A Systematic Overview of Anomalous Coronary Anatomy and an Examination of the Association With Sudden Cardiac Death

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Coronary artery anomalies are a frequently neglected topic in cardiology. However, they are an important cause of cardiac ischemia and sudden death. Anomalous pulmonary artery and anomalous aortic origins of the coronaries are 2 major types of coronary artery anomalies. This article will examine the occurrence of these anomalies in the absence of associated congenital defects. A review of the literature reveals that the anomaly most commonly associated with sudden death is a left main coronary artery arising from the right aortic sinus of Valsalva with an interarterial course. The increased risk of sudden death with this anomaly has been associated with any of 4 high-risk features: slit-like ostium, acute-angle take-off, intramural aortic segment, and interarterial course.

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Coronary artery anatomy is not constant. Variation in the number, origin, or course of coronary arteries is present in approximately 0.2% to 1.2% of the population.[1] Coronary anomalies in the young are a major cause of morbidity and mortality. Such anomalies cause approximately 12% of the deaths among American high school and college athletes.[2,3]

Normally, both coronary arteries arise from the aortic sinuses of Valsalva. The right coronary artery (RCA) arises from the right aortic sinus of Valsalva (RASV), and the left main coronary artery (LMCA) arises from the left aortic sinus of Valsalva (LASV). The LMCA divides almost immediately into the left anterior descending artery and the circumflex artery. The left anterior descending artery gives off branches that supply blood to the ventricular septum. The circumflex artery travels around the base of the left ventricle, terminating in a posterior descending branch.[4]

Anomalies are generally classified into 7 major categories developed by the Society of Thoracic Surgeons (Table 1): anomalous pulmonary artery (PA) origin of the coronaries, anomalous aortic origin of the coronaries, congenital atresia of the LMCA, coronary artery fistulas, coronary artery bridging, coronary aneurysms, and non-atherosclerotic coronary stenosis.[5] This article reviews anomalous pulmonary and anomalous aortic origins of the coronaries in the absence of associated congenital defects, with a focus on the lethal LMCA arising from the RASV with an interarterial course.

Anomalous PA Origin of the Coronaries

Anomalous Origin of the LMCA from the PA

Anomalous origin of the LMCA from the PA accounts for 90% of cases of anomalous PA origins.[6] This anomaly, first described by Abbott in 1908,[4] is present in approximately 1 in 300,000 live births.[5] The anomaly presents with a bimodal age distribution.[6] Approximately 80% of cases are recognized in infancy. Insufficient oxygen delivery to the papillary muscles and anteroseptal wall leads to congestive heart failure, mitral regurgitation, myocardial ischemia, and death.[6,7] This clinical spectrum is known as the Bland-White-Garland syndrome.[6] If sufficient collaterals develop from the RCA, infants may survive into adolescence and even adulthood.[7] A "steal" phenomenon may occur in which blood passes in a retrograde fashion from the RCA through the LMCA and into the PA, resulting in a left-to-right shunt.[6]

Anomalous Origin of the RCA from the PA

An anomalous origin from the PA is much less common for the RCA than for the LMCA. The RCA anomaly, first described by Brooks in 1885,[4] is considered to have a better prognosis than its LMCA counterpart.[8] However, the RCA anomaly is not entirely benign, and angina and sudden death have been reported in adults.[8]

Other Anomalous Origins from the PA

Anomalous origin of the left anterior descending artery arising from the PA has been reported in at least 8 patients.[4] There are also reports of

