Congenital Coronary Artery Obstruction Associated with Aortic Valve Anomalies in Children: Report of Two Cases

RICHARD GIBSON, M.D., MICHAEL R. NIHILL, M.D., CHARLES E. MULLINS, M.D., DENTON A. COOLEY, M. D., FRANK M. SANDIFORD, M.D., AND DAN G. MCNAMARA, M.D.

SUMMARY Coronary arterial obstruction associated with congenital aortic valve disease is rare in childhood. We studied two children with aortic valve disease and symptoms of coronary insufficiency. Cineangiography revealed localized obstruction of the proximal left coronary artery. In one patient, a membrane-like structure partially covered the left sinus of Valsalva, almost isolating the ostium of the left coronary artery; the other patient had severe stenosis of the proximal left coronary artery and a congenitally hypoplastic left coronary cusp. Surgical repair was successful in both cases. Symptoms of myocardial ischemia in children with aortic valve disease should be investigated promptly to exclude obstruction of the coronary arteries.

SURGICAL REPAIR of significant coronary anomalies in children is common in most cardiovascular centers. Evaluation of coronary arteries in children with manifestations of coronary insufficiency, such as angina, syncope, unexplained left ventricular failure or myocardial infarction, is mandatory and should be accomplished by aortic root contrast angiography and, if necessary, selective coronary cineangiography. In this report, we describe two children with anatomic obstruction of the proximal left coronary artery associated with aortic valve disease and symptoms of coronary insufficiency. In both cases, angiographic studies led to the correct diagnosis and successful surgical repair.

Case 1

MT is an 8-year-old Egyptian boy with a history of a cardiac murmur since age 1 year. He was asymptomatic, but cardiac catheterization at 4 years of age at another institution revealed subaortic stenosis and a peak systolic pressure gradient of 80 mm Hg across the left ventricular outflow tract. At surgery, a discrete subaortic membrane was resected; the aortic leaflets appeared thick and an abnormal fibrous band arose from the wall of the aorta above the left sinus of Valsalva and bridged both commissures of the left coronary cusp, leaving a small orifice into the left coronary sinus. This membrane was removed, freeing the leaflet of the left coronary cusp and revealing a normal left coronary ostium. He had a satisfactory recovery and remained asymptomatic until 4 years later, when he had two episodes of severe chest pain associated with strenuous exercise. He was evaluated in our clinic soon afterward. There was a systolic thrill over the base of the heart that radiated to the neck, corresponding to a grade IV/VI aortic ejection murmur. This was followed by a grade I/VI high-pitched diastolic murmur of aortic regurgitation; no click was heard and the remainder of the physical examination was normal. He had no clinical features of William's syndrome or symptoms in infancy to suggest hypercalcemia.

The chest roentgenogram and the resting ECG were normal. The M-mode echocardiogram showed concentric hypertrophy of the left ventricular wall and fine systolic vibrations of the aortic valve leaflets with mild early semiclosure of the anterior aortic valve leaflet. The left ventricular shortening fraction was 44%.

A treadmill exercise test (2.5 mph, 12% grade) was abnormal, with 3–4 mm of ST-segment depression after 4 minutes of exercise (fig. 1). Cardiac catheterization revealed a 40-mm Hg peak systolic gradient across the aortic valve and 2+ aortic insufficiency. Aortic root angiography showed an abnormal membrane-like structure that partially isolated the left coronary sinus (fig. 2), a normal proximal left coronary artery and a dominant left coronary artery system. There were no other radiographic signs of supravalvar aortic stenosis.

Operation was performed under cardiopulmonary bypass in September 1977. A residual subaortic membrane was resected. As described at his first operation, a fibrous membrane covered the left coronary sinus. The membrane was attached to the rim of the sinotubular junction above the origin of the left coronary artery and bridged the commissural edges of the left coronary cusp. The inferior edge of the membrane extended to the base of the left coronary cusp (fig. 2), with incomplete attachment or fenestrations, causing partial obstruction of blood flow to the left coronary artery. There was no commissural fusion or attachment of the free edge of the left coronary cusp to the membrane or the wall of the aorta. The fibrous membrane was removed, mobilizing the left coronary cusp and exposing a normal orifice of the left coronary artery. He recovered from the operation and has remained asymptomatic. One year after his last opera-
tion he ran on a treadmill for 15 minutes, with no chest pain or ST-segment depression.

