# Marfan Syndrome Is Associated With Recurrent Dissection of the Dissected Aorta

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*Background.* **Recurrent dissection of a previously dissected aortic segment has been reported as a rare, late complication in single case reports. The infrequency of this event makes informed risk assessment in an individual patient challenging.**

*Methods.* **To investigate this issue we examined the database of the Massachusetts General Hospital Thoracic Aortic Center between January 1, 2003 and December 31, 2012. A retrospective review was performed to identify patients with both (1) an acute aortic dissection after a prior aortic dissection and (2) evidence of a new dissection within a previously dissected aortic segment creating a triple lumen aorta. Data were reviewed to identify factors predisposing to dissection of a previously dissected aortic segment.**

*Results.* **Over a 10-year period we identified 5 cases of aortic dissection within a previously dissected aortic segment presenting as a new acute aortic syndrome. On average, the recurrent dissection occurred 1 decade after the first aortic dissection (mean = 9.8 ± 1.9 years). Patients identified in this series were significantly younger at first dissection and more likely to carry the diagnosis of Marfan syndrome. Aortic aneurysm diameter was quantified before and after the new dissection event and demonstrated a marked increase in aneurysmal size (mean increase = 1.6 ± 0.3 cm).**

*Conclusions.* **We conclude that medial degeneration, as seen in the Marfan aorta, represents a predisposing factor for recurrent dissection of the dissected aorta. Our data indicate that double aortic dissections cause significant arterial destabilization and a low threshold for surgical intervention is appropriate.**

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Aneurysmal degeneration is the most worrisome late complication of thoracic aortic dissection. Late aneurysmal degeneration and the subsequent need for aortic replacement complicate at least 40% of all dissections, irrespective of initial medical or surgical treatment [1–10]. Between 10% and 20% of patients who have undergone surgical repair for type A dissection require repeat aortic repair within 10 years of the first procedure [11]. Recurrent dissection of a previously dissected segment of the aorta is an extremely rare occurrence, and only a handful of cases have been reported in the literature [12–15]. Risk factors for such recurrent dissections are not clearly defined from individual case reports, although connective tissue disorders are suspected as the affected patients have been young and carry a diagnosis of Marfan syndrome (MFS) [16]. In this series, we describe 5 cases of recurrent aortic dissection occurring within a previously dissected segment. All patients presented with a recurrent acute aortic syndrome accompanied by a significant increase in aortic size that prompted operative repair.

## Material and Methods

All cases of acute aortic dissection at Massachusetts General Hospital are entered into a Thoracic Aortic Center database. We examined all records in the database over the 10-year period from January 1, 2003 through December 31, 2012 for evidence of recurrent aortic dissections. For all cases identified as having recurrent dissection, imaging studies were reviewed to exclude cases of dissection within a new anatomic territory or as an extension of a prior dissection. Iatrogenic aortic dissections were excluded. Patients were identified who had evidence of recurrent dissection of a previously dissected aortic segment. The demographics and characteristics of these 5 patients were collected and compared with remaining aortic dissection patients. Due to the small number of cases, a nonparametric statistical model was utilized. Continuous variables were compared with the Mann-Whitney $U$ test while the Fisher exact test was utilized for categoric data sets.

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*Table 1. Characteristics of Double-Dissection Patients Versus Other Patients in Our Registry of Type A and Type B Dissections*

| Characteristics | Double-Dissection (n = 5) | Type A (n = 222) | *p* Value | Type B (n = 156) | *p* Value |
|---|---|---|---|---|---|
| Age at first dissection, in years (mean ± SD) | 37.4 ± 5.5 | 59.8 ± 16.1 | $5.4 \times 10^{-4}$ | 61.5 ± 14.65 | $1.5 \times 10^{-4}$ |
| Male sex | 4 (80%) | 164 (74%) | 1.0 | 99 (64%) | 0.65 |
| Marfan syndrome | 4 (80%) | 15 (7%) | $1.7 \times 10^{-4}$ | 7 (5%) | $5.9 \times 10^{-5}$ |
| Hypertension | 1 (20%) | 138 (62%) | 0.08 | 109 (70%) | 0.03 |
| Atherosclerosis | 0 (0%) | 91 (41%) | 0.08 | 71 (46%) | 0.05 |
| Diabetes | 0 (0%) | 11 (5%) | 1.0 | 11 (7%) | 1.0 |
| Smoking history | 2 (40%) | 97 (44%) | 1.0 | 91 (58%) | 0.65 |