Table 1
Types of Coronary Artery Anomalies

Type of Anomaly	Frequency	Description
Anomalous pulmonary origin	1 in 300,000 live births	Coronary artery arises from pulmonary artery; may cause myocardial infarction, congestive heart failure, or death in early infancy
Anomalous aortic origin	Unknown	Coronary artery arises from wrong aortic sinus; considered benign, except for origin of the left main coronary artery from the right aortic sinus of Valsalva and origin of the right coronary artery from the left aortic sinus of Valsalva
Congenital atresia of the left main coronary artery	40 cases described in literature	Single left or right coronary artery provides blood flow to the entire heart
Coronary artery fistulas	1 in 50,000 live births	Abnormal communications between coronary arteries, other cardiac vessels, or chambers
Coronary artery bridging	5.4% to 85.7% in general population	Segment of coronary artery is intramural or intramyocardial
Coronary aneurysms	0.3% to 4.9% in general population	Abnormal dilatation of coronary artery
Coronary stenosis	Unknown	Non-atherosclerotic coronary artery narrowing

Data extracted from Dodge-Khatami A et al.[5]

anomalous circumflex arteries arising from the PA in both children and adults.[4,6] Although both coronary arteries can arise from the PA, death usually occurs within weeks of birth due to myocardial ischemia. Survival to the age of 7 years, however, has been reported.[8]

Anomalous Aortic Origin of the Coronaries

Anomalous Origin of the LMCA from the RASV

An anomalous origin of the LMCA from the RASV accounts for only 1.3% of all coronary anomalies.[9] However, it is an anomaly of significant hemodynamic consequence. Approximately 59% of individuals with an LMCA arising from the RASV die from this disorder, most before age 20 years.[2] Eighty-one percent of the deaths are associated with exercise.[2] Anomalous LMCAs arising from the RASV may be subdivided based on the coronary arterial course in relation to the great vessels[5]:

- Septal (through the ventricular septum beneath the right ventricular infundibulum)
- Anterior to the aorta
- Posterior to the aorta
- Interarterial (between the aorta and pulmonary trunk) (Figure 1)

The septal course, the most common, is generally asymptomatic. An anterior course is also usually benign, although chest pain and myocardial infarction can occur. The posterior course has been associated with syncope and myocardial infarction. The interarterial course is the most symptomatic and fatal variant.[10]

Patients with the interarterial course (Figure 2) usually present with angina, syncope, dyspnea, dizziness, or sudden cardiac death.[11] It has been postulated that an interarterial course can cause sudden death during exercise as a result of increased pressures

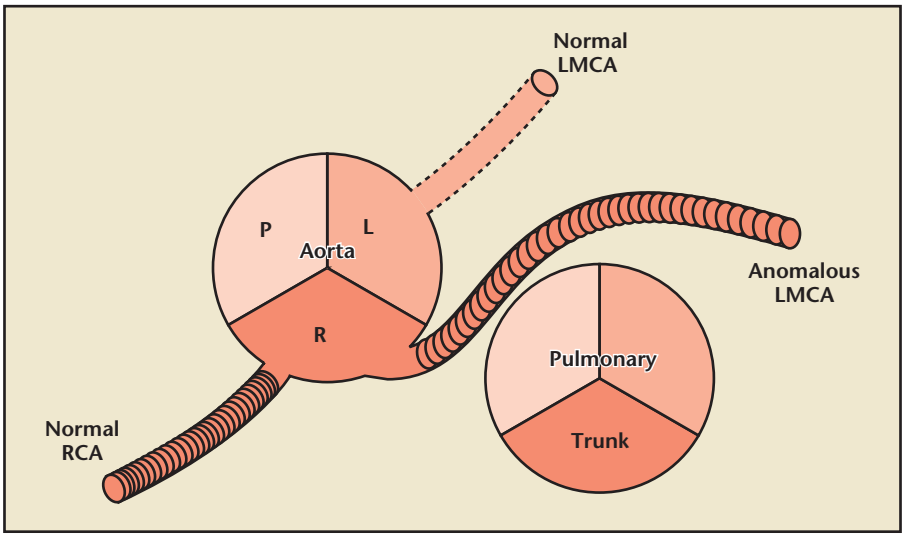

Figure 1. *Diagram showing an anomalous origin of the LMCA from the right aortic sinus of Valsalva with a course between the aorta and pulmonary trunk. LMCA, left main coronary artery; P, posterior aortic cusp; L, left aortic cusp; R, right aortic cusp; RCA, right coronary artery.* www.medreviews.com

in the aorta and pulmonary trunk that lead to expansion of vessels and compression of the artery between them. This mechanism of high pressure and expansion results in decreased anomalous coronary blood flow and cardiac ischemia.[10]

