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Sudden Cardiac Death Associated With Isolated Congenital Coronary Artery Anomalies

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Objectives and Background. Congenital coronary anomalies are associated with sudden death and exercise-related death. Clarification of the risk and mechanisms of sudden death in patients with coronary anomalies may aid in decisions on intervention.

Methods. The clinicopathologic records of 242 patients with isolated coronary artery anomalies were reviewed for information on mode of death and abnormalities of the initial segment (acute angle takeoff, valvelike ridges or aortic intramural segments) and course of the anomalous coronary artery.

Results. Cardiac death occurred in 142 patients (59%); 78 (32%) of these deaths occurred suddenly. Of sudden deaths, 45% occurred with exercise. Sudden death (28 of 49, 57%) and exercise-related death (18 of 28, 64%) were most common with origin of the left main coronary artery from the right coronary sinus. Anomalous origin of the right coronary artery from the left coronary sinus was also commonly associated with exercise-related sudden death (6 of 13 sudden deaths, 46%). High risk anatomy involved abnormalities of the initial coronary artery segment or coursing of the anomalous artery between the pulmonary artery and aorta. Younger patients (≤30 years old) were significantly more likely than older patients (≥30 years old) to die suddenly (62% vs. 12%, $p = 0.0001$) or during exercise (40% vs. 2%, $p = 0.00001$) despite their low frequency of significant atherosclerotic coronary artery disease (1% vs. 40%, $p = 0.00001$).

Conclusions. Younger patients (≤30 years old) with an isolated coronary artery anomaly are at risk of dying suddenly and with exercise. Therefore, greater effort for early detection and surgical repair of these lesions is warranted.

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Coronary artery anomalies are found in approximately 1% of all patients undergoing coronary angiography (1,2) and in approximately 0.3% of patients undergoing autopsy (3). Although coronary anomalies can be an incidental autopsy finding, they are also a well known cause of sudden death (4–10) and are found relatively more frequently in young persons who die during exercise (11). Identification of high risk subsets of anomalies may aid clinicians in decisions on intervention.

The objective of the present study was to determine the frequency and mechanisms of sudden cardiac death in patients with a major congenital coronary artery anomaly not associated with a congenital anomaly of the heart and great vessels. We report the mode of sudden cardiac death and the associated clinical and pathologic findings in 242 patients with isolated congenital coronary anomalies at the Armed Forces Institute of Pathology.

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Methods

Study cases. We reviewed the clinicopathologic records of patients with coronary anomalies referred to the Armed Forces Institute of Pathology between 1967 and 1991. The vast majority of cases were referred from military health care facilities and the patients included active duty military personnel, military retirees and dependents. A fair number were submitted from civilian hospitals and medical examiners. The cases were electively referred on the basis of the pathologic observations from all sources. When available, gross pathologic specimens were reviewed to confirm reported findings.

Two hundred fifty-six patients with coronary anomalies in the absence of congenital anomalies of the heart and great vessels were identified. The coronary anomalies that we included in this analysis are listed in Table 1. Additional anomalies that we observed included left anterior descending and left circumflex coronary arteries from separate ostia (12 cases; this number is underrepresented because most pathologists consider this condition to be a normal variant rather than an anomaly) and accessory coronary artery from the pulmonary trunk (two cases; this anomaly is often overlooked by pathologists). Both of these anomalies have no functional significance. The information uniformly obtained in each case included the clinical presentation; symptoms or signs (angina pectoris, syncope, dyspnea, myocardial infarction, congestive heart failure or arrhythmias, when

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Table 1. Classification of Coronary Artery Anomalies

I. Anomalous origin of ≥1 CA from pulmonary trunk
  A. LMCA or LAD from pulmonary trunk
  B. Both CAs from pulmonary trunk
  C. RCA from pulmonary trunk
II. Anomalous origin of ≥1 CA from aorta
  A. LMCA and RCA from R Ao sinus
  B. LMCA and RCA from L Ao sinus
  C. Anomalous origin of LCx from R Ao sinus or RCA
  D. Anomalous origin of CAs from posterior Ao sinus
  E. RCA and LAD from R Ao sinus
III. Single CA ostium from Ao
  A. Single RCA ostium
  B. Single LCA ostium
IV. Congenital hypoplastic CAs
V. CA fistula

Ao = aorta or aortic; CA = coronary artery; L = left; LAD = left anterior descending coronary artery; LCA = left coronary artery; LCx = left circumflex coronary artery; LMCA = left main coronary artery; R = right; RCA = right coronary artery.

present); mode of death; time of death; the origin and course of the anomalous coronary artery, and the presence of atherosclerotic coronary artery disease, myocardial fibrosis or necrosis.

