Single-center experience with thoracoabdominal aortic replacement in patients with Marfan syndrome

Saad Rustum, MD, Osama Zahlout, MD, Andreas Martens, MD, Tim Kaufeld, MD, Heike Krüger, RN,
Linda Rudolph, MS, Axel Haverich, MD, PhD, Malakh Shrestha, MBBS, PhD, and
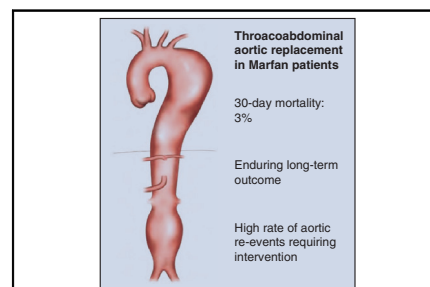Erik Beckmann, MD, PhD

ABSTRACT

Objectives: Patients with Marfan syndrome are usually not suitable for endovascular repair of the thoracoabdominal aorta. This study was designed to analyze our center's experience with open surgical thoracoabdominal aortic replacement in Marfan patients.

Methods: This was a retrospective study with prospective follow-up. Between January 1995 and September 2021, a total of 648 patients underwent thoracoabdominal aortic replacement at our center. Of these, 60 had Marfan syndrome and were included in this study.

Results: The mean age was 39.5 ± 10.7 years, and 36 (60%) were male. Ten (17%) had aortic aneurysm, 4 (7%) acute/subacute dissection, and 46 (77%) chronic dissection. Patients presented with the following extent of aortic disease according to the Crawford classification: I-17 (28%), II-18 (30%), III-22 (37%), IV-2 (3%), and V-1 (2%). The mean cardiopulmonary bypass time was 173.9 ± 84.7 minutes. Four (7%) patients required stent graft extraction. Postoperatively, 5 (8%) patients required rethoracotomy and 6 (10%) tracheostomy. One (1.7%) patient had permanent paraplegia and 2 (3%) permanent paraparesis. Two (3%) patients had stroke. One (1.7%) patient was discharged with dialysis. The 30-day mortality was 3% (n = 2). Median follow-up time was 21.5 (range, 9.4-33.6) years. The 1-, 5-, and 10-year survival rate was 87%, 80%, and 68%, respectively. There were 16 aortic reinterventions in 9 patients during follow-up.

Conclusions: Thoracoabdominal aortic replacement remains a complex procedure but can be done extremely safely in Marfan patients. Perioperative mortality rates are very low, and the long-term outcomes are enduring. Because endovascular aortic repair is not recommended for patients with connective tissue disease, open surgery remains an important cornerstone of therapy. (JTCVS Open 2022;12:13-9)

Throacoabdominal aortic replacement in Marfan patients
30-day mortality: 3%
Enduring long-term outcome
High rate of aortic re-events requiring intervention

Summary of the main findings of this study.

CENTRAL MESSAGE

Open surgical thoracoabdominal aortic replacement in Marfan patients can be performed with very low perioperative risk and provides enduring outcome.

PERSPECTIVE

Open surgical aortic replacement remains the gold standard for Marfan patients with thoracoabdominal aortic disease, even in the age of endovascular therapy. Thoracoabdominal aortic replacement can be performed with acceptable risk and enduring long-term outcome.

▶ Video clip is available online.

Marfan syndrome is a well known autosomal dominant inherited connective tissue disorder with multisystem features involving the cardiovascular system.[1,2] These cardiovascular pathologies including aneurysms and dissections represent the leading cause of increased morbidity and mortality in patients with Marfan syndrome.[3] Aortic root or ascending repair is the most common lesion affecting 75%-85% of these patients due to dilatation or type A dissection,[4,5] and survivors of aortic root replacement, especially those with aortic dissection, might need distal aortic repair.[6,7]

According to high failure rates at the mid- and long-term in Marfan patients, endovascular aortic repair (EVAR) is currently not recommended as a standard option for these patients.[8,9] Endovascular repair has a high frequency of

Abbreviations and Acronyms
EVAR = endovascular aortic repair
TEVAR = thoracic endovascular aortic repair

reintervention. Open thoracoabdominal aortic replacement remains the gold standard treatment for aneurysms and chronic dilatations after aortic dissection in Marfan syndrome patients.[1]

However, open surgical replacement of the thoracoabdominal aorta is a complex procedure and carries some risk for morbidity and mortality. Previously published studies report perioperative mortality rates ranging from 1.3% to 17%.[10-12] Perioperative complications, including paraplegia, stroke, renal failure, and vocal cord paralysis, remain also a matter of concern.[13,14] The aim of this study was to present our short- and long-term results and clinical outcome of open thoracoabdominal aortic replacement in patients with Marfan syndrome.

