Acute Aortic Dissection Determines the Fate of Initially Untreated
Aortic Segments in Marfan Syndrome

Running title: Schoenhoff et al.; Primarily non-treated aortic segments in MFS

Florian S. Schoenhoff, MD1; Silvan Jungi, MD1; Martin Czerny, MD1; Eva Roost, MD1;
David Reineke, MD1; Gabor Matyas, PhD2,3,4; Beat Steinmann, MD2; Juerg Schmidli, MD1;
Alexander Kadner, MD1; Thierry Carrel, MD1

1Dept of Cardiovascular Surgery, University Hospital Berne, Berne; 2Division of Metabolism
and Molecular Pediatrics, University Children’s Hospital Zurich; 3Institute of Medical Genetics,
Division of Medical Molecular Genetics and Gene Diagnostics, University of Zurich; 4Center for
Cardiovascular Genetics and Gene Diagnostics, Zurich, Switzerland

Address for Correspondence:
Thierry P. Carrel, MD
Department of Cardiovascular Surgery
University Hospital Berne
3010 Berne, Switzerland
Tel: +0041-31-6322111
Fax: +0041-31-6324443
E-mail: thierry.carrel@insel.ch

Journal Subject Codes: Cardiovascular (CV) surgery:[35] CV surgery: aortic and vascular
disease, Cardiovascular (CV) surgery:[41] Pediatric and congenital heart disease, including
cardiovascular surgery
Abstract:

**Background**—Aim of the current study was to investigate incidence and etiology of surgical interventions in primarily non-treated aortic segments after previous aortic repair in patients with Marfan syndrome (MFS).

**Methods and Results**—Retrospective analysis of 86 consecutive MFS patients fulfilling Ghent criteria that underwent 136 aortic surgeries and were followed at this institution in the past 15 years. Mean follow-up was 8.8±6.8y. Thirty-day, 6-months, 1-year and overall mortality was 3.5%, 5.8%, 7.0% and 12.8%, respectively. Ninety-two percent of patients initially presented with aortic root, ascending aortic or arch lesions, whereas 8% presented with descending aortic or thoraco-abdominal lesions. Primary presentation was acute aortic dissection (AAD) in 36% [77% type A, 23% type B] and aneurismal disease in 64%. Secondary complete arch replacement had to be performed in only 6% of patients without AAD, but in 36% with AAD (p=0.0005). In patients without AAD, 11% required surgery on primarily non-treated aortic segments [5 out of 6 patients suffered from type B dissection during follow-up], whereas in patients after AAD, 48% underwent surgery of initially non-treated aortic segments [42% of patients with type A and 86% of those with type B dissection] (p=0.0002).

**Conclusions**—The need for surgery in primarily non-treated aortic segments is precipitated by an initial presentation with AAD. Early elective surgery is associated with low mortality and re-intervention rates. Type B dissection in patients with MFS is associated with a high need for extensive aortic repair even if the dissection is being considered uncomplicated by conventional criteria.

**Key words**: aortic dissection, Marfan syndrome, aortic surgery
Introduction

Marfan syndrome (MFS) is an autosomal dominant disorder affecting about 1 in 5,000 individuals. The phenotypic changes of MFS are imposed by mutations in the gene encoding for the extracellular matrix protein fibrillin-1. Although patients with MFS exhibit skeletal, ocular and cardiovascular manifestations, acute aortic dissection due to aortic aneurysms still determines morbidity and mortality in this patient population. In a multi-institutional series of 675 patients, 30-day mortality for elective, urgent or emergency repair was 1.5%, 2.6%, and 11.7% respectively.

Low morbidity and mortality rates in patients undergoing elective root surgery and an increasing awareness for patients with connective tissue disease have continuously lowered the threshold for surgical interventions on the proximal aorta. The concept of prophylactic aortic root surgery in preventing acute dissection is well established and has contributed to the improved survival of MFS patients over the past decades.

However, it is not very well understood how this affects the need for further surgical interventions on primarily non-treated aortic segments in the long-term. Several studies suggested a shift of morbidity and mortality towards the distal aorta. Analysis of the Euro Heart Survey database revealed that 31% of interventions in patients with MFS have been performed on the distal aorta and Finkbohner and colleagues noted that 18% of primary interventions were due to lesions on the distal aorta. It is unclear whether the increased life expectancy in patients with MFS just makes an event on the distal aorta more likely or if elective root surgery itself might trigger downstream aortic events. In an in vitro model it was suggested that wall tension in the residual aorta increases after prosthetic replacement of the ascending aorta.

