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POSTMORTEM STUDIES

Acute Takeoffs of the Coronary Arteries Along the Aortic Wall and Congenital Coronary Ostial Valve-Like Ridges: Association With Sudden Death

RENU VIRMANI, MD,* PATRICK K. C. CHUN, MD, FACC,† ROBERT E. GOLDSTEIN, MD, FACC,‡ MAX ROBINOWITZ, MD, FACC,§ HUGH A. McALLISTER, MD, FACC§

Washington, D.C. and Nashville, Tennessee

Congenital coronary artery anomalies have been associated with sudden death. Twenty-two patients who were victims of sudden death (mean age 46) and who had no significant anatomic cause of death were examined at autopsy and compared with 19 patients who died of known causes (control group). The hearts of these 41 patients were examined for abnormalities of acute angle takeoff of the coronary artery and presence of ostial valve-like ridges. Of the 22 patients who died suddenly, 13 (59%) had acute angle takeoff of the coronary artery and 9 (41%) had ostial valve-like ridges. Of the 19 control subjects, 4 (21%) had acute angle takeoff and only 2 (11%) had an ostial valve-like ridge. The difference was statistically significant ($p = 0.015$ and 0.031, respectively).

It is suggested that aortic root dilation may compress coronary arteries with acute angle takeoff and that ostial valve-like ridges may act as occlusion valves. Thus, either may cause acute obstruction of the proximal coronary artery and lead to sudden death. A very lethal combination for sudden death would be the presence of severe coronary artery disease, an acute angle takeoff and an ostial valve-like ridge.

Congenitally abnormal coronary arteries have been associated with sudden death (1–4). Cheitlin et al. (1) were the first to report a large series of patients with coronary arteries arising either as a single or double vessel from the same sinus of Valsalva; sudden death occurred only in patients in whom both coronary arteries arose from the right sinus of Valsalva. Cheitlin et al. postulated that the flap-like closure of the ostium of the coronary artery occurred with expansion of the aorta as well as with pull on the left coronary artery. Recently, Roberts et al. (5) reported clinical and autopsy findings in patients in whom origin of the right coronary artery from the left coronary sinus caused sudden death. They postulated that diminished flow into the anomalous right coronary artery may cause sudden death owing to a slit-like orifice that is further accentuated with dilation of the aorta.

We examined the hearts from 22 patients who died suddenly and compared the findings with data on 19 control subjects who died of known causes. The hearts of these 41 patients were examined for abnormalities of coronary artery takeoff in relation to the aortic wall and for the presence of coronary ostial valve-like ridges. We postulate that aortic dilation may compress coronary arteries with acute angle takeoff and that ostial valve-like ridges may act as occlusion valves, thus predisposing to sudden death.

Methods

Patients. *Study group.* We studied retrospectively the hearts of patients with sudden death from 1980 to 1981. All hearts had been submitted from medical examiners and fulfilled the following criteria: 1) occurrence of death within 24 hours in a previously healthy patient, and performance of autopsy within 24 hours after death; 2) absence of "noncardiac" cause of death; that is, no other disease process

From the §Armed Forces Institute of Pathology, Washington, D.C.; ‡Uniformed Services University of the Health Sciences, Bethesda, Maryland; †Tripler Army Medical Center, Honolulu, Hawaii; and *Vanderbilt University Medical Center, Veterans Administration Medical Center, Nashville, Tennessee. Manuscript received August 23, 1983; revised manuscript received October 4, 1983, accepted October 7, 1983.

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Address for reprints: Renu Virmani, MD, Department of Pathology, Vanderbilt University Medical Center, Veterans Administration Medical Center, Nashville, Tennessee 37232.

