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ORIGINAL ARTICLE

Should aortic arch replacement be performed during initial surgery for aortic root aneurysm in patients with Marfan syndrome?†

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Abstract

OBJECTIVES: The aim of this study was to investigate whether total arch replacement (TAR) during initial surgery for root aneurysm should be routinely performed in patients with Marfan syndrome (MFS).

METHODS: Retrospective analysis of 94 consecutive MFS patients fulfilling Ghent criteria who underwent 148 aortic surgeries and were followed at this institution during the past 16 years.

RESULTS: The mean follow-up interval was 8.8 ± 7 years. Initial presentation was acute aortic dissection (AAD) in 35% of patients (76% Type A and 24% Type B) and aneurismal disease in 65%. TAR was performed in 8% of patients during initial surgery for AAD (otherwise a hemi-arch replacement was performed) and 1.6% in elective root repair. Secondary TAR had to be performed in only 3% of patients without, but in 33% following AAD (33% Type A and 33% Type B; $P = 0.0001$). Thirty-day, 6-month, 1-year and overall mortalities were 3.2, 5.3, 6.4 and 11.7%, respectively. Operative and 30-day mortalities in secondary aortic arch replacement were zero. Secondary TAR after AAD did not increase the need for the replacement of the entire thoracoabdominal aorta during follow-up compared with patients without secondary TAR (37 vs 40%, $P = 1.0$).

CONCLUSIONS: MFS patients undergoing elective root repair have small risk of reinterventions on the aortic arch, and primary prophylactic replacement does not seem to be justified. In patients with AAD, the need for reinterventions is precipitated by the dissection itself and not by limiting the procedure to the hemi-arch replacement in the emergency setting. Limiting surgery to the aortic root, ascending aorta and proximal aortic arch is associated with low mortality in MFS patients presenting with AAD.

Keywords: Aortic surgery • Marfan syndrome • Aortic arch • Acute aortic dissection • Connective tissue disease

INTRODUCTION

Aneurysm of the aortic root is the hallmark feature of Marfan syndrome (MFS), an autosomal dominant disorder imposed by mutations in the gene encoding for the extracellular matrix protein fibrillin-1 [1, 2]. Although patients with MFS exhibit skeletal, ocular and cardiovascular manifestation, aortic aneurysm determines morbidity and mortality in this patient population [3].

Aortic aneurysm in MFS is typically pear-shaped and involves progressive dilatation of the sinus of Valsalva. In most of the MFS patients presenting with aortic root aneurysm on a non-emergent basis, the aneurysm is limited to the root and proximal ascending aorta and shows almost normal diameter at the level of the brachiocephalic trunk.

In MFS patients presenting with acute aortic dissection (AAD) Stanford Type A, the dissection frequently progresses beyond the ascending aorta through the arch into the descending and abdominal aorta as seen in non-MFS patients. The low morbidity and mortality rates in MFS patients undergoing elective root surgery have fostered the concept of prophylactic aortic surgery to prevent AAD and its sequelae [4]. Life expectancy in MFS patients has dramatically improved over the past decades through the prevention of AAD [5].

As several recent studies suggested a shift of morbidity and mortality towards the distal aorta [6–8], the question remains to what extent the aorta should be replaced during initial surgery for either elective root repair or emergency surgery for Type A dissection.

In the acute setting, the surgeon has to weigh the increased operative risk associated with a total arch replacement (TAR) against the risk of future reintervention and interstage mortality. Unfortunately, there are little data available to estimate the true additional burden in terms of myocardial injury or neurological complications regarding primary TAR in elective patients or those presenting with Type A dissection.

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Aim

The aim of this study was to investigate whether TAR during initial surgery for root aneurysm should be routinely performed in patients with MFS. Since it can be assumed that additional primary TAR carries a certain extra risk for the patient, we also compared outcomes in patients with and without secondary TAR in order to analyse whether primary TAR prevents further interventions aside from arch replacement.