Definitions. Sudden cardiac death was defined as 1) the occurrence of death within 6 h of the onset of symptoms in a previously healthy patient; 2) the absence of other disease processes that could have caused death; and 3) the exclusion of death from accident, suicide or homicide. Cardiac death occurring >6 h after the onset of symptoms was classified as nonsudden cardiac death. Exercise-related death was defined as death occurring during exercise or within 1 h of engaging in a sport-related activity.

Pathologic analysis. The right and left epicardial coronary arteries were dissected to define the angulation of the first 1.5 cm of the coronary artery relative to the aortic wall. Acute angle takeoff was diagnosed if the angle between the proximal coronary artery and the aortic wall was <45°. The coronary ostia were evaluated for the presence of valvelike ridges. An ostial valvelike ridge was considered significant if the surface area of the ridge was >50% of the coronary ostial lumen area (12). Severe coronary atherosclerosis was defined as narrowing ≥75% in cross-sectional area of at least one of the four major epicardial coronary arteries (left main, left anterior descending, left circumflex and right). Hypoplastic coronary arteries were defined as diminutive coronary arteries insufficient to supply the posterior wall of the left ventricle. The ventricular myocardium was examined for evidence of acute or healed myocardial infarction. Myocardial infarction was defined as any pale area of scarring or necrosis >1 cm in any direction.

Coronary artery anomalies were classified according to a four-group descriptive classification (Table 1). This descriptive scheme is a modification of the classification proposed by Roberts (6) in 1986.

Statistical analysis. Statistical comparisons of the older (≥30 years) and younger (<30 years) patients were performed with use of Pearson's chi-square test. A p value < 0.05 was considered significant.

Results

Observed coronary artery anomalies. Table 2 shows the distribution, ages and gender of 242 autopsy patients with isolated coronary artery anomalies. Patients ranged in age from 1 day to 87 years, with a male gender predominance probably due to our military referral population. Origin of the right and left main coronary arteries from the left coronary sinus was the most common anomaly observed. The rates of sudden, nonsudden cardiac and exercise-related death are displayed in Table 3. Overall, cardiac death occurred in 142 (59%) of the 242 cases; sudden death occurred in 78 (32%). No diagnosis was made before death except in three cases of left main coronary artery arising from the pulmonary trunk.

Table 2. Distribution of Isolated Coronary Artery Anomalies in 242 Autopsy Cases

Anomaly	No.	Age Range (mean)	F/M
I. Anomalous origin of ≥1 CA from pulmonary trunk			
A. LMCA or LAD from pulmonary trunk	37	3 days to 17 yr (2)	13/24
B. Both CAs from pulmonary trunk	3	1 to 7 days (3 days)	1/2
C. RCA from pulmonary trunk	1	34 yr	0/1
II. Anomalous origin of ≥1 CA from aorta			
A. LMCA and RCA from R Ao sinus	49	2 to 87 yr (30)	5/44
B. RCA and LMCA from L Ao sinus	52	0 to 82 yr (39)	4/48
C. LCx and RCA from R Ao sinus	21	20 to 82 yr (49)	4/17
D. RCA and/or LMCA from posterior Ao sinus	17	1 to 67 yr (39)	3/14
E. RCA and LAD from R Ao sinus	1	20 yr	0/1
III. Single CA ostium from Ao	44	1 to 80 yr (47)	8/36
IV. Congenital hypoplastic CAs	13	1 to 69 yr (20)	2/11
V. CA fistula	4	1 day to 7 yr (2)	1/3
Total	242	1 day to 87 yr	41/201

F = female; M = male; other abbreviations as in Table 1.