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could reasonably have been expected to cause death; 3) no death from unnatural causes, that is, accident, suicide or homicide; 4) absence of acute myocardial infarction; 5) no acute coronary occlusion (thrombosis); 6) no in-hospital death; and 7) no documented prior coronary heart disease. A total of 22 patients fulfilled these criteria. These 22 patients were further classified into two groups: 1) 11 patients with severe coronary atherosclerosis (defined as narrowing of one or more of the four major epicardial coronary arteries [right, left main, left anterior descending and left circumflex] of 75% or more in cross-sectional area by atherosclerotic plaque), and 2) 11 patients without severe coronary atherosclerosis.

Control subjects. The 22 patients were compared with 19 control subjects who had died suddenly. Fifteen of the control subjects had either an accidental death or had committed suicide (noncardiac death) and four died in the hospital with known coronary artery disease; that is, they had a documented acute myocardial infarction.

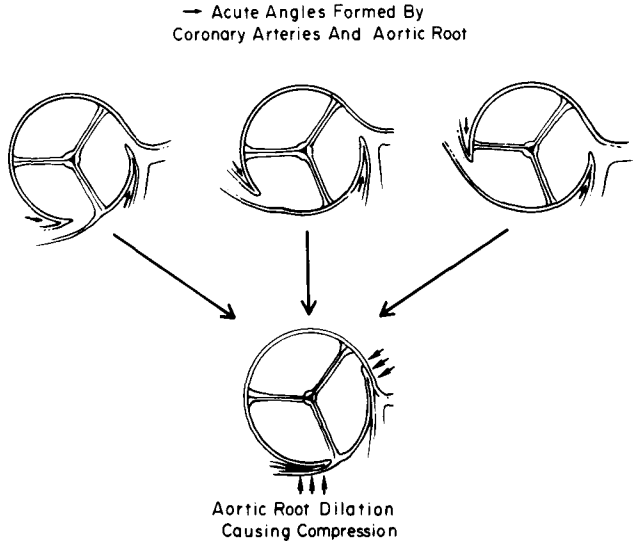
Examination of the coronary artery takeoff and coronary ostial valve-like ridges. The hearts of all patients and control subjects included in the study were examined in a similar manner. The aortic valve was exposed by removing the ascending aorta at the sinotubular junction, and the right and left epicardial coronary arteries were dissected to define the angulation of the first 1.5 cm of the coronary artery with the aortic wall (Fig. 1 and 2). The presence or absence of coronary ostial valve-like ridges at the coronary ostia was also evaluated (Fig. 3 and 4). Each heart was photographed (Fig. 2 and 4) and then three of us examined and evaluated the photographs separately for the presence or absence of acute angle takeoffs and ostial valve-like ridges. All evaluators were unaware of the patients' autopsy findings and their clinical course. The findings of these observers were then collated.

Definition of acute angle takeoff and ostial valve-like ridge. A coronary artery was considered to have an acute angle takeoff if the angle between the proximal coronary artery and the aortic wall was less than 45°. An ostial valve-like ridge was considered to be present and significant enough to contribute to sudden death if the surface area of the ridge exceeded 50% of the coronary ostial luminal area. Each observer was required to make a "yes" or "no" decision as to the presence or absence of an acute angle takeoff, or ostial valve-like ridge, or both.

Examination of coronary arteries and myocardium. The four major coronary arteries in all patients and control subjects were examined in a similar manner. The epicardial coronary arteries were cut at 5 mm intervals perpendicular to the long axis and examined for the degree of severe coronary atherosclerosis, that is, greater than 75% cross-sectional area luminal narrowing by an atherosclerotic plaque. The coronary arteries were examined for the type of dominance, that is, origin of the posterior descending coronary artery from the right coronary artery (right dominance) or left coronary artery (left dominance) or from neither or both arteries (co-dominance) (6).

The right and left ventricular myocardium were sliced at 1 to 1.5 cm intervals parallel to the posterior atrioventricular groove, such that at least four to five ventricular slices were available for examination for the presence or absence of acute or healed myocardial infarction. Myocardial infarction was defined as any pale area of scarring or necrosis greater than 3 cm in any direction (7).

