Commentary: Beyond the edge of the map

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In this issue of the *Journal*, Schoenhoff and colleagues¹ present their analysis of 100 consecutive patients with Marfan syndrome, all of whom fulfilled the revised Ghent criteria.² These patients were followed up for more than a decade and underwent 34 interventions related to nonaortic arterial segments, in addition to 158 thoracic, thoracoabdominal, and abdominal aortic interventions. The authors point out that little is known regarding the incidence and clinical relevance of nonaortic arterial aneurysms in patients with Marfan syndrome, but that these aneurysms may be more common than many realize. Peripheral aneurysms such as these have received more attention in patients with other connective tissue disorders (eg, Loeys-Dietz syndrome, Ehlers-Danlos syndrome). Notably, there are few data on the size of peripheral arteries, even in healthy individuals. Whereas efforts have been made to determine normal ranges of aortic diameter by age, gender, body surface area, and other parameters, this depth of information is simply not available for peripheral artery dimensions. The authors relied on arterial reference values provided by Johnston and colleagues,³ working on behalf of the subcommittee on reporting standards for arterial aneurysms for the Society of Vascular Surgery. Because this source did not include reference values for the dimensions of the subclavian artery, the authors turned to the work of Thrush and Hartshorne,⁴ who chiefly used peripheral vascular ultrasonography to obtain these values.

It is well known that the aortopathy of Marfan syndrome, as well as other connective tissue disorders, requires life-long surveillance and presents a strong possibility of future need for intervention. In recent decades, and largely because of successful efforts to replace the aortic root prophylactically, the lives of patients with Marfan syndrome have been prolonged such that issues beyond the root, such as concerns related to the peripheral arteries, have become more important. In particular, Schoenhoff and colleagues¹ describe their experience regarding 48 patients with aortic dissection, 33 of whom had 66 related or

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Central Message

Nonaortic disease sometimes occurs in patients with Marfan syndrome. Commonly, it manifests as dissection of the iliac arteries, but aneurysm of any peripheral artery remains a possibility.

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unrelated dissections in nonaortic arterial segments. The majority of these dissections affected the iliac arteries (40/66, 61%). During the follow-up period, the authors carried out 34 interventions in these nonaortic arterial segments in 19 patients. The majority (19/34; 56%) of these interventions were for the iliac arteries. Although the methods of intervention were not clearly specified, interventions were performed by open or endovascular approaches (only the use of coiling was clearly described). The authors importantly point out that in addition to there being little information on normal arterial dimensions, there is virtually no information on diameter-based thresholds for intervention. Notably, such thresholds should be set according to the cause, anatomic location, growth rate, and structure of the aneurysm, as well as other patient-specific factors. Despite the general lack of information, ultimately, the authors state that they operate on peripheral arterial aneurysms in patients with Marfan syndrome once the aneurysm has reached a diameter of 2.5 to 3 cm, and largely regardless of its specific location.

Importantly, this article highlights the paucity of information on peripheral arterial aneurysms for virtually all forms of connective tissue disorder. Hopefully, it will stimulate further investigation into the size of normal peripheral arteries, as well as the natural history of peripheral aneurysm in patients with and without connective tissue disorders and those with and without dissection. Expert consensus documents generally contraindicate the use of endovascular

aortic repair in patients with a connective tissue disorder.^{5,6} Much less is known about the endovascular treatment of peripheral arterial aneurysm in patients with Marfan syndrome, a technique that has proved valuable in treating atherosclerotic and other forms of peripheral aneurysm.

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