Original Article

Total arch replacement and frozen elephant trunk for type A aortic dissection after Bentall procedure in Marfan syndrome

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Background: We seek to report the long-term outcomes of the total arch replacement and frozen elephant trunk (TAR + FET) technique for type A aortic dissection (TAAD) following prior Bentall procedure in patients with Marfan syndrome (MFS).

Methods: Between 2003 and 2015, we performed TAR + FET for 26 patients with MFS who developed TAAD following a prior Bentall procedure. Mean age at FET 36.9±9.7 years and 24 were males. TAAD was acute in 8 (30.8%, all new dissections from precious root aneurysm) and chronic in 18 (69.2%, 15 residual and 3 new). The interval from Bentall procedure to FET averaged 6.4±5.8 years, which was significantly longer in the acute group (10.3±6.3 *vs.* 4.6±4.9, P=0.021). The early and long-term outcomes were compared between two groups and risk factors identified for late adverse events.

Results: Operative mortality was 11.5% (3/26). Stroke, lower limb ischemia and reexploration for bleeding occurred in 1 patient each (3.8%). Follow-up was complete in 100% (23/23) at mean 5.1±2.3 years (range, 0.9–11.2 years). The maximal diameter (DMax) of distal aorta in the chronic group was significantly greater at the unstented descending aorta [DA, (56.4±15.5 *vs.* 35.6±12.2 mm, P=0.006)] compared to acute patients. The false lumen was obliterated in 95.7% across the FET and 56.5% in the unstented DA. Distal aortic dilation occurred in 13 patients (11 chronic, 68.8%). Of those 11 patients, 4 underwent an open thoracoabdominal aortic repair and 3 died of distal aortic rupture. Late death occurred in 7 patients at mean 3.9±2.5 years. At 6 years, the incidence was 18% for death, 11% for distal aortic reoperation, and 71% for reoperation-free survival. Survival did not differ between two groups (75.0% *vs.* 71.3%, P=0.851), while acute patients had significantly higher freedom from late rupture and reoperation at 6 years (100% *vs.* 61.9%, P=0.046). Hypertension was the sole risk factor for distal aortic dilatation [hazard ratio (HR) =7.271; 95% confidence interval (CI), 1.814–29.143; P=0.005]. Risk factors for late adverse events were hypertension (HR =6.712; 95% CI, 1.201–37.503; P=0.030) and age <35 years (HR =6.760; 95% CI, 1.154–39.587; P=0.034).

Conclusions: The TAR and FET technique was feasible and efficacious for TAAD following previous Bentall procedure in patients with MFS. Early and late survival did not differ with acute and chronic dissections, while freedom from late rupture and reoperation is significantly higher in patients with acute TAAD. Patients with hypertension and aged <35 years are at higher risk for late distal aortic dilation, reoperation and death.

Keywords: Type A aortic dissection (TAAD); Marfan syndrome (MFS); total arch replacement (TAR); frozen elephant trunk (FET); reoperation; Bentall procedure

Submitted Dec 23, 2017. Accepted for publication Mar 08, 2018.
doi: 10.21037/jtd.2018.03.79
View this article at: http://dx.doi.org/10.21037/jtd.2018.03.79

Introduction

Aortic root dilatation is the most common cardiovascular manifestation in patients with Marfan syndrome (MFS) because of their inherently weakened aortic wall (1). Aortic root aneurysm and aortic dissection followed by aortic rupture are the major causes of morbidity and mortality. Prophylactic aortic root surgery has been effective in preventing acute dissection and rupture (2-4), with low operative mortality and excellent long-term outcomes (2,3,5,6). However, many patients develop aneurysm or dissection beyond the aortic root after prophylactic root surgery (1,4,6). According to recent data from the International Registry on Acute Aortic Dissection (4), acute type A dissection (TAAD) in Marfan patients is usually managed with an emergency root and/or ascending replacement, which is associated with low operative risks but leaves the aortic arch and distal aorta untreated (1). Consequently, reoperation is required to manage the patent false lumen (FL), seen in 29–78% of patients (7-12), and the expanding aneurysm in the distal aorta (7,13,14). In addition, reoperation for arch dissection was required in 40.9–49% of patients (15,16). As a result, the more extensive total arch replacement (TAR) (15) or in combination with the frozen elephant trunk (FET) has been advocated as an approach to TAAD involving the aortic arch in patients with MFS during initial surgical treatment (17-19), despite concerns on the mortality and paraplegia risks and limited imaging follow-up (20,21). Although our previous study has proved the efficacy and durability of TAR + FET for TAAD in MFS (22), data is scarce regarding the long-term outcomes of this technique for TAAD in Marfan patients who have undergone a Bentall operation previously. The present study aims to evaluate the early and long-term outcomes of the FET + TAR techniques for such patients, including mortality and morbidity, late survival and reoperation, changes in the distal aorta, and risk factors for late adverse events.