Figure 1. Diagram showing acute angles (**small arrows**) between the aortic wall and right and left main coronary arteries at different position of origin of the coronary arteries (**upper panel**). **Lower panel** shows suggested mechanism of ostial closure with aortic root dilation.



Results

Clinical and pathologic features. There were 22 patients in the sudden death group, ranging in age from 20 to 79 years (mean 44); 16 were men and 6 were women (Table 1). Of these 22 patients, 11 had severe coronary atherosclerosis (one or more of the four major coronary arteries narrowed more than 75% in cross-sectional area by atherosclerotic plaque) and 11 did not have severe atherosclerosis. These 22 patients with sudden death were compared with 19 control subjects whose ages ranged from 19 to 81 years (mean of 56); 15 were men and 4 were women. Fifteen control subjects died of noncardiac causes such as accidents or suicide, and four had known coronary artery disease and died in the hospital (Table 1).

Table 2 summarizes the findings of heart weight, healed myocardial infarction and the number of patients with severe narrowing of one or more coronary arteries. Of the 11 patients with sudden death and no coronary artery disease, 1 (9%) had a healed myocardial infarct compared with 7 (64%)

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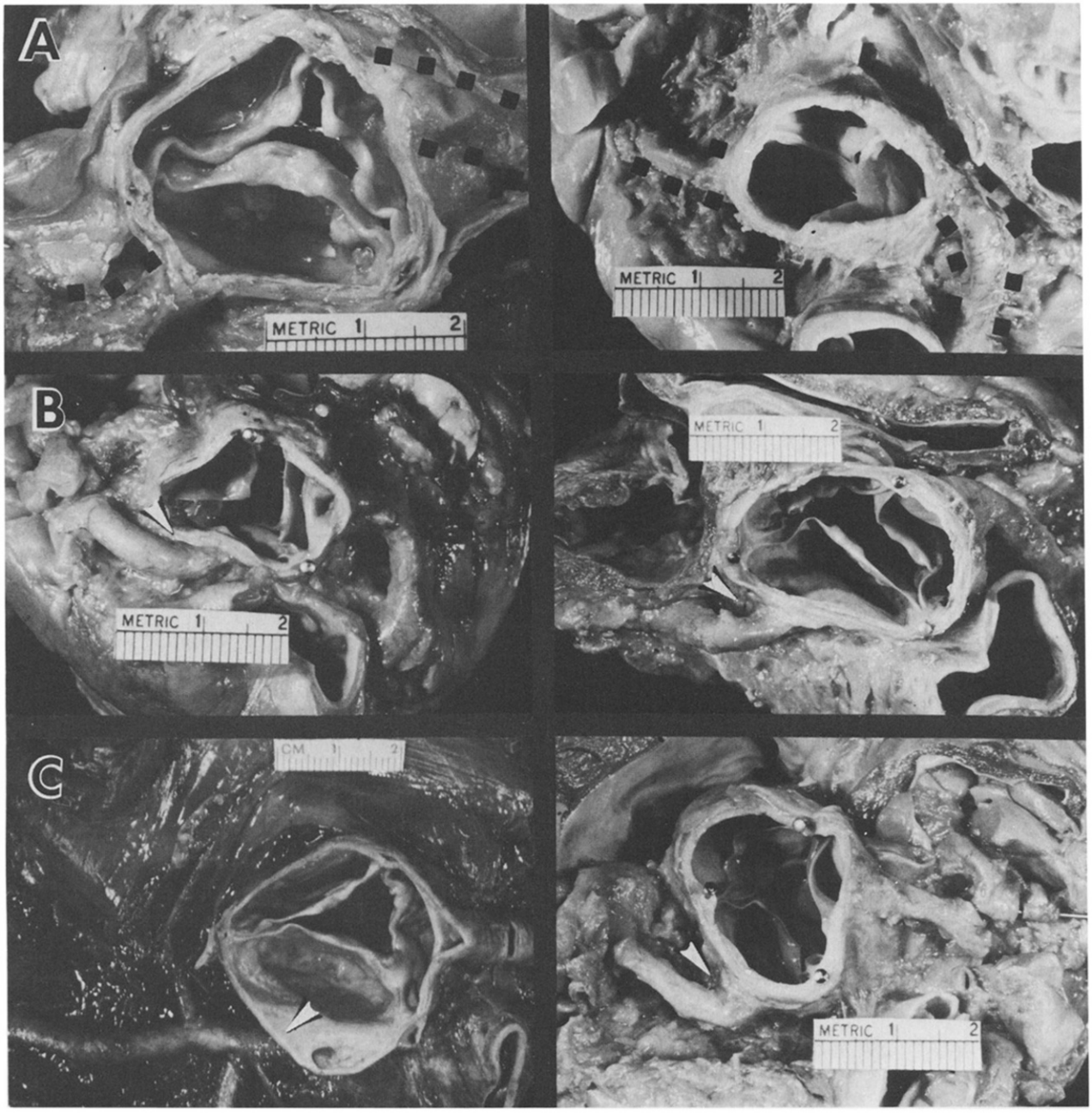
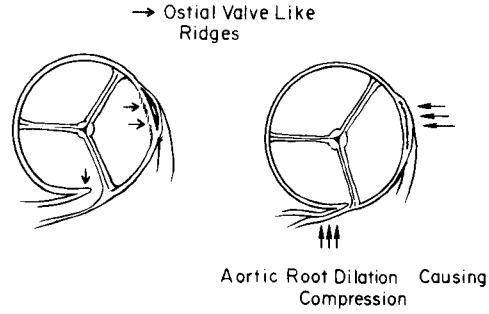