Table 3. Mode of Cardiac Death in 242 Autopsy Cases

	All Deaths	Sudden Deaths (%)	NSCD (%)	Exercise* (%)
I. Anomalous origin of ≥1 CA from pulmonary trunk				
A. LMCA or LAD from pulmonary trunk	37	14 (38)	23 (62)	2 (14)
B. Both CAs from pulmonary trunk	3	3 (100)	0 (0)	0 (0)
C. RCA from pulmonary trunk	1	0 (0)	1 (100)	0 (0)
II. Anomalous origin of ≥1 CA from aorta				
A. LMCA and RCA from R Ao sinus	49	28 (57)	8 (16)	18 (64)
B. RCA and LMCA from L Ao sinus	52	13 (25)	2 (4)	6 (46)
C. LCx and RCA from R Ao sinus	21	2 (10)	6 (29)	0 (0)
D. RCA and/or LMCA from posterior Ao sinus	17	5 (29)	4 (24)	2 (40)
E. RCA and LAD from R Ao sinus	1	1 (100)	0 (0)	0 (0)
III. Single CA ostium from aorta				
A. Single RCA ostium	22	4 (18)	5 (23)	2 (50)
B. Single LCA ostium	22	2 (9)	8 (36)	1 (50)
IV. Hypoplastic CAs	13	5 (38)	4 (31)	3 (60)
V. CA fistula	4	1 (25)	3 (75)	0 (0)
Total	242	78 (32)	64 (26)	34 (44)

*Exercise-related sudden deaths expressed as percentage of sudden deaths. Patients not accounted for in totals died from noncardiac causes. NSCD = nonsudden cardiac death; other abbreviations as in Table 1.

Anomalous origin of coronary artery from the pulmonary trunk. When the left main coronary artery arises from the pulmonary trunk, the most common site of origin (95%) is the left pulmonary sinus. The wall of the left main coronary artery is veinlike, and extensive collateral vessels arise from the right coronary artery. The heart is enlarged and there is extensive scarring and thinning in the anterolateral wall of the left ventricle with marked scarring of the anterolateral papillary muscle (Fig. 1). Compensatory hypertrophy is observed in the posterobasal portion of the left ventricle. Endocardial fibrosis may be prominent in older children.

We observed this anomaly in 37 patients, with death occurring in 25 before the age of 1 year. Fourteen patients (38%) died suddenly, with exercise-related sudden death observed in 2 (14%) of these patients. All 37 patients died of cardiac causes. The diagnosis was made before death in only three cases, and all three patients died at the time of attempted surgical repair, at the age of 5 weeks in one case and 4 months in two cases.

We observed four cases of origin of both coronary arteries from the pulmonary trunk. All four patients were symptomatic with congestive heart failure and died suddenly within days of birth. We also observed one case in which origin of the right coronary artery from the pulmonary trunk was an incidental finding at autopsy.

Anomalous origin of one or more coronary arteries from the aorta. In 49 cases the left main coronary artery arose from the right coronary sinus or as a first branch of the right coronary artery. This anomaly was classified further according to the course of the left main coronary artery: 1) left main artery anterior to the pulmonary trunk (four patients), 2) left main artery between the aorta and pulmonary trunk (28 patients) (Fig. 2A), 3) left main artery posterior to the aorta (four patients) (Fig. 2B), and 4) left main artery posterior to the right ventricular outflow tract and within the ventricular septum (five patients). Of these 49 patients, 28 (57%) died suddenly and 18 (64%) died during or soon after exercise (Fig. 3). Sudden death is particularly common when the left coronary artery courses between the aorta and pulmonary trunk. Twenty-three (82%) of the 28 patients with this anomaly died suddenly.

Sudden death occurs less frequently with other anomalous origins of coronary arteries from the aorta (Table 3). Anomalous origin of the right coronary artery from the left coronary sinus was the second most frequently observed anomaly, with sudden death occurring in 13 (25%) of 52 cases (Fig. 4). Two patients with anomalous origin of the left circumflex artery from the right coronary sinus or right coronary artery died suddenly. In one, the death was believed to be due to severe coronary atherosclerosis; in the other, it was attributed to dilated cardiomyopathy with congestive heart failure.

