Editorial

Recurrent aortic dissection: a challenging but rare dilemma

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Acute aortic dissection (AD) is a potentially catastrophic disease process, which has undergone significant improvement in treatment options leading to increased overall survival; however recurrent aortic dissection (RAD) is a rare phenomenon that may require a multidisciplinary approach and more complex treatment strategies. Recent observations from the International Registry of Aortic Dissection (IRAD) investigators published in the October 2016 issue of *Circulation* evaluated the demographic, operative, and anatomic characteristics of patients in the database who had a prior diagnosis of AD when they presented with their index dissection (1).

Within the known limitations of a retrospective database review, Isselbacher and colleagues comprehensively compared those patients in the IRAD dataset who had documentation of either the presence or absence of prior AD when they were treated for their index dissection. Of 3828 patients over a nearly 20-year period, only 204 (5%) carried a diagnosis of prior AD. While the cohort of initial AD patients consisted of 67% Stanford Type A dissections, the RAD cohort was slanted towards more Type B dissections (52.5% Type B versus 47.5% Type A). However, the glaring omission here is that since operative details of the first AD in the RAD group are not available, we cannot assume that the Type B "recurrent" dissections are truly new acute Type B dissections and not simply residual Type B dissection with persistence of false lumen blood flow after an initial Type A dissection repair. This seems particularly possible since the investigators noted that recurrent AD patients were more likely to present symptom-free.

The pattern of recurrence (Type A then Type B) was markedly more pronounced in the cohort of patients diagnosed with Marfan Syndrome (MFS). Initial AD in the entire cohort was Type A in 66%, and RAD was Type B in 53%, but in the MFS group of patients, 79% of initial AD was Type A and 62% of RAD was Type B. This finding is consistent with prior reports of patients with MFS who initially present with Type A AD in that a significant portion of this patient cohort will ultimately need the remaining thoracic aorta replaced at some point for either progressive aneurysmal disease or recurrent distal dissection (2).

When the authors examined detailed anatomic, operative, and demographic characteristics of a group of patients with recurrent AD from a three-institution database, the age at presentation was younger for patients who progressed from proximal to distal as compared those who progressed from distal to proximal (initial to recurrent AD). This effect was pronounced and significant in the MFS cohort, and present, although not significant, in the non-MFS group. This implies that in patients who present with AD at a young age, and especially those who present with an initial Type A AD and have a recurrent Type B dissection, may have an underlying vascular connective tissue disorder even if they do not carry a diagnosis of MFS or Loeys-Dietz. Isselbacher and others have thus advocated for formal cardiovascular genetics testing and consultation in any patient who fits into this cohort.

Clearly, RAD is a significant event for the rare individual since it carries a significantly greater risk of aortic reintervention and all-cause mortality at 5 years compared to initial AD. As shown in review of the IRAD database

however, this is luckily a rare event, at most 5% of patients with AD. Unfortunately, what is not collected in the IRAD database and evaluated here in this study is the fate of the aortic root in those patients (often young ones) who presented with Type A dissections. While some groups have shown that the aortic root remains normal after acute Type A repair with root preservation, need for subsequent root replacement carried a 17% mortality risk. Furthermore, 11% of patients had root dilatation meeting criteria for replacement but were not operated on in that study, and patients with MFS were at higher risk for needing reoperation for root replacement if it was not intervened on at the time of initial Type A dissection (3).

As a result of this dilemma, we have advocated for either conservative root repair or valve-sparing root replacement at the time of initial Type A dissection in the appropriate group of patients (4). Previous extensive studies demonstrated the safety of root replacement at the time of AD when the root tissue was involved (5,6). Since these patients tend to be younger, they have the most benefit to gain from preserving the native aortic valve when possible at the time of Type A repair and root replacement. We showed that in appropriately selected patients with good ventricular function, no evidence of malperfusion, and indications for root intervention (sinus destruction, severe valve dysfunction, or root aneurysm >4.5 cm) at the time of AD presentation, David V valve-sparing root replacement is a safe and durable choice with 94% freedom from 2+ aortic insufficiency and 100% freedom from valve replacement at nearly 4 years follow-up (7). Finally, we showed that in a similar group of patients, valve-sparing root replacement conferred a significant mid-term survival advantage with no added operative morbidity or mortality as compared to patients who underwent root replacement with mechanical valve-conduit (8).

Taken together in light of the findings by Isselbacher and colleagues from the IRAD database, a comprehensive management strategy for patients with AD has started to materialize. In young, hemodynamically stable patients presenting with Type A acute AD, valve-sparing root replacement should be the primary surgical option when any valve or root pathology is concomitant. These patients should also be screened for underlying connective tissue disorders to guide future aortic surveillance and screening of family members. The IRAD database also suggests that in any patient with an initial Type A, and to a lesser-extent Type B, dissection, life-long surveillance of the remaining aorta is needed to identify and allow for early intervention on recurrent dissection since these are often asymptomatic.

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Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

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