PATIENTS AND METHODS

Data from 94 MFS patients (mean age at initial surgery 43 ± 16 years, range 8–69 years, 55% male patients) fulfilling Ghent criteria who underwent 148 major aortic operations and were followed at this institution since 1995 were retrospectively analysed. 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 evaluated using electrocardiography-gated, computed tomography (CT) angiography to plan surgery, as a follow-up in patients with dissections and in patients undergoing surgery on an emergent basis. In benign cases or after uneventful elective surgery, magnetic resonance imaging was performed to reduce cumulative radiation exposure.

Furthermore, 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 were performed outside our institution. Thus, a 99% completeness of follow-up was achieved.

This study was approved by the institutional review board and individual informed consent from the patient or, in the case of minors, the parent or the legal guardian was obtained.

Statistical analysis

The values are given in mean ± standard deviation, when appropriate. In addition to descriptive statistics, data underwent a Kaplan–Meier survival analysis, with either reoperation 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 or with or without secondary TAR. Analysis was performed with SPSS version 15.0 software (SPSS, Inc., Chicago, IL, USA).

Indication for surgery and surgical techniques

Indications for surgery generally followed the 2010 AHA guidelines [9] for the treatment of thoracic aortic disease. Since 15% of patients with MFS dissect at a diameter of <50 mm, aortic root surgery was considered at a diameter of 45–50 mm or progressive dilatation of >5 mm per year [5, 6, 10]. If aortic regurgitation was present and aortic root size was <45 mm, indication for surgery depended on the extent of regurgitation and hence left ventricular dimensions. Prophylactic root replacement was suggested in women wishing to conceive if aortic root size exceeded 40 mm. Aortic root replacement by means of a modified Bentall procedure or valve-sparing root replacement (VSRR) in suitable candidates was the treatment of choice [11, 12] in the present study. If the aorta at the level of the brachiocephalic trunk was 35 mm or larger, repair was extended into the arch by performing a hemi-arch replacement.

Surgical repair of the aortic arch and descending aorta was considered if the diameter exceeded 55–60 mm or in the case of rapid enlargement, e.g. after Stanford Type B dissection. In patients presenting with acute Stanford Type A dissection, the distal anastomosis was performed by removing the concavity of the aortic arch using moderate hypothermic circulatory arrest with bilateral antegrade cerebral perfusion [13]. If TAR was necessary, separate reimplantation of the supra-aortic branches using a vascular graft with multiple side branches was preferred.

While the hemi-arch replacement was considered standard of care in patients presenting with Type A dissection, primary TAR was only done if needed in order to perform a sufficiently stable distal anastomosis or to prevent neurological damage by obstruction of the supra-aortic branches. For the purpose of this study, TAR was defined as a circular anastomosis between the brachiocephalic trunk and the left subclavian artery with reimplantation of at least one supra-aortic branch.

Management of cardiopulmonary bypass

The management of cardiopulmonary bypass and circulatory arrest has improved over the course of the study [13, 14]. Most notably, the routine use of bilateral selective antegrade cerebral perfusion began in 2004. In elective cases scheduled for aortic root replacement, standard aortic and right atrial cannulation were performed, and cardiopulmonary bypass was conducted in moderate hypothermia (32°C). Patients with acute Stanford Type A aortic dissection were cannulated through the right axillary artery whenever possible and cooled to 20°C tympanic and 26° C core temperature. Patients with chronic ascending aortic aneurysms involving the proximal aortic arch are cooled to 26°C tympanic and 30°C core temperature. Bilateral selective antegrade cerebral perfusion was either performed through perfusion catheters in both common carotid arteries or via the right axillary arterial cannula (using the arterial return line) and an additional perfusion catheter in the left common carotid artery with perfusion pressure not exceeding 50–60 mmHg. Cerebral perfusion was monitored using continuous bilateral near-infrared oximetry. Algorithmic analysis of electroencephalogram data allowed the confirmation of burst suppression before the administration of sodium thiopenthal and the initiation of circulatory arrest.