Methods

Between April 2003 and August 2015, we performed TAR and FET for 26 patients who developed TAAD involving the aortic arch after a prior Bentall procedure. Patients were divided into two groups according to acuity, acute in 8 (30.8%, all new dissections) and chronic in 18 patients (69.2%, 15 residual and 3 new dissections). Indication for prior Bentall was TAAD in 15 (57.7%) and root aneurysm in 11 (42.3%). The diagnosis of MFS was confirmed by the Ghent and/or revised Ghent criteria (23,24). Mean age at TAR + FET was 36.9±9.7 years (range, 17–50 years). Hypertension was seen in 13 (50.0%) patients. The interval between Bentall and FET averaged 6.4±5.8 years, which was significantly longer in patients with acute TAAD (10.3±6.3 *vs.* 4.6±4.9 years; P=0.021). The preoperative clinical profiles are listed in *Table 1*.

All dissections were confirmed by computed tomographic angiography (CTA). Before the FET, the mean maximal diameter (DMax) of the aortic arch was 51.0±16.5 mm, which was larger in chronic TAAD patients, but without significant difference (52.9±18.3 *vs.* 46.9±11.5 mm; P=0.402); the mean DMax in the chronic group were significantly larger compared to acute patients at the proximal descending aorta (DA) (54.8±11.6 *vs.* 43.9±11.6 mm; P=0.036), diaphragmatic hiatus (DH) (44.2±11.4 *vs.* 31.3±8.0 mm; P=0.008) and renal arteries (RA) (37.1±10.1 *vs.* 28.3±7.0 mm, P=0.034), respectively. Coronary artery endoleak was detected in 4 patients with chronic TAAD.

Surgical technique

Our selection criteria for TAR + FET for TAAD in Marfan patients are as follows: (I) dissection with an entry tear located in the transverse arch or DA; (II) aneurysm of the aortic arch or distal aorta (>40 mm in diameter); (III) dissection, aneurysm, or occlusion of the brachiocephalic arteries.

All patients underwent this reoperative procedure through a redo sternotomy. Before sternotomy, cardiopulmonary bypass (CPB) was instituted first by cannulation of the femoral artery and vein. The aortic arch, proximal DA and arch vessels were dissected on CPB before circulatory arrest. The surgical technique, known as the Sun's procedure, has been previously described in detail (17,25-27). Briefly, right axillary artery cannulation is used for CPB and unilateral selective antegrade cerebral perfusion under moderate hypothermic circulatory arrest at 25 ℃ is utilized. The aortic arch is transected between the left carotid and left subclavian arteries (26). After the FET (Cronus®, MicroPort Medical, Shanghai, China) is deployed in the DA, TAR is performed with a 4-branched vascular graft (Maquet, Rastatt, Germany). To minimize the time of cerebral, myocardial and spinal cord ischemia, distal reperfusion is initiated once the distal anastomosis is completed, and the left carotid artery is reconstructed first (after which rewarming is started and the brain is perfused bilaterally), followed by the ascending aorta (to resume

Table 1 Preoperative clinical profile

Variables	Total (n=26)	Acute TAAD (n=8)	Chronic TAAD (n=18)	P value
Age (years)	36.9±9.7	42.0±7.5	34.7±9.9	0.074
Gender (male)	24 (92.3%)	7 (87.5%)	17 (94.4%)	0.540
Hypertension	13 (50.0%)	2 (25.0%)	11 (61.1%)	0.089
Family history	16 (61.5%)	6 (75.0%)	10 (55.6%)	0.347
Initial root aneurysm	11 (47.3%)	8 (100%)	3 (16.7%)	<0.001
Time from Bentall to FET (year)	6.4±5.8	10.3±6.3	4.6±4.9	0.021
Entry tear				
Ascending end graft	6 (23.1%)	1 (12.5%)	5 (27.8%)	0.393
Aortic arch	22 (84.6%)	8 (100%)	14 (77.8%)	0.147
Proximal descending aorta	15 (57.7%)	4 (50.0%)	11 (61.1%)	0.597
Maximal aortic diameter (mm)				
Aortic arch	51.0±16.5	46.9±11.5	52.9±18.3	0.402
Proximal descending aorta	50.9±12.5	43.9±11.6	54.8±11.6	0.036
Mid-descending aorta	41.0±10.2	33.1±7.5	44.9±9.1	0.004
Diaphragmatic hiatus	40.4±11.9	31.3±8.0	44.2±11.4	0.008
Renal arteries	34.1±9.7	28.3±7.0	37.1±10.1	0.034
Thoracoabdominal aorta	52.3±12.1	44.5±10.8	56.1±11.1	0.023

