AORTIC REPAIR IN MARFAN SYNDROME: LET'S NOT FORGET THE ARCH WHEN TALKING ABOUT THE ROOT

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To the Editor:

We read with great interest the editorial commentary by the David group regarding our article about aortic root repair in patients with Marfan syndrome.[1,2] It was indeed Dr David's honest criticism and kind advice that motivated us to continue and to modify the patient-tailored root repair (PTRR) technique, as he and others similarly did with the original David procedure.[1,3,4] Even if we are still not able to compare the long-term results between our PTRRs and those reimplantation technique cases initiated more than 20 years ago, we can look at ever increasing follow-up data. Meanwhile, the combined 12-year freedom of aortic replacement and relevant aortic insufficiency (>2+) in 289 patients who received a successful PTRR in the years between 2000 and 2010 is 96.1 ± 1.2 (unpublished data). Nevertheless, we are reluctant to generalize any comparisons because the series of true Marfans is small, the pathologic and clinical presentations are very different, and consequently, the reports are biased by patient and surgeon selections.

There is another aspect we would like to discuss because the reviewers of our article have already asked us why so many series patients had received a complete aortic arch replacement. We answered the reviewers, but we were not able to discuss this topic in the article because it would go beyond its scope.[2] Our experience and the evaluation of our data revealed to us that it was not the aortic root or the valve that was responsible for surgical problems after primary valve-sparing surgery. The sequelae of limited root surgery are not comparable with those after limited aortic repair.[E1,5] Replacement of the aortic valve or even the entire root after previous ascending aorta repair is neither life-threatening nor a surgical challenge. Replacement of an acutely dissected aortic arch is. Consequently, we concomitantly replace even a nondilated arch in patients with Marfan syndrome who have a positive family history or who present with any signs of distal aortic pathology (dilatation, dissection). Figure 1 demonstrates an angio-computed tomography of a female patient with

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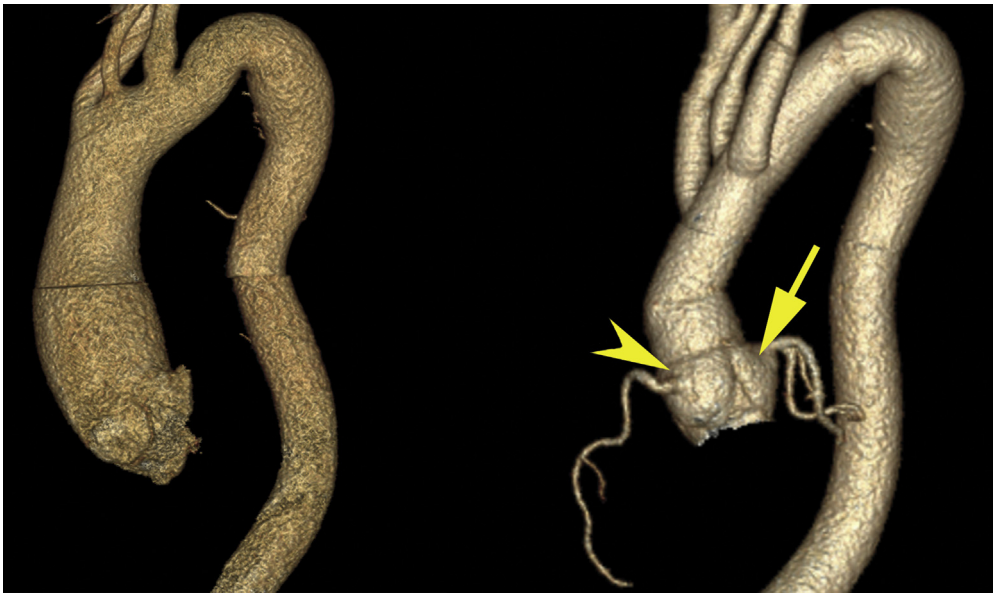

FIGURE 1. Preoperative and postoperative 3-dimensional volume rendering technique reconstructions of contrast-enhanced computed tomography scans (computed tomography angiography) of 52-year-old woman with confirmed diagnosis of Marfan syndrome who received patient-tailored aortic root repair with replacement of noncoronary and right coronary (marked with *arrowhead*) sinus, leaving the left coronary sinus (marked with *arrow*) untouched. A total arch replacement was performed concomitantly because the patient presented with dissection history of abdominal aorta and positive familial history.

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Marfan syndrome who fulfilled these criteria and received arch replacement concomitantly to PTRR with replacement of only 2 sinuses. In contrast, Figures E1-E3 demonstrate an odyssey that patients with Marfan not infrequently experience after limited aortic surgeries. Yet, we are aware that any generalization of the indications for arch replacement can be problematic, especially because the reported risk of conventional aortic arch replacement differs substantially between surgical centers.[E2] Even if this does not seem credible to all members of our community,[E3] it is a fact, however, that elective aortic arch replacement performed by experienced surgeons using modern, albeit simplified, techniques can offer excellent results.[E4] Having reliable data about the surgical risk of elective arch replacement in Marfan syndrome (preferably from multicenter registries) and about the natural history of acute dissection after previous root repair would make it possible to develop evidentiary recommendations for aortic arch surgery, taking into account patient- and surgeon-related risks, as has been provided for decades for asymptomatic carotid stenosis surgery.[E5]

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REPLY TO "AORTIC REPAIR IN MARFAN SYNDROME: LET'S NOT FORGET THE ARCH WHEN TALKING ABOUT THE ROOT": Reply to the Editor:

Urbanski and colleagues' letter on aortic valve sparing in patients with Marfan syndrome (MFS) brought up 2 issues. One was in support of their selective approach to replace 1, 2, or all 3 aortic sinuses[1] during remodeling of the aortic root. Most surgeons with experience in treating patients with MFS and other connective tissue disorders would disagree with this approach because if one follows them long enough, the aortic annulus continues to dilate and aortic insufficiency ensues.[2] The second issue was how radical one should be with the distal aorta during aortic root surgery in patients with MFS. In their series of 42 patients they "replaced the arch" in 10 patients, in 1 patient because of acute dissection during aortic cannulation and 4 in patients presenting with acute dissection. It is unclear how many of the remaining 5 patients had an aneurysmal or dissected aortic arch. Only 3 patients had total arch replacement; the remaining patients had "hemiarch replacement."

Complications of the aortic root aneurysm are the leading causes of death in untreated patients with MFS, and in our opinion complete aortic root replacement with reimplantation of the aortic valve remains the most effective and durable approach to prevent proximal aortic dissection and death in these patients. The need for distal intervention in patients with MFS is far greater in those initially presenting with dissection compared with aneurysmal disease.[3] Replacement of the aortic arch (hemiarch or total arch) does not guarantee prevention of dissection of the remaining thoracic aorta. In an observational study on 600 patients with MFS from a Dutch registry, den Hartog and colleagues[4] examined the issue of distal aortic dissections, which occurred in 54 patients for an annualized rate of 1.5%. Of note, no arch dissections occurred in this group. Replacement of the proximal aorta with a noncompliant graft may result in greater pulsatile forces in the distal arch and proximal descending thoracic aorta, increasing the risk of dissection. Currently available data do not support prophylactic replacement of the aortic arch in patients with MFS with a nondilated arch or positive family history dissection.

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