RESULTS

Primary interventions on the aortic root

Initial presentation was AAD in 35% of patients and aneurismal disease in 65%. In patients undergoing elective repair of the root and ascending aorta, 46% received a modified Bentall procedure, 44% a VSRR, in 3% only the ascending aorta was replaced and 5% primarily received other procedures (heart transplantation, the use of homograft for aortic root replacement, etc.). In the group of patients presenting with AAD, 76% presented with Stanford Type A and 24% with Stanford Type B dissection. In patients with Type A dissection, a modified Bentall procedure was performed in 68%, and in 32% of patients only the ascending aorta was replaced since the diagnosis of MFS was not

established at the time of surgery (mainly in the early years of the observation period). In the group of patients primarily presenting with Type B dissection, 25% suffered from Type A dissection during follow-up and underwent a modified Bentall procedure.

Primary aortic arch interventions

Primary TAR was only performed in 1.6% of patients undergoing elective aortic root repair. Simultaneous TAR was performed in 8% of patients with Type A dissection and 7% if including those patients who initially presented with Type B dissection that later evolved into Type A dissection.

Secondary aortic arch interventions

Mean time from initial surgery to secondary TAR was 8 ± 6 years. Secondary TAR after primary elective proximal repair had to be performed in only 2 (3%) patients. One patient needed TAR 6 years after a Bentall procedure and 1, 3 years after replacement of the ascending aorta followed by Type B dissection, replacement of the descending aorta and finally replacement of the aortic root by a modified Bentall procedure.

In patients after successful repair of Type A dissection, secondary TAR became necessary in 33% of patients ($P = 0.0001$). There was no significant difference in secondary TAR regarding the type of proximal procedure at initial surgery. Reintervention became necessary in 38% of patients after replacement of the ascending aorta and in 35% after a Bentall procedure. In those 2 patients with Type A dissection after initial presentation with Type B dissection, 1 needed secondary TAR. Interestingly, secondary TAR had to be performed in only 27% of cases at the time of the first reintervention (Fig. 1).

Implantation of an elephant trunk to prepare for later surgery on the descending aorta was performed in 27% of patients during secondary TAR, including 1 patient who presented with Type B dissection and a dilated descending aorta, but had to undergo proximal repair first due to a very large root aneurysm.

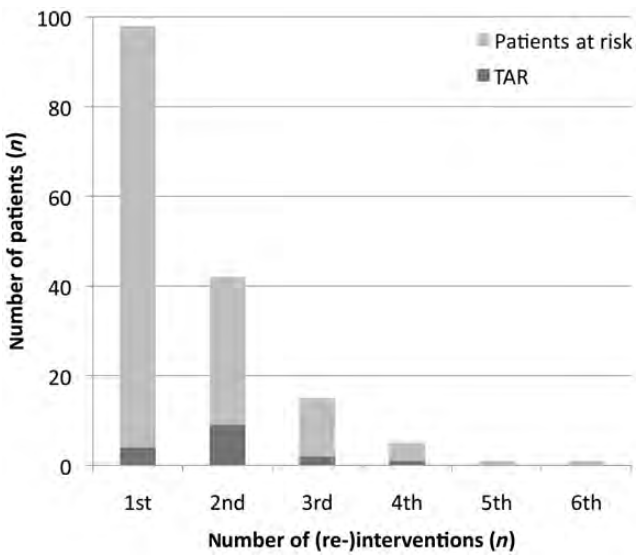

Figure 1: Graph depicting the number of total arch replacements compared with all reinterventions performed.

Reoperations in patients after secondary total arch replacement

Patients initially presenting with AAD who underwent secondary TAR during follow-up had a higher rate and number of reoperations compared with the group that never had the aortic arch replaced (78 vs 43%, $P = 0.12$; 1.5 vs 3.4 surgeries per patient).

