Sequestration of the Left Coronary Artery From the Aorta

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A patient with the left coronary artery isolated from the ascending aorta is discussed. This is the seventh case described and the third diagnosed preoperatively. In contrast to other patients she had predominant aortic incompetence and was much older (52 years compared with the other patients, all younger than 25 years). She also had two angiograms demonstrating progressive isolation of the coronary artery with time. The patient was managed by aortic valve replacement. It is our belief that the condition is acquired and resulted from fusion of the free edge of the aortic valve leaflet to the supravalvar ridge secondary to an inflammatory process.

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A 45-year-old woman was admitted initially in 1983 with dyspnea and chest pain. There was a history of rheumatic fever as a child and previous treatment for pulmonary tuberculosis. Investigations including cardiac catheterization confirmed aortic valve regurgitation and almost complete isolation of the left coronary artery, except for a small communication, from the aorta. Surgical correction was advised but was declined by the patient.

In 1990, when 52 years old, she was readmitted with similar symptoms. She was undistressed with no features of infective endocarditis. The pulse was regular and the blood pressure was 150/60 mm Hg. The apex of the heart was laterally displaced. The first heart sound was soft with an ejection click present. A systolic ejection murmur and a soft early diastolic murmur were heard at the base of the heart and along the left sternal border.

The electrocardiogram demonstrated sinus rhythm, right bundle-branch block, left-axis deviation, and signs of left atrial enlargement. The cardiac silhouette was enlarged on chest roentgenogram.

A 10-mm Hg peak to peak gradient across the aortic valve was measured at catheterization. The valve was not calcified, and the aortic root was dilated. Severe aortic regurgitation was evident, but the ventricle contracted well and was not dilated. The left coronary artery could not be entered. The right coronary artery was a widely patent dominant vessel. A very large conal branch arising from the proximal right coronary artery filled the unobstructed left coronary artery system. Retrograde filling of the left coronary artery showed delineation of a rudimentary left valvar sinus (Fig 1).

During cardiopulmonary bypass with moderate hypothermia and crystalloid cardioplegic arrest the anatomy was defined through a transverse aortotomy. Large right and noncoronary cusps with slightly thickened edges were found. In the position of the left coronary cusp a domed structure, which appeared to be thickened valve tissue, was completely fused with the supravalvar ridge (Fig 2). No egress of blood or cardioplegia could be demonstrated. An incision exposed a valve sinus with a small but patent left coronary artery ostium at the base. No thrombus was present. The valve was replaced with a 25-mm tilting disc prosthesis. Her postoperative course was uneventful.

Comment

Adhesions of the free edge of an aortic valve leaflet to the aortic wall have been well documented. However, this is mostly associated with supraaortic stenosis. In fact, cusp abnormalities were present in more than a third of cases reported in a review of supraaortic stenosis by Rastelli and associates [1]. Underdevelopment of the left coronary cusp was noticed most commonly, but partial cusp adhesions were seen frequently by Flaker and colleagues [2].

Complete isolation of the coronary cusp and the coronary artery in patients with supraaortic stenosis has been described four times [3–6]. Two left and two right coronary cusps were involved. Only one [5] was recognized preoperatively on angiography, the other three being noted at autopsy.

Two further cases have been described [7, 8]. Waxman and co-workers [7] reported isolation of the left coronary artery in a patient with aortic stenosis and regurgitation. A 16-year-old girl presented with syncope and chest pain. There was no history of an inflammatory process. Cardiac catheterization revealed moderate aortic valve regurgitation and a 20-mm Hg gradient across the aortic valve. An isolated left coronary sinus filled retrogradely through the right coronary artery and collaterals. The indication for operation was angina, and at operation the cusp was freed. Follow-up examination and catheterization were normal. Although it was not histologically proven, Waxman and co-workers speculated that their case was a forme fruste of supravalvular aortic stenosis.