Single coronary artery ostium. When compared with anomalous aortic origin of one or more coronary arteries, single coronary artery ostium was less frequently associated with sudden death (Table 3). Overall, sudden death occurred in 6 (14%) of 44 patients. Sudden cardiac death was more common when the single coronary artery originated in the right coronary sinus (18%) versus the left coronary sinus (9%).

Hypoplastic coronary arteries. Hypoplastic coronary arteries (the condition when neither the right nor the left circumflex coronary artery goes beyond the lateral borders of the heart) were diagnosed in 13 patients. Five patients (38%) died suddenly; three of these deaths were related to exercise. Cardiac death occurred in four other patients, as a result of coronary atherosclerosis in three and of aortic stenosis in one.

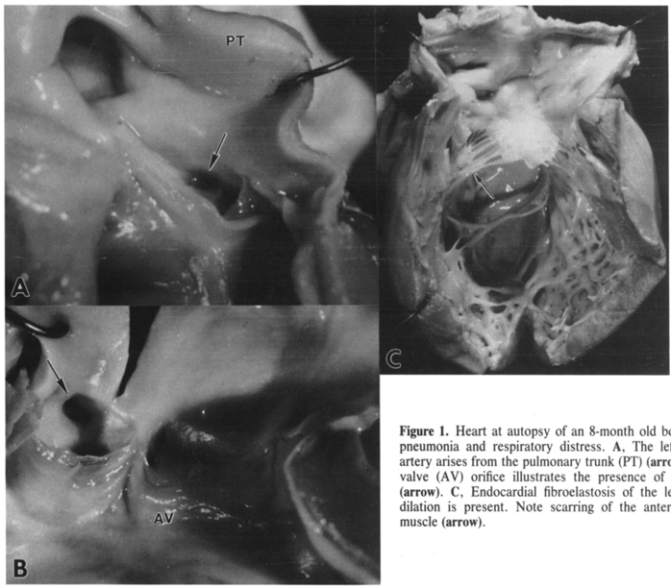

Figure 1. Heart at autopsy of an 8-month old boy who died with pneumonia and respiratory distress. **A**, The left main coronary artery arises from the pulmonary trunk (PT) (**arrow**). **B**, The aortic valve (AV) orifice illustrates the presence of only one ostium (**arrow**). **C**, Endocardial fibroelastosis of the left ventricle with dilation is present. Note scarring of the anterolateral papillary muscle (**arrow**).

Coronary artery fistula. A major coronary artery fistula that is capable of producing a significant shunt can be identified at autopsy, although smaller fistulas such as those commonly identified at cardiac catheterization in adults are likely to be missed. Sudden death can occur, as we observed in one 7-year old child who had acute pulmonary edema after general anesthesia.

Presence of acute angle takeoff and intramural coronary segments. Acute angle takeoff and intramural (within the aortic wall) coronary segments were observed only in anomalous origin of one or more coronary arteries from the aorta, particularly when the left coronary artery arose from a separate ostium in the right coronary sinus and vice versa. The anomalous artery most commonly traversed the area between the aorta and the pulmonary trunk (Table 4). In contrast, acute angle takeoff was not observed in cases involving a single coronary artery ostium (Table 4).

In patients with origin of the left or right coronary artery from the contralateral coronary sinus, all instances of sudden death involved the presence of an acute angle takeoff, an intramural coronary segment or coursing of the anomalous coronary artery between the pulmonary trunk and aorta. The risk was particularly great for origin of the left coronary artery from the right coronary sinus; 84% of such patients died suddenly when the anomalous artery coursed between the pulmonary trunk and aorta.

Sudden death during exercise. Exercise-related sudden death (Table 3) was seen most frequently when the left coronary artery arose from the right coronary sinus, followed by anomalous origin of the right coronary artery from the left coronary sinus. Single coronary ostium, anomalous origin of the left main artery from the right posterior sinus, left main artery arising from the pulmonary trunk and hypoplastic coronary arteries were infrequently seen and, when seen, were often associated with exercise-related sudden death (11% to 60%). In all cases exercise consisted of aerobic activity, such as running, marching, calisthenics, basketball and soccer.