METHODS
Study Design
We set up a single-center retrospective clinical study with follow-up. The main findings of this study are summarized in Figure 1 and in Video 1. The study was approved by our institution's ethics committee (10425_BO_K_2022). Patients were asked to give informed consent. We retrospectively analyzed our center database to identify patients with Marfan syndrome, who received thoracoabdominal aortic replacement.

From January 1995 to September 2021, a total of 648 patients underwent open thoracoabdominal aortic replacement at our center. Of these,

60 patients had Marfan syndrome. The Crawford classification and the DeBakey classifications were used to specify aortic pathology.

Study Definitions and Follow-up
We examined data related to patient characteristics, surgical procedures, postoperative outcome, and survival. Operative mortality was defined as death occurring within 30 days of the operation.

Operative complications were considered temporary if present at the time of hospital stay and re-treated at the discharge or within follow-up. Stroke was defined as a new-onset clinically and/or radiographically evident permanent brain injury. Paraplegia was defined as a complete loss of motor strength of the lower extremity, and paraparesis as a partial motor lower extremity deficit. Renal failure was defined as an increase of the serum creatinine ≥ 2.5 mg/dL.

Follow-up was done according to the common guidelines.[15] Patients were contacted via telephone, and seen in our clinic. Primary care physicians were contacted to obtain examination results.

Surgical Technique
This sections describes the current protocol at our institution. Patients at high risk for spinal cord injury underwent preoperative placement of a cerebrospinal fluid drainage. In this study, 13 patients received an intrathecal catheter the day before the operation and cerebrospinal fluid was drained if pressure increased to >10 mm Hg intra- and postoperatively. To enable the collapse of the left lung, intubation was performed with a double-lumen endotracheal tube. Patients were placed in the left helical position on a vacuum beanbag. A left lateral thoracolaparotomy with diaphragmatic detachment and retroperitoneal dissection was used to obtain access. After systemic heparinization, extracorporeal circulation was established via femoro–femoral access, and the patient was cooled to 32 °C. The descending aorta was reconstructed from proximal to distal with a Dacron graft prosthesis (Unigraft; Braun). During the early years of our experience, the thoracoabdominal aorta was replaced using a straight Dacron prosthesis and the visceral arteries were reimplanted as an island. More recently, we prefer a 4-branched Dacron prosthesis to reconnect the visceral arteries

Open surgical thoracoabdominal aortic replacement in Marfan patients

METHODS
- Retrospective study with follow-up
- Procedure: thoraco abdominal aortic replacement
- Study time: 1995-2021
- Total patients: n = 648
- Marfan patients: n = 60

RESULTS

Years	0	1	5	10	15	20
Patients at risk	60	47	35	26	12	6
Survival	100%	86%	78%	68%	58%	44%

- 30-Day mortality: 3%
- Median follow-up time: 21.5 (9.4-33.6 years
- Survival: 87% at 1 year 80% at 5 years 68% at 10 years
- 16 aortic reinterventions during follow up in 9 patients