These findings are contrary to our clinical experience in which patients who underwent
elective root replacement seemed to have a more favorable long-term outcome compared to patients with a history of acute aortic dissection.

**Aim**

Aim of the current study was to determine incidence and substrate for intervention in initially non-treated aortic segments in patients with MFS.

**Patients and Methods**

Data from 86 MFS patients (mean age 35 years, range 8-69 years, 57% male patients) fulfilling Ghent criteria that underwent 136 major aortic operations and were followed at this institution in the past 15 years was retrospectively analyzed. Patients are followed-up in our MFS clinics 3, 6 and 12 months after surgery and then depending on the findings, at least once per year. Patients were generally evaluated using ECG-gated, CT angiography to plan surgery, as a follow-up in patients with (residual) dissections and in the acute setting. In benign cases or after uneventful elective surgery MR imaging was performed to reduce cumulative radiation exposure.

Additionally, a phone interview was conducted according to a standardized questionnaire that was sent to the patients in advance. Individual informed consent was obtained and patients were asked if we were allowed to contact their primary care provider regarding recent developments, changes in medication or CT scans that have been performed outside our institution. Hereby, a 99% completeness of follow-up was achieved. For analysis, patients were divided in 4 groups (Fig. 1): group (1), patients that initially presented without acute aortic dissection; group (2), patients that initially presented with acute aortic dissection; group (3), patients that never experienced acute aortic dissection during follow-up and group (4), patients that suffered from acute aortic dissection initially or at any time during follow-up. This study
was approved by the institutional review board and individual informed consent from the patient or, in case of minors, the parent respectively the legal guardian was obtained.

**Statistical analysis**

In addition to descriptive statistics, data underwent a Kaplan-Meier survival analysis, with either re-operation or death as an event, followed by a log rank test to compare the event risk for patients with or without history of aortic dissection. Analysis was performed with SPSS version 15.0 software (SPSS, Inc, Chicago, Ill).

**Indication for surgery and surgical techniques**

In patients presenting with aortic root aneurysm, surgery was considered at a diameter of 45-50mm or progress of more than 5mm per year as an indication for surgery since 15% of patients with MFS dissect at diameters of less than 50mm. If aortic regurgitation was present and aortic root size was less than 45mm, indication for surgery depended on the extent of regurgitation and hence left ventricular dimensions. In younger women with MFS who want to become pregnant, prophylactic root replacement was considered if aortic root size exceeded 40mm. Surgical repair of the aortic arch and descending aorta was considered if the diameter exceeded 55 to 60mm or in case of rapid enlargement, e.g. after Stanford type B dissection. There are several procedures available for the treatment of the aortic root in MFS patients; complete root replacement by means of a modified Bentall procedure or a valve sparing root replacement (VSRR) was the treatment of choice in the current series.

In patients with more advanced stage of disease where dilation of the aorta extended into the aortic arch and descending aorta, arch replacement with or without implantation of an elephant trunk was performed. In patients with acute Stanford type A dissection, the distal anastomosis was performed at least as a hemi-arch replacement using moderate hypothermic
circulatory arrest with bilateral antegrade cerebral perfusion. If complete arch replacement was necessary, separate re-implantation of the supra-aortic branches using a vascular graft with multiple side-branches was preferred.

**Results**

Seventy-eight patients (91%) initially presented with aortic root, ascending aortic or aortic arch lesions, whereas 7 patients (8%) primarily presented with descending aortic or thoraco-abdominal lesions; 1 patient (1%) with aortic root aneurysm and dilative cardiomyopathy underwent heart transplantation as a primary procedure. Etiology at initial presentation was acute dissection in 36% and chronic dilative disease in 64%. In the group of patients presenting with acute aortic dissection, 77% of patients presented with Stanford type A dissection and 23% with Stanford type B dissection. In patients with acute Stanford type A dissection, 67% received a composite graft whereas in 33% of patients only a supracoronary replacement was performed since the diagnosis of MFS was not established at the time of surgery. In the latter group, 2 out of 8 patients needed re-do surgery on the aortic root. No VSRR was performed in patients with acute dissection. In patients undergoing elective surgery, 43% of patients received a composite graft, 50% underwent VSRR. Reasons not to perform VSRR in patients with MFS were bicuspid valve, asymmetric aortic valve leaflets and large fenestrations of the leaflets with partial detachment of the commissures.