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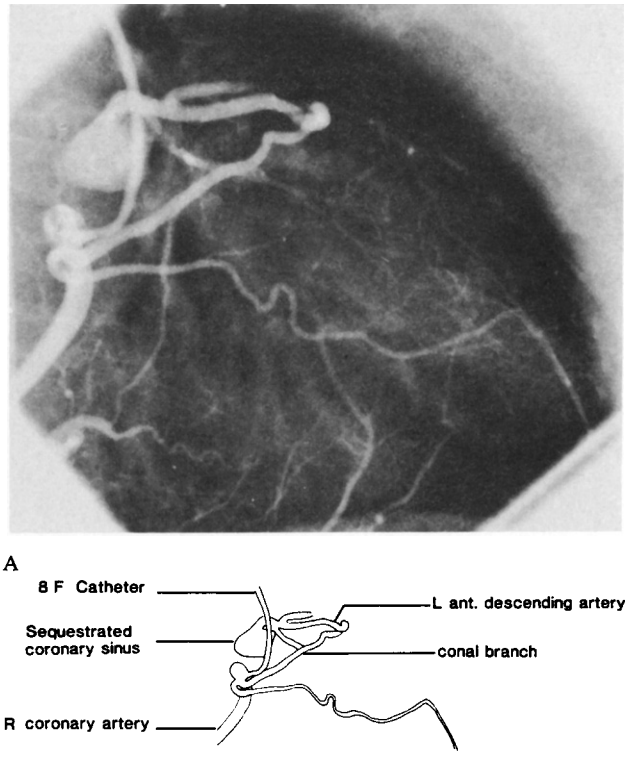

Fig 1. Catheterization of right coronary artery demonstrating filling of the left coronary system and left sinus of Valsalva through a large caval branch.

The question still remains whether isolation of the left coronary ostium is acquired or congenital. The absence of initial signs and the thickened cusps tend toward a gradual inflammatory process involving the valve cusps with adhesions to the aortic wall, but also may be ascribed to a congenital abnormality of the leaflet tissue. In two reports [7, 8] the consensus leaned toward a developmental abnormality. We believe that in our patient it was probably acquired. A heart murmur was noticed 25 years before, and she became symptomatic late in life. There is a history of rheumatic fever as a child, although in isolated aortic valve disease one should also consider nonrheumatic aortic root disease. Over a 7-year period the left coronary ostium progressed from a small communication between the aortic root and left coronary sinus to a complete seal. Unfortunately, the first angiogram was inadvertently destroyed, but the catheterization report is adamant in the description of the left coronary artery filling retrogradely from the right and communicating with the aortic root. One of us (R.F.) performed the first angiogram. Varying degrees of adherence of valve cusps to the aortic wall have been described [2]. Histologic examination of the thickened cusps in our case was unhelpful.

Under normal circumstances one would expect a blind-ending vessel to thrombose and obliterate, but compression of the sinus during systole with to-and-fro movement of blood in and out of the sinus during the cardiac cycle probably prevented stasis formation. This was noticed by Waxman and co-workers [7] as well.

Adhesions of aortic valve cusps are common in the rare condition of supravalvular aortic stenosis. Complete isolation has been described in 4 patients with supravalvular aortic stenosis. This phenomenon was also seen in a patient with quadricuspid aortic valve and could represent a congenital defect [8]. Although the patient of Waxman and co-workers had associated aortic regurgitation, operation was for angina and syncope. Our patient was operated on for pure regurgitation with late onset of symptoms and signs, and no evidence of stenosis. A preoperative diagnosis was possible and a relatively simple operation gave good results.

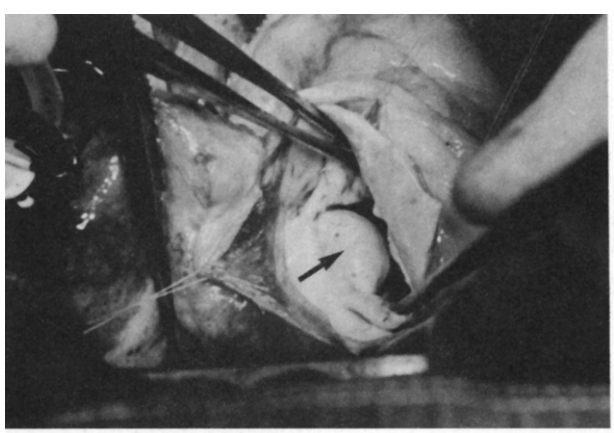

Fig 2. Surgeon's view of domed structure in left sinus of Valsalva.

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