Anomalous Aortic Origin of a Coronary Artery: Surgical Repair With Anatomic- and Function-Based Follow-Up

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Background. **Anomalous aortic origin of the coronary artery (AAOCA) with an interarterial (IAC) course is an uncommon congenital anomaly. Surgical indications and repair techniques have evolved. We have managed 259 adult patients with AAOCA over 40 years. Our management strategy includes anatomic- and function-based surveillance to select surgical candidates. We reviewed our surgical cohort and analyzed anatomic and functional outcomes.**

Methods. **We queried our heart center databases to obtain the names of all patients with AAOCA managed at our institution between 1974 and 2014. We performed a retrospective chart review.**

Results. **Two hundred fifty-nine patients were managed for AAOCA. Sixty-one underwent surgical intervention. Twenty-six with associated coronary atherosclerosis were excluded. Thirty-one who underwent surgical repair were analyzed. Mean age was 42.5 ± 2.7 years. Twenty-four patients (77.4%) had right AAOCA. Six (19.4%) had left AAOCA. One (3.2%) had bilateral coronary anomalies. Repair techniques included 21 unroofing procedures (67.7%), 6 translocations (19.4%), and 4 coronary artery bypass grafting (CABG) procedures (12.9%). Mean follow-up was 3.8 ± 0.8 years. Thirteen patients underwent follow-up anatomic testing with computed tomography. Twelve of these patients had widely patent coronary arteries, and 1 patient had mild coronary artery stenosis. Seventeen patients underwent functional testing. Fifteen of these patients had no evidence of ischemia. One patient had reversible ischemia after CABG, and 1 had subclinical ischemia after unroofing. There was 1 late mortality from endocarditis.**

Conclusions. **Our multidisciplinary program uses a treatment algorithm to select patients with AAOCA for surgical intervention. Only a small subset requires an operation, and we favor unroofing and translocation techniques. With this paradigm, outcomes are excellent, as validated with anatomic- and function-based testing.**

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Coronary artery anomalies compose a small subset of congenital heart defects, which manifest with a wide range of clinical presentations. Importantly, anomalous aortic origin of coronary arteries (AAOCA) is the second most common cause of sudden cardiac death (SCD) in young individuals [1]. In the remote past, the diagnosis was commonly made at autopsy [2]. Although the danger of SCD in AAOCA has been known for decades, the rarity of this condition and the frequent lack of a prodrome have limited the ability to prospectively manage affected patients. Even with a known diagnosis, conventional methods of coronary artery revascularization have been disappointing.

Improvements in imaging modalities combined with increased diagnostic testing and awareness have greatly increased the number of patients diagnosed with AAOCA. Some of these patients present with signs and symptoms of myocardial ischemia. Many patients identified with AAOCA are asymptomatic or have negative provocative testing results for myocardial ischemia (or both), which may or may not be reassuring. Surgical techniques to manage patients with coronary artery anomalies have anecdotally improved, leading several centers to liberalize the recommendation for surgical intervention [3]. We have developed a management strategy using a multidisciplinary team approach to selectively manage patients based on clinical presentation, anatomic risk factors, and encouraging surgical results. The objective of this study was to identify patients referred for surgical management and to assess their clinical, functional, and anatomic outcomes.

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Patients and Methods

We queried the Massachusetts General Hospital Catheterization Lab Database, the Cardiac Surgical Database, the Cardiac Radiology Database, and the Adult Congenital Heart Disease Program Database to identify patients with AAOCA referred from 1974 to 2014. We then performed a retrospective chart review. This study was approved by our institutional review board, and individual patient consent for the study was waived.

This cohort included all patients with anomalous origin of the left main coronary artery (LMCA) or the right coronary artery (RCA), or both, arising from the opposite sinus of Valsalva; patients with a single coronary artery; and patients with a coronary artery arising abnormally high in the sinus of Valsalva or ascending aorta with an angled takeoff or slit-like orifice, or both. Excluded from the analysis were individuals with anomalous coronary arteries arising from the pulmonary artery and patients with concurrent complex congenital heart defects. We further identified all patients who had any surgical procedure to treat the anomalous coronary artery. We excluded patients with AAOCA who underwent coronary artery bypass grafting (CABG) for concomitant atherosclerotic coronary artery disease in the affected coronary artery.