IMPLICATIONS
- The perioperative risk is low
- Long-term outcome is enduring
- Marfan patients have a high frequency of aortic reoperation
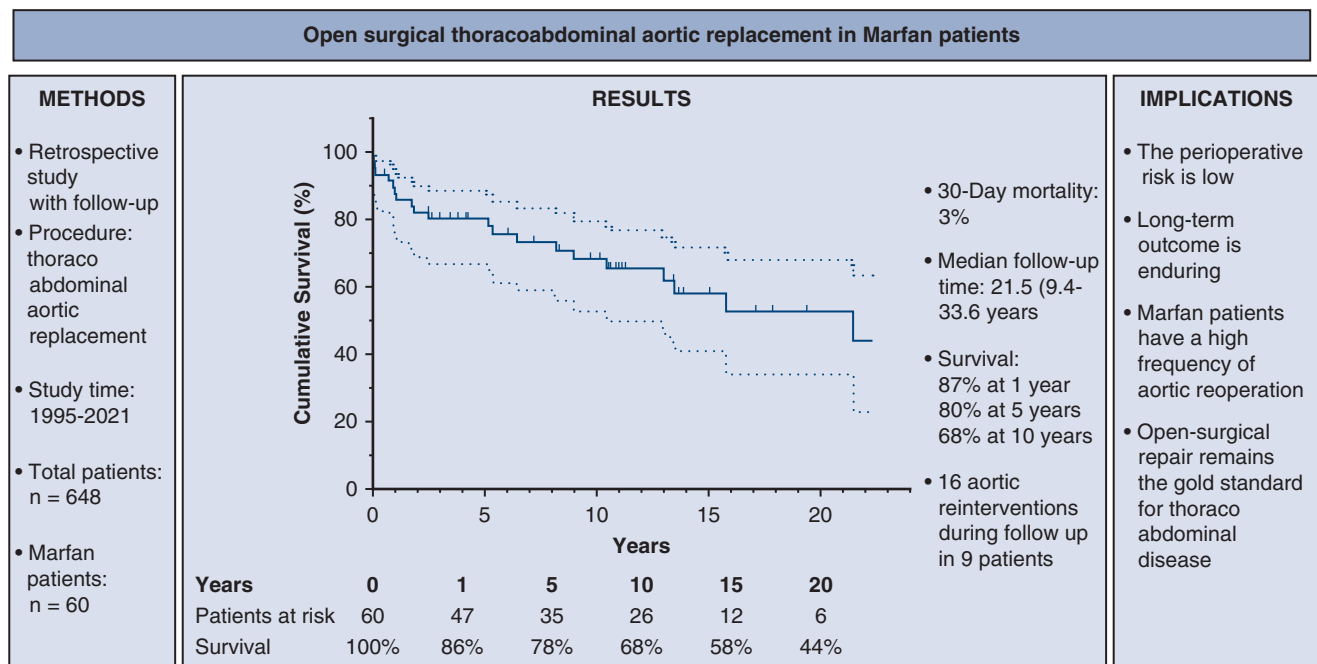- Open-surgical repair remains the gold standard for thoraco abdominal disease

FIGURE 1. Summary of the main findings of the study.

Single-Center Experience with Thoracoabdominal Aortic Replacement in Patients with Marfan Syndrome

Saad Rustum, Osama Zahlout, Andreas Martens, Heike Krüger, Linda Rudolph, Axel Haverich, Malakh Shrestha, Erik Beckmann

Department of Cardiothoracic, Transplantation & Vascular Surgery
Hannover Medical School, Germany

VIDEO 1. The video summarizes the study, including background information, methods, results and conclusions. Video available at: https://www.jtcvs.org/article/S2666-2736(22)00315-1/fulltext.

individually. Intercostal arteries were reimplanted depending on the size and backflow. After the operation, patients were transferred to the intensive care unit. Patients underwent early wake-up trial for neurological assessment. If low-risk patients who did not undergo preoperative cerebrospinal fluid drainage placement showed symptoms of spinal cord injury, they received emergent cerebrospinal fluid drainage.

Statistical Analysis

Data analysis was done using SPSS 26 Statistics software (IBM Corp). Normal distribution of variables was analyzed via the Kolmogorov–Smirnov test. Normally distributed continuous variables are stated as mean ± standard deviation, whereas continuous variables without normal distribution are stated as median and interquartile range. Kaplan–Meier analysis was used for evaluating survival and aortic reoperation.

RESULTS

Preoperative Patient Demographic Characteristics

The preoperative patient characteristics are presented in Table 1. The mean age of patients at the time of surgery was 39.5 ± 10.7 years. Most of our cohort was male (n = 36; 60%). Of our cohort, 49 patients (82%) had previously underwent cardiac surgery and 3 (5%) patients presented with endoleak after previous thoracic EVAR (TEVAR) or EVAR. Aortic dissection was present in 50 (83%) patients, and aortic aneurysm in 10 (17%). Fifty-seven procedures (95%) were scheduled electively and 3 patients (5%) underwent an emergency surgery. The extent of aortic repair is summarized in Table 2.