Complete thoracoabdominal aortic replacement (TAAR) in patients initially presenting with AAD became necessary in 44% of patients with secondary TAR and in 24% ($P = 0.21$) of those without secondary TAR. Interestingly, if including those patients who suffered from AAD at any point during follow-up, the rate of TAAR in patients with secondary TAR was 40% compared with 37% ($P = 1.0$) in patients without aortic arch replacement.

Circulatory arrest in total arch replacement and neurological complications

Selective antegrade cerebral perfusion was performed in all patients since 2004. The duration of circulatory arrest was 17 ± 11 min during initial surgery for proximal repair, including those cases with primary TAR. In patients with secondary TAR, circulatory arrest time was 25 ± 11 min.

Cerebrovascular complications occurred in 6 patients (4% of performed surgeries) of the overall study population, all of them except 1 in those with a history of AAD. In 3 patients, symptoms completely resolved during follow-up, including 1 patient after secondary TAR 16 years after a Bentall procedure. Two patients showed residual hemiparesis, including 1 with already pre-existing neurological impairment after a modified Bentall procedure and primary TAR due to Type A dissection. The patient was referred from an outside hospital for progressive dilatation of the residual aortic tissue surrounding the supra-aortic branches. The patient underwent redo arch surgery without new neurological events. One patient died after suffering from stroke during elective coronary angiography 6 months after surgery for Type A dissection.

Therefore, there was no patient who suffered from persistent neurological impairment after primary or secondary TAR in our institution.

Follow-up and mortality

The mean follow-up was 8.8 ± 7 years. Thirty-day, 6-month, 1-year and overall mortalities were 3.2, 5.3, 6.4 and 11.7%, respectively. Operative mortality in patients presenting with acute Type A dissection was zero. Thirty-day mortality was 7%, with 1 patient suffering from rupture of the descending aorta 6 days after initial surgery for Type A dissection and 1 patient experiencing major perioperative stroke. Operative and 30-day mortalities in secondary TAR were zero.

Freedom from reoperation and survival

There were no significant differences in survival between patients who underwent primary, secondary or no TAR. Survivals at 5, 10, 15 and 20 years were 91, 91, 87 and 77% in patients without TAR, 100% in patients with primary TAR at 5 and 10

years and 100, 100, 80 and 80% in patients with secondary TAR, respectively (Fig. 2).

Freedom from secondary arch replacement in patients initially presenting with AAD was significantly worse compared with patients without AAD (log rank $P = 0.002$). Freedom from secondary TAR at 5, 10, 15 and 20 years were 96, 96, 96 and 72% in patients without initial dissection and 80, 67, 53 and 42% at 5, 10, 15 and 20 years in patients initially presenting with AAD (Fig. 3).

Freedom from reoperation at 5, 10 and 15 years in patients presenting initially with AAD and needing secondary TAR during follow-up were 51, 30 and 8% compared with 77, 65 and 29% in patients without secondary TAR during follow-up. Freedom from reoperation at 5, 10 and 15 years in patients initially presenting without AAD and needing secondary TAR during follow-up were 67, 44 and 44% compared with 88, 61 and 32% in patients without secondary TAR during follow-up (Fig. 4).

DISCUSSION

The additional burden of replacing the aortic arch as an adjunct to elective or emergent proximal repair is not very well defined and makes comparisons with patients undergoing secondary TAR difficult. Most of the papers reporting on outcomes after surgery for Type A dissection or those dealing with reintervention after proximal repair do not discuss arch-related morbidity and mortality separately [15–17]. Although most of the authors report their general experience, it can be assumed that most larger series contain a significant number of MFS that may influence the results, but again, this is not reflected in the discussion. On the other hand, even in larger series reporting results from patients with MFS, the number of those undergoing aortic arch surgery is small, and reports on mortality or neurological outcome are mostly anecdotal [18].