FET, frozen elephant trunk; TAAD, type A aortic dissection.

myocardial perfusion), then the left subclavian artery (LSA), and finally, the innominate artery. Proximal procedures (coronary artery anastomotic leak repair and mitral valve surgery) are performed during the cooling phase.

Patient follow-up

Operative survivors were recommended to take β-blockers as routine medication after discharge, even if they had normal blood pressure.

CTA of the entire aorta was performed at discharge or 1, 3 and 6 months, 1 year and annually henceforth to assess the FL, TL, maximal aortic size (DMax) and growth rate, and complications (endoleak, migration, etc.). The statuses of the FL were classified as: completely thrombosed (obliterated) if no flow was present; partially thrombosed if both flow and thrombus were present; and patent if flow was present in the absence of thrombus (28). The maximum aortic diameter (DMax) was measured from the outer contours of the aortic wall in the axial plane and analyzed at four different distal aortic levels: the stented proximal DA (FET), the unstented DA, the DH and renal arteries. The segmental aortic growth rate was assessed in patients who had ≥2 CT scans postoperatively with an interval of at least 6 months apart. The growth rate was calculated by dividing the diameter differences between the initial and last CTs by the interval in between (mm/year). Dilatation was defined as a maximum diameter ≥50 mm (45 mm in patients with a family history of aortic surgery or rupture) or an average growth rate of ≥5 mm/year.

Follow-up was complete in 100% (23/23) by June 2017 for a mean duration of 5.1±2.3 years (range, 0.9–11.2 years).

Statistical analysis

Statistical analysis was performed using SPSS for Windows 19.0 (SPSS Inc., Chicago, IL, USA), Stata 15.1 for Mac (StataCorp, College Station, TX, USA) and Prsim 7.0d for Mac (GraphPad, La Jolla, CA, USA). Data are expressed as the mean ± standard deviation or number (percentage) and were compared using the Student *t*-test or Pearson chi-square test for normal distributions, and the Mann-Whitney

U test for abnormal distributions, as appropriate. Risk factors for distal aortic dilatation, late distal adverse events (reoperation and rupture) were identified with Cox regression model. Variables considered in multivariate analysis included age, hypertension, preoperative descending aortic diameter, FL patency, annual growth rate and maximum diameter of thoracoabdominal aorta. Survival and freedom from dilatation and reoperation were estimated using the Kaplan-Meier method, and intergroup comparisons made with the log-rank test. Competing risks of death and reoperation were analyzed with the Fine and Gray proportional hazards model. All statistical tests were 2-sided and a P value of <0.05 was considered statistically significant.

Results

Operative data

The entry tears were located in the aortic arch in 22 (84.6%), proximal DA in 15 (57.7%), and the ascending aorta (distal to the Bentall graft) in 6 (23.1%) patients. Multiple entry tears were seen in 13 (50%) patients. The extent of dissection was to the iliac artery in 24 (92.3%) patients and to the distal descending thoracic aorta in 2 (7.7%) patients.

An entry tear in the arch was found in 100% (8/8) patients of the acute group (all with prior root aneurysm) and in 77.8% (14/18) patients of the chronic group (P=0.147). In the chronic group, there were 15 residual dissections from previous TAAD; among these, 1 (6.7%) patient underwent FET for an entry tear in the arch 9 years after Bentall. In his prior Bentall procedure, no intimal tear was found in the arch and DA. The other 14 (14/15, 93.3%) patients had intimal tears located in the aortic arch or proximal DA that were not resected during initial surgery and underwent FET for dilation at mean 2.3±1.3 years, with an annual growth rate of 8.6 mm in the DA and 13 mm in the DH.

The times of CPB, cross-clamp and cerebral perfusion were 184±42, 88±32, 23±6 minutes, respectively. CPB time was longer in chronic patients (191.3±37.2 *vs*. 168.8±49.4 minutes, P=0.208), but without significant difference between two groups. Concomitant procedures included mitral valve surgery in 2 (7.7%), ascending aortic to femoral bypass in 1 (3.8%), and coronary leakage repair in 4 (15.4%) patients, respectively.

