

### Timing of the surgery in patients with Marfan syndrome and definition of the aortic aneurysm

To the Editor,

We read the paper "Case images: huge ascending aortic aneurysm" by Demirkol et al.<sup>[1]</sup> with great interest. Given the central role that Marfan syndrome (MS) plays in the progression of ascending aortic aneurysm, the question as to whether earlier surgery might favorably modify this disease process is an important one. The authors lead the readers to believe this, since the patient has a known history of MS. However, it is difficult to understand why the physicians did not refer the patient to surgery until the aneurysm reached 10 cm in diameter. According to many authors, the timing of the surgery is important because it is possible to carry out valve-sparing surgery in the earlier stages of the disease.<sup>[2]</sup> This significantly reduces the risk of mortality, since elective aortic-root replacement has a low operative mortality. In contrast, emergency repair, usually for aortic rupture or acute dissection, is associated with a much higher mortality.<sup>[3]</sup>

The kind of surgical technique used in an aortic root of that size is another point that arouses interest. It was stated in the manuscript that only the aneurysm was excised, and the aortic valve was replaced with a valved conduit. Considering the size of the aneurysmal sac and the aortic root, it is almost impossible to replace the valve in such conditions without aortic root replacement.

In addition, the aortic aneurysm was regarded as greater than 10 cm in diameter in the paper.<sup>[1]</sup> A nor-

mal diameter for the ascending aorta has been defined as 20 to 37 mm depending on the height, weight, body surface area, and body mass index. Many authors define aneurysm as a vessel diameter larger than 1.5 times the normal diameter.<sup>[3]</sup>

In conclusion, MS is a multisystemic disease with cardiovascular complications that are recognized to be the major cause of morbidity and mortality. Improvement in life expectancy in MS patients noted over the past 30 years is largely due to early recognition of aortic aneurysmal disease and prophylactic aortic root replacement. Thus, it is important to consider the diagnosis and perform serial cardiovascular imaging studies as an essential part of systematic follow-up.

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### Authors reply

To the Editor,

We would like to thank the authors for their valuable suggestions about our case image.<sup>[1]</sup> The authors asked why the physicians did not refer the patient to the surgery until the aneurysm had reached 10 cm in diameter. Our case was diagnosed with Marfan syndrome at the age of 10 years, and had not undergone cardiac evaluation until the time of surgery.

Aortic root disease is the main cause of morbidity and mortality in patients with Marfan syndrome. It can be detected in 50-60% of adults and 50% of children. Echocardiography has an important role in the early diagnosis of aortic aneurysm and can also assess other cardiovascular manifestations of Marfan syndrome. Therefore, it is used in monitoring patients during follow-up. Because of the increased risk for aortic dissection, prophylactic surgery is recommended when the diameter of the aortic sinuses of Valsalva reaches 5.0 cm. We did not define the aortic aneurysm