Symptoms. Sudden death was frequently not heralded by the presence of symptoms or signs such as dyspnea, chest pain, congestive heart failure or syncope (Table 5). Of the 78 cases of sudden cardiac death, 62% occurred in asymptomatic patients. This rate was comparable to that of patients who did not die suddenly, of whom 68% were free of cardiac

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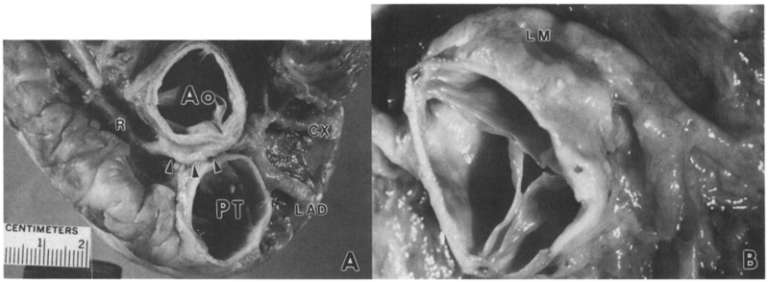

Figure 2. **A,** Superior view of the aorta (Ao) and pulmonary trunk (PT) with both coronary arteries arising from the right sinus. Note the left main coronary artery (**arrowheads**) coursing between the aorta and pulmonary trunk before dividing into the left anterior descending (LAD) and left circumflex (CX) arteries. R = right coronary artery. **B.** Superior view of the aorta with the left main (LM) and right coronary arteries arising from the right coronary sinus and the left main artery coursing posterior to the aorta before dividing into the left anterior descending and left circumflex arteries.

symptoms. In anomalies with the highest rate of sudden death (left or right coronary artery from the contralateral coronary sinus), 66% of patients were asymptomatic at the time of death. Similar rates of death with exercise and at rest were observed in symptomatic and asymptomatic patients.

In patients with origin of the left main coronary artery from the right coronary sinus, reported symptoms included angina (20%), congestive heart failure (14%), syncope (14%) and myocardial infarction (6%). In comparison, symptoms were less common in origin of the right coronary artery from the left coronary sinus and included angina (10%), congestive heart failure (8%) and myocardial infarction (2%).

Comparison of patients ≤30 and >30 years old. The majority of cases of origin of the left main artery from the right coronary sinus were observed in young patients; otherwise, anomalies were evenly distributed among patients ≤30 and >30 years old. (Patients with origin of the left main artery from the pulmonary trunk, both coronary arteries from the pulmonary trunk and coronary artery fistulas were not considered here, because all patients in these groups were ≤30 years old.) The frequency of symptoms in these two groups (≤30 and >30 years old) was similar (Table 6). However, patients ≤30 years old had higher rates of sudden death and exercise-related death, but a lower rate of atherosclerotic coronary artery disease. Atherosclerotic coronary artery disease was seen in only one patient ≤30 years old. This patient had origin of the right coronary artery from the left coronary sinus and died after acute myocardial infarction with left main coronary artery thrombosis.

Figure 3. View of left ventricular outflow tract and aorta showing left (L) and right (R) coronary ostia arising in the right coronary sinus of a 14-year old boy who died suddenly while playing soccer.

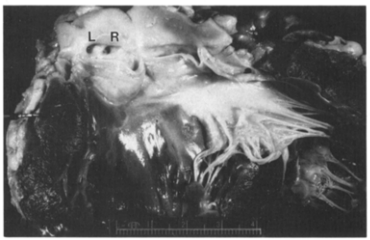

Figure 4. View of the left ventricular outflow tract and aorta showing left (L) and right (R) coronary arteries arising from the left coronary sinus in a 19-year old man who died during exercise.

