

Non-Dissecting Distal Aortic and Peripheral Arterial Aneurysms in Patients With Marfan Syndrome

BACKGROUND: In Marfan syndrome (MFS), an aortic or peripheral arterial dilatation is usually the consequence of aortic dissection. Non-dissecting distal aortic and peripheral aneurysms (DAPA) are barely described. We sought to determine the incidence and prognostic impact of non-dissecting DAPA, requiring a surgical repair in a large population of patients with MFS.

METHODS: The patients referred to the French MFS reference center were included in a prospective database, and the patients treated for a non-dissecting DAPA between 2013 and 2020 were retrospectively reviewed. The first-line therapy was open surgery. The patients unfit for open repair or experiencing life-threatening complications underwent endovascular repair.

RESULTS: Among 1,575 patients with MFS, 19 (1.2%) were operated for 25 non-dissecting DAPA. The mean age was 42.4 ± 11.5 years. Non-dissecting DAPA involved the subclavian or axillary artery ($n = 12$), the descending or thoracoabdominal aorta ($n = 6$), the abdominal aorta and iliac arteries ($n = 6$), and the popliteal artery ($n = 1$). Open and endovascular repairs were performed in 22 and three cases, respectively. After a median follow-up of 54.2 months, no local recurrence was noticed and no secondary procedure was performed. Eight patients presented a new aortic event, including two aortic dissections and seven new aortic surgeries. Compared to the overall MFS population, the non-dissecting DAPA group presented a significantly higher risk of an aortic event (100 vs. 28%, $p < 0.0001$), a higher risk of aortic dissection (53 vs. 8%, $p < 0.0001$), and a higher rate of pejorative genetic mutations (68 vs. 40%, $p = 0.011$).

CONCLUSION: Among the patients with MFS, the diagnosis of non-dissecting DAPA is infrequent but is associated with a significant adverse outcome, thus, advocating for a specific follow-up.