Intraoperative Data

Intraoperative data are summarized in Table 2. The mean operation time was 374.1 ± 126.8 minutes. Of all cases, 2 (3%) involved left heart bypass and 10 (17%) involved hypothermic circulatory arrest. Cerebrospinal fluid drainage was used in 13 patients (22%) and selective renal artery perfusion in 22 cases (36.7%). Lumbal artery reinsertion was performed in 32 patients (53%) and visceral artery bypass was used in 9 patients (15%). Extraction of previous stent graft was performed in 4 (7%) patients (2 TEVAR, 2 EVAR) and completed with aortic replacement.

TABLE 1. Preoperative patient demographic characteristics (N = 60)

Characteristic	Value
Male sex	36 (60)
Age, y	39.5 ± 10.7
Height, cm	186 (180-190)
Weight, kg	87.7 ± 22.6
Endoleak	3 (5)
Stent dislocation	0 (0)
Previous cardiac surgery	49 (82)
Extent of disease	
Crawford I	17 (28)
Crawford II	18 (30)
Crawford III	22 (37)
Crawford IV	2 (3)
Crawford V	1 (2)
Aortic pathology	
Aneurysm	10 (17)
Acute dissection	2 (3)
Subacute dissection	2 (3)
Chronic dissection	46 (77)
Type of dissection	
DeBakey I	26 (43)
DeBakey II	16 (27)
Status of operation	
Elective	57 (95)
Emergent	3 (5)

Data are presented as n (%), median (interquartile range), or mean ± SD.

Early Postoperative Outcome

The postoperative outcome is shown in Table 3. Prolonged mechanical ventilation (>96 hours) was necessary in 10 patients (16.7%), and 6 (10%) patients required tracheostomy. Postoperative bleeding complications requiring surgical revision occurred in 5 patients (8.3%). Permanent paraplegia was present in 1 (1.7%) patient, and permanent paraparesis in 2 (3.3%). Two patients suffered from stroke (3%). Six patients (10%) developed acute renal failure necessitating temporary dialysis in 3 patients (5%). At discharge, only 1 (2%) patient required dialysis. The 30-day mortality rate was 3% (n = 2). The first patient who died was a 40-year-old woman with mega-aortic syndrome. She underwent composite aortic root replacement and total aortic arch replacement with elephant trunk as a cardiac redo operation initially in our hospital, and had acute rupture of the descending aorta on the sixth postoperative day. She was brought emergently to the operating room with cardiopulmonary reanimation and thoracoabdominal aortic replacement was carried out. However, the patient died on the same day due to myocardial failure. The second patient was a 32-year-old woman with severe obesity (body mass index of 44) and status postaortic valve-sparing root replacement (David) at our center at the age of 20 years.

TABLE 2. Intraoperative data (N = 60)

Characteristic	Value
Operation time, min	374.1 ± 126.8
CPB time, min	173.9 ± 84.7
Crossclamp time, min	111.7 ± 66.6
Extent of repair	
I	19 (32)
II	14 (23)
III	21 (35)
IV	4 (7)
V	2 (3)
Reversed elephant trunk	2 (3)
(Frozen) elephant trunk completion	9 (15)
Lumbar artery reinsertion	32 (53)
Visceral artery bypass	9 (15)
Cerebrospinal fluid drainage	13 (22)
Left heart bypass	2 (3)
Hypothermic circulatory arrest	10 (17)
Extraction thoracic stent graft	2 (3)
Extraction abdominal stent graft	2 (3)

Data are presented as n (%) or mean ± SD. *CPB*, Cardiopulmonary bypass.

TABLE 3. Early postoperative outcome

Characteristic	Value
Intensive care unit stay, d	3 (2-7)
Mechanical ventilation time, d	0.8 (0.5-2.0)
Tracheostomy	6 (10)
Stroke	2 (3)
Paraplegia	
Temporary	1 (1.7)
Permanent	1 (1.7)
Paraparesis	
Temporary	0 (0.0)
Permanent	2 (3.3)
Chylothorax	0 (0)
Left vocal cord paralysis	3 (5)
Bleeding	5 (8)
Acute kidney failure	6 (10)
Dialysis	
Temporary	3 (5)
Permanent	1 (1.7)
Myocardial infarction	0 (0)
Extracorporeal membrane oxygenation	1 (1.7)
30-Day mortality	2 (3)

Data are presented as n (%) or median (interquartile range).

