Type A Aortic Dissection in Marfan Syndrome
A Case for More Aggressive and Extensive Surgery at the Time of the Initial Surgical Operation

Marzia Leacche, MD; John G. Byrne, MD

In the article by Rylski et al in this issue of Circulation, the authors investigate the long-term outcomes of repair for type A aortic dissection in patients with Marfan syndrome.

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Marfan syndrome is one of the genetic syndromes associated with thoracic aortic aneurysms and aortic dissection. The syndrome is the result of fibrillin-1 gene mutation (FBN-1), which is an autosomal-dominant mutation with variable expression. Fibrillin-1 is a large glycoprotein that serves as a structural component of calcium-binding microfibrils. These microfibrils are present in elastic connective tissue such as the medial layer of the ascending aorta and in nonelastic connective tissue throughout the entire body.

The criteria for Marfan syndrome include the presence of clinical findings in the cardiovascular, ocular, and skeletal systems and a positive family history, even though ~25% of patients have a negative family history and FBN-1 mutations status.

Patients with Marfan syndrome are predisposed to have thoracic aortic aneurysms involving the aortic root, ascending aorta, arch, and descending thoracic aorta. The majority of the patients with Marfan syndrome will present with dilation of the aortic root and ascending aorta, and the degree of severity of the disease increases with the extension of the dilation beyond the ascending aorta.

Current guidelines recommend surgical repair for an external diameter of 5.0 cm, unless rapid growth, defined as >0.5 cm/y has occurred or in the presence of significant aortic regurgitation or a family history of aortic dissection. In these situations, a diameter <5.0 cm may trigger the decision to operate. Furthermore, the decision to offer surgical repair often involves other important factors such as age, sex, and body size.

Most patients with Marfan syndrome have proximal aortic root disease. In a recent study, among 86 consecutive patients with Marfan syndrome studied over a 15-year period undergoing aortic surgery, 92% of patients presented with proximal aorta pathology involving the aortic root/ascending aorta/arch, whereas only 8% presented without proximal aortic involvement. Of these 86 patients, 36% presented with acute aortic dissection, 77% with type A and 23% with type B, and 64% had aneurysmal disease.

Although about a third of the patients with Marfan syndrome will present with an aortic dissection, few data exist on the long-term implications of surgical repair. Rylski et al have attempted to provide such data by putting together one of the largest series to date of patients with Marfan syndrome undergoing repair of type A aortic dissection. The data analysis spans 25 years of surgical experience at 2 high-volume centers for aortic surgery, 1 center in Europe and 1 in the United States.

Although the guidelines for repair of aortic root/ascending aorta in patients with Marfan syndrome changed over those 25 years, with treatments becoming more aggressive and the threshold size for surgical repair being lowered, this series provides a unique opportunity to assess the extent and implications of surgical repair for aortic dissection in this unique patient population.

Among 1324 consecutive patients, 74 (18%) underwent surgical repair for aortic dissection. The location of the dissection was proximal (type I DeBakey) in 68% of patients and distal (type II DeBakey) in 32% of patients. Proximal repair techniques included composite valved conduit (55%), supracoronary conduit (30%), and valve-sparing aortic root replacement (15%). Distal repair included hemi-arch replacement in the majority of patients (88%) and total arch replacement in only 12% of patients.

The principal finding of this study is that, at the time of surgery for the type A dissection in patients with Marfan syndrome, a more aggressive surgical approach seems to be superior to a “less is more” strategy. Rylski et al found that patients who underwent full aortic root replacement in addition to ascending aorta replacement experienced superior freedom from reintervention compared with those who underwent ascending aorta replacement alone (95±3%, 88±5%, and 79±5% versus 83±9%, 60±13%, 20±16% at 5, 10, and 20 years, respectively). The median interval between the initial surgery and the reintervention was 8 years (range, 2.2–12.7 years). In other words, leaving the root alone at the time of type A repair in patients with Marfan syndrome may be a formula for need for late reoperation. Interestingly, in the ascending aorta only group, one third of the patients required late redo surgery, not necessarily as a result of progressive aortic root aneurysm but because of significant aortic regurgitation despite aortic valve resuspension at the time of the initial surgery. It is not known whether these patients left the operating room with a completely intact aortic valve or with some degree of aortic insufficiency, necessitating reintervention several years later.
One of the pitfalls of this study is the lack of more specific data on the size of the aortic root at the time of surgical repair, raising the question of whether leaving any aortic root tissue behind is a risk factor for future need of reintervention or if there are more specific characteristics to determine whether the aortic root can be safely left behind at the time of the surgery.

Although more extensive surgery is certainly beneficial to avoid reintervention, it carries more surgical risk for the need of coronary reimplantation in the setting of less-than-ideal tissue resulting from the dissection, in addition to the need for potentially replacing a normally functioning intact aortic valve with either a mechanical valve or a tissue valve unless a David procedure can be performed. Valve-sparing root surgery replacement is certainly an option in a few high-volume centers, but it may not be practical in the majority of the cardiac centers worldwide. Moreover, in younger patients, lifelong anticoagulation would be necessary unless a biological valved conduit is constructed at the time of the surgery or a valve-sparing operation can be performed.

Reintervention for aortic root surgery after aortic dissection repair carried an operative mortality of 22%, whereas operative mortality for the initial surgery was only 3%. These data highlight that aortic dissection repair can be carried out with low operative mortality in experienced centers. The question the authors cannot answer is what the mortality would have been had they implemented the aggressive full root replacement and arch approach from the beginning of the series. Common sense would suggest that it would perhaps have been higher than 3%.

Replacing the entire aortic arch at the time of the initial surgery reduced the percentage of arch reinterventions compared with replacing the hemi-arch (freedom from arch reintervention at 10 years was 100% in the total arch compared with 85±6%). This modest 15% difference seems to confirm the general consensus that total arch replacement should be performed only if strictly necessary. Reducing the time of deep circulatory hypothermic arrest at the initial surgery is probably more important than reducing the rate of future arch reinterventions, because so few are needed.

In conclusion, Rylski et al provide new insight into the surgical approach for the patient with Marfan syndrome at the time of aortic dissection. Confirmation of these findings may help shape future guidelines for surgical treatment of aortic dissection in patients with Marfan syndrome.

Disclosures

None.

References


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