Peripheral aneurysms in Marfan patients are common and are related to age and advanced aortic disease.

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BACKGROUND
Peripheral aneurysms are not included in the diagnostic criteria for Marfan syndrome (MFS); however, their real prevalence in MFS is unknown. Furthermore, they are commonly seen in other genetic entities such as Loeys-Dietz syndrome.
We aimed to investigate the prevalence of peripheral aneurysm in Marfan syndrome.

METHODS
Patients with clinical criteria of Marfan syndrome and identified FBN1 mutation were evaluated. Only patients with either MRI or CT angiography assessing peripheral vessels were included in this study. MRI and CT angiography studies were retrospectively evaluated to detect the presence of peripheral aneurysms. Aortic dissection-related arterial dilations were excluded. Aortic events and those related to aneurysm complications were collected during follow-up.

RESULTS
Two hundred and nine patients with Marfan and FBN1 mutation were evaluated. Of these 136 (65.1%) had undergone either MRA or CTA with peripheral artery study during follow-up. Mean age at the last follow-up visit was 42.4±14.1yrs; 54.4% were men, and mean follow-up 7.3±3.1 years. Sixty-six aneurysms were identified in 42 (30.9%) patients. The most common locations were the iliac arteries in 23. The rest were: renal (7), vertebral (5), splenic (5), coeliac (3), brachiocephalic (1), subclavian (3), carotid (3), axillary (2), internal mammary (3), femoral (2), hypogastric (3), bronchial (2), coronary (1), hepatic (1), lumbar (1), gastroduodenal (1) and popliteal (1). Twenty-six patients (61.9%) had more than one peripheral aneurysm, and only 4 required surgery.

Patients with peripheral aneurysms were older (47.2±14.3yrs vs 40.2±13.6yrs, p=0.06) and more frequently men (69.0% vs 47.9% p=0.026). Although patients with peripheral aneurysms did not more frequently have aortic dissection (16.7% vs 17.0%, p=0.586), they did more frequently have aortic surgery (73.8% vs 47.9% p=0.05).

CONCLUSIONS
Peripheral aneurysms are present in one third of Marfan syndrome patients and are related to age and more advanced aortic disease. Systematic use of whole-body vascular assessment in Marfan patients can provide a comprehensive evaluation of the entire arterial system, identifying other sites of vascular involvement at risk of potential complications, and the subgroup of patients with more aggressive vascular disease expression.