Management Implications for Anomalous Aortic Origin of Coronary Arteries*

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The report by Lorber et al. (1) in this issue of *iJACC* is a large, important, and timely review of the ultrasound diagnosis and delineation of clinically pertinent characteristics in 159 pediatric patients with anomalous aortic origin of the coronary artery (AAOCA), including 89 who underwent surgical revision. Patients were managed at 24 institutions. Ultrasound interpretations by the individual hospital laboratories were compared with reinterpretation by a central expert core laboratory, and then, in those undergoing surgery, with intraoperative findings. Significant discrepancy as well as deficiency of data pertinent to patient management were found between the individual and the core laboratory evaluations, and between both, and intraoperative surgical evaluation. The study provides helpful practical detailed diagnostic and delineation guidelines for patients with these anomalies and offers uniform comprehensive reporting protocols for both transthoracic and transesophageal evaluation. It also clarifies and illustrates the still too often confused terminology used to describe essential anatomic features, notably: interarterial, intramural, ostial morphology, high coronary takeoff relative to the sinotubular junction, acute angle take off and its association with intramural course, and subpulmonic (intracoronary coronary course). Importantly, this study provides new data that underscores the need for congenital coronary artery registries to validate diagnostic and management guidelines and track patient outcomes both natural and post-operative. Although focused on ultrasound, its intent as mentioned was not to exclude other imaging modalities as needed.

I will address my thoughts regarding AAOCA to: 1) what we presently know with confidence; 2) what aspects are still controversial; and 3) what is still unknown but likely will be resolved by efforts such as this study. By design, this study addressed a pediatric population with entry age cutoff at 30 years, targeting the highly charged precocious, exertion-related sudden death aspect encountered mostly in younger individuals (1). I would urge the society to extend this window to include those at all ages, to provide a broader overall understanding of this entity, as increasingly AAOCA is incidentally recognized by ultrasound, angiography, magnetic resonance imaging (MRI), and computed tomography angiography (CTA) through midlife and in the elderly where it presents its own management issues but is still haunted by its malignant reputation at younger ages.

My oldest patient is 84 years old and had a large proximally unobstructed ectopic left coronary artery and unrelated arteriosclerotic heart disease. For several decades, I’ve followed numerous others with unobstructed nondominant ectopic right or left intra-arterial arteries who were identified past age 50 years, including some whose younger lifestyle included extreme professional or recreational exertion. Clearly highest sudden death risk exists from adolescence through the 30s, it occurs in the 40s but far less often, and is distinctly rare beyond 50 years of age. During my 5-decade experience managing these entities, I have not seen a related sudden death beyond age 50 years. This does not imply that longevity imparts sudden death immunity, but it does underscore the need for late outcomes to help guide lifelong management. A large Arm Forces Institute of Pathology study by Taylor also identified octogenarians with both ectopic left and right coronary arteries (2). AAOCA is rarely noted in infancy, although it may occur, and I have seen a 9-month old who clutched her chest while nursing on a hot July New England day, arrested, and at autopsy had a dominant ectopic right coronary with severe ostial stenosis and early histologic ischemic changes in the perfusion territory (3).
Ascertaining the true prevalence of AAOCA in the population is challenging, although a recent MRI screening study of 1,836 middle school students by Angelini et al. (4) identified 13 with AAOCA, 11 (6%) with ectopic right, and 2 (.1%) with ectopic left coronary arteries. These findings roughly approximate reported incidences in postmortem, catheterization, CTA, and MRI laboratory populations. Overall, ectopic right coronary artery is more commonly diagnosed, but ectopic left accounts for more sudden deaths (5). AAOCA accounts for approximately 15% of deaths in young competitive athletes (6), and in more than 6 million recent military recruits accounted for 21 deaths during 6 weeks of basic training (7). Unexpected sudden cardiac death in young individuals with AAOCA typically occurs during or shortly after extreme exertion. It is most common with high dynamic sports including basketball, soccer, track, and football. There have been several excellent definitive reviews describing the clinical profile of sudden death in highly trained athletes originating in both the United States and Italy (5,6). Although controversial, I have long had a low threshold for targeted ultrasound screening.

Common pathologic findings in those at high sudden death risk include the presence of congenital obstruction in the proximal ectopic coronary artery, and its presence generally mandates surgical revision. Obstruction may occur at several sites including the ostium, which may be hypoplastic and appear slit like (2); or within the intramural segment as the coronary obliquely traverses the aortic wall, for variable length, from ostium through media. These often, but not only, occur with high right coronary origin, and more acute angle takeoff relative to the aorta. A graduating tubular obstruction as the coronary enters the proximal portion of the interarterial space may also exist and appear as oval flattening on bread loaf CTA coronary cross sectioning. Whereas ultrasound contributes to identifying and delineating the morphology of proximal obstruction, CTA and MRI also have important defining roles, as may intravenous ultrasound in selected circumstances.

Although still less than totally clear, when exertional sudden death occurs in AAOCA, it is likely secondary to acute ischemia (2) which triggers ventricular tachyarrhythmia and fibrillation. Larger size/dominance of the ectopic coronary and the muscle mass it perfuses likely increase sudden death risk, as does the magnitude of underlying proximal coronary obstruction. As postulated in 1974 by Cheitlin (8), increased cardiac output with strenuous physical exertion may acutely add to underlying coronary obstruction by aortic dilation and torsion, which may accentuate ostial kinking and intramural segment compression precisely when myocardial demand is increased. Of note, chronic fibrosis may coexist in the ischemic region and may have late arrhythmic consequences, although to date, these have been rare; MRI late gadolinium uptake may have a role. My threshold for beta blockers in some has been low, and recognizing that advice regarding unrestricted physical exertion must always be carefully approached. I have been an advocate of golf for many of these patients.

Advances in surgical management for those at high sudden death risk with very low short-term morbidity and near zero mortality as in this report and others (9) has lowered the current bar for surgical revision in those with compelling clinical presentation and high-risk anatomy and age. As longer-term outcome data accumulates, these advances will no doubt further define optimal patient management. The authors are to be complimented for this important contribution (1).

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KEY WORDS anomalous coronary artery, best practices, echocardiography