In addition to an interarterial course, 3 other high-risk features of an anomalous artery have been identified: slit-like ostium, acute-angle take-off (Figure 3), and intramural aortic segment.[12,13] These features probably increase the risk of sudden death by further limiting coronary blood flow. A slit-like ostium and acute-angle take-off of the narrow lumen diameter may lead to vessel

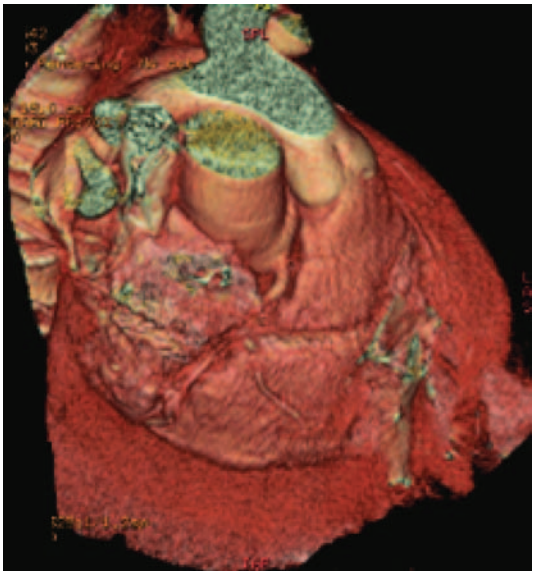

Figure 2. *Computed tomography angiogram showing an anomalous left main coronary artery coursing between the aorta and the pulmonary trunk.*
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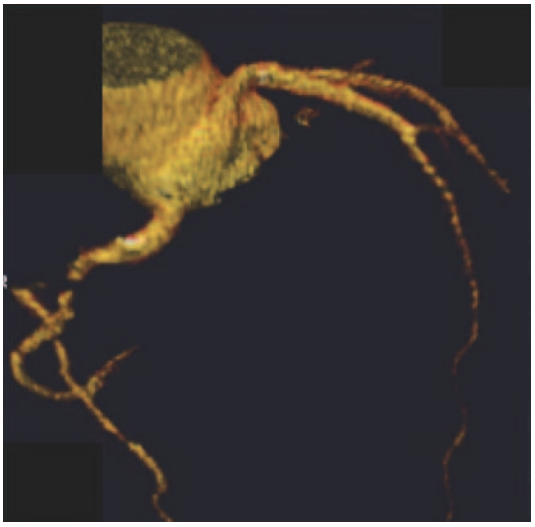

Figure 3. *Computed tomography angiogram demonstrating an anomalous left main coronary artery arising from the right aortic sinus of Valsalva at an acute angle.*
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collapse.[12,14] Likewise, an intramural segment may be compressed during ventricular systolic contraction.[12,15]

Anomalous Origin of the RCA from the LASV

Anomalous origin of the RCA from the LASV is more common than the LMCA equivalent.[4] Anomalous origin of the RCA from the LASV with passage between the aorta and pulmonary trunk is generally considered less dangerous than its LMCA counterpart because the RCA has a less dominant role in myocardial perfusion.[8] However, the RCA anomaly has been associated with myocardial ischemia and sudden death, presumably through the same mechanisms discussed above for the LMCA.[7]

Anomalous Origin of the Circumflex Artery from the RASV

An anomalous origin of the circumflex artery from the RASV generally does not cause myocardial ischemia. In a series of nearly 3000 patients, this anomaly was found in 0.67%. The anomalous circumflex artery generally arises posterior to the RCA and courses inferiorly and posteriorly to the aorta to enter the left atrioventricular groove. An interarterial course is uncommon.[7]

