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

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In the manuscript by Rylski et al.\(^1\) in this issue of *Circulation*, the authors investigate the long-term outcomes of repair for Type A aortic dissection in Marfan’s patients.

Marfan’s syndrome (MFS) is one of the genetic syndromes associated with thoracic aortic aneurysms and aortic dissection. The syndrome is the results of fibrillin-1 gene mutations (FBN-1) which is an autosomal dominant mutation with variable expression. Fibrillin 1 is a large glycoprotein that serve 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 as well as in non-elastic connective tissue trough ought the entire body.

Marfan’s criteria include the presence of clinical findings in the cardiovascular, ocular and skeletal systems, and a positive family history, even though about 25% of patients will have a negative family history as well as FBN1 mutations status\(^2\).

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

Current guidelines\(^3\) recommend surgical repair for an external diameter of 5.0 cm, unless rapid growth has occurred, defined as >0.5 cm per year, or presence of significant aortic regurgitation or 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, gender and body size.

Most Marfan’s patients have proximal aortic root disease: In a recent study\(^4\), among 86 consecutive Marfan’s patients studied over a 15 year period undergoing aortic surgery, 92% of patients presented with proximal aorta pathology involving aortic root/ascending aorta/arch,
while only 8% presented without proximal aortic involvement. Of these 86 patients, 36% presented with acute aortic dissection, with 77% type A and 23% type B, and 64% with aneurysmal disease.

While about a third of Marfan’s patients will present with an aortic dissection, few data exists on the long-term implication of surgical repair. Rylski et al have attempted to answer this question by putting together one of the largest series to date of Marfan’s patients undergoing repair of type A aortic dissection. The data analysis spans 25 years of surgical experience at two high volume centers for aortic surgery, one in Europe and one in the United States.

Notwithstanding the fact that the guidelines for repair of aortic root/ascending aorta in Marfans’ patients have changed over the 25 years, with treatments becoming more aggressive and lowering the threshold size for surgical repair, this series gives an 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%), supra-coronary 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 principle finding of this study is that, at the time of surgery for the type A dissection in Marfans patients, 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 re-intervention compared to
those who underwent ascending aorta replacement alone (95 ± 3%, 88 ± 5, 79 ± 5% at 5, 10, 20 years vs. 83 ± 9%, 60 ± 13%, 20 ± 16%). The median interval between the initial surgery and the re-intervention was 8 years (range 2.2 years-12.7 years). In other words, leaving the root alone at the time of type A repair in Marfans patients may be a formula for need for late reoperation. Interestingly, in the ascending aorta only group, 1/3rd of patients required late redo surgery not necessarily due to progressive aortic root aneurysm but because of significant aortic regurgitation despite undergoing aortic valve re-suspension at the time of the initial surgery. It is not known if these patients left the operating room with a completely intact aortic valve or with some degree of aortic insufficiency and thus the need of re-intervention 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 if leaving any aortic root tissue behind is a risk factor for future need of re-intervention or there are more specific characteristics, to determine if the aortic root can be safely left behind at the time of the surgery.

While more extensive surgery is certainly beneficial to avoid re-intervention, it carries more surgical risk for the need of coronary re-implantation in the setting of less than ideal tissue because of the dissection, but also the need of 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, life-long anticoagulation would necessary unless a biological valved conduit or valve sparing operation is constructed at the time of the surgery.

Re-intervention for aortic root surgery after aortic dissection repair carried an operative mortality of 22%, while 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. What 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 have perhaps been higher than 3%.

Replacing the entire aortic arch at the time of the initial surgery reduced the percentage of arch re-interventions compared to replacing the hemi-arch (freedom from arch re-intervention at 10 years was 100% in the total arch compared to 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 re-interventions, since so few are needed.

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

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References:


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