MFS groups. Another interesting and clinically relevant observation was that 5 patients in the MFS group and 4 patients in the LDS group still developed type A aortic dissection after aortic root replacement.

Based on the findings in this study, along with similar findings from Hopkins group⁴ and a report of 441 patients with *TGFBR1* and *TGFBR2* mutations from the Montalcino Aortic Consortium,² we would agree that as much of the ascending aorta as possible should be resected during prophylactic aortic root replacement in patients with LDS or MFS. More arch replacement could be performed in patients with LDS, but it is still not clear whether either a hemiarch or total arch replacement should be routinely performed. Just like MFS, not every patient with LDS behaves the same due to different gene mutations (*TGFBR1* vs *TGFBR2*) and mutations at different sites in the same gene. More aggressive prophylactic aortic root and arch replacements should be considered in patients with LDS with arterial/aortic tortuosity and severe extra-aortic features,

which may serve as biomarkers of more severe and aggressive vascular disease.² A less-aggressive surgical management strategy could be pursued in the absence of any familial event or rapid increase in aortic diameter, especially in women harboring *TGFBR1* mutations who on average have later onset aortic disease than men with *TGFBR1* mutations.²

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Commentary: Prophylactic total arch replacement in Loeys–Dietz syndrome: Perfect may be the enemy of good

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In this issue of *The Journal*, Seike and colleagues¹ compared the surgical fate of 123 patients with 2 genetically distinct heritable aortopathies (HAPs): Marfan syndrome (MFS) and Loeys–Dietz syndrome (LDS). The authors

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CENTRAL MESSAGE

In experienced hands, prophylactic total arch replacement may reduce the need for reoperation in patients with Loeys–Dietz syndrome.

should be commended for their outstanding surgical skills, which resulted in no operative mortality or permanent neurologic deficit in this complex cohort. The reoperation rate for native aortic segments at 10-year follow-up was greatest for total arch replacement (TAR) in patients with

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LDS (22% vs 12%, $P < .01$). There was no difference between groups with respect to reoperations for the aortic root or distal aorta. When dissection was the reason for the index procedure, the risk for reoperation was greater for the entire study cohort. When acute dissection was not present, patients with LDS incurred both greater late dissection and reoperation rates during follow-up. On these grounds, the authors recommend prophylactic TAR in all patients with LDS and in patients with MFS presenting with acute type A aortic dissection (ATAAD).

However, there are a number of concerns for the conclusions drawn from this comparison of 2 HAP groups. Besides the fact that the LDS group (24 patients, 19.5%) was notably under-represented compared with the MFS group (99 patients, 80.5%), their complement of index procedures was also profoundly different. Distal aorta replacement was performed in a significantly greater percentage of patients with LDS (37.5% vs 10.1%, $P < .01$), whereas aortic root replacement was preponderant in MFS (85.9% vs 50%, $P < .01$).

The authors' proposition that ATAAD is a predictor of late reoperations, particularly TAR, in HAP is corroborated by recent literature.^{2,3} The latter also reiterates that the occurrence of ATAAD elevates the risk of reoperation in MFS, equalizing it to the one observed in LDS.³ Although upon these considerations experienced aortic centers may be enticed to consider prophylactic TAR during ATAAD repair,⁴ prudence is required in cajoling the broader surgical community into such endeavors. Perfection may, in fact, be the enemy of good in critical contingencies such as the ones epitomized by ATAAD. Indeed, even in experienced hands,

prophylactic TAR increases the already-substantial operative mortality of ATAAD repair.⁵ Conversely, a generous hemiarch replacement during ATAAD repair provides a freedom from TAR of 85% at 10-year follow-up in HAP.⁶ The outcomes of ATAAD repair are highly dependent on relative volume of the center,⁷ and adding prophylactic TAR will likely exacerbate the gap in both surgeon and institutional experience. Fortunately, it is likely that an eventual reoperative TAR will be performed electively in an aortic center with a low surgical risk.⁸

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