Operative mortality and morbidity

The operative mortality rate was 11.5% (3 of 26), occurring in 1 patient with acute TAAD and 2 with chronic TAAD (12.5% *vs*. 11.1%, P=0.919). All 3 expired patients were with a root aneurysm; 2 had a markedly dilated arch (65 and 150 mm in size) with a CPB time of 246 and 153 minutes, respectively. The cause of death was multiorgan failure, hemorrhagic stroke and distal aortic rupture, in 1 each (3.8%). No spinal cord injury occurred. Lower limb ischemia and reexploration for bleeding occurred in 1 patient each (3.8%) (*Table 2*).

Distal aortic dilation and reoperation

Obliteration of the false lumen was observed in 95.7%, 56.5% and 26.1% at the levels of FET, DA and DH, respectively. Patients with acute TAAD had higher rates of FL thrombosis (*Figure 1*) at the levels of FET (100% *vs*. 93.8%, P=0.499) and DA (71.4% *vs*. 50.0%, P=0.340); this difference was significant at the level of DH (57.1% *vs*. 12.5%, P=0.025).

On the latest follow-up CT, the DMax of distal aorta in the chronic group were greater compared to patients with acute TAAD (*Figure 2*), which differed significantly at the levels of FET (65.9±22.6 *vs*. 41.1±12.5 mm, P=0.015) and DA (56.4±15.5 *vs*. 35.6±12.2 mm, P=0.006). However, the DMaxs did not differ significantly at the levels of DH (52.6±16.9 *vs*. 37.6±13.7 mm, P=0.056) and RA (41.4±13.1 *vs*. 32.3±11.0 mm, P=0.133).

Distal aortic dilatation occurred in 13 patients during follow-up, 2 in the acute and 11 in the chronic groups (28.6% *vs*. 68.8%, P=0.074). In the chronic group, 3 patients died of distal aortic rupture and 4 underwent an open thoracoabdominal aortic repair (TAAAR). The median interval from FET to TAAAR was 0.5 years. The average DMax at TAAAR was 83.2±21.2 mm (range, 70–115 mm).

The freedom from reoperation on the distal aorta was 86.96% [95% confidence interval (CI), 64.8–95.6%] at 1, 3 and 6 years, respectively (*Figure 3*); and 72.5% (95% CI, 33.8–90.9%) at 7 years.

Late survival

Late death occurred in 7 patients. The cause of death was distal aortic rupture from residual dissection in 3 patients, all in the chronic TAAD group, and non-cardiac reasons in 4 patients (*Table 3*). Survival was 84.6% (95% CI, 64.0–93.9%), 76.9% (95% CI, 55.7–88.9%) and 72.4% (95% CI, 50.5–85.8%) at 1, 3, and 6 years, respectively (*Figure 4*). At 6 years after FET, survival did not differ between two

Table 2 Operative outcomes

Variables	Total (n=26)	Acute TAAD (n=8)	Chronic TAAD (n=18)	P value
Operative data				
Cardiopulmonary bypass time (min)	184.4±41.7	168.8±49.4	191.3±37.2	0.208
Cross-clamp time (min)	87.9±32.0	86.0±37.5	88.8±30.3	0.843
Cerebral perfusion time (min)	22.9±6.4	22.9±6.2	22.8±6.7	0.988
Concomitant procedures				
Mitral valve surgery	2 (7.7%)	0	2 (11.1%)	0.326
Ascending aortic to femoral bypass	1 (3.8%)	1 (12.5%)	0	0.126
Anastomotic leak repair	4 (15.4%)	0	4 (22.2%)	0.147
Frozen elephant trunk				
Diameter (mm)	26.8±1.5	27.3±1.5	26.6±1.5	0.286
Length (mm)	101.5±7.8	105.0±9.3	100.0±6.9	0.199
Postoperative data				
Operative mortality	3 (11.5%)	1 (12.5%)	2 (11.1%)	0.919
Operative complications	5 (19.2%)	3 (37.5%)	2 (11.1%)	0.115
Spinal cord injury	0	0	0	–
Low cardiac output	1 (3.8%)	0	1 (5.6%)	0.497
Stroke and cerebral hemorrhage	1 (3.8%)	0	1 (5.6%)	0.497
Visceral and lower limb ischemia	1 (3.8%)	1 (12.5%)	0	0.126
Distal aortic rupture	1 (3.8%)	1 (12.5%)	0	0.126
Reexploration for bleeding	1 (3.8%)	1 (12.5%)	0	0.126

TAAD, type A aortic dissection.

groups (75.0% *vs.* 71.3%, P=0.851; *Figure 5*), while freedom from distal aortic reoperation and rupture were significantly higher in the acute group at 6 years (100% *vs.* 61.9%, P=0.046; *Figure 6*).