Figure 2. A, Normal coronary artery takeoff from the aortic root. Note that the coronary arteries do not form an acute angle with the aortic wall. **B** and **C,** The aorta in four patients who died suddenly with acute takeoff of the right coronary artery along the aortic wall (**arrows**).

of those with severe coronary atherosclerosis and sudden death. Only 1 (6%) of the 15 control subjects with a non-cardiac cause of death, had a healed myocardial infarct compared with all 4 control subjects with severe coronary artery disease (Table 2).

Extent of coronary artery atherosclerosis. Nine (81%) of the 11 patients with severe coronary atherosclerosis and

Figure 3. Diagram illustrating ostial valve-like ridges (**left**) and the proposed mechanism of ostial valve-like ridges causing ostial compression with aortic root dilation (**right**).



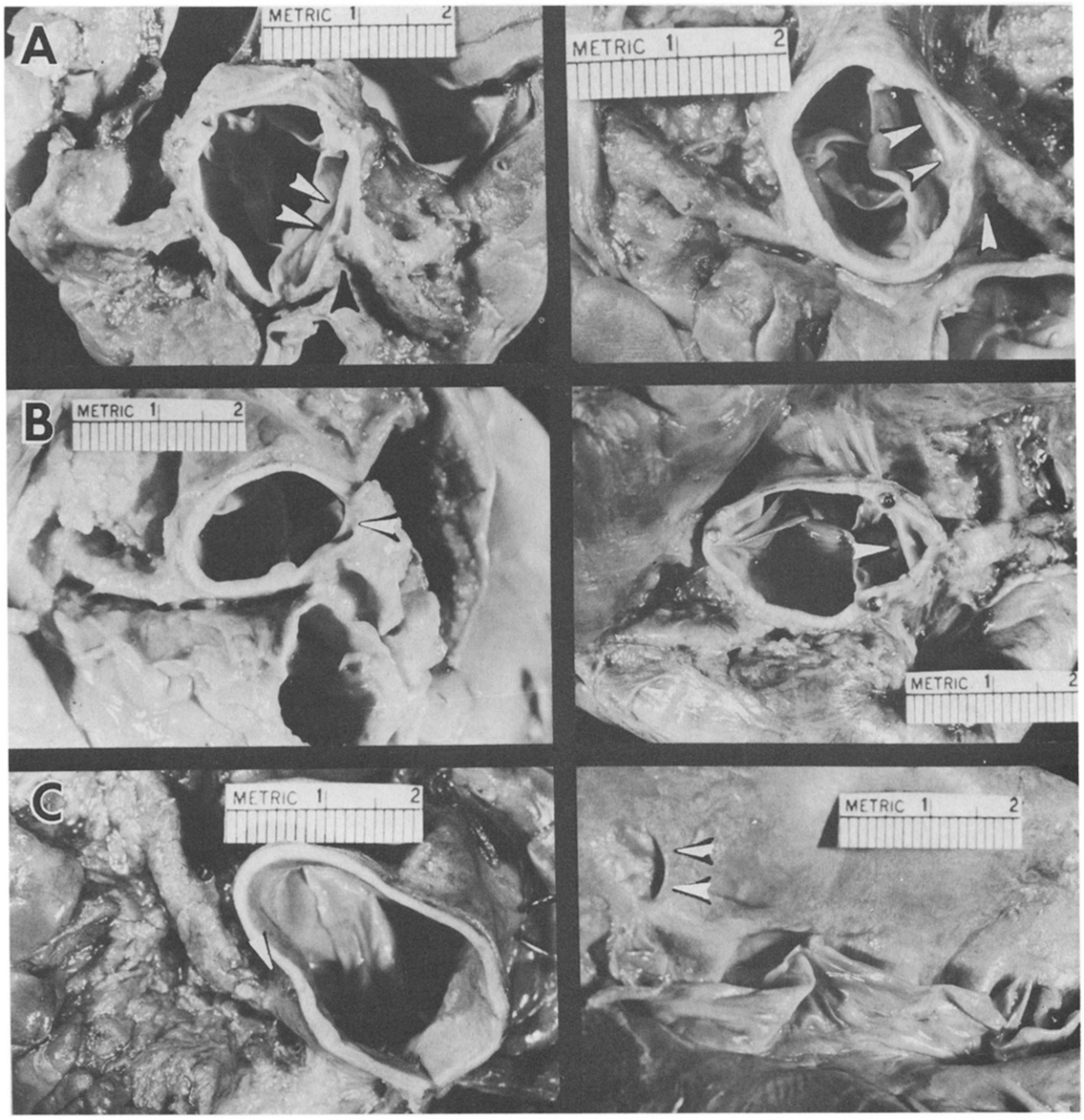


Figure 4. A and **B,** Ostial valve-like ridges (**arrows**) at the origin of the left main (LM) coronary artery in four patients who died suddenly. **C,** Acute angle takeoff of the right coronary artery (**arrow**) on the **left**; on the **right** is the opened aortic valve with the right coronary ostium (**arrows**) appearing slit-like because of the presence of crescent-shaped ostial valve-like ridge.

sudden death had two or three coronary arteries narrowed more than 75% in cross-sectional area (Table 3), as did all 4 control subjects with coronary artery disease. The type of coronary dominance was similar in the patients with sudden death and the control subjects (Table 3).

Prevalence of acute angle takeoff. Acute angle takeoff of coronary arteries along the aortic wall was evaluated in

Table 1. Number, Age and Sex of Patients Who Died Suddenly and Control Subjects

Patient Group	No.	Age Range (mean) (yr)	Sex M:F
Sudden death	22	20–79(44)	16:6
No CAD	11	20–70(37)	6:5
CAD	11	39–71(52)	10:1
Control subjects	19	19–81(56)	15:4
Noncardiac death	15	19–81(54)	12:3
CAD	4	60–74(64)	3:1

CAD = coronary artery disease; F = female; No. = number of patients; M = male.