She underwent thoracoabdominal aortic replacement and infrarenal aorto–bi-iliac replacement. She suffered from intracranial hemorrhage due to cerebral aneurysm rupture and died on postoperative day 3.

Late Outcome

The follow-up was complete for 95% of all patients and comprised a total of 512 patient-years. The Kaplan–Meier survival curve is shown in Figure 2. The 1-, 5-, and 10-year survival rates were 87%, 80%, and 68%, respectively. The causes of late deaths were: 1 patient underwent redo mitral valve surgery 357 days after descending aortic replacement and died postoperatively due to cardiac failure. One patient died because of hemorrhagic shock due to gastrointestinal bleeding 337 days after descending aortic replacement, and 1 patient died from multiorgan failure 635 days after the index surgery. The causes of death in all other patients are unknown.

During a median follow-up time of 21.5 (range, 9.4-33.6) years, a total of 16 aortic reinterventions were performed in 9 patients. The Kaplan–Meier survival curve for freedom from reintervention is shown in Figure 3. The freedom from aortic reintervention at 1, 5, and 10 years was 96%, 70%, and 68%, respectively. The aortic reinterventions were: 1 ascending aortic replacement combined with aortic valve replacement, mitral valve surgery and left internal mammary artery bypass, 1 conventional total aortic arch replacement, 4 total aortic arch replacements with the frozen elephant trunk procedure, 3 descending aortic replacements, 2 TEVAR, 1 infrarenal aortic replacement, 2 abdominal EVAR, 1 renal bypass due to insufficiency of the anastomosis, and 1 aorta–subclavian bypass.

DISCUSSION

Thoracoabdominal aortic replacement remains a complex procedure but this study showed that this operation can be done extremely safely in Marfan patients. Our study showed very low perioperative mortality rates, and enduring long-term outcomes. Many Marfan patients require aorta-related reinterventions, however. Because EVAR is not recommended for patients with connective tissue disease, open surgery remains an important cornerstone of therapy.

Marfan patients have an increased risks for morbidity and mortality if history of complex aortic disease is present.[3] Open thoracoabdominal aortic repair remains the treatment of choice with good outcomes and associated with a low reintervention.[16]

Coselli and colleagues[10] reported 4% mortality in 127 patients with Marfan syndrome who underwent thoracoabdominal aortic repair. Similarly, Omura and colleagues[17] reported no in-hospital mortality in their experience of thoracoabdominal aortic repair in 20 patients with Marfan syndrome. Ghanta and colleagues[18] reported no deaths with in 30 days of operations. With an early mortality rate
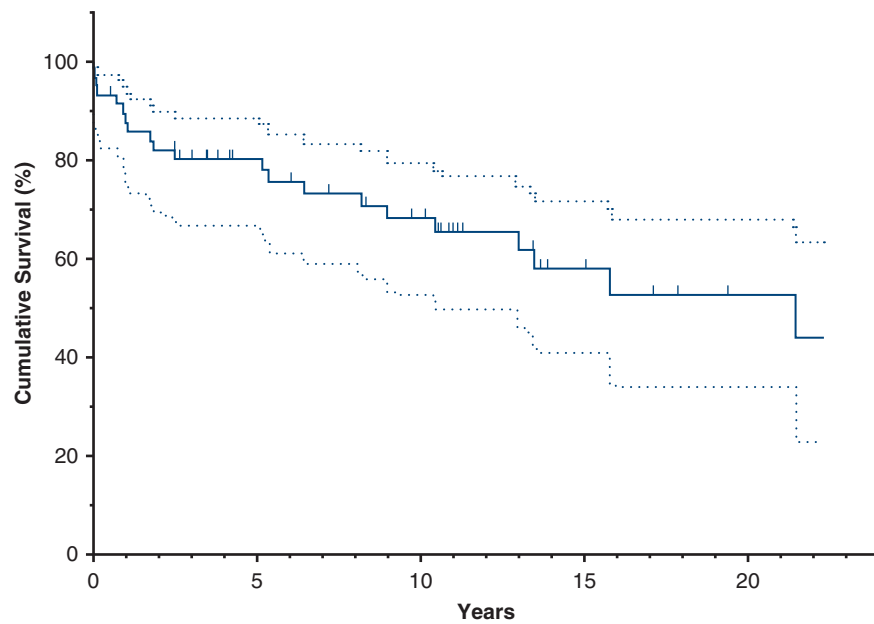
FIGURE 2. Kaplan–Meier survival curve of Marfan patients after thoracoabdominal aortic replacement. X-axis denotes time after surgery. Curve includes 95% confidence intervals.