In competing risks analysis, the incidence of was 18% for late death, 11% for distal aortic reoperation, and 71% for event-free survival at 6 years after FET (*Figure 7*).

Risk factors for late adverse events

Hypertension was the sole risk factor for distal aortic dilatation [hazard ratio (HR) =7.271; 95% CI, 1.814–29.143; P=0.005]. Risk factors for late adverse events (aortic rupture and reoperation) were hypertension (HR =6.712; 95% CI, 1.201–37.503; P=0.030) and age <35 years (HR =6.760; 95% CI, 1.154–39.587; P=0.034) (*Table 4*).

Discussion

In surgical management of type A dissection for patients with MFS, the extent of surgery (limited proximal versus extensive total arch repair) has to be balanced between the operative mortality risk and the unique challenges in those patients, such as residual and new dissections, and expanding aneurysms in downstream aorta (1,14,16). This is particularly important in the surgical management of Marfan patients sustaining TAAD after a Bentall procedure. Besides the resternotomy and adhesions, technical complexities in this reoperative setting include the multiple residual intimal tears in the arch or proximal DA, usually with a huge arch aneurysm, and significantly dilated proximal distal aorta. Dissecting the brachiocephalic arteries, especially the left subclavian is much more technically demanding. A recent study has found that at the time of initial repair for TAAD in MFS, a more aggressive

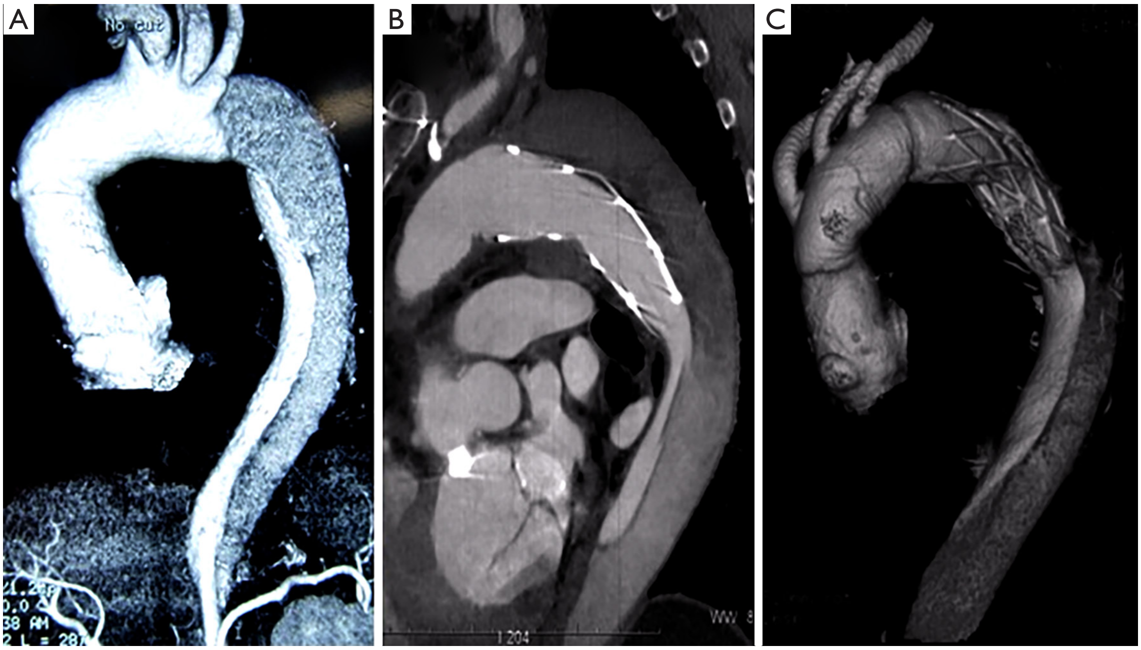

Figure 1 A 40-year-old male with Marfan syndrome developed acute type A dissection involving the aortic arch 10 years after a Bentall procedure for root aneurysm (A). CTA at 6 months (B) and 6 years (C) after FET showed false lumen thrombosis and aortic wall remodeling around the FET and in the descending aorta (C). CTA, computed tomographic angiography; FET, frozen elephant trunk.

