Proximal aortic aneurysms in patients with conotruncal anomalies: Size matters...but so do other things

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"All's well that ends well"

— William Shakespeare

Two decades ago, Coady and colleagues¹ published their seminal article regarding the appropriate aortic size for thoracic aortic aneurysm (TAA) surgical intervention in the *Journal of Thoracic and Cardiovascular Surgery*. At that time, much had been published on how to do TAA operations, but little was available about the natural history and timing of intervention. They found that the hinge point for complications (dissection or rupture) of an ascending aortic aneurysm was a size of 6 cm. This led to recommending surgical intervention for asymptomatic proximal aortic aneurysms at 5.5 cm. Importantly, they pointed out that age of patient and coexisting disease, as well as the experience of the surgical team should all have a bearing on the decision to operate.

Twenty years later, in this issue of the *Journal*, Dr Vricella² has provided the readership with an excellent review of the key articles supporting the current advice for surgical intervention in nonsyndromic subsets of patients with proximal TAA. The subsets include those with a bicuspid aortic valve (BAV), nonsyndromic young patients with inherited TAA, and patients with root aneurysms after repair of congenital conotruncal anomalies. A strong argument is presented for surgical intervention at 5 cm in patients with BAV with a family history of dissection or a growth rate of 0.5 cm per year. Patients with nonsyndromic TAA are discussed in detail and the literature presented supports a more aggressive timeline to surgery. With regard to the patients with conotruncal TAA, he points out the rarity of proximal aortic complications as well as the limited data and advocates a more conservative approach.

As physicians who treat adult congenital cardiac patients, this latter subset is of particular interest. As noted, it is not uncommon for patients with conotruncal anomalies to have early dilation of the proximal aortic root. Theories for this enlargement include increased aortic blood flow before

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Central Message

The decision to operate on an aortic aneurysm in patients with conotruncal anomalies is dependent on size as well as other factors.

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complete repair and intrinsic vascular structural abnormalities.³ This preexisting dilation is just one of several issues that support a more conservative approach to patients with conotruncal anomalies. It is important to remember that these patients have often had multiple cardiac operations. Performing an aortic root replacement in an adult patient with multi-redo congenital cardiac anomalies is likely associated with a greater surgical risk than patients with nonconotruncal anomalies. Surgical concerns include not only when to operate for aortic enlargement, but more commonly, should the aorta be addressed at the time of operation for another issue, such as pulmonary valve replacement or pulmonary artery reconstruction. Adding an aortic root replacement to a low-risk pulmonary valve replacement in a patient with tetralogy of Fallot certainly adds to the risk. Whether or not adhesions related to multiple cardiac operations is protective is up for debate. Furthermore, the fact that so very few dissections or ruptures have been reported, despite the high number of currently living adult patients with conotruncal anomalies, makes it hard to support an aggressive tactic. Other factors that may play a role in the decision making include aortic aneurysm metabolic behavior on positron emission tomography scan,

aortic mechanical properties on echocardiography, or the presence of specific biomarkers.⁴⁻⁷

In our practice, we have recommended annual imaging with magnetic resonance imaging (MRI) to assess aortic size in patients with conotruncal abnormalities who are found to have aortic dimensions greater than 5 cm on echo. If the patient is unable to have an MRI, we recommend 1-time computed tomography (CT) angiogram to assess aortic dimension. If the patient has similar measurements on echocardiogram, follow-up with annual echocardiography is recommended with repeat CT angiogram only if there is significant change in aortic measurement by echocardiography. Patients who are thought to have rapid expansion of aortic dimension or who have other risk factors for aortic catastrophe warrant more frequent CT angiography. We recommend surgical referral for aortic enlargement when the aorta is 5.5 cm or larger if the patient has no other indication for surgery. After multidisciplinary review, most will be surgical but some may be managed expectantly. If the patient is referred to surgery for a

nonaortic indication, discussion regarding risks and benefits of aortic replacement or reduction should be undertaken when the aorta is 4.5 cm or larger.

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