

**Incidence and Etiology of Secondary Surgical Interventions on the Aorta in Marfan Syndrome**

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**Objectives:**

Patients with Marfan syndrome (MFS) frequently require interventions on the distal aorta. Aim of the current study was to determine incidence and etiology of secondary surgical interventions in downstream aortic segments in MFS after previous aortic repair.

**Patients and Methods:**

Data were prospectively collected from 86 MFS patients fulfilling Ghent criteria that underwent a total of 136 operations and were followed at this institution between 1995 and 2010.

**Results:**

Mean follow-up of survivors was  $8.8 \pm 6.8$ y, mean time-to-re-operation was  $5.5 \pm 4.6$ y. Thirty-day, 6 months, 1 year and late mortality was 3.5%, 5.8%, 7.0% and 12.8%, respectively. Four out of these 11 deaths (36%) were due to aortic rupture during late follow-up.

Seventy-eight patients (91%) primarily presented with root, ascending or arch lesions, whereas 7 patients (8%) presented with thoracoabdominal lesions. Etiology at primary presentation was acute dissection in 36% [24 (77%) type A, 7 (23%) type B] and chronic dilative disease in 64%. Secondary arch replacement had to be performed in only 6% of patients in the non-dissected, but in 36% of the dissection group ( $p=0.0005$ ).

In the non-dissection group, 11% of patients underwent surgery in downstream aortic segments [5 out of 6 patients suffered from type B dissection in the meantime], whereas in patients after acute dissection, 48% patients had to undergo surgery on the distal aorta [42% of patients with type A and 86% of those with type B dissection] ( $p=0.0002$ ).

**Conclusion:**

In a contemporary cohort of patients with MFS, a third of the patients still presents initially with acute dissection despite the wide availability of prophylactic surgery. The current data suggests that in patients with MFS, the need for repeated surgery in downstream aortic segments is not determined by the segment of the aorta that is initially involved but rather the presence or absence of acute dissection.