Diagnosis

The diagnosis of coronary artery anomalies can be challenging and may require multiple imaging modalities. X-ray coronary angiography has been the only method of detection for several decades and remains the gold standard.[16] However, this modality may be unable to identify the proximal course the vessel. In addition, it provides only a 2-dimensional image.[16,17]

Magnetic resonance angiography (MRA) is an adjunct to coronary angiography in identifying coronary anomalies. MRA is noninvasive, does not involve radiation or contrast agents, and provides 3-dimensional images. MRA is often better than x-ray coronary angiography at determining the proximal course of the anomalous coronary artery.[18] It is reported to have 92% sensitivity and 100% specificity in diagnosing anomalies.[16]

An alternative to MRA is computed tomography (CT) coronary angiography (Figures 2 and 3). CT angiography may be necessary for patients with defibrillators or other implanted metal objects. CT involves shorter examination times and less complicated monitoring but exposes patients to both radiation and intravenous contrast.[19]

Repeatedly compromised coronary blood flow associated with anomalous vessels can lead to recurrent ischemia and myocardial hibernation. During hibernation, myocardial contractility is downregulated, thereby reducing oxygen demand and preserving myocyte viability. Positron emission tomography has been shown to have a positive predictive value of 78% to 85% in identifying hibernating myocardium.[20]

Treatment

Once a symptomatic coronary anomaly is diagnosed, the treatment is generally surgical correction. Coronary artery bypass grafting is often the preferred method. When the LMCA arises from the RASV, the left internal thoracic artery is usually anastomosed to the left anterior descending artery. If anatomy allows, surgeons may excise the ostium of the anomalous vessel with a "button" of surrounding aorta and reimplant the ostium at a different site.[21]

Literature Search

We conducted a computerized search of Medline (January 1, 1966, to January 1, 2005), EMBASE (1988 to January 2005), and Web of Science (1993 to January 2005) to identify articles describing cases of LMCA arising from the RASV with an interarterial course. The search was performed using the terms "coronary vessel anomalies," "sinus of Valsalva," and "pulmonary artery," in English-language abstracts, titles, and text; the bibliographies of all relevant articles were also searched. Articles were identified as being relevant by a priori criteria (ie, randomized trials, cohort studies, case series, or case reports). We excluded cases using the descriptor "single coronary artery."

Table 2 summarizes the 104 cases found. In addition, we considered

Table 2
Reported Cases of Anomalous Left Main Coronary Arteries Arising From the Right Aortic Sinus of Valsalva