Surgical Techniques

All surgical operations were performed through a median sternotomy using cardiopulmonary bypass, aortic cross-clamping, and intermittent antegrade and retrograde cold-oxygenated blood cardioplegic arrest. Patients were treated with intramural coronary artery unroofing, coronary artery translocation, or CABG. The latter was performed using either pedicled internal mammary artery grafts or saphenous vein aortocoronary bypass grafts with fine polypropylene sutures.

Patients managed by coronary artery unroofing or translocation had their aortic root explored through a partial transverse aortotomy above the sinotubular junction, being careful to identify and avoid an anomalously high coronary artery origin. The coronary ostia were inspected for location within the sinus of Valsalva relative to the aortic annulus and commissures, morphologic characteristics of the orifice (eg, slit-like features and angulation), and for an intramural course (Fig 1). To assess the width and length of an intramural course, graduated coronary artery probes (1.5–5 mm) were carefully passed into the native coronary ostium and through the intramural segment until the probe emerged in the coronary artery within the epiaortic fat. In patients with a long intramural segment, coronary artery unroofing was preferentially performed. Over a coronary probe, the aortic layer separating the intramural coronary lumen from the aortic lumen was excised entirely until epiaortic fat was identified. A neo-ostium was constructed using interrupted 7–0 polypropylene sutures securing full-thickness bites of the coronary artery to all layers of the aortic wall (Fig 2). When the intramural course passed behind the aortic valve commissure, unroofing of the intramural segment was performed on either side of the commissure, leaving the commissure intact. We reinforced the commissure with a pledgeted suture.

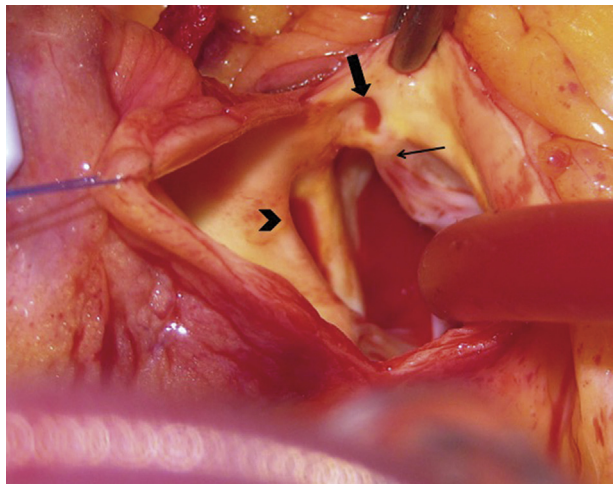

Fig 1. Intraoperative photograph of an anomalous right coronary artery (RCA) arising above and to the left of the intercoronary commissure. The RCA has an angulated slit-like ostium. (Thick arrow = anomalous RCA ostium; thin arrow = intercoronary commissure; arrowhead = left main coronary artery [LMCA]).

In patients with a short intramural course or when coronary unroofing would result in the coronary artery maintaining an interarterial course, coronary artery translocation was performed. After careful identification and mobilization of the proximal coronary artery in the epicardial fat, the coronary artery was transected just as it emerged from the aortic wall. The proximal end was oversewn at its exit site. Using an aortic punch, a new proximal site was created within the proper sinus of Valsalva. The transected end of the coronary artery

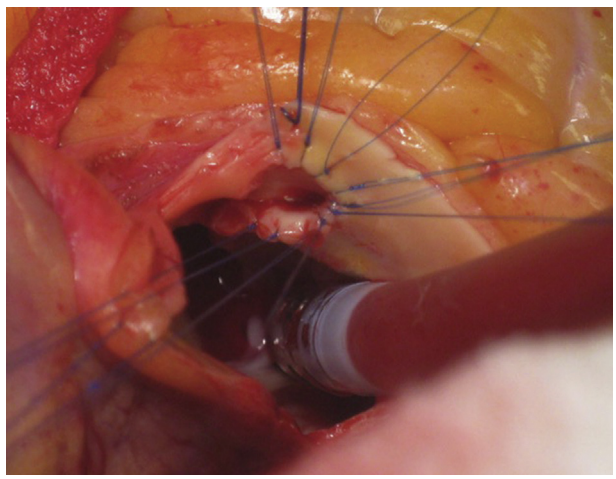

Fig 2. Intraoperative photograph of neo-ostium creation after unroofing. The right coronary artery (RCA) neo-ostium is patulous and no longer possesses the angulated slit-like orifice.