**Re-Intervention on Aortic Root and Aortic Arch**

Twenty-one David operations and 5 Yacoub procedures were performed. The rate of reoperation was 1 out of 21 patients in the David and 3 out of 5 in the Yacoub group, which represents a significantly higher reoperation rate in the re-modeling compared to the re-implantation group.
(p=0.01). All reoperations became necessary due to development of moderate to severe aortic regurgitation. Secondary replacement of the complete aortic arch became necessary in only 6% of patients in group (1), but in 36% of the patients in group (2) (p=0.0005) (Fig. 2, panel A and B).

**Distal Aorta**

In group (1), 11% of patients underwent surgery in downstream aortic segments, whereas in group (2), 48% patients had to undergo surgery on the distal aorta. Interestingly, 5 out of the 6 patients undergoing re-interventions in group (1) suffered from type B dissection in the meantime (Fig. 3, panel A and B).

Acute type B dissection occurred in 19% (16/86) of patients. Mean diameter at the time of dissection was 37±8.5mm (range 19-50mm). Except for 1 case the maximum diameter was at the proximal descending aorta. Mean diameter at time of surgery was 64±10.6mm (range 41-80mm). This translates into an estimated average growth rate of 16±11.4mm/6months after onset of pain. All but 1 patient presented with primarily uncomplicated type B dissection. One patient developed severe pseudocoarctation with visceral ischemia and underwent surgery shortly after presentation. Mean time between development of type B dissection and surgery was 2±2.7years, but 67% of patients underwent surgery within the 1st year after the event. Only 1 patient could be identified where type B dissection evolved from the distal anastomosis 3 months after elective complete arch replacement.

In the group of patients with a history of dissection, patients suffering from Stanford type B dissection are significantly more likely to undergo surgery on the distal aorta than those with type A dissection (86% vs. 42%, p=0.0002).

**Freedom-from-Reoperation and Survival**
Freedom-from-reoperation in group (1) was 86%, 69% and 27% at 5, 10 and 15 years and 65%, 45% and 20% at 5, 10 and 15 years, respectively, in group (2) (Fig. 4). In group (3), freedom from reoperation was 92%, 86% and 64% at 5, 10 and 15 years compared to 65%, 42% and 16% at 5, 10 and 15 years, respectively, in group (4) (Fig. 5).

Furthermore, history of dissection at any time during follow-up (group (3)) was not only associated with a higher rate of re-operation but also a predictor of survival in patients with MFS. Survival in group (1) was 93%, 93% and 93% at 5, 10 and 15 years and 90%, 90% and 72% at 5, 10 and 15 years, respectively, in group (2) (Fig. 6). In group (3), survival was 98%, 98% and 98% at 5, 10 and 15 years compared to 85%, 85% and 71% at 5, 10 and 15 years, respectively, in group (4) (Fig. 7).

**Other Risk Factors for Re-Intervention**

There were no differences in the incidence of surgical interventions of initially non-treated aortic segments between smokers and non-smokers (p=0.59). There were no differences in the incidence of surgical interventions of downstream aortic segment between patients who were treated with ACE-inhibitors or ATII-receptor-blockers and patients without such a treatment (p=0.47). Furthermore, there was no significant influence of beta-blockers (BB) or calcium channel blockers (CaB) on either re-operation rate (BB, p=0.26; CaB, p=0.14) or the likelihood of suffering from dissection (BB, p=0.66; CaB, p=0.5).

In patients with high blood pressure (at initial presentation) 12 out of 28 patients (43%) needed secondary surgical interventions of the distal aorta, compared to 10 out of 58 patients (17%) in those patients without hypertension (p=0.02).

**Follow up and Complications**

Mean follow-up of survivors was 8.8±6.8 years, mean interval between initial and redo-surgery
was 5.5±4.6 years. Thirty-day, 6 months, 1 year and late mortality was 3.5%, 5.8%, 7.0% and 12.8%, respectively. Intraoperative mortality during the index operation was zero but 1 patient (1.2%) died during emergency surgery for rupture of the descending aorta 6 days after surgery for type A dissection. The majority of deaths (9 out of 11, 82%) occurred in patients with a history of dissection, including 3 patients that suffered from type B dissection during follow-up after uneventful elective surgery for aortic root disease. Two patients (18%) without a history of dissection died during follow-up. One was a patient with aortic root aneurysm and severe dilative cardiomyopathy that underwent heart transplantation and died due to Non-Hodgkin lymphoma 15 years after the initial surgery. The other patient was a young man with a severe form of MFS and concomitant dilative cardiomyopathy who underwent aortic root replacement, concomitant mitral valve replacement and tricuspid valve repair and died of a malignant arrhythmia during the post-operative course.