Study	Year	Patient Age (y)	Patient Sex	Ostium	Clinical Presentation	Treatment/Outcome
Bunce NH[25]	2003	56	Male	—	Angina	Surgery
		77	Male	—	Angina	Surgery
		18	Male	—	Syncope	Surgery
Frommelt PC[26]	2003	13.5	Female	—	Syncope	Surgical unroofing
		14.9	Male	—	Syncope	Surgical patch augmentation at artery origin
		14.7	Male	—	Asymptomatic	Surgical unroofing
		4.5	Male	—	Asymptomatic	No surgery
		20.1	Male	—	Angina	Surgical unroofing
Khouzam R[27]	2003	57	Female	Common	Angina	Single-vessel internal thoracic artery to LADA bypass
Frommelt PC[28]	2001	13	Female	—	Syncope, angina	Unroofing of intramural segment through anterior aortic wall, creating a large opening between aorta and anomalous coronary
		15	Male	Separate	Asymptomatic	Unroofing of proximal intramural course
		4	Male	—	Asymptomatic	No surgery
Frescura C[29]	1998	11	Male	—	—	Sudden death during exertion
		11	Female	—	—	Sudden death during exertion
		32	Female	—	—	Sudden death during exertion
		15	Male	—	Syncope	Sudden death during exertion
Wang A[30]	1997	39	Male	Separate	Angina	Aortoplasty: proximal, intramural portion of LMCA unroofed, with relocation of its ostium to left sinus of Valsalva and resuspension of aortic commissure between right and left sinuses of Valsalva; RCA bypassed using right internal thoracic artery
Mousseaux E[31]	1996	1	—	—	MI	—
Doorey AJ[32]	1994	1	—	—	Symptomatic	—
Selig MB[10]	1994	57	Female	—	Unstable angina	Single aorto-coronary reversed saphenous vein graft placed to LADA
Fernandes F[33]	1993	49	Male	—	Chest pain	—
		45	Male	—	Chest pain	—
		70	Male	—	MI	—
Thomas D[34]	1991	40	Female	—	Angina	Aorto-coronary bypass between aorta and LADA
Kragel AH[35]	1988	29	Male	Separate	Angina, syncope	—
		50	Female	Common	—	—
Barth CW[36]	1986	13	Female	—	Syncope	Sudden death shortly after exertion
		14	Male	—	Angina, syncope	Sudden death shortly after exertion
		19	Male	—	Asymptomatic	Sudden death shortly after exertion
		64	Female	—	MI, dyspnea	Death from congestive heart failure
		81	Male	—	Asymptomatic	Death from alcoholism
Ishikawa T[37]	1985	73	Female	Separate	Angina, MI	—
Kimbiris D[38]	1985	16-71 (6 patients)	5 Male, 1 Female	—	All had angina	—
Topaz O[39]	1985	15	Female	Separate	—	Sudden death during exertion
Grey DP[40]	1984	33	Female	Separate	Angina	LMCA ostium sphincteroplasty
Donaldson RM[41]	1983	15	Male	Separate	Symptomatic	Ostium split and LMCA fixed to aorta
		21	Male	Separate	Symptomatic	Ostium split and LMCA fixed to aorta
		32	Male	Separate	Symptomatic	Ostium split and LMCA fixed to aorta
Tsung SH[42]	1982	14	Male	Common	—	Sudden death during exertion
		18	Male	Common	—	Sudden death during exertion
Mustafa I[43]	1981	12	Male	Separate	Angina	Ostium split and LMCA fixed to aorta
Lynch P[44]	1980	20	Male	—	—	Sudden death during exertion
		19	Male	—	—	Sudden death during exertion
Maron BJ[45]	1980	17-22 (4 patients)	—	—	—	Sudden death during or shortly after exertion

(*Continued*)

Table 2
(Continued)