of 3%, our study showed comparable results. We believe that the relatively young age at the time of the operation contributed to the low mortality rate. It has been previously shown that younger patients have a lower perioperative risk when undergoing thoracoabdominal aortic repair.[11]

Although neurological complications including stroke, paraplegia, and paraparesis represent catastrophic events, most studies show that the incidence of these complications is relatively low after thoracoabdominal aortic repair in Marfan syndrome patients.[5,17,19,20] Our study showed a very low incidence of permanent paraplegia (n = 1; 1.7%) and permanent paraparesis (n = 2; 3.3%) due to spinal cord ischemia. These results seem to be acceptable, especially against the background that some studies report spinal cord ischemia of up to 14% after descending aortic/thoracoabdominal aortic replacement.[21] The incidence of stroke (n = 2; 3%) was low as well in our study. Neurological complications can be an issue, because previous studies reported stroke rates of up to 5%.[22]

Acute renal failure is not uncommon after complex thoracoabdominal aortic repair. Six patients in our study (10%) developed acute renal failure postoperatively. Four patients required dialysis (6.6%) and 1 patient (1.7%) required permanent dialysis. These results are comparable with other studies published previously.[10] We think that renal protection is of great importance to minimize the risk of renal failure.

Our study investigated the long-term survival, too. The 1-, 5-, and 10-year survival rates were 87%, 80%, and 68%, respectively. Although our patient cohort had a relatively young age at the time of initial surgery, Marfan patients have a reduced life expectancy. This might explain the acceptable but reduced survival rates. The causes of death are known in 3 patients, but are unknown in the remaining patients. This is because of the retrospective character of this study and represents another minor limitation.

In our study, 16 aortic reinterventions were necessary during follow-up. This underlines that patients with connective tissue disease are not cured after aortic replacement, but have a lifelong elevated risk for vascular (re-)operation with associated periprocedural risks. One might have expected an even higher number for aortic reoperation at other aortic segments. With regard to this, we mention that most of our patient cohort had already undergone a cardio (vascular) procedure in the past, mostly for acute aortic type A dissection. In these patients, the aortic root/ascending aorta had already been addressed. Also, some of the patients in the present study might have had an enlarged distal aorta, but died before reaching a diameter necessitating aortic repair.

EVAR in patients with Marfan syndrome is still controversial with little experience and lack of mid- and long-term follow-up studies. Kölbel and colleagues[4] presented a series of 24 Marfan patients who underwent endovascular repair or hybrid procedures with a follow-up time of
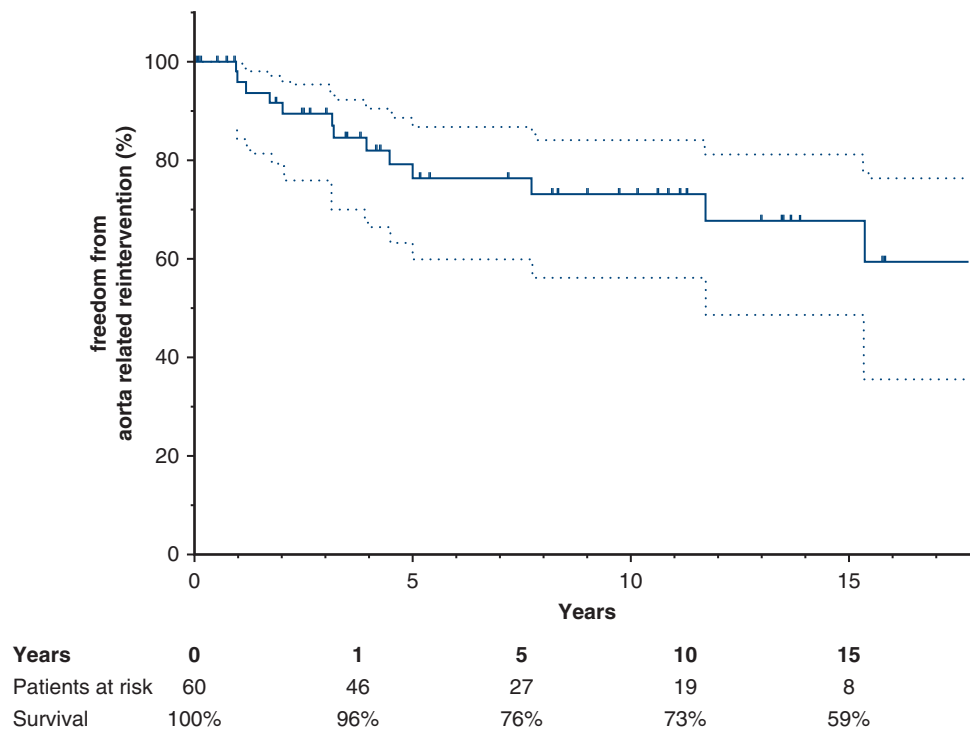
FIGURE 3. Kaplan–Meier curve for freedom from aortic reintervention of Marfan patients after thoracoabdominal aortic replacement. X-axis denotes time after initial surgery. Curve includes 95% confidence intervals.