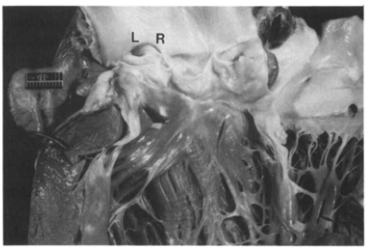

Table 4. Presence of Acute Angle Takeoff and Course of Anomalous Coronary Arteries in 242 Autopsy Cases

Anomaly	No.	Acute Angle	Within Ao Wall	Course of CA			
				A	B	P	I
LMCA and RCA from R Ao sinus	49	25	11	4	28	4	5
Single CA from R Ao sinus	22	0	0	4	7	7	3
Total	71	25	11	8	35	11	8
LMCA and RCA from L Ao sinus	52	16	5	0	24	11	0
Single CA from L Ao sinus	22	0	0	0	28	20	0
Total L Ao sinus	74	16	5	0	52	31	0

A = anterior; B = between aorta and pulmonary trunk; I = intraseptal; P = posterior to aorta; other abbreviations as in Table 1.

Discussion

Present study. Anomalies of coronary artery origin encompass a wide range of potential pathologic alterations that are matched by the variability in their clinical presentations (6). In their mildest form, the abnormalities are asymptomatic and without clinical consequence. For example, separate origin of the left circumflex coronary artery from the left coronary sinus is commonly identified at cardiac catheterization but is not associated with sudden death. Other coronary anomalies may be more frequently fatal, usually in the absence of warning symptoms.

Our series has demonstrated that anomalous origin of the left main coronary artery from the right coronary sinus was the abnormality most commonly associated with sudden death. Sudden death was most likely to occur in this anomaly when the anomalous left coronary artery coursed between the aorta and pulmonary trunk. Other anomalies that are important causes of sudden death include origin of the right coronary artery from the left sinus, origin of the right or left coronary artery, or both, from the posterior sinus, single coronary artery ostium and hypoplastic coronary arteries.

Mechanism of sudden death. Anomalous coronary arteries are believed to result in sudden death from several different mechanisms. Cheitlin et al. (4) first described the importance of a slitlike orifice in anomalous origin of the left coronary artery from the right coronary sinus. Of our 49 patients with the left main and right coronary arteries arising from the right aortic sinus, 25 had acute angle takeoff with valvelike ridges. The oblique takeoff of the anomalous artery causes a slitlike orifice in the aortic wall and is capable of collapsing in a valvelike manner, especially when the initial portion of the vessel remains within the wall of the aorta. Compression of the coronary artery occurs at the onset of diastole with aortic root distension. This takeoff compression may explain the increased frequency of sudden death when the left coronary artery courses between the aorta and the pulmonary trunk and originates from two ostia as opposed to a single ostium. It has been postulated that exer-

Table 5. Symptoms and Exercise-Related Sudden Death

	Sudden Deaths (no.)			
	Symptomatic		Asymptomatic	
	Ex	Rest	Ex	Rest
I. Anomalous origin of ≥1 CA from pulmonary trunk				
A. LMCA from pulmonary trunk	1	5	1	7
B. Both CAs from pulmonary trunk	0	3	0	0
C. RCA from pulmonary trunk	0	0	0	0
II. Anomalous origin of ≥1 CA from Ao				
A. LMCA and RCA from R Ao sinus	9	5	9	5
B. RCA and LMCA from L Ao sinus	0	0	6	7
C. LCx and RCA from R Ao sinus	0	1	0	1
D. RCA and/or LMCA from posterior Ao sinus	0	1	2	2
E. RCA and LAD from R Ao sinus	0	0	0	1
III. Single CA ostium from Ao				
A. Single RCA ostium	1	1	1	1
B. Single LCA ostium	0	0	1	1
IV. Congenital hypoplastic CAs	1	1	2	1
V. CA fistula	0	1	0	0
Total	12	18	22	26
Percent*	15	23	28	33

*Values do not total 100% because of rounding of decimal points. Ex = exercise; other abbreviations as in Table 1.

