Guidelines should bother us, not comfort us

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Anomalous aortic origin of a coronary artery (AAOCA)—specifically one that takes an interarterial course—is an infrequent anomaly (prevalence, 0.2%-0.8%) that is associated with a small incidence of sudden cardiac death, especially in athletes. The combination of high public visibility, low prevalence of the anomaly, and apparent low incidence of death caused by it, has been a cause célèbre for many publications promoting diagnostic algorithms and management schemes.

In this issue, Brothers and colleagues report findings and recommendations of The American Association for Thoracic Surgery–sanctioned AAOCA Guidelines Writing Committee on the diagnosis and management of this disorder. This review was sorely needed, and the authors have done a laudable job executing it. Additionally, this cyclopean effort has the collateral advantage of stimulating more thought and debate on this challenging anomaly. For example, consider the following:

The authors make a Class I, Level B recommendation for surgical treatment for any operative patient with anomalous aortic origin of the left coronary artery (AAOLCA), with or without symptoms. Table 1 displays data extracted from 12 contemporary case studies and 50 case reports on operations for AAOLCA (representing most of the published articles from which data on AAOLCA could be extracted). None of these studies is a comparative study.

All data refer only to the subset of patients with anomalous aortic origin of the AAOLCA. Symptoms include all of those recorded as “cardiac” in origin by each study. Provocative tests include exercise echocardiogram testing and/or stress echocardiography. Deaths exclude those attributable to associated anomalies or to the extremity of the preoperative state. The denominators are the number of subjects evaluated for each column. AAOLCA, Anomalous aortic origin of the left coronary artery; NA, value could not be calculated using the data reported in the study.
The total number of reported cases is 176. The average of the mean durations of follow-up is about 2.6 years. There were no deaths. Seems like perfect results, but how do they compare with the natural history of untreated AAOLCA? Brothers and colleagues\(^1\) estimated that the cumulative risk of death over a 20-year period was about 6.3% for AAOLCA patients participating in competitive sports. Taking this percentage as an upper limit (because most patients do not participate in competitive sports), one would expect 1 to 2 deaths during the average 2.6-year follow-up among the 176 patients in this collection of studies. At least on the basis of this comparison, with such short-term average follow-up, there does not seem to be strong evidence for the superiority of surgical versus not-surgical treatment.

Now consider, in this same set of 176 patients, the evidence that surgical operation relieves cardiac ischemia in AAOLCA. To test this, one must know how many patients had evidence of preoperative ischemia. As shown in Table 1, the evidence of preoperative ischemia is scant—most studies did not quantitatively evaluate it. This leaves symptoms as the only evidence of ischemia—the reliability of which (except for sudden cardiac collapse) is among the most hotly debated questions in those studying this disorder. Furthermore, the quantitative evaluation of postoperative ischemia is also scant, with the largest single study (Mainwaring and colleagues\(^2\)) not reporting postoperative testing at all. Thus, whether surgery on AAOLCA results in a lower incidence of persistent or not-surgical treatment.

Figure 2 reproduces an August 2015 refinement of the American College of Cardiology/American Heart Association Clinical Practice Guidelines grading system, more granular than that used in the present article.\(^3\) Do we have objective evidence to strongly recommend surgery under all circumstances for AAOLCA (Class I), given that the margin in the above ad hoc comparative analysis of all of the Table 1 reports was only 1 or 2 deaths? Do we have 1 or more well-designed, well-executed studies with moderate quality evidence (Level B-NR), when we only have case reports or case studies subject to publication bias, when we average a few years’ follow-up, and when we infrequently report quantitative ischemic tests for AAOLCA? We might consider being more humble so that we know we still have a problem here.

Perhaps it is time, as many have suggested, to focus on anatomic and clinical risk stratification of both AAOLCA and anomalous aortic origin of a right coronary artery, as well as accumulate greater length of follow-up. The Congenital Heart Surgeons’ Society AAOSA registry, initiated in 2009, containing almost 600 patients and with 70 to 80 new enrollments per year, may provide the best opportunity to reach the next class of recommendation and level of evidence, because the cohort is to be followed for a lifetime. Studies based on the registry must be carefully crafted to provide prospective, serial, pre-, and postoperative quantitative evaluation of these patients by the Congenital Heart Surgeons’ Society member institutions. How that is executed is still an organizational and regulatory challenge, but will be worth it. In the meantime, the current guidelines for surgical management of AAOCA, driven by the treatable anatomic nature of the anomaly and the Housmanian tragedy of sudden death in the young,\(^4\) represent an important update of our knowledge base and expert opinion. The status of our evidence base should continue to bother us.

### References


TABLE 1: Characteristics of the Five Cases with CTA Images

<table>
<thead>
<tr>
<th>Case</th>
<th>Patient Age (y)</th>
<th>Lesion Location</th>
<th>Lesion Length (cm)</th>
<th>Lesion Type</th>
<th>Treatment Modality</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>12</td>
<td>Right coronary artery</td>
<td>2.5</td>
<td>Type A</td>
<td>Stent placement</td>
</tr>
<tr>
<td>B</td>
<td>16</td>
<td>Left coronary artery</td>
<td>3.0</td>
<td>Type B</td>
<td>Bypass surgery</td>
</tr>
<tr>
<td>C</td>
<td>20</td>
<td>Right coronary artery</td>
<td>4.0</td>
<td>Type C</td>
<td>Ablation</td>
</tr>
<tr>
<td>D</td>
<td>23</td>
<td>Right coronary artery</td>
<td>5.0</td>
<td>Type D</td>
<td>Open surgery</td>
</tr>
<tr>
<td>E</td>
<td>25</td>
<td>Left coronary artery</td>
<td>6.0</td>
<td>Type E</td>
<td>Ablation</td>
</tr>
</tbody>
</table>


FIGURE 2. American College of Cardiology/American Heart Association revised 2016 recommendation scheme for clinical practice guidelines. Reprinted with permission. 16