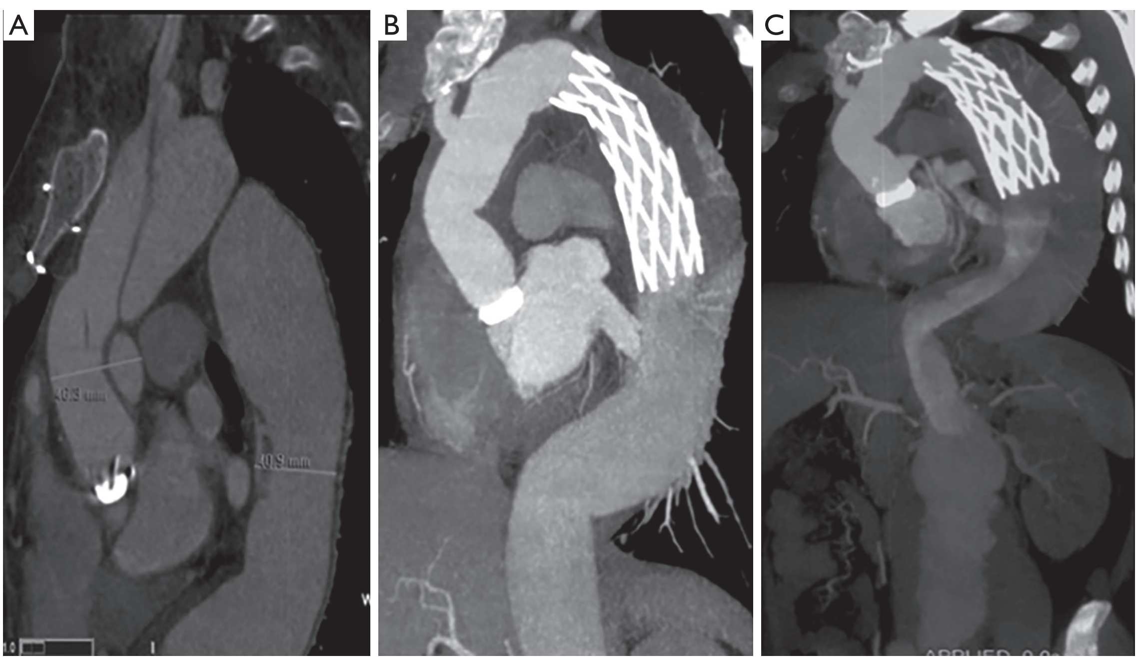

Figure 2 A male Marfan patient underwent a Bentall procedure for type A dissection at the age of 20 years. At 16 months postoperatively, he sustained a chronic dissection involving aortic arch (A), which was managed with FET. At 1 year (B) and 4 years after FET (C), CTA detected thrombosis of false lumen and aortic wall remodeling across the FET and in the descending aorta. However, distal dilatation was observed on CTA at 4 years after FET (C). CTA, computed tomographic angiography; FET, frozen elephant trunk.

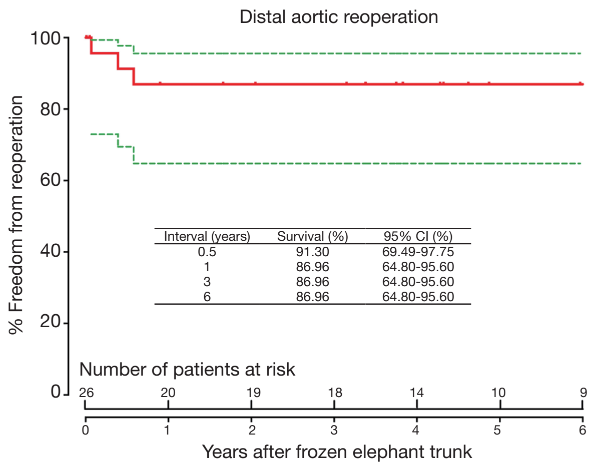

Figure 3 Freedom from reoperation after frozen elephant trunk.

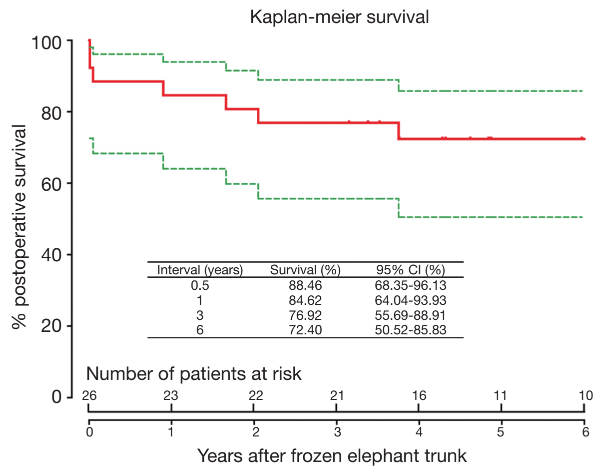

Figure 4 Survival after frozen elephant trunk.

