“Sawfish” Systolic Narrowing of the Left Anterior Descending Coronary Artery: An Angiographic Sign of Hypertrophic Cardiomyopathy

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SYSTOLIC NARROWING (“milking” or “bridging”) of the left anterior descending coronary artery (LAD) at angiography is a well-known phenomenon. It occurs both as an isolated finding at cardiac catheterization and in patients with arteriosclerotic coronary artery disease, left ventricular hypertrophy and hypertrophic cardiomyopathy. The angiographic morphology of systolic narrowing of the LAD has not been analyzed in relation to the underlying heart disease, and it is not known whether the same pathologic basis accounts for systolic narrowing of the LAD in patients with or without heart disease. We studied the morphologic aspect at angiography of systolic narrowing of the LAD in six patients without hypertrophic cardiomyopathy, left ventricular hypertrophy or any other form of heart disease and in eight patients with hypertrophic cardiomyopathy.

Methods

Patients

We reviewed the records of 520 patients who underwent cardiac catheterization and coronary angiography. In six patients with otherwise normal coronary arteries and no associated cardiovascular abnormalities part of the LAD had an intramural course that produced systolic narrowing (group A, patients 1–6). In eight of 15 patients with hypertrophic cardiomyopathy and otherwise normal coronary arteries, systolic narrowing of the LAD was demonstrated by selective coronary arteriograms (group B, patients 7–14).

Summary The morphologic characteristics at coronary arteriography of systolic narrowing of the left anterior descending coronary artery (LAD) were evaluated in 14 patients. Six patients had systolic narrowing of the LAD not associated with other cardiovascular abnormalities (group A) and eight patients had systolic narrowing of the LAD associated with hypertrophic cardiomyopathy (group B). Patients in group A showed a smooth and progressive constriction of the vessel up to the point of maximal stenosis, giving it a “rat-tail” appearance. There was no systolic narrowing of septal branches or of other epicardial vessels in this group. In patients of group B, systolic narrowing of the LAD had a “saw-fish” appearance. Seven patients had systolic narrowing of the septal branches, and five had systolic narrowing of other epicardial vessels. These data indicate that systolic narrowing of the LAD in patients with hypertrophic cardiomyopathy differs angiographically from systolic narrowing due to an intramural course of a part of the vessel (as in group A patients). We postulate that in patients with hypertrophic cardiomyopathy, fiber hypertrophy and disarray in the vicinity of the coronary vessels is responsible for the morphology and the widespread distribution of systolic narrowing.

Hypertrophic cardiomyopathy was diagnosed from the characteristic echocardiogram, left ventriculogram and an intraventricular pressure gradient at rest or after provocation by induced extrasystoles. In one patient with characteristic electrocardiographic and angiographic findings of hypertrophic cardiomyopathy and no gradient at rest, provocation was not attempted. In one patient, an intraventricular gradient was not demonstrated even after provocation. In this patient, hypertrophic cardiomyopathy was diagnosed at autopsy. The severity of systolic narrowing of the LAD was arbitrarily graded according to the severity of narrowing observed in grade 3 (greater than 75% systolic narrowing), grade 2 (50–75%) and grade 1 (less than 50%). The morphology of the LAD during systolic narrowing was evaluated in all available projections, including angulated craniocaudal and caudocranial views, and the findings in the two groups were compared. Associated systolic narrowing of other epicardial vessels was also analyzed. The severity of systolic narrowing of the septal perforator was arbitrarily graded as grade 3 (systolic disappearance of the entire septal artery), grade 2 (systolic disappearance of the lower two-thirds) or grade 1 (systolic disappearance of the lower third).

Results

There were 13 males and one female. The six patients in group A were 25–65 years old and the eight patients in group B were 15–68 years old.

Systolic Narrowing of the LAD and Other Coronary Arteries

In group A, two patients had grade 2 systolic narrowing of the LAD and four had grade 3 systolic narrowing. In all six patients, systolic narrowing of the LAD occurred in its middle third (fig. 1). No patient in group A had systolic narrowing of the septal perforator or other epicardial branches.

In group B, three patients had grade 1 systolic nar-
“SAWFISH” SYSTOLIC NARROWING OF THE LAD/Brugada et al.