Stroke occurred in 4.2% of patients, all of them in patients with a history of acute aortic dissection. There was no case of paraplegia but 1 patient with paraparesis. Other complications are shown in table 1, causes of death are shown in table 2.

Discussion

Aortic root disease predisposes MFS patients for the occurrence of life-threatening aortic dissection. Despite the wide availability of screening and prophylactic surgery, 36% of MFS patients operated on in our institution initially presented with acute dissection.

The rate of acute type A dissections in the current series (28%) is comparable to recent studies, where 16% to 28% of the patients present with acute type A dissection. Bentall procedures at initial presentation were performed in 47% of patients compared to 40% in a
similar study\textsuperscript{19}. Cameron and colleagues from Johns Hopkins Hospital recently reported a series of 372 patients over the last 30 years where almost 90\% of patients in the last 5 years received VSRR\textsuperscript{17}. In our cohort, half of the patients in the non-dissection group received a valve-sparing root replacement. Compared to the Hopkins experience, the percentage of patients presenting with acute type A dissection in our institution was much higher; 4.4\% vs. 28\%. In the present series none of the patients with acute dissection underwent a VSRR procedure. We are not opposed to VSRR in patients presenting with Stanford type A dissection, but would only consider performing VSRR in young patients who present with uncomplicated type A dissection without signs of myocardial or visceral ischemia or neurologic impairment. Otherwise, a composite graft, which gives predictable long-term results and limits time on cardiopulmonary bypass, is performed.

In 8 patients in our early experience that suffered from Stanford type A dissection where the diagnosis of MFS was not established at the time of surgery there was no significant aortic root dilation and in an emergency situation the procedure was limited to isolated replacement of the ascending aorta. The need for re-operation due to aortic regurgitation in our series is significantly higher in the Yacoub than in the David group. As reported by several groups, the rate of annular dilation and valve incompetence is higher with the Yacoub technique than after a David procedure\textsuperscript{17}. The remodeling technique represents our early experience with VSRR and was abandoned in favor of the re-implantation technique.

Although endovascular surgery is generally not recommended for MFS patients because of poor tissue quality and ongoing dilation even after technically successful stentgraft implantation\textsuperscript{20}, 1 patient with MFS underwent TEVAR to bridge a short aneurismal segment between 2 polyester grafts.
It is difficult to estimate whether root replacement actually triggers disease progress in downstream aortic segments as has been suggested recently. Re-interventions on the distal aorta after proximal repair due to type A dissection are mostly precipitated by residual dissection in the downstream aorta. Therefore, no statement regarding the rate of new dissections after successfully repaired type A dissection can be made. There is only 1 patient in the study population with a truly unrelated type B dissection after completely excluded type A dissection. We can only state that in the present series, re-operation on the distal aorta was significantly more frequent in patients with a history of dissection than in patients undergoing elective aortic root surgery.

For MFS patients undergoing elective root repair, type B dissection poses a serious threat throughout follow-up. All except 1 patient in the present study undergoing interventions on the distal aorta after elective proximal aortic repair suffered from type B dissection. Only in rare cases there seems to be a direct correlation in terms of a clamp injury or a dissection originating at the distal anastomosis.

Therefore, even if elective root replacement by a prosthesis would constitute a risk factor for downstream aortic dissection (due to the loss of the elastic properties of the root or clamp injuries of the aorta) it is obviously outweighed by the risk of type A dissection if timely proximal repair is not performed.

