic examinations. It may therefore be necessary to diagnose, or at least suspect, the presence of these lesions from left ventriculograms or supravalvular aortograms. If the lesions themselves cannot be clearly shown by such studies, the most reliable clue to their presence is visualization of an enlarged coronary artery. Disproportionate enlargement of one coronary artery may result from the requirement to supply blood to a high flow fistula or to supply collaterals to a contralateral coronary artery which either arises from the low pressure main PA or is significantly narrowed by one of the congenital diseases referred to above. If significant enlargement of a coronary artery is seen during the course of pediatric angiocardiacographic studies, a hemodynamically significant anomaly should be suspected and selective arteriography of the affected vessel should be obtained.

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