42 months. The early mortality was 4%, and the survival was 87% at 24 months. The freedom from reintervention at 12 months was 77%. On the contrary, a small study with 6 patients and a follow-up time of 51 months showed that open surgical repair was necessary in 2 patients, and a third patient was considered for surgery. Further, 1 patient died 12 months after endovascular repair.[23] These data show that endovascular therapy is not an enduring solution for Marfan patients. As endovascular therapies improve, it might be an option in the future but not at present. However, it can be a bailout strategy in emergent situations. For instance, our group has published a study on the surgical management of patients with native and prosthetic graft infection of the thoracic aorta.[24] In this study, some patients underwent emergent endovascular stent graft placement because of aortic rupture or fistula formation as a bridge to open surgical repair. Similarly, we think that endovascular therapy could be an option in urgent situations for patients with connective tissue disorders. For these reasons, we think that open surgical replacement remains the preferred choice of therapy for Marfan patients with thoracoabdominal pathologies.

CONCLUSIONS

We believe that open surgical repair remains the gold standard of treatment for the thoracic and thoracoabdominal aorta in patients with Marfan syndrome. This study showed that open surgical repair in modern times can be performed with extremely low perioperative risks. We think that endovascular repair in connective tissue disease can be considered for emergent bailout situations, for instance, as bridge to open repair. However, endovascular repair still remains to show enduring long-term outcome in Marfan patients.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References

1. Pellenc Q, Girault A, Roussel A, De Blic R, Cerceau P, Raffoul R, et al. Optimising aortic endovascular repair in patients with Marfan syndrome. *Eur J Vasc Endovasc Surg*. 2020;59:577-85.
2. Ugurlucan M, Onal Y, Ali Sayin O, Ekiz F, Oztas DM, Basaran M, et al. Endovascular thoracoabdominal replacement after total abdominal aortic debranching. *Aorta (Stamford)*. 2018;6:43-5.
3. Miyahara M, Okita Y. Overview of current surgical strategies for aortic disease in patients with Marfan syndrome. *Surg Today*. 2016;46:1006-18.
4. Kölbel T, Eleshra A, Aldag M, Rohlffs F, Debus SE, Honig S, et al. Endovascular treatment of aortic pathologies in patients with Marfan syndrome: single-center experience. *J Endovasc Ther*. 2022;29:602-10.
5. Mommertz G, Sigala F, Langer S, Koeppel TA, Mess WH, Schurink GWH, et al. Thoracoabdominal aortic aneurysm repair in patients with Marfan syndrome. *Eur J Vasc Endovasc Surg*. 2008;35:181-6.
6. Schoenhoff FS, Jungi S, Czerny M, Roost E, Reineke D, Matyas G, et al. Acute aortic dissection determines the fate of initially untreated aortic segments in Marfan syndrome. *Circulation*. 2013;127:1569-75.

7. Kari FA, Russe MF, Peter P, Blanke P, Rylski B, Euringer W, et al. Late complications and distal growth rates of Marfan aortas after proximal aortic repair. *Eur J Cardiothorac Surg*. 2013;44:163-71.

8. Gagné-Loranger M, Voisine P, Dagenais F. Should endovascular therapy be considered for patients with connective tissue disorder? *Can J Cardiol*. 2016; 32:1-3.