Table 2. Heart Weight and Presence or Absence of Myocardial Infarction and Severe Coronary Atherosclerosis

Patient Group	No.	Heart Weight Range (mean) (g)	Healed MI	Patients With Severe CAD*
Sudden death	22	170–600(386)	8(36%)	11(50%)
No CAD	11	170–540(342)	1(9%)	0
CAD	11	240–600(430)	7(64%)	11(100%)
Control subjects	19	300–625(429)	5(26%)	7(37%)
Noncardiac death	15	300–500(405)	1(6%)	3(13%)
CAD	4	390–625(516)	4(100%)	4(100%)

*Greater than 75% cross-sectional narrowing of one or more coronary arteries. CAD = coronary artery disease; m = mean; MI = myocardial infarction; no. = number of patients.

each group (Table 4, Fig. 1 and 2) by three observers. Of the 22 patients who died suddenly, 13 (59%) had acute angle takeoff of either the right or both the right and the left coronary artery. Of these 13 patients, 6 had no coronary artery disease and no other cause of death and 7 had coronary artery disease but had died suddenly of no known prior heart disease. Ten of the 13 patients had acute angle takeoff of the right coronary artery and only 3 had right and left acute angle takeoffs. Of the 19 control subjects, 4 (21%) had an acute angle takeoff of the right or left coronary artery. Three of these four had died of noncardiac causes and one had coronary artery disease. The differences in the two groups were statistically significant ($p = 0.015$) by Fisher's exact test. Interobserver differences occurred in assessment of acute angle takeoff in only two instances; at all other times there was a consensus among the three observers.

Prevalence of ostial valve-like ridges. Ostial valve-like ridges were present in 9 (41%) of 22 patients who died suddenly and of these, 5 had coronary artery disease (Table 4, Fig. 3 and 4). Of the 19 control subjects, only 2 (11%) had ostial valve-like ridges and both had died of noncardiac causes. Again, this difference, as evaluated by Fisher's exact test, was statistically significant ($p = 0.031$). There were no differences among the three observers in assessment of presence of ostial valve-like ridges.

Discussion

Previous reports on congenital coronary artery anomalies. Congenital origin of both coronary arteries from either the right or the left coronary artery as a cause of sudden death has been well accepted (1–5). However, these anomalies do not always cause sudden death. In the study of Cheitlin et al. (1), of 51 patients whose coronary arteries arose either as a single or a double vessel from the same sinus of Valsalva, sudden death occurred in only 9 patients (18%) and in all of these, the left coronary artery arose from the right sinus of Valsalva. Of 10 patients, examined by Roberts et al. (5), whose right coronary artery arose from the left sinus of Valsalva, 3 (30%) died suddenly. Thus, the congenital coronary artery anomaly of either the right or the left coronary artery arising as single or double vessels from the same sinus of Valsalva is not uniformly fatal; this difference is not well understood. Both these studies postulate that acute left or rightward passage of the coronary artery along the aortic wall causes the ostium to be slit-like and that aortic and pulmonary trunk distension may further compress the coronary artery at its point of takeoff.

Proposed mechanism of cardiac dysfunction. We propose that the coronary artery forms an acute angle at its origin with the aortic wall as it follows the contour of the aorta (Fig. 1) and that the ostial valve-like ridge functions so as to compress the ridge against the coronary artery wall (Fig. 3) as the aortic root dilates. In addition, it is possible that diminished flow occurs into the abnormally angled coronary artery and that the ostial valve-like ridge also impedes flow into the coronary artery by obstructing the ostium.

When both coronary arteries arise from their respective sinuses of Valsalva, these arteries can still form an acute angle with the aortic wall. This is especially true of the

Table 3. Number and Percent of Coronary Arteries With Severe Disease*

Group	No.	Arteries With Severe Disease				Coronary Artery Dominance		
		0	1	2	3	R	L	Co
Sudden death	22	11(50%)	2(9%)	5(23%)	4(18%)	18(82%)	3(14%)	1(4%)
No CAD	11	11(100%)	0	0	0	9	1	1
CAD	11	0	2(18%)	5(45%)	4(36%)	9	2	0
Control subjects	19	12(63%)	1(5%)	5(26%)	1(5%)	15(79%)	3(16%)	1(5%)
Noncardiac death	15	12(80%)	1(7%)	2(13%)	0	12	2	1
CAD	4	0	0	3(75%)	1(25%)	3	1	0

*Greater than 75% narrowing of cross-sectional area. Co = codominant; L = left; No. = number of patients; R = right.