In a recently published cohort of MFS patients with a history of aortic dissection, Mimoun and colleagues observed a clinical event in 52% of the patients within a follow-up period of 9.8 years. In our present series, re-operations on the distal aorta were significantly frequent in patients with a history of dissection compared to those without a history of dissection (48% vs. 11%). While this has already been shown by Girdauskas and colleagues to be true for
patients presenting with Stanford type A dissection (44% vs. 10%), we were surprised about the high rate of re-operations (86%) in patients with Stanford type B dissection in our cohort. It seems that in these patients the need for subsequent surgery is driven by rapid enlargement rather than the absolute diameter. Therefore, MFS patients presenting with type B dissection should remain under close surveillance even if the dissection is considered uncomplicated by conventional clinical and imaging criteria. Notably, in our patient population, the need for re-operations on the distal aorta in patients with prior elective root replacement was driven (5 out of 6) by the development of type B dissection although the descending aorta in these patients was not necessarily severely dilated at that time.

Although treatment with the AT2 receptor antagonist losartan seems a very promising option in patients with MFS, in the present study it did not affect the need for re-intervention on the distal aorta in the present series. However, numbers were small, there was no defined treatment regimen and, in many cases, the need for anti-hypertensive treatment was driven by the presence of dissection.

In the current study population, 79% of patients underwent genetic testing and 79% of these carried pathogenic mutations. There were no significant differences regarding rate of dissection (32% vs. 43%; p=0.53) or later repair of non-treated aortic segments (26% vs. 29%; p=1.0) between patients with or without pathogenic mutations. Even when looking at these groups of patients separately, the main determinant for later repair of previously non-treated aortic segments was history of acute dissection. In the group of patients with confirmed pathogenic mutations, 76% of patients with a history of dissection underwent later repair of precisely non-treated aortic segments, whereas this was the case in 67% of patients without a pathogenic mutation. Of course this observation is biased by the fact that many patients with a
clear phenotype, being known for having MFS since childhood have not been genotyped and mostly undergo elective surgery before acute dissection occurs.

Our 30d, 6mo and 1y (3%, 6% and 7%, respectively) mortality compares favorably with other published reports, especially considering the higher rate of patients with acute dissection. Our overall mortality is still quite favorable with 13% after a mean follow-up of 8.8±6.8 years. As expected, survival was better in the group without history of dissection, compared to the group with a history of dissection. We think that analyzing late mortality is crucial in order to optimize treatment of patients with MFS. Although it is not surprising that the majority of deaths are patients with a history of dissection, it seems important to point out that it is not only the direct perioperative mortality that is increased in these patients. Causes of death in patients with a history of dissection included aortic rupture shortly after acute type B dissection, perioperative stroke in type A dissection, intracerebral hemorrhage 4 months after aortic root surgery due to type A dissection, stroke during coronary angiography 6 months after surgery for type A dissection, aortic rupture during femoral hernia repair, aortic rupture 11 years after surgery for type A dissection, myocardial infarction 3 months after surgery for type A dissection and rupture of descending aorta 6 days after root surgery for type A dissection.

Conclusion
In conclusion, 1/3 of MFS patients still present with acute dissection, despite the increasing awareness of connective tissue disease, improved screening and the wide availability of elective surgery. Furthermore, our study confirmed that true long-term follow-up is mandatory to avoid adverse aortic events in patients with MFS. The current data suggests that in patients with MFS, the need for surgery on initially non-treated aortic segments is primarily determined by an initial
presentation with acute dissection. Furthermore, in patients with MFS, type B dissection constitutes a strong predictor for the need of re-intervention in primarily non-treated aortic segments even if the dissection is being considered uncomplicated by conventional criteria.

**Acknowledgements:** We would like to thank Brigitta Gahl, MSc, University Hospital Berne, for statistical assistance.

**Conflict of Interest Disclosures:** None.

**References:**


957.


Table 1. Complications.

<table>
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<th>Complications</th>
<th>n (%)</th>
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<tbody>
<tr>
<td>Tachyarrhythmia</td>
<td>7 (4.9)</td>
</tr>
<tr>
<td>Hemorrhage needing intervention</td>
<td>6 (4.2)</td>
</tr>
<tr>
<td>Stroke</td>
<td>6 (4.2)</td>
</tr>
<tr>
<td>Pneumonia</td>
<td>3 (2.1)</td>
</tr>
<tr>
<td>Recurrent laryngeal nerve paralysis</td>
<td>3 (2.1)</td>
</tr>
<tr>
<td>Phrenic nerve injury</td>
<td>3 (2.1)</td>
</tr>
<tr>
<td>Fever of unknown origin</td>
<td>2 (1.4)</td>
</tr>
<tr>
<td>Pericardial effusion</td>
<td>2 (1.4)</td>
</tr>
<tr>
<td>Pleural effusion</td>
<td>1 (0.7)</td>
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<tr>
<td>Pericarditis</td>
<td>1 (0.7)</td>
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<tr>
<td>Perioperative MI</td>
<td>1 (0.7)</td>
</tr>
<tr>
<td>Postcardiotomy syndrome</td>
<td>1 (0.7)</td>
</tr>
<tr>
<td>Paraparesis</td>
<td>1 (0.7)</td>
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Table 2. Causes of death.