9. Waterman AL, Feezor RJ, Lee WA, Hess PJ, Beaver TM, Martin TD, et al. Endovascular treatment of acute and chronic aortic pathology in patients with Marfan syndrome. *J Vasc Surg*. 2012;55:1234-40; discussion: 1240-1.

10. Coselli JS, Green SY, Price MD, Hash JA, Ouyang Y, Volguina IV, et al. Results of open surgical repair in patients with Marfan syndrome and distal aortic dissection. *Ann Thorac Surg*. 2016;101:2193-201.

11. Tanaka A, Leonard SD, Sandhu HK, Afifi RO, Miller CC III, Charlton-Ouw KM, et al. Open descending and thoracoabdominal aortic repairs in patients younger than 50 years old. *Ann Thorac Surg*. 2019;108:693-9.

12. Frankel WC, Song HK, Milewski RK, Shalhub S, Pugh NL, Eagle KA, et al. Open thoracoabdominal aortic repair in patients with heritable aortic disease in the GenTAC registry. *Ann Thorac Surg*. 2020;109:1378-84.

13. Coselli JS, LeMaire SA, Preventza O, de la Cruz KI, Cooley DA, Price MD, et al. Outcomes of 3309 thoracoabdominal aortic aneurysm repairs. *J Thorac Cardiovasc Surg*. 2016;151:1323-37.

14. Coselli JS, LeMaire SA, Conklin LD, Köksoy C, Schmittling ZC. Morbidity and mortality after extent II thoracoabdominal aortic aneurysm repair. *Ann Thorac Surg*. 2002;73:1107-15; discussion: 1115-6.

15. Akins CW, Miller DC, Turina MI, Kouchoukos NT, Blackstone EH, Grunkemeier GL, et al. Guidelines for reporting mortality and morbidity after cardiac valve interventions. *Eur J Cardiothorac Surg*. 2008;33:523-8.

16. Afifi RO, Tanaka A, Yazji I, Safi HJ, Estrera AL. Thoracoabdominal aortic aneurysm repair in Marfan syndrome: how we do it. *Ann Cardiothorac Surg*. 2017;6: 709-11.

17. Omura A, Tanaka A, Miyahara S, Sakamoto T, Nomura Y, Inoue T, et al. Early and late results of graft replacement for dissecting aneurysm of thoracoabdominal aorta in patients with Marfan syndrome. *Ann Thorac Surg*. 2012;94: 759-65.

18. Ghanta RK, Green SY, Price MD, Arredondo CC, Wainwright D, Preventza O, et al. Midterm survival and quality of life after extent II thoracoabdominal aortic repair in Marfan syndrome. *Ann Thorac Surg*. 2016;101:1402-9; discussion: 1409.

19. Dardik A, Krosnick T, Perler BA, Roseborondo GS, Williams GM. Durability of thoracoabdominal aortic aneurysm repair in patients with connective tissue disorders. *J Vasc Surg*. 2002;36:696-703.

20. Kalkat MS, Rahman I, Kotidis K, Davies B, Bonser RS. Presentation and outcome of Marfan's syndrome patients with dissection and thoracoabdominal aortic aneurysm. *Eur J Cardiothorac Surg*. 2007;32:250-4.

21. Bavaria JE, Appoo JJ, Makaroun MS, Verter J, Yu ZF, Mitchell RS. Endovascular stent grafting versus open surgical repair of descending aortic aneurysms in low-risk patients: a multicenter comparative trial. *J Thorac Cardiovasc Surg*. 2007; 133:369-77.

22. Keschenau PR, Kotelis D, Bisschop J, Barbati ME, Grommes J, Mees B, et al. Editor's choice - open thoracic and thoraco-abdominal aortic repair in patients with connective tissue disease. *Eur J Vasc Endovasc Surg*. 2017;54:588-96.

23. Ince H, Rehders TC, Petzsch M, Kische S, Nienaber CA. Stent-grafts in patients with Marfan syndrome. *J Endovasc Ther*. 2005;12:82-8.

24. Rustum S, Beckmann E, Martens A, Krüger H, Arar M, Kaufeld T, et al. Native and prosthetic graft infection of the thoracic aorta: surgical management. *Eur J Cardiothorac Surg*. 2021;60:633-41.