Morphologic Aspect of Systolic Narrowing of the LAD

The morphologic aspect of systolic narrowing of the left anterior descending coronary artery (LAD) is shown in Figure 1. Patient 5 had high-grade systolic narrowing of the LAD, with proximal narrowing and a "rat-tail" appearance. The vessel proximal to the stenosis has a "rat-tail" appearance.

FIGURE 1. Patient 5. (A) Diastolic and (B) systolic left coronary arteriograms are shown in the craniocaudal left anterior oblique projection. Systolic compression of the left anterior descending coronary artery develops progressively and smoothly. The vessel proximal to the stenosis has a "rat-tail" appearance.

Narrowing of the middle third of the LAD, two patients had grade 2 narrowing and three patients had grade 3 narrowing. The septal perforator was narrowed in seven patients (figs. 2-4) and narrowing was absent in one patient (fig. 5). Three of the seven patients had grade 2, two grade 1 and two grade 3 septal perforator narrowing. In these patients, there was no apparent relation between the degree of LAD systolic narrowing and the degree of septal perforator compression with the grading systems used. Patient 7 had systolic narrowing of the diagonal, intermediate and marginal branches, the circumflex artery and the distal third of the right coronary artery (fig. 2). Patient 8 had systolic narrowing of the second diagonal branch, patient 10 of a marginal branch, patient 13 (fig. 3) of a second diagonal, intermediate branch and right coronary artery, and patient 14 (fig. 4) of the marginal and first diagonal branches.

FIGURE 2. Patient 7. Left coronary arteriograms in the right anterior oblique projection. (A) Diastolic and (B) systolic frames are shown. Note widespread high-grade systolic narrowing of the coronary arteries, distal disappearance of all small branches and systolic narrowing of the septal perforator branches, left anterior descending coronary artery and the circumflex and marginal branches.
described the angiographic appearance of this anomaly in 1960. \(^1\) Since then, systolic compression of the LAD on coronary angiograms has been considered the result of an intramural course of the constricted segment. This thesis has been confirmed in patients undergoing surgical debridging of the intramural LAD to alleviate myocardial ischemia. \(^8\)-\(^11\)

The results of our study do not show whether the systolic narrowing of the LAD in patients with hypertrophic cardiomyopathy and that in patients with an intramural LAD and without hypertrophic cardiomyopathy have a different pathologic basis. Our results do show, however, that systolic narrowing of the LAD is morphologically different in patients with and in patients without hypertrophic cardiomyopathy. In patients with hypertrophic cardiomyopathy, systolic compression of the vessel had an irregular aspect, giving it a "saw-fish" appearance (figs. 3-5). Narrowing occurred in both the middle and distal parts of the vessel along a long segment. Furthermore, in patients with hypertrophic cardiomyopathy, systolic narrowing of the LAD was associated with widespread systolic narrowing.

**Discussion**

The intramural course of part of the LAD at autopsy was first described by Crainianu in 1922. \(^7\) Porstmann
compression of the septal and of other epicardial vessels (figs. 2-4). We have not observed the "saw-fish" systolic narrowing of the LAD in any patient with severe hypertrophy of the heart due to valvular (n = 30) or supravalvular aortic stenosis (n = 1).

Systolic narrowing of the LAD was present in one patient with hypertrophic cardiomyopathy in the patients studied by Noble et al.1 Two of the 13 patients with hypertrophic cardiomyopathy and septal perforator compression reported by Pichard et al.12 had systolic narrowing of the LAD. Possible morphologic differences in the systolic narrowing of the LAD were not discussed in this article.

The characteristic histologic finding in hypertrophic cardiomyopathy is disorganized arrangements of hypertrophied muscular fibers. Cellular disorganization is widely distributed in the ventricular free wall.13 The angiographic aspect of the systolic narrowing of the LAD in patients with hypertrophic cardiomyopathy, and the associated systolic narrowing of the septal tree and other epicardial branches, suggest that the disorganized hypertrophy present in this disease might play a role in the type of systolic narrowing observed. The indentations during systolic narrowing of the vessel might result from the effect of the contracting hypertrophied and disorganized muscular fibers in the vicinity of the coronary arteries, and not from an intramural course of the vessels.

We realize the limitations imposed by the small number of patients in our series, but we believe that this postulate deserves further evaluation not only in larger angiographic series, but also by observations from the pathologist and surgeon in patients with hypertrophic cardiomyopathy and systolic narrowing of the LAD.

References
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Circulation. 1982;66:800-803
doi: 10.1161/01.CIR.66.4.800

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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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:
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