Although, in our series, there was no persistent neurological impairment after either primary or secondary TAR, the low rate of primary TARs performed makes a valid comparison difficult. The same applies for mortality since survival in both primary and secondary TAR was 100% at 10 years. One could argue that the number of patients in need of secondary TAR may have died during follow-up, but that is not the case since there was no significant difference in mortality compared with those patients who never had the arch replaced.

The major risk factor for the need of reintervention on the aortic arch and distal aorta after repaired Type A dissection is a patent false lumen [17]. Therefore, several groups recently began to advocate TAR and implantation of a frozen elephant trunk (FET) in addition to proximal repair in Type A dissection. Sun *et al.* [19] recently published their experiences with 44 MFS patients including 57% of patients with chronic Type A dissection in which they performed primary TAR with implantation of FET. They reported 4.8% mortality over a mean follow-up of 3 years, only 1 stroke and no other neurological event. In contrast to other major surgical series involving MFS patients, the rate of reinterventions

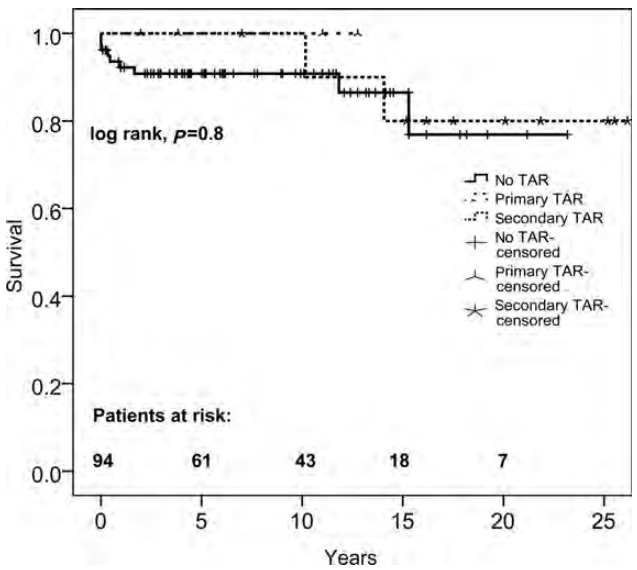

Figure 2: Kaplan–Meier curve showing no significant differences regarding survival in patients who underwent primary, secondary or no TARs.

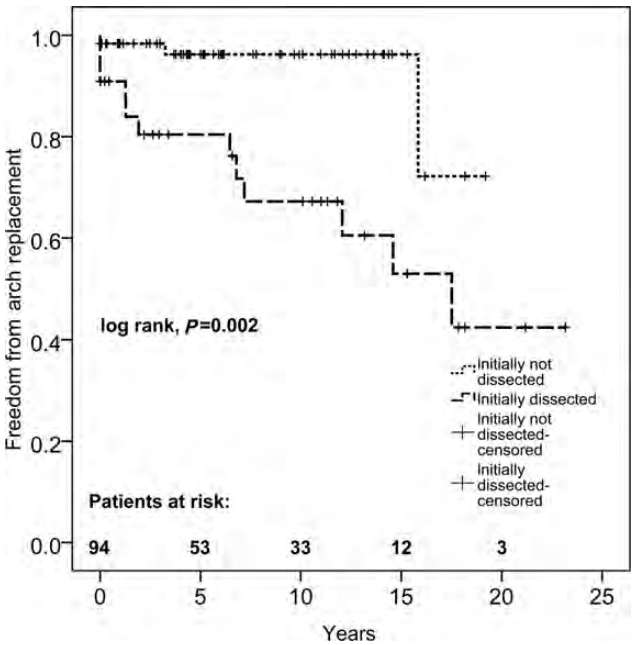

Figure 3: Kaplan–Meier curve showing significantly worse freedom from secondary TARs in patients initially presenting with AAD compared with those without AAD.