Case 2

GV is a 9-year-old Italian boy with an aortic insufficiency murmur first detected at the age of 7 years. During the preceding 10 months he had experienced four episodes of severe precordial pain, accompanied by pallor and sweating, triggered by mild physical activities. During the last episode, 4 months before our evaluation, he developed severe pulmonary edema requiring intensive anticongestive therapy and hospitalization for 3 weeks in Italy. Because of the discrepancy between the degree of the aortic regurgitation and the severity of symptoms, further evaluation was recommended. There was no history of rheumatic fever or endocarditis.

Physical examination revealed a diffuse and prominent systolic impulse over the apex of the heart. Auscultation of the heart revealed a grade III/VI aortic regurgitation murmur and a grade II/VI aortic systolic ejection murmur. The chest roentgenogram showed mild cardiomegaly and the ECG (fig. 3) revealed left anterior hemiblock and abnormal Q waves that suggested old anterolateral myocardial infarction. The M-mode echocardiogram showed a dilated left ventricular cavity and diastolic fluttering of the anterior leaflet of the mitral valve. The left ventricular shortening fraction was 35%. A thallium myocardial perfusion scan revealed dilatation of the left ventricular cavity and diminished concentration of the tracer at the apex.

At cardiac catheterization, an aortic root angiogram revealed 3+ aortic regurgitation and a dysplastic left coronary cusp, but the left coronary system was not visualized (fig. 4). Selective right coronary artery angiography revealed retrograde filling of the left anterior descending coronary artery and circumflex coronary artery up to the proximal left main coronary artery (fig. 5). A left ventriculogram showed mild cavity dilatation and anteroapical hypokinesis.

During cardiopulmonary bypass, exploration of the aortic root revealed a distorted and hypoplastic left coronary cusp and severe stenosis of the left coronary ostium; the right and the posterior coronary cusps appeared normal. Competence of the valve was restored by approximating the "free commissures" of the right and noncoronary cusps, resulting in a bicuspid aortic valve. A saphenous vein bypass graft was interposed between the aorta and the proximal left anterior descending coronary artery. Two weeks after surgery, a thallium myocardial perfusion scan showed reduction of left ventricular cavity size, but no change in tracer distribution. The ECG was unchanged.

Discussion

We present two patients with anatomic obstruction of the left coronary artery and coexistent congenital aortic valve disease. To our knowledge, this association has not been stressed in the literature.3, 9-11, 14, 15

The obstructive fibrous membrane over the left coronary ostium appeared normal and the coronary ostia were patent. The proximal bifurcation of the right coronary artery appeared hypoplastic, and the left circumflex coronary artery had a slight narrowing. The left anterior descending coronary artery was partially occluded by a fibrous membrane, which could not be excised. The patient was discharged from hospital 2 weeks later on antiplatelet agents and 9 months later, he was asymptomatic and had a normal ECG.
coronary ostium in our first patient could represent a forme fruste of supravalvar aortic stenosis. Abnormal bands bridging the free edges of the aortic valve cusp have been reported; these bands were called "chorda tendinea congenita in aorta" by Rohrer. Generally, this anomaly causes no significant hemodynamic change or cardiac symptoms.

Coronary obstruction often occurs with supravalvar aortic stenosis. Peterson et al. reported three types of coronary involvement in 20 of 33 cases of supravalvar aortic stenosis he reviewed. This consisted of localized stenosis at or just distal to the coronary ostium; diffuse thickening of the medial layer of the coronary arteries, with intimal fibrosis and atherosclerosis, probably related to chronic localized hypertension of the coronary arteries due to obstruction of the aorta distal to the coronary ostium; and isolation of the coronary artery ostium due to fusion of the free edge of the coronary cusp to the wall of the aorta at the site of the supravalvular stenosis. The findings in our first patient are not the same as those described by Peterson et al. and there was no evidence of supravalvar aortic stenosis by hemodynamic measurements, angiography or at surgery. The distortion of the left coronary cusp in our second patient resembles the third type of pathology described by Peterson et al., but there was no evidence of supravalvar aortic stenosis. We found no report of proximal left coronary artery obstruction due to an obstructive membrane in association with subvalvar aortic stenosis.