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Table 6. Symptoms and Mode of Death in Patients ≤30 and >30 Years Old (n = 198*)

	Age		
	≤30 Years	>30 Years	p Value
Patients	73 (37)	125 (63)	
Symptoms	15 (41)	38 (31)	0.12
ASCAD	1 (1)	50 (40)	0.00001
Sudden death†	45 (62)	15 (12)	0.00001
Exercise-related death	29 (40)	2 (2)	0.00001
Nonsudden cardiac death	4 (5)	34 (27)	0.0004
Noncardiac death	24 (33)	76 (61)	0.0001

*Patients with origin of the left main or left anterior descending coronary artery from the pulmonary trunk, patients with origin of both right and left coronary arteries from the pulmonary trunk and patients with a coronary artery fistula were excluded from this analysis because all patients in these groups were <30 years old. †Includes exercise-related deaths. All data are expressed as number and (percent) of patients. ASCAD = severe atherosclerotic coronary artery disease (≥75% cross-sectional area lumen narrowing).

cise-induced expansion of the pulmonary artery and aortic root may further compress the coronary lumen, leading to the propensity for exercise-related death (9). A similar mechanism may be responsible for sudden and exercise-related death in anomalous origin of the right coronary artery from the left coronary sinus. Abnormal initial coronary segments were observed in 16 of the 52 cases of the latter anomaly.

Not all patients with coronary anomalies and sudden death have abnormal coronary artery takeoff. Specifically, acute angle takeoff was not observed in patients with a single coronary artery ostium or in cases where the left coronary artery arose as a branch of the right coronary artery and coursed between the aorta and the pulmonary trunk. This finding suggests other mechanisms for sudden death, such as a coronary artery compression between the aorta and the pulmonary trunk or inadequate coronary flow reserve during exercise (9). Most sudden deaths related to coronary anomalies are probably the result of regional myocardial ischemia.

The relative importance of these mechanisms in sudden death is uncertain. All of the sudden deaths that we observed in association with origin of the left or right coronary artery from the contralateral coronary sinus occurred in the presence of either or both abnormal initial coronary segments or course of the anomalous coronary artery between the pulmonary artery and the aorta. An abnormal orifice and takeoff of the coronary artery may alone be sufficient to result in sudden death. Evidence for this comes from data on the importance of acute angle takeoff and ostial valvelike ridges in patients who die suddenly and do not have coronary anomalies (12).

Myocardial ischemia resulting from concomitant atherosclerotic coronary artery disease is another possible cause of sudden death. Severe atherosclerotic coronary artery disease was present almost exclusively in the older patients with coronary artery anomalies. Our data demonstrate that sudden death in young patients (≤30 years old) with coronary artery anomalies is most likely due to the coronary anomaly and not to severe atherosclerotic coronary artery disease. However, in older patients with coronary anomalies, the extent of atherosclerotic coronary artery disease may be accelerated by turbulent coronary blood flow as a result of an acute angle takeoff. High coronary flow rates in single coronary ostium and hypoplastic coronary arteries may also predispose to atherosclerotic coronary artery disease through endothelial injury.

Symptoms and sudden death. Although most patients were reported to have no symptoms, a significant number of patients (38%) were symptomatic before death. Although such symptoms may be a warning sign of underlying cardiac disease, they do not predict risk for sudden death. In fact, more sudden deaths occurred in asymptomatic than in symptomatic patients (62% vs. 38%).

Limitations. Our study is limited by its retrospective design. However, many patients with coronary artery anomalies are asymptomatic and unlikely to be identified prospectively. Therefore, controlled data on these subjects will be difficult to obtain. The number of patients reported to have cardiac symptoms or signs preceding death probably represents the lower limit of the true value because symptoms were likely underreported. Therefore, clinical conclusions based on such information should be carefully interpreted. Among the many causes of sudden death, it should be noted that coronary artery anomalies account for relatively few deaths. Highlighting this point are two reports, one from Israel (13) reporting a single instance of coronary artery anomaly among 162 cases of sudden death and another from our laboratory (11) with only 1.2% of sudden cardiac deaths in patients with an anomalous coronary artery (8 of 656 cases).

Conclusions. The management of patients with anomalous coronary artery origin, regardless of the presence of symptoms, may include surgical correction (6). For the clinician who identifies an anomaly at cardiac catheterization, data on those anomalies that are associated with an increased risk of sudden death can be an aid in determining whether to perform surgical revascularization. This is particularly important in cases of anomalous origin of the left coronary artery from the aorta, which carries a high risk of sudden death. Our recommendation is based on the potential for sudden death and the knowledge that death may not be heralded by symptoms.

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