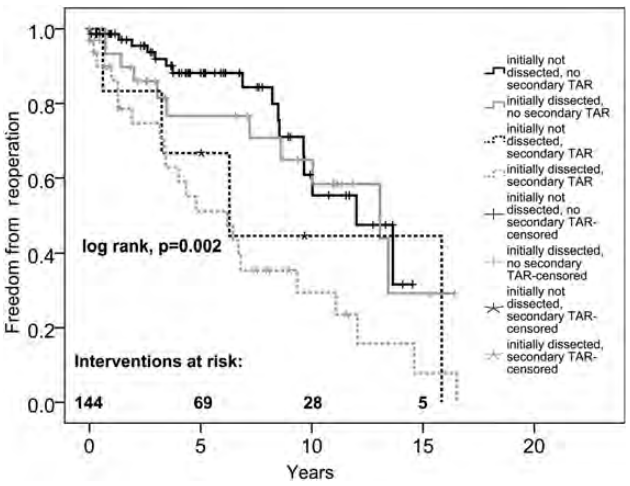

Figure 4: Kaplan–Meier curve depicting freedom from reoperation in patients initially presenting with or without AAD as well as those with or without secondary TARs.

was surprisingly low. Although primary technical success seems feasible, there is evidence that continued dilatation of the aorta around the stent graft will limit the durability of the repair [20]. We only use TEVAR in very selected cases in MFS to bridge a short aneurismal segment between two polyester grafts.

Interestingly, groups based on Asia tend to advocate a more aggressive approach and mostly recommend TAR during initial surgery for Type A dissection. It has been discussed whether this is also due to a more favourable anatomy in the Asian population and a more pronounced atherosclerotic burden in Western countries, which increases the risk for stroke during TAR.

In 2009, Uchida *et al.* [21] published one of the very few reports comparing the hemi-arch replacement with an open distal anastomosis to TAR with implantation of an FET. In 120 patients presenting with acute Type A dissection, mortality was only 4% with no new cerebral events and a survival of 95% at 5 years in the FET and 69% in the hemi-arch group.

Although pseudoaneurysm or dehiscence at the level of the distal anastomosis has been described as a frequent cause for reoperation [16], it was a rare event in our series. Obviously, MFS patients do have a more fragile tissue, and our strategy to use glue and bovine pericardium to reinforce all anastomosis, including those of the coronary buttons, may have positively influenced the outcome in this series.

Although the need for replacing the entire thoracoabdominal aorta was somewhat higher in the group with secondary TAR, this did not reach statistical significance and the difference completely vanishes when including those patients who suffered from acute dissection at any point during follow-up.

These data suggest that it is the dissection itself that drives the need for reoperations in these patients and that the aortic arch is only one of the many segments that has to be repaired over the years. In a large series of MFS patients, Tagusari *et al.* [18] showed that there was no significant difference regarding the rate of reoperation in patients with persisting dissection in the descending aorta after TAR compared with those without. The rate for reinterventions was 50% in both groups at 10 years. Nevertheless, the rate of reoperation was higher in patients with a dissection in the aortic arch where only the ascending aorta was replaced compared with those patients without a dissected arch. Therefore, in the rare cases, where the dissection is confined to the aortic arch, complete exclusion of the dissection may reduce the need for reinterventions and should be attempted.

In large series of 95 patients with MFS, 9.7% required reinterventions on the distal aorta after elective aortic root surgery, compared with 44% after surgery for acute Type A dissection, including 4 (15%) with secondary TAR [15]. Although the rate of secondary TAR in our population was higher, comparisons are difficult since the paper does not specify the percentage of primary TAR during the initial proximal repair.

Bachet *et al.* [22] reported that, in their experience with MFS patients, secondary TAR had to be performed in 16% after elective root surgery and in 73% of patients after Type A dissection compared with 3 and 33% of patients, respectively, in the current series. In-hospital mortality was 13 with 9.2% major neurological events. In the subgroup of patients with secondary TAR, mortality was considerably higher than in our series with 9 and 22% of patients suffering from neurological events, although there was no mortality and no neurological event after the initial surgery.