Study	Year	Patient Age (y)	Patient Sex	Ostium	Clinical Presentation	Treatment/Outcome
Moodie DS[46]	1980	17	Male	—	Syncope	Left internal thoracic artery to LADA and saphenous vein graft between aorta and circumflex
		72	Male	—	Syncope, dyspnea	Left internal thoracic artery to LMCA and saphenous vein grafts to circumflex and RCA
		57	Male	—	Syncope, angina	Left internal thoracic artery to LADA and saphenous vein graft to RCA
		58	Male	—	—	No surgery on anomalous vessel
Liberthson RR[47]	1979	1	Male	Separate	Asymptomatic	Sudden death shortly after exertion
		11	Male	Separate	Angina	Sudden death shortly after exertion
		17	Male	Separate	Asymptomatic	Sudden death during exertion
		36-70 (6 patients)	5 Male, 1 Female	—	All had angina	4 patients had medical management; 2 patients underwent bypass grafting to LADA and left circumflex
Kimbiris D[3]	1978	2 patients	—	Separate	Both had angina	—
		16	Male	—	Angina, MI	—
Sher RF[48]	1978	63	Male	Common	Angina	Double saphenous vein bypass surgery to LADA and to marginal branch of left circumflex
Sacks JH[49]	1977	20	Male	Common	Angina, syncope	Saphenous vein graft from ascending aorta to LMCA and LADA
Chaitman BR[50]	1976	49	Female	—	Asymptomatic	—
		39	Male	—	Angina	—
		70	Female	—	Angina	—
Chaitman BR[51]	1975	14	Female	Separate	Asymptomatic	No intervention
Adams RC[52]	1974	11	Male	Separate	Angina	Sudden death during exertion
Cheitlin MD[14]	1974	48	Male	Separate	—	—
		40	Male	Separate	—	—
		66	Male	Separate	—	—
		67	Male	Separate	—	—
		42	Male	Separate	—	—
		17	Male	Separate	Syncope	Sudden death shortly after exertion
		14	Male	Separate	—	Sudden death shortly after exertion
		18	Male	Separate	—	Sudden death during exertion
		17	Male	Separate	—	Sudden death during exertion
		18	Male	Separate	—	Sudden death during exertion
		22	Male	Separate	—	Sudden death during exertion
		20	Male	Separate	—	Sudden death during exertion
		22	Male	Separate	—	Sudden death during exertion
		49	Male	Separate	MI	MI
		49	Male	Separate	MI	MI
		64	Male	Separate	—	—
		69	Male	Separate	MI	MI
		82	Male	Separate	—	—
		14	Male	Separate	Syncope	Surgery
Liberthson RR[53]	1974	11	Male	Separate	Angina	Sudden death shortly after exertion
		17	Male	Common	Asymptomatic	Sudden death shortly after exertion
Benson PA[54]	1970	54	Male	Separate	Angina	Sudden death not related to exertion
Benson PA[15]	1968	13	Male	Separate	Asymptomatic	Sudden death shortly after exertion
		13	Male	Separate	Asymptomatic	Sudden death shortly after exertion
Cohen LS[55]	1967	11	Male	Separate	MI	Sudden death shortly after exertion
Jokl E[56]	1966	16	Male	Separate	Syncope, dyspnea	Sudden death during exertion
Jokl E[57]	1962	14	Male	Common	Asymptomatic	Sudden death shortly after exertion
Alexander RW[58]	1956	67	Female	Separate	Asymptomatic	Death from tuberculosis
		53	Male	Separate	Asymptomatic	Death from hemorrhage

LADA, left anterior descending artery; LMCA, left main coronary artery; RCA, right coronary artery; MI, myocardial infarction. Dash (—) indicates not reported.

data from a case of an anomalous LMCA arising from the RASV with an interarterial course in a surviving adult.

Results

In the 40 years of data, no adult with all of the 4 high-risk features associated with cardiac death survived. Patient and clinical characteristics are shown in Table 3. Of the cases found, 77 (74%) occurred in males and only 19 (18%) in females. The anomaly occurred in patients of all ages, from 4.5 years to 77 years (mean, 33 years). Many articles did not identify whether the anomalous LMCA arose from a separate ostium from the RCA or if both vessels arose from a common ostium. Of those articles that did specify the type of ostium, the vessels arose from a separate ostia in 44 (42%) and from a common ostium in 8 (8%). Sixty patients (58%) with the anomaly were symptomatic and only 16 (15%) were asymptomatic (symptom status was not indicated for the remainder). Symptoms included angina, dyspnea, syncope, and myocardial infarction. Thirty-six patients (35%) experienced sudden death, and 26 (25%) underwent surgical correction of their anomaly.