Table 3 Late outcomes

Variable	Total∗ (n=23)	Acute TAAD (n=7)	Chronic TAAD (n=16)	P value
Follow-up complication	3 (13.0%)	0	3 (18.8%)	0.219
Proximal stent leakage	2 (8.7%)	0	2 (12.5%)	0.328
Distal end stent entering false lumen	1 (4.3%)	0	1 (6.2%)	0.499
Late adverse events				
Dilatation	13 (56.5%)	2 (28.6%)	11 (68.8%)	0.074
TAAAR	4 (17.4%)	0	4 (25.0%)	0.146
Late death	7 (30.4%)	2 (28.6%)	5 (31.2%)	0.898
Distal aortic rupture	3 (13.0%)	0	3 (18.8%)	0.219
Non-cardiac cause	4 (17.4%)	2 (28.6%)	2 (12.5%)	0.349
TAAAR and late rupture	7 (30.4%)	0	7 (43.8%)	0.036
Maximal diameter at last CT (mm)				
Frozen elephant trunk	57.7±22.9	41.1±12.5	65.9±22.6	0.015
Unstented descending aorta	49.4±17.4	35.6±12.2	56.4±15.5	0.006
Diaphragmatic hiatus	47.6±17.1	37.6±13.7	52.6±16.9	0.056
Renal arteries	38.3±12.9	32.3±11.0	41.4±13.1	0.133
Thoracoabdominal aorta	62.1±22.4	46.7±12.4	69.7±22.5	0.022

Excluding 3 operative deaths. TAAAR, thoracoabdominal aortic aneurysm repair; TAAD, type A aortic dissection.

surgical approach seems to be superior to limited repair (29). However, there is scarce data on the long-term outcomes of such extensive repair for TAAD in such patients (19,22,30).

The current cohort expanded the special subset of MFS patients in our experience (17,22,31), who, following a prior Bentall procedure, developed TAAD involving the arch, which was repaired in a reoperative setting with the extensive FET technique. Compared to other MFS patients in our experience (22), the proximal procedure (Bentall) had already been performed in this cohort, which avoids longer

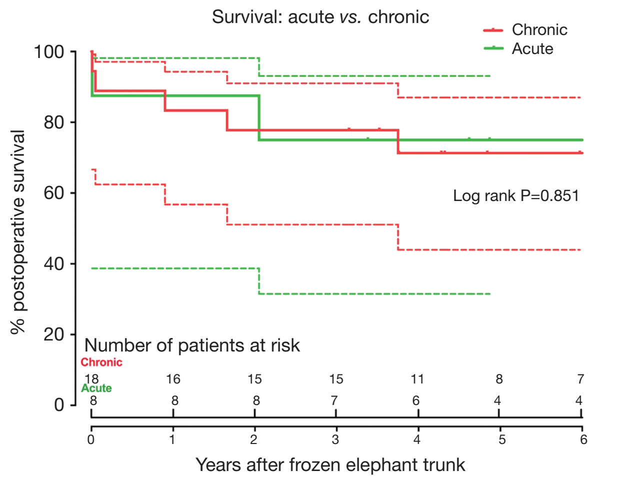

Figure 5 Survival in patients with acute and chronic type A aortic dissection.

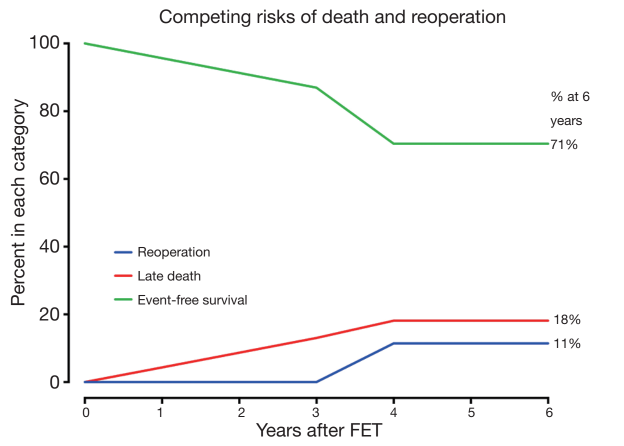

Figure 7 Competing risks of death and reoperation.