In case 1, a membrane-like structure was removed at surgery 4 years earlier, suggesting that the bridging membrane over the left coronary artery ostium may regrow with time. In case 2, hypoplasia of the left coronary cusp producing aortic regurgitation was associated with stenosis of the left coronary ostium with normal main left, left anterior descending and circumflex coronary arteries. Line et al. reported a similar case. The late appearance of symptoms in their patient, a 44-year-old man, could have been caused by the existence of collateral circulation to the left coronary artery from a conal artery. To our knowledge, our case 2 is the first reported case of hypoplastic cusp and stenosis of the left coronary artery ostium in a child.

Isolated atresia of the origin of the left coronary artery is a rare anomaly. Mullins et al. suggested that failure of canalization of the proximal segment of the left coronary anlage was responsible for this lesion. Although case 2 had atresia of the proximal segment of the left coronary artery, the coexistent deformity of the left coronary artery cusp may suggest a localized teratogenic effect late in embryogenesis or in fetal life as a more likely explanation for this lesion. Neither patient had a history of

**Figure 3.** Case 2. Preoperative resting ECG showing abnormal Q waves in precordial leads I and aVL that suggest left anterior hemiblock and old anterolateral myocardial infarction.

**Figure 4.** Case 2. Aortic root angiogram showing moderate aortic insufficiency, an abnormal left coronary cusp and no filling of the left coronary artery.
teratogen exposure. The clinical and catheterization findings were similar to patients with anomalous origin of the left coronary artery from the main pulmonary artery. Clues to the correct diagnosis were a
dysplastic left coronary cusp seen on the aortogram and visualization of the proximal main left coronary artery after selective injection of the right coronary artery in proximity with the aorta rather than main pulmonary artery.

Among the congenital coronary anomalies that produce coronary insufficiency in children, anomalous origin of left coronary artery from the pulmonary artery is the most frequently reported, and most of the experience and advances in coronary surgery in children have been developed from experience with this anomaly. Origin of the left coronary artery from the right sinus of Valsalva has caused myocardial ischemia and death in some children and teenagers during strenuous physical activity. Anomalous origin of the left anterior descending coronary artery from the right coronary artery may result in great morbidity and mortality if transected during repair of tetralogy of Fallot. Other congenital anomalies include congenital atresia or stenosis of the coronary arteries, either isolated or associated with supravalvar, valvar or subvalvar aortic disease. Occasionally, coronary insufficiency may occur as a result of coronary artery fistula.

The acquired causes of coronary obstruction in children include mucocutaneous lymph node syndrome, calcific coronary sclerosis, Marfan's syndrome, homocystinuria, progeria, rubella syndrome, hypercholesterolemic xanthomatosis, Hurler's syndrome, Friedreich's ataxia, idopathic aortitis and Pseudoxanthoma elasticum.

Symptoms and signs of myocardial ischemia may occur as a result of reduced coronary flow reserve in patients with significant aortic stenosis or regurgitation and normal coronary arteries. Despite the close anatomic relation of the aortic valve to the coronary ostia, simultaneous involvement of the coronary arteries in congenital aortic valve disease appears to be very uncommon or infrequently recognized in children.

In conclusion, anatomic obstruction of the coronary arteries can result from a congenitally abnormal aortic valve. Therefore, the coronary arteries should be carefully evaluated in children with aortic stenosis or regurgitation who have symptoms of coronary insufficiency, regardless of the severity indicated by the hemodynamics.

References

Figure 5. Case 2. Selective right coronary angiograms in the posteroanterior projection (A) and left anterior oblique projection (B) showing retrograde filling of the left coronary artery and visualization of the main left coronary artery in close proximity with the left aortic sinuses of Valsalva.


23. Kucera J: Epidemiology and phenotype of congenital coronary artery anomalies analysis of 80 cases found in necropsies of stillborn fetuses and infants. Cor Vasa 20: 369, 1978


R Gibson, M R Nihill, C E Mullins, D A Cooley, F M Sandiford and D G McNamara

_Circulation._ 1981;64:857-861
doi: 10.1161/01.CIR.64.4.857

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1981 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/64/4/857

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in _Circulation_ can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to _Circulation_ is online at:
http://circ.ahajournals.org//subscriptions/