The search for the Holy Grail: Risk stratification in anomalous aortic origin of a coronary artery

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Sudden cardiac death continues to shatter families and communities at large, despite its rarity. Anomalous aortic origin of a coronary artery (AAOCA) from the opposite sinus of Valsalva, the second most common cause of sudden cardiac death in young patients,¹ claims young lives every year. Despite a wealth of information on anatomic types, course, degree of stenosis on dynamic assessment, advanced imaging (Figure 1), and evaluation of myocardial ischemia,^{2,3} we remain devoid of defined measures to stratify those at risk.

In this issue of the *Journal*, Sachdeva and colleagues⁴ report a single-center experience with the unroofing procedure in AAOCA during a 17-year period. One of the salient and disturbing findings of this series was that 3 patients suffered a sudden cardiac arrest (SCA) 2-6 years after the unroofing procedure, 2 of whom had an initial presentation of SCA. Given the retrospective nature of the study and the heterogeneity of preoperative and postoperative anatomic and functional imaging, the reasons for these occurrences are unknown. However, they highlight one of the inherent limitations of our counseling of families on surgical intervention (or not) for AAOCA. Does surgical intervention alter the risk of SCA in a particular patient? Were all anatomic culprits addressed in these patients at the time of surgery or was the unroofing procedure incomplete or inadequate? Does presenting with SCA translate into a greater risk for a recurrent event? Do patients presenting with SCA have myocardial scarring that predisposes them to recurrent SCA despite surgical intervention? Should we place automated internal cardiac defibrillators in those presenting with SCA? What tests can we use to risk-stratify patients?

These are just a few of many unanswered questions in AAOCA. We recently described a basketball player who had an unroofing procedure for an anomalous left coronary artery after SCA but presented with a recurrent episode of SCA 20 months later.⁵ Postoperative stress cardiac magnetic resonance imaging had shown no perfusion abnormalities and no scarring, but further imaging showed

Computed tomography showing an anomalous right coronary artery with an intramural segment.

Central Message

Despite advances in diagnostic and surgical techniques for patients with anomalous aortic origin of a coronary artery, there are no good methods to identify those at risk for sudden cardiac death.

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residual narrowing of the left coronary artery due to a procedure that failed to address compression of the vessel by the intercoronary pillar.

Although meaningful information is provided by this surgical series,⁴ there are inherent limitations that are common to similar retrospective series of AAOCA without standardized management, namely, short and incomplete follow-up, the lack of a true denominator (all patients with AAOCA regardless of whether they had surgery or not), inconsistent preoperative and postoperative evaluation, and the inclusion of patients with other significant congenital heart diseases and impaired ventricular function.

Unquestionably, this study urges the scientific community to unite in acquiring consistent prospective long-term data on these patients through efforts such as the Congenital Heart Surgeons' Society Registry on AAOCA,⁶ standardization of workup within each center,⁷ and multi-institutional collaboration for development of consistent approaches on imaging, functional assessment, and longitudinal follow-up. We may not see the blossomed fruits of these efforts in our lifetime, but we ought to be planting the seeds for our children and grandchildren to benefit in the future.

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FIGURE 1. Computed tomography showing an anomalous right coronary artery (arrow) with an intramural segment before (A) and after (B) unroofing.

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