Table 4. Prevalence of Acute Takeoff of Coronary Arteries and Congenital Coronary Ostial Valve-like Ridges

Patient Group	No.	CA Acute Angle Takeoff				Ostial Valve-like Ridges		
		R or L	R	L	R + L	R or L	R	L
Sudden death	22	13(59%)*	10(45%)	0	3(14%)	9(41%)*	1(5%)	8(36%)
No CAD	11	6(55%)	5	0	1	5(45%)	0	5
CAD	11	7(64%)	5	0	2	4(36%)	1	3
Control subjects	19	4(21%)*	2(11%)	2(11%)	0	2(11%)†	1(5%)	1(5%)
Noncardiac deaths	15	3(20%)	1	2	0	2	1	1
CAD	4	1(25%)	1	0	0	0	0	0

*p = 0.015 for patients with sudden death compared with control subjects; †p = 0.035 for patients with sudden death compared with control subjects by Fisher's exact test. CA = coronary artery; CAD = coronary artery disease; L = left; No. = number of patients; R = right.

right coronary artery. Of the 41 patients whose hearts was examined for the presence of acute takeoff of the coronary artery, this anomaly was observed in 17 patients, with the incidence being higher in patients who died suddenly (13 [59%]) than in control subjects (4 [21%]). Also, of the 13 patients who died suddenly 6 had none of the four major coronary arteries severely narrowed by atherosclerotic plaque and 7 had severe coronary atherosclerosis. The 11 patients with severe coronary artery disease had not been symptomatic before the manifestation of sudden death, yet at autopsy, 8 (36%) had a healed myocardial infarct. Healed infarcts occurred much more frequently in patients with severe coronary atherosclerosis. Therefore, the combination of an acute angle takeoff and severe coronary atherosclerosis was fatal in seven of our patients with sudden death.

We examined hearts not only for the presence of acute angle takeoff of the coronary arteries but also for ostial valve-like ridges. Ostial valve-like ridges are created by the aortic wall and the coronary artery wall as they meet and form the coronary ostial orifice (Fig. 3 and 4). The presence of ostial valve-like ridges was noted more frequently in patients who died suddenly (9 [41%]) than in control subjects (2 [11%]). The distribution of ostial valve-like ridges was similar in patients who died suddenly with and without severe coronary atherosclerosis. However, in those who had severe coronary artery atherosclerosis, the concurrent finding of ostial valve-like ridges was fatal.

Importance of the angle of takeoff of coronary arteries and ostial valve-like ridge. From our study, as well as those of Roberts (5) and Cheitlin (1), and their coworkers, we conclude that it is important to study the angle of takeoff of the coronary arteries from the aorta and to note the presence or absence of ostial valve-like ridges. These findings are of concern not only when there is anomalous origin of the coronary arteries from the sinus of Valsalva, but also when the arteries arise from their normal sinuses of Valsalva. These relations may be a triggering mechanism of angina and sudden death.

It has been well documented that 5 to 10% of patients with angina or myocardial infarction do not have severe coronary atherosclerosis (8). Coronary artery spasm has been shown to be the cause of Prinzmetal's variant angina pectoris in patients with and without coronary atherosclerosis (9). There is also evidence (10,11) to suggest that coronary artery spasm with or without coronary atherosclerosis may contribute to the development of unstable angina pectoris, acute myocardial infarction and sudden death. The role of acute angle takeoffs and ostial valve-like ridges in coronary artery spasm and sudden death should also be considered. Confirmation of these findings may be possible with two-dimensional echocardiography, which permits visualization of coronary artery takeoffs at their origin from the aorta, but the right coronary artery is more difficult to visualize than the left.

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