## Results

There were 383 patients in the registry diagnosed with aortic dissection during the 10-year period. Of these, 26 (7%) represented a recurrent aortic dissection and in 5 of these 26 the recurrent dissection occurred within a previously dissected aortic segment; these 5 patients represent the study group. All 5 study patients presented initially with a type A aortic dissection, so we chose the remaining patients in our database with type A dissection (n = 222) as the control group. The study patients were significantly younger, on average, at the time of initial dissection than were the other patients in the control group, at 37.4 ± 5.5 versus 59.8 ± 16.1 years ($p = 5.4 \times 10^{-4}$) (Table 1). The average interval between the initial dissection and the recurrent dissection within the previously dissected segment was 9.8 ± 1.9 years. In addition, the study group had a trend toward less hypertension and atherosclerosis than the control group. There were similar differences observed between the study group and those who had type B aortic dissections (Table 1).

The characteristics of the study group are similar to what is seen among dissection patients with syndromic aortic disease [16, 17]. In fact, 4 of 5 patients in this series carried the diagnosis of MFS (cases 2, 3, 4, and 5) compared with 15 of 222 in the control group, a significant difference (80% vs 7%, $p = 1.7 \times 10^{-4}$). When stratified for MFS diagnosis, age at first dissection in this series was similar to age at first dissection for other MFS patients in our database (38.8 ± 8.7 vs 37.4 ± 5.5 years) as well as previously reported ages in the literature (35 ± 12 years) [16]. Patients in this series were indistinguishable from other Marfan patients in our aortic dissection database with respect to hypertension, atherosclerosis, diabetes, or smoking (Table 2).

All 5 patients in the study group had first presented with a type A dissection that was managed with ascending aortic replacement, in keeping with the surgical guidelines at the time. A list of specific aortic events and surgical therapies for patients in the series is presented in Table 3. The double dissection event was a type B dissection in 3 patients and a type A dissection in 2 patients. In case 1 the patient experienced an initial type A dissection and presented 12 years later with an additional type A dissection (Fig 1A to 1D, and Fig 2). Three patients had major clinical aortic events between their initial dissection and the double dissection event (cases 2, 3, and 5). In case 2, the patient experienced extension of his thoracic aortic dissection into the abdominal aorta with a contained

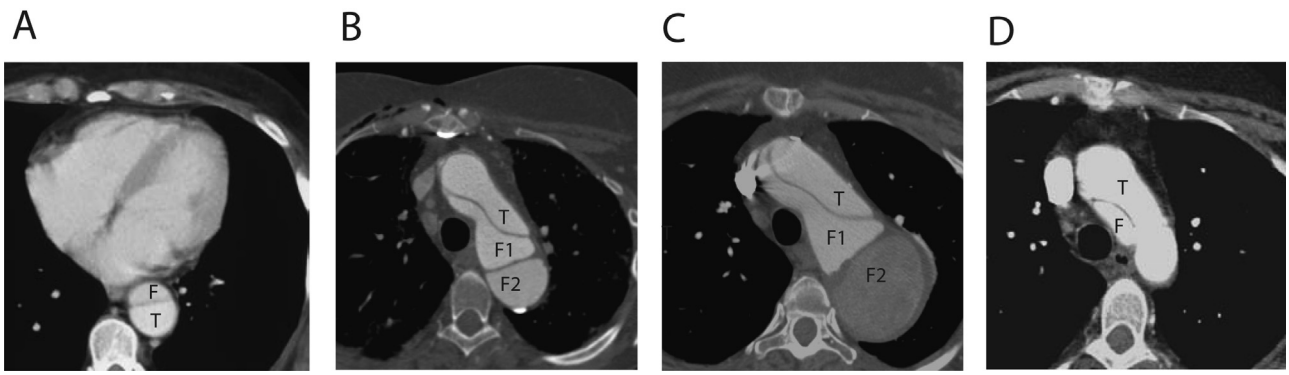


*Fig 1. Computed tomography angiography images of the evolving aortic dissection anatomy in the patient from case 1. (A) At age 36, a single dissection flap separating the true (T) and false (F) lumens in a descending aorta that is only mildly dilated. (B) At age 46, an image taken when the patient presented with recurrent symptoms, which now demonstrates 2 dissection flaps that separate the true lumen (T), original false lumen (F1), and second false lumen (F2). (C) At age 47, a similar flap configuration is present but the F2 lumen has expanded, resulting in a large thoracoabdominal aneurysm. (D) One year after surgical repair, an intimal flap persists in the aortic arch.*