Authors have nothing to disclose with regard to commercial support.

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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 sinuses¹ 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 colleagues⁴ 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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TECHNIQUE OF CIRCULATORY ARREST MAKES A DIFFERENCE

To the Editor:

I enjoyed the recent article by Damberg and colleagues¹ and the accompanying editorial commentary by Kouchoukos² regarding the ongoing differences of opinion regarding the application of deep hypothermic circulatory arrest during aortic surgery in adults. The same controversy continues among congenital surgeons. However, I was surprised to see both in the article by Damberg and colleagues¹ as well as the editorial commentary by Kouchoukos that the only technical detail of circulatory arrest that was discussed was its duration. I was also surprised to see that the Yale group continues to use the alpha-stat pH strategy despite good evidence that this provides less neuroprotection than the alternative pH stat strategy (ie, CO₂ addition) and in fact is likely to shorten the safe maximal duration of circulatory arrest.

The alpha-stat strategy is appropriate for normal flow cardiopulmonary bypass in adults because it reduces the microembolic load to the brain by more closely linking cerebral blood flow with cerebral oxygen requirement.³ However, cerebral blood flow has 2 functions for the patient undergoing deep hypothermic circulatory arrest. In addition to supplying oxygen and other substrates, cerebral blood flow also cools the brain to deep hypothermia. Cerebral blood flow is reduced by use of the more-alkaline alpha-stat strategy so that the total duration of cooling must increase to compensate. In fact, the total volume of blood (ie, CBF in mL/min × time) needed to cool the brain to the desired deep hypothermic temperature will be the same with either alpha-stat or pH-stat.

There are additional reasons why pH-stat should be preferred.

1. pH-stat is known to suppress cerebral metabolism.⁴ Studies in our laboratory⁵ confirmed that pH-stat lengthens the safe duration of deep hypothermic circulatory arrest for a given temperature and hematocrit.
2. pH-stat improves oxygen availability by counteracting the leftward shift of oxyhemoglobin induced by hypothermia. This is particularly important in the early cooling phase, when the brain is warm but the blood is cold.
3. The only prospective, randomized clinical trial of pH strategy during deep hypothermic bypass that included patients undergoing deep hypothermic circulatory arrest demonstrated an improved perioperative clinical outcome with pH-stat.⁶ Although this trial was in infants, in view of the aforementioned points, we believe the conclusions can be extrapolated to adults undergoing deep hypothermic circulatory arrest.

In addition to the importance of pH strategy, our laboratory studies over many years as well as 2 randomized prospective clinical trials in infants demonstrated that hematocrit is also critical in determining the safe maximum duration of circulatory arrest. Interestingly the Yale article fails to mention the level of hematocrit that was applied. There is no question that the outdated dogma that hemodilution improves the safety of circulatory arrest is unsupported by any current data. In fact, our clinical trials demonstrated that a hematocrit greater than 23.5% was required to avoid neurodevelopmental impairment in infants undergoing deep hypothermic bypass with or without circulatory arrest. Furthermore, our laboratory studies demonstrated an additive impact of low hematocrit, alkaline pH strategy (alpha-stat), greater temperature, and longer duration of circulatory arrest. At a minimum, adult centers undertaking deep hypothermic circulatory arrest in addition to those proposing alternative methods of neuroprotection should undertake a prospective randomized study with careful cognitive assessment before and after surgery to investigate optimal pH strategy and hematocrit during deep hypothermic circulatory arrest. We need to move beyond a simple debate of safe maximal duration.

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