Table 3
Reported Clinical Characteristics of Patients With Anomalous Left Main Coronary Arteries Arising From the Right Aortic Sinus of Valsalva

	Number of Patients (% total)
Sex	
Male	77 (74%)
Female	19 (18%)
Not reported	8 (8%)
Ostium	
Common	8 (8%)
Separate	44 (42%)
Not reported	52 (50%)
Clinical presentation	
Symptomatic	60 (58%)
Asymptomatic	16 (15%)
Not reported	28 (27%)
Treatment/outcome	
Sudden death	36 (35%)
Surgical correction	26 (25%)
Nonsurgical or medical management	8 (8%)
Death from myocardial infarction	3 (2.5%)
Death from noncardiac causes	4 (3.5%)
Not reported	27 (26%)

Discussion

Coronary artery anomalies are important clinical phenomena. They can be diagnosed by CT, MRA, or coronary angiography. Treatment is surgical and usually involves bypass grafting of the anomalous vessel. Anomalous origins from the PA are often associated with myocardial ischemia and death in infancy. Anomalous origins from the aorta tend to be benign, with the important exception of the LMCA arising from the RASV. This anomaly is often symptomatic and can be fatal.

A review of reported cases reveals that patients with an anomalous left main coronary artery often present with angina, dyspnea, syncope, myocardial infarction, or sudden death.

A review of reported cases reveals that patients with an anomalous LMCA often present with angina, dyspnea, syncope, myocardial infarction, or sudden death. Symptoms frequently occur during or shortly after physical exertion and are related to cardiac ischemia. Although the exact mechanism of ischemia is unclear, certain anatomic features are associated with a higher risk of sudden death, presumably through increased risk of cardiac ischemia. Slit-like ostium, acute-angle take-off, intramyocardial segment, and course between the great vessels all likely lead to compromised coronary flow. Our review suggests that the majority of cases of anomalous LMCA arising from the RASV occur in men.

This review has several limitations. Asymptomatic cases of anomalous LMCA tend not to be identified or reported, and given this bias, reports may not represent the entire spectrum of cases. In addition, the distinction between a single coronary artery and a common ostium was often blurred, and several cases may have been overlooked or misdiagnosed. As radiologic imaging improves, more anomalies will be diagnosed and their exact origin and route will be more easily identified.

Recommendations

In general, we recommend that anomalous LMCA arising from the RASV with an interarterial course be surgically corrected in all symptomatic patients. We believe that bypass revascularization is the best surgical option, and we favor the use of internal thoracic arteries in young patients. We recommend complete assessment of cardiac viability, since anomalies can result in chronic low-flow states leading to hibernating myocardium. In addition, we recommend that asymptomatic individuals, especially pre-adolescents and adolescents, be vigorously assessed for ischemia or arrhythmia; if present, surgical revascularization is recommended.

Anomalies of the LMCA are not just of academic interest—they have important implications for public health. Data indicate that approximately

5.6% of Americans[2,22] possess some type of coronary anomaly. According to the American Heart Association,[23] approximately 19% of sudden deaths in young athletes are related to coronary anomalies. US Census Bureau statistics[24] indicate that of approximately 281.4 million Americans, 57.6% (162.0 million) are age 39 years or younger, and 36.2% (101.8 million) are age 15 years to 39 years. If we assume that 5.6% of these age groups have an anomalous coronary artery and 19% of those with anomalies have sudden death, approximately 1.72 million such deaths would result, or, more conservatively, 1.08 million deaths in the age range of 15 years to 39 years. This represents a significant public health and socioeconomic burden. A national registry of coronary anomalies is therefore needed to assist in evidence-based recommendations for the diagnosis and treatment of this potentially lethal condition. ■

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Main Points

- An anomalous left main coronary artery arising from the right aortic sinus of Valsalva with an interarterial course is associated with significant morbidity and mortality.
- Symptoms include angina, myocardial infarction, syncope, or dyspnea; sudden death associated with exercise is common.
- Anatomic features of the anomalous artery associated with higher risk of sudden death include slit-like ostium, acute angulation, intramyocardial segment, and interarterial course.
- The proposed mechanism of morbidity is compromised coronary blood flow and cardiac ischemia; recurrent ischemia can lead to myocardial hibernation.
- Anomalies can be detected with magnetic resonance imaging, computed tomography, or angiography.
- The treatment is surgical correction. Coronary artery bypass grafting is often the preferred method.

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