In our current series, the need for secondary TAR in patients after elective proximal repair was very low such that primary TAR cannot be recommended. Nevertheless, we advocate liberal use of a short circulatory arrest to perform the hemi-arch replacement if there is evidence of dilatation at the level of the brachiocephalic trunk.

Replacing the aortic arch during initial surgery for AAD obviously spares the patients secondary TAR, but it does not protect the patient from reoperations on primarily non-treated aortic segments, ultimately leading up to replacement of the entire aorta. Furthermore, two-thirds of patients after AAD will never need additional arch procedures if the proximal arch was addressed during the initial surgery.

Considering the advances in aortic surgery over the past decade, even complex reoperations seem to carry a moderate risk if performed in an elective setting. Therefore, we suggest delaying major additional procedures during initial surgery for AAD since they can be performed more safely under elective circumstances. In our current study, there was no 'interstage' mortality, new neurological events or operative or 30-day mortality after secondary TAR.

Therefore, we suggest that, within the spectrum of repairing acute Type A dissection ranging from isolated replacement of the ascending aorta to root replacement with TAR and implantation of a FET, our strategy to perform aggressive root replacement and a hemi-arch procedure with replacement of the concavity of the aortic arch seems to balance the need for further reinterventions and the operative risk.

CONCLUSION

MFS patients undergoing elective root repair have small risk of reinterventions on the aortic arch, and primary replacement does not seem to be justified.

In patients with AAD, the need for reinterventions is precipitated by the dissection itself and not by limiting the procedure to the hemi-arch replacement in the emergency setting. This strategy is associated with low mortality in MFS patients presenting with AAD.

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Conflict of interest: none declared.

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APPENDIX. CONFERENCE DISCUSSION

Dr T. Dessing *(Nieuwegein, Netherlands)*: I think in Antonius Hospital in Nieuwegein we have the same strategy and same approach for these kinds of patients. I have one question for you. What is your opinion on replacement of the total arch with a frozen elephant trunk in Marfan patients with a type A dissection?

Dr Schoenhoff: We were very hesitant to perform any type of endovascular procedure in this patient population. In a report by Norwood and colleagues it was shown that the aorta dilates around the stented segment. So we only use stent grafting to bridge a short aneurysmal segment between two already replaced aortic segments so that we can use the Dacron as a landing zone, but we won't do a frozen elephant trunk in a Marfan patient.

Dr W. Harringer *(Braunschweig, Germany)*: May I just have a brief last question. In my experience, the worst type of Marfan patients are the Marfan type II, the Loeys-Dietz syndrome patients, which is rather difficult, of course, to assess preoperatively. But this might be the only group where maybe a more aggressive strategy could be recommended because of the huge progression of the disease and the malignant course. What are your thoughts on that? Are you meanwhile doing routine testing for this type of Marfan?

Dr Schoenhoff: In our Marfan population, 80% of patients underwent genetic testing and 80% of these were mutation positive. And actually, in retrospect, we also had some Loeys-Dietz patients that were primarily treated as having Marfan syndrome. Interestingly, many of the recent publications just report on children or young adolescents with very aggressive forms of this disease, but we forget that we all already carried out surgery on these patients in past. I think that what has been published now is just the tip of the iceberg, and I'm not sure if the majority of Loeys-Dietz patients are very much different from Marfan patients with regard to the aortic arch.

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EDITORIAL COMMENT

Total aortic arch replacement in Marfan patients: caution or boldness?

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Keywords: Aortic surgery • Marfan syndrome • Aortic arch • Acute aortic dissection • Connective tissue disease

In their article titled 'Should aortic arch replacement be performed during initial surgery for aortic root aneurysm in patients with Marfan syndrome?' Schoenhoff *et al.* raise again a very important question which, after several decades of aortic surgery in this particular group of patients, still remains unresolved.