<table>
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<tbody>
<tr>
<td>Aortic rupture shortly after acute type B dissection *</td>
</tr>
<tr>
<td>Malignant arrhythmia after mitral valve replacement, aortic root surgery and</td>
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<tr>
<td>tricuspid valve repair</td>
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<tr>
<td>Perioperative stroke in type A dissection *</td>
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<td>Intracerebral hemorrhage 4 months after aortic root surgery due to type A dissection *</td>
</tr>
<tr>
<td>Stroke during coronary angiography 6 months after surgery for type A dissection *</td>
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<tr>
<td>Aortic rupture during femoral hernia repair *</td>
</tr>
<tr>
<td>Aortic rupture 11 years after surgery for type A dissection *</td>
</tr>
<tr>
<td>Multiorgan failure in Non-Hodgkin lymphoma</td>
</tr>
<tr>
<td>Myocardial infarction 3 months after surgery for type A dissection *</td>
</tr>
<tr>
<td>Rupture of descending aorta 6 days after root surgery for type A dissection *</td>
</tr>
<tr>
<td>Congestive heart failure 5 years after last surgery *</td>
</tr>
</tbody>
</table>

Causes of death; an asterisk (*) marks patients with a history of dissection at primary presentation or at any time during follow up. Each cause represents a single patient.
Figure Legends:

Figure 1. Allocation of patients to groups 1 to 4

Figure 2. Panel A shows a significantly higher rate of reoperations in patients after VSRR with the re-modeling compared to the re-implantation technique. Panel B depicts a significantly higher rate of secondary full-arch replacements in patients with a prior history of acute dissection.

Figure 3. The rate of re-interventions on the distal aorta is significantly higher in patients with a history of acute dissection (panel A). Interestingly, 5 out of 6 patients in the group of patients without prior dissection suffered from Stanford type B dissection in the meantime. Panel B demonstrates the high rate of patients in need for interventions on the distal aorta after type B dissection, even compared to patients with a history of type A dissection.

Figure 4. Kaplan-Meier curve depicting freedom-from-reoperation in group (1) (primarily non-dissected) compared to group (2) (primarily dissected). Vertical lines truncate Kaplan-Meier graph at 2/3 of patient population in each group.

Figure 5. Kaplan-Meier curve depicting freedom-from-reoperation in group (3) (never dissected) compared to group (4) (dissection primarily or later). Vertical lines truncate Kaplan-Meier graph at 2/3 of patient population in each group.
**Figure 6.** Kaplan-Meier curve depicting survival in group (1) (primarily non-dissected) compared to group (2) (primarily dissected). Vertical lines truncate Kaplan-Meier graph at 2/3 of patient population in each group.

**Figure 7.** Kaplan-Meier curve depicting survival in group (3) (never dissected) compared to group (4) (dissection primarily or later). Vertical lines truncate Kaplan-Meier graph at 2/3 of patient population in each group.
Figure 1

86 patients

Group 1
Initial presentation without AAD
n=48
Group 3
Never experienced AAD

Group 2
Initial presentation with AAD
n=7
Group 4
AAD at initial presentation or Fup
n=31

n=55
n=31
Reoperations after valve-sparing root replacement

Secondary total arch replacement

Figure 2
Reinterventions on the distal aorta after acute dissection

Figure 3
Figure 4

Number of operations:

<table>
<thead>
<tr>
<th></th>
<th>136</th>
<th>61</th>
<th>25</th>
<th>3</th>
</tr>
</thead>
</table>
| Log rank, p    | 0.018

Initial presentation

- Primarily dissected
- Primarily non-dissected
- Primarily dissected-censored
- Primarily non-dissected-censored
Figure 5

Number of operations:

136             61    25                                 3

log rank, \( p<0.0001 \)
Fig. 6

Patients at risk:

86               49     34                                13

log rank, p=0.47

Figure 6
Figure 7

log rank, p=0.072
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Circulation, published online March 14, 2013;
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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