Major Coronary Artery Anomalies in the Pediatric Population

We read with interest the article by Davis et al. (1). The investigators nicely address the problems related to the identification and treatment of these rare coronary anomalies. Although agreeing with the indications for intervention, we believe that aortocoronary bypass grafting may not be the optimal treatment. The long-term patency of arterial grafts (78% at 10 years for internal thoracic artery grafts in the treatment of young patients after Kawasaki disease) (2) may not be satisfactory for children. In addition, given that most of the time the flow through the affected coronary is unimpeded, the graft sets up a competitive flow, which might jeopardize long-term patency. We have taken a different approach, adapted from the translocation of intramural coronary arteries during the arterial switch operation for transposition of the great arteries. The inner wall of the sinus of Valsalva along the intramural portion of the artery is excised to enlarge the ostium. An autologous pericardial patch is used to enlarge the proximal portion of the artery on its external aspect. This is readily accomplished by transection of the aorta and pulmonary artery to obtain a good exposure. This strategy eliminates the acute takeoff angle and external compression of the anomalous coronary, while avoiding competitive flow through a graft, and thus may result in better long-term patency than aortocoronary bypass. We offer this as an alternative to the use of aortocoronary bypass or internal thoracic artery grafting.

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REFERENCES


Reply

The letters from Drs. Davidson and Karl and Dr. Phoon highlight the controversy and difficulties inherent in planning intervention for anomalous origin of the coronary arteries (AOCA). The principal goal of our study was to estimate the prevalence of these anomalies in a relatively normal and asymptomatic population (1). This represents only the first step in defining a disease process. Phoon correctly states that the true natural history of these coronary artery anomalies is unknown. The sporadic nature of AOCA causes difficulties in determining the natural history of these abnormalities when they present as an incidental finding. Conceivably, a national or international registry for such patients could be established with the goal of defining both the natural and surgical history of these patients.

We concur with the management outline described in the editorial comment by Pelliccia (2). Certainly, surgical intervention is mandatory in any young patient with symptoms; or those with AOCA as an incidental finding and signs of myocardial perfusion abnormalities identified after rigorous laboratory examination. Pelliccia states that uncertainty exists regarding patients without symptoms or signs of myocardial ischemia. Therefore, it seems reasonable that these patients are not exposed to risk for sudden death, and surgical management should be considered. One can take issue with Phoon’s statement that “activity can be modified, and therefore risk can be reduced.” Behavioral modification might be possible, but at what price? Our patients and their families are routinely provided the option of reduced physical activity as an alternative to operative intervention. In this population of otherwise healthy young persons, lifestyle considerations tend to direct the families to choose surgical treatment.

The surgical approach depends on the specific anatomy and circumstances. Davidson and Karl suggest that aortocoronary bypass grafting may not be the optimal procedure for AOCA. We agree that arterioplasty involving the ostium is appropriate and perhaps preferable, when the right coronary artery with anomalous origin (ARCA) follows an intramural course. We have performed this operation in two patients with good immediate results. In two other patients, the ARCA did not demonstrate an intramural course, and bypass seemed preferable. We have operated on three patients with left coronary arteries arising from the right sinus, all without intramural component and all were bypassed. All of these grafts have remained patent up to three years postoperatively.

Furthermore, two of the three grafts were performed without cardiopulmonary bypass, thus avoiding possible neurological complications. The long-term results including graft patency duration for these surgical procedures remain unknown. Extrapolation of data regarding Kawasaki disease patients to AOCA patients may not be appropriate (3). Kawasaki disease is a vasculitic syndrome associated with increased risk for thrombosis (4). Vascular remodeling can continue for years (5) and possibly promote graft occlusion. Such vascular pathology does not exist in AOCA patients.