“Cui periculum”—Who is at risk?

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Mery and colleagues¹ have described their experience in 44 patients with anomalous aortic origin of a coronary artery (AAOCA) who underwent surgical repair at their institution over a 4.5-year time period. An additional 17 patients were offered surgery, with 10 declining, 5 waiting, and 2 who underwent surgery at another institution. During this same time frame, there were 101 patients who were newly diagnosed but were not offered surgery. Thus, 38% of the cohort had a recommendation for surgical intervention and 30% have received that treatment.

The real question is...Who is at risk? or “Cui periculum?” It is evident that AAOCA is a heterogeneous diagnosis, and the natural history (and risk) will vary considerably on the basis of the underlying anatomy. We have previously made the case that there are 2 separate risks that should be thought of differently: (1) The risk of sudden cardiac death (SCD) and (2) the risk of ischemic events that are not SCD.² SCD has been associated with the presence of an anomalous left coronary artery, although it also can occur with an anomalous right coronary artery (ARCA). The caveat is that SCD represents only 2% to 3% of patients who are currently being discovered with the diagnosis of AAOCA.

The overwhelming majority (97%-98%) of patients with AAOCA are diagnosed by a mechanism other than SCD

	Anomalous Right	Anomalous Left	Single Ostium, Eccentric
Intramural	63 (87%)	23 (75%)	
Not Intramural	13 (46%)	14 (28%)	2 (50%)

Diagram demonstrating the anatomy and prevalence of symptoms for patients with AAOCA.

Central Message

Anomalous coronary arteries have been associated with myocardial ischemia and SCD. Although the risk factors for SCD are well documented, the risk factors for those without SCD remain unclear.

See Article page 305.

(Figure 1). Some will have “ischemic” symptoms such as exertional chest pain, nonexertional chest pain, syncope, and presyncope. Many of these symptoms are the bane of a pediatric cardiologists clinic and thus are treated with some skepticism. However, sometimes they prove to be “real,” as documented by the fact that half of the patients in our series with ischemic symptoms had evidence of having sustained myocardial damage. This objective evidence includes troponin leaks, electrocardiogram changes, wall motion abnormalities on echocardiography, perfusion defects, or catheter documentation.³ Admittedly, there is also a large cohort of patients with AAOCA who are genuinely “asymptomatic” and are diagnosed in the process of evaluating innocent murmurs or other screening measures.

FIGURE 1. Pie chart demonstrating the mode of presentation for the 115 patients who underwent surgical repair of AAOCA. Reprinted with permission.⁴

	Anomalous Right	Anomalous Left	Single Ostium, Eccentric
Intramural	63 (87%)	23 (75%)	
Not Intramural	13 (46%)	14 (28%)	2 (50%)

FIGURE 2. Two-by-two box diagram demonstrating the distribution of anatomy for the 115 patients who underwent surgical repair of AAOCA. Shown in parentheses are the percentage of patients in each subgroup who presented with symptoms of myocardial ischemia. Modified with permission.⁴

FIGURE 3. Algorithm for the surgical management of the 115 patients with AAOCA. AAOCA, Anomalous aortic origin of a coronary artery; PA, pulmonary artery. Reprinted with permission.⁴

The risk factors for “ischemic” symptoms not associated with SCD have drawn far less attention than the risk factors for SCD and often are viewed as interchangeable. However, the evidence would suggest that the underlying anatomy is completely different. In our surgical series, 85% of patients with an intramural coronary artery had associated symptoms of ischemia, whereas only 41% of patients with a non-intramural coronary artery had symptoms (Figure 2). There were no differences in symptoms based on ARCA versus anomalous left coronary artery. On the basis of these data, we proposed that intramural anatomy is the key risk factor associated with ischemic symptoms and believe that all intramural coronaries should have surgical repair.^{3,4} This association between intramurality and ischemic symptoms has been observed in the current article and by numerous other groups.

The conflation of these 2 risk factor concepts has resulted in significant misconceptions and in effect has prevented the correct identification of the risk factors for patients without SCD. It is almost certain that there are subgroups at increased risk of developing ischemic symptoms (ARCA, intramural), and conversely, there are subgroups that are likely at lower risk (ARCA, nonintramural). At the present time, despite more than 40 published articles on AAOCA over the past 15 years, we still do not know definitively whether there are any subgroups that are safe to follow.⁵

Returning to the article at hand, I would agree with Dr Mery and colleagues¹ on many of their observations. We follow the same algorithm, where the surgical procedure is driven by the anatomy and not the other way around (Figure 3). I would like to underscore one specific concern that they have raised regarding patients with a “short intramural course.” These patients have historically undergone a “short unroofing,” and Dr Mery and colleagues¹ reported a patient who had recurrent SCD in this setting. We too have had failures in the setting of a short intramural course and

concur that the unroofing procedure must move the coronary ostia well beyond intercoronary pillar into the correct sinus of Valsalva. When this cannot be accomplished by a simple unroofing, then the coronary should be surgically translocated. We have been performing more coronary translocations in recent years after our unfavorable experience with “short unroofings.”

Finally, there has been much consternation regarding the efficacy of AAOCA surgery.⁶⁻⁸ One of the most widely misinterpreted and misquoted articles on the subject contains a graph showing only 20% of patients free from cardiac symptoms at 9 years after AAOCA repair.⁹ This actuarial curve has been cited by many authors as proof that the surgical repair does not work as well as we would like (or think). However, the authors of the original article were inclusive of any abnormal findings as a clinical event. Many of these findings were transient, and almost certainly not indicative of a failure of surgery. The authors of this article acknowledge that all of their patients are alive and “the subjects continue to experience average or higher than average quality of life without exercise restriction.” The experience of Dr Mery and colleagues¹ is consistent with our own experience that the surgical repair of AAOCA is highly efficacious in relieving the symptoms of myocardial ischemia.

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