Anomalous aortic origin of a coronary artery: 90% of optimal therapy is 50% improved technology

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Anomalous aortic origin of the coronary artery describes a heterogeneous group of coronary anomalies in which both major coronary arteries arise from the same coronary sinus. The subsequent course taken by the anomalous artery varies, and the interarterial subtype has been associated with sudden cardiac death.¹ The rate of diagnosis has increased dramatically as a result of improved imaging techniques² and heightened clinical awareness. The mechanism of sudden cardiac death is thought to be arrhythmogenic as a result of ischemia related to inherent anatomic features of the coronary artery.³⁻⁵ Unfortunately, much of what is currently guiding clinical therapy is “logical conjecture,” heretofore limited by the lack of large, protocol-driven analyses that are based on reliable diagnostic imaging and functional studies. As a result, surgical repair is likely overused, and results have been imperfect. Multicenter clinical trials are underway, but the results are likely years in the offing.

The study by Mery and colleagues⁶ in this issue of the *Journal* used a standardized management algorithm informed by the best diagnostic and functional tests available and guided by best evidence and expert opinion-based guidelines, with the goal of analyzing patient outcomes. To create the study population, 162 were evaluated; of these, 44 were prospectively treated and had their results analyzed. As a consequence of this thoughtful process, diagnostic criteria were developed, logical surgical therapies were used, and excellent patient outcomes were achieved. In relatively short-term follow-up, only 1 patient was found to have persistent ischemic symptoms of somewhat uncertain etiology. Mery and colleagues⁶ are to be commended, as they have taken an important first step toward standardizing therapy and gathering information.

The quandary here is that much of this is predicated on multiple uncertainties. Symptoms appear of little use in stratifying risk.⁷ We believe that there are certain malignant subtypes of interarterial coronaries; those with slitlike ostia, acutely angulated, intramural, and crossing the restrictive substance of the intercoronary pillar. Our most frequently

In AAOCA, 90% of optimal patient care is 50% improved technology.

Central Message

Much of our understanding of anomalous aortic origin of a coronary artery is speculative. Only with improved anatomic and functional studies can we optimize care.

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used imaging studies—echocardiography, computed tomographic angiography, and magnetic resonance imaging—are unable to delineate these subtleties reliably.⁸ Preoperative and postoperative measures of ischemia are essential, but our functional studies—exercise testing, stress nuclear perfusion imaging, cardiac magnetic resonance imaging, and computed tomographic angiography—are often nonspecific and unreliable.⁹

Most patients born with anomalous aortic origin of the coronary artery live out their lives unaffected. Recent reports describe more aggressive surgical therapies designed to ensure that all potential malignant anatomic elements are addressed. As a result, reports of significant complications are emerging. Perhaps most disconcerting, some patients are developing more significant ischemia after surgery.¹⁰⁻¹² We need improved imaging and functional technology to bring clarity to the nature and anatomic causes of ischemia, as well as proof of its remedy. Perhaps technology such as intravascular ultrasound⁴ and catheter-based fractional flow reserve¹³ can be made suitable for use in children. Alternatively, improved computed tomographic imaging techniques, computed tomography-based fractional flow reserve,¹⁴ and cardiac stress magnetic resonance imaging⁹ may improve and become clinical viable. Until then, although we “can observe a lot by watching,” we lack the tools to conquer this anomaly.

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