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Commentary: Tissue may not be the issue in patients with Marfan syndrome and acute type A aortic dissection: Lessons from the German Registry for Acute Aortic Dissection Type A

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One of the most robust acute type A aortic dissection registries is the German Registry for Acute Aortic Dissection Type A (GERAADA), which over the years (2006-2015) has helped us evaluate outcomes in these patients.^{1,2} In a new report from the GERAADA, Farag and colleagues³ identified, from a total of 3385 patients who underwent surgery for acute type A aortic dissection, 117 patients (3.5%) diagnosed with Marfan syndrome (MFS). Although this sample may be too small to definitively answer questions regarding early outcomes in patients with MFS, the importance of this study is the multicenter registry data.

The study brings to light several important points. For one, no differences were observed in early outcomes between the patients with MFS and the remaining patients in matched or nonmatched cohorts. The intraoperative surgical challenges that are commonly encountered with friable tissue in patients with MFS were not enough to increase significantly their operative mortality, perhaps because the patients with MFS were younger and had fewer comorbid conditions than the patients without MFS. On

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CENTRAL MESSAGE

Data are limited regarding surgical outcomes in patients with Marfan syndrome and acute type A aortic dissection, so multicenter registry data such as these are valuable.

review of the International Registry of Acute Aortic Dissection data, de Beaufort and colleagues⁴ reported no significant difference in hospital mortality between patients with and without MFS and MFS.

Another point that Farag and colleagues' study³ highlights is the differences among the surgical procedures performed. The extent of aortic resection varied, but the results and the study's 30-day outcomes did not differ by extent of resection. Proximally, in the matched patients (ie, all patients with MFS and a set of matched patients without MFS), a supracoronary graft was placed in one-half of the patients, whereas the other half underwent some sort of root repair or replacement. The number of patients with MFS who had more extensive distal procedures involving part or all of the aortic arch was higher than expected, being approximately equal to the number in the patients without MFS, both in the matched and the nonmatched cohorts. This is an interesting finding given that patients without MFS generally have a larger root diameter proximally but a relatively normal aortic diameter distally. One would expect a higher percentage of patients with MFS to have root repair or replacement and possibly more ascending aorta replacement than arch replacement. A caveat is that the definition of "arch repair" included anything from open distal anastomosis to partial arch replacement ending at different levels of the supra-aortic vessels.

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Regarding the preserved aortic root after treatment of acute type A aortic dissection, root preservation and ascending aortic replacement lead to low reoperation and root dilatation rates except in patients who are aged less than 60 years or have MFS.⁵ Freedom from arch repair in patients with MFS with residual arch dissection after aortic root replacement was 49% at 15 years in one of the earlier studies.⁶ In our own experience, over a period of 8.5 years in 137 consecutive patients (approximately one-third of whom had heritable thoracic aortic disease) with prior acute type A aortic dissection who underwent proximal and total aortic arch procedures, we found respectable morbidity and survival.⁷

The brain protection strategy, with approximately one-third of patients receiving only hypothermia as the main adjunct for brain protection, was another compelling point.³ This is particularly interesting given that in a survey conducted a few years ago, the typical aortic arch repair in Europe was done with moderate hypothermia and bilateral antegrade cerebral perfusion.⁸ According to another, more recent report from 2 aortic referral centers in Germany, in a large cohort of patients undergoing surgery for acute type A aortic dissection, unilateral or bilateral cerebral perfusion under mild or moderate hypothermia was used.⁹ It is our strategy to use bilateral antegrade cerebral perfusion in addition to moderate hypothermia.⁷ By default, in all patients with acute type I aortic dissection, we always use open distal anastomosis with a range of surgical aortic arch resection and repair depending on the anatomy.

Regarding the aortic root, we are more inclined to repair or replace the root in patients with MFS, either because it is dilated to some repair threshold or to prevent further dilatation in the future.⁷

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