Editorial:
The Intramural Coronary Artery: Another Cause for Sudden Death with Exercise?

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Sudden death, the ultimate complication, is generally thought of in the context of coronary artery disease. Most studies of the total experience with death that occurs suddenly and unexpectedly and is not associated with trauma conclude that about 90% are associated with pathologically significant coronary atherosclerosis.1

Sudden death in young people, especially while engaging in strenuous physical activity, is a rare and always shocking, tragic event. The causes of sudden unexpected death in young people are more varied and appear to be increasing. In this issue of Circulation, Morales, Romanelli and Boucek report three cases of sudden death that occurred during severe exercise. Autopsy revealed evidence of previous myocardial fibrosis and necrosis, which they attributed to myocardial bridging of the left anterior descending coronary artery and, in the third case, also of the right coronary artery.

So we add to the short list of known causes of sudden death in young people the myocardial muscle bridge, sometimes referred to as an intramural coronary artery. The evidence presented to support this explanation, with the exception of the first case, is based entirely on the pathologic findings at autopsy. Unequivocally, all three patients had a portion of their left anterior descending coronary artery pass intramurally and all three had evidence of myocardial necrosis and fibrosis in the areas subserved by the bridged coronary vessels consistent with both old and recent myocardial injury, probably ischemic.

The first case is most interesting because the patient had had an extensive medical work-up for chest pain, atypical for angina in that it was precipitated by emotional stress but not by exercise. The major clinical evidence for myocardial ischemia was a "strongly positive" Master two-step exercise test and progressive ECG changes consistent with myocardial septal fibrosis. Cardiac catheterization 8 years before death showed normal coronary arteries and, retrospectively, a "milking" of the left anterior descending coronary artery consistent with a myocardial bridge.

But the first case has other findings which soften the conclusion that the muscle bridge was causally related to the sudden death. At autopsy there was coronary atherosclerosis of the left anterior coronary artery just proximal to the muscle bridge, estimated to reduce the lumen to 50% of normal, as well as a similar but longer lesion just proximal to the takeoff of the posterior descending coronary artery. There was also 50% narrowing of the atrioventricular nodal artery by intimal hyperplasia, a finding associated with sudden death. The patient was 54 years old, and although these lesions might not have been critical obstructions to flow at ordinary activity, with jogging and the consequent large increase in myocardial oxygen demand, it is possible that the atheromatous obstructions resulted in myocardial ischemia. Another complicating issue in the first case is the short PR interval, and although no accelerated atrioventricular conduction pathway was demonstrated at electrophysiologic study 8 years before death, the possibility that one existed and was associated with a resultant tachyarrhythmia that caused ventricular fibrillation cannot be entirely excluded. Finally, there was a "thick left ventricle" described on angiocardiography that resulted in the diagnosis in this patient of hypertrophic cardiomyopathy, a disease certainly associated with the complication of sudden death.

The problem with causally linking the intramural coronary artery with sudden death is that myocardial bridges are found in 5.4–85.5%2 of autopsied hearts.3 In pooling all reports, Morales et al. conclude that 27% of consecutive autopsies will show some evidence of dipping of the coronary artery intramurally. The chance association of autopsy evidence of myocardial bridges found in patients who die suddenly for some other reason is therefore quite possible. However, the evidence in the literature that myocardial bridges do indeed cause angina and myocardial ischemia, although limited, is compelling. In spite of the autopsy frequency of myocardial bridges, the evidence of systolic contraction of a portion of a coronary artery at coronary arteriography is much smaller, estimated at from 0.5–1.6%.4,5 Morales et al. review briefly the evidence for myocardial bridges causing myocardial ischemia, citing Noble et al.,6 who reported 11 cases of myocardial bridging by coronary arteriography where lactate production was observed with pacing stress in four of five patients with severe systolic compression of the left anterior descending coronary, and the reports of relief of angina by the surgical removal of the myocardial bridge.6,7

All three patients had dominant left coronary artery systems and diminutive right coronary arterial supply

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to the diaphragmatic surface of the heart, thus increasing the volume of the left ventricle supplied by the left coronary system. If systolic compression caused ischemia in the muscle supplied by the left anterior descending coronary in these patients, the volume of myocardium affected might be quite large, and thus form the basis for sudden death. Severe exercise with tachycardia, and therefore marked decreased time for diastolic filling, might seriously compromise myocardial blood supply through an epicardial coronary artery that is compressed during systole by the overlying, vigorously contracting ventricular muscle. With the increased myocardial oxygen demand caused by exercise this could result in critical imbalance between demand and supply for myocardial blood flow, resulting in myocardial ischemia and arrhythmia and leading to sudden death.

As stated, the known causes of sudden death in young people are few and include congenital coronary anomalies, such as anomalous origin of the left coronary from the pulmonary artery and origin of the left coronary from the anterior sinus of Valsalva, hypertrophic cardiomyopathy, aortic stenosis and atherosclerotic coronary artery disease. Even rarer causes are prolapsed mitral valve and dissection of the aorta, with rupture into the pericardium, coronary emboli from atrial myxoma and occlusion of the coronary ostium by vegetations and benign papillary fibroelastoma of the aortic valve. Electrophysiologic problems include accelerated atrioventricular conduction with atrial fibrillation, causing ventricular fibrillation and prolonged QT-interval syndromes.

In a review of sudden death in athletes in this issue, Maron et al. concluded that hypertrophic cardiomyopathy is the most frequent cause. How often myocardial bridging is seen in those cases was not mentioned, but a recent report shows compression of the septal branches of the anterior descending coronary artery to occur frequently in many diseases associated with left ventricular hypertrophy. Whether this compression plays a role in sudden death in hypertrophic cardiomyopathy, hypertension, and aortic stenosis is not clear; the possibility may have not been raised. Another report of sudden death in joggers confirms that the most common cause of death is coronary atherosclerosis.

The report of Morales et al. indicates that muscle bridges found at coronary arteriography have an ominous significance. Long-term clinical follow-up of patients found at coronary arteriography to have systolic compression of a coronary artery is necessary to evaluate the frequency of myocardial infarction and sudden death. If sudden death occurs it would be interesting to see whether a dominant left coronary system with a diminutive right coronary artery is a necessary adjunct. Until then, the significance of muscle bridges found at coronary arteriography must remain unclear.

And yet . . . .

References

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_Circulation_. 1980;62:238-239
doi: 10.1161/01.CIR.62.2.238

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1980 American Heart Association, Inc. All rights reserved.
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:
http://circ.ahajournals.org/content/62/2/238.citation

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