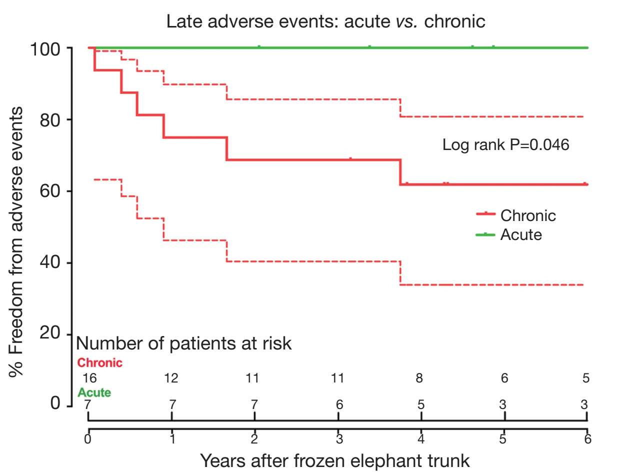

Figure 6 Freedom from distal aortic reoperation and rupture was significantly higher in patients with acute type A aortic dissection.

CPB time, a risk factor for operative mortality (32,33). In this series, operative mortality was 11.5%; at 6 years late death occurred in 18%, 11% underwent a distal aortic reoperation, and 71% of patients were alive without reoperation. These early and late outcomes are comparable or superior to most series of arch reoperations (1,15,34). In the report of Bachet and associates (15), 35 patients underwent 45 reoperations and 7 died before discharge (13%). Girdauskas and colleagues (1) reported a cohort of 15 Marfan patients undergoing reintervention after root surgery and the mortality rate was 6.7%. However, long-term data is lacking in both reports. These results of the current study show that the FET technique is a feasible and efficacious approach to TAAD after a prior Bentall procedure in Marfan patients in a reoperative setting.

The incidence of reoperations is significantly higher in

Table 4 Risk factors for distal aortic dilatation and late adverse events

Endpoint/risk factor	Hazard ratio	95% confidence interval	P value
Distal aortic dilatation			
Hypertension	7.271	1.814–29.143	0.005
Late adverse events (aortic rupture and reoperation)			
Hypertension	6.712	1.201–37.503	0.030
Age <35 years	6.760	1.154–39.587	0.034

patients who presented initially with acute TAAD than in those with dilatation only (1,34,35). In the present study, the entry tear was not resected during previous surgery in 15 of 26 patients, which exposed them to the risk of chronic dissection with residual tears, aggravating dilation of distal aortic segments (36). MFS patients with chronic dissection tend to have a very narrowed true lumen, which is compressed by an enlarged false lumen as a result of residual dissections untreated in the prior Bentall operation. And many of them have a thoracoabdominal aortic aneurysm, as evidenced by the significantly larger maximal distal aortic sizes before surgery. These chronic dissection patients may be at the more severe end of the disease spectrum (22). Following the FET, the rates of false lumen obliteration were significantly lower across the FET, quick dilation of downstream aorta, re-dissection and rupture and the need for distal reoperation are not unexpected in these patients. This largely accounts for the lower freedom from late rupture and distal reoperation in the chronic group as compared to the patients with acute TAAD.

When should the aortic arch be replaced in Marfan patients and whether TAR should be performed during initial surgery are controversial (1,7,8,11,15). Few reports have addressed the issue of the extent of the aortic replacement (15,16,35). In the present study, the significantly longer duration between the two procedures in acute TAAD group proved the durability of the Bentall operation for patients with root aneurysm. Although it is not clear how and when a new dissection would happen to effect on primarily nontreated aortic segments, prophylactic arch replacement is not necessary during initial root replacement (16). In this study, favorable long-term outcomes have been achieved using TAR + FET for the root aneurysm patients, with no late rupture and reoperation. To decrease the mortality risk of reoperation, we recommend that secondary TAR + FET be performed for root aneurysm patients when the maximal size of the aortic arch and/or proximal DA reached 4.0 cm or when the growth rate exceeds 5 mm/year.

Aortic dissection during the initial root procedure has been identified as a significant risk factor for distal aortic reoperations (1,12). Bachet *et al.* (15) reported that 73% of secondary TAR and 53% of TAAAR were required after Bentall procedure in patients with initial TAAD, and therefore proposed more aggressive approach toward the aortic arch during initial surgery. However, it remains unclear how long the interval is safe enough between the root and arch surgery to prevent rupture of the distal aneurysm. In some patients with residual dissection and significantly dilated arch and thoracoabdominal aorta, the choice of surgical access would be another dilemma. Based on our experience (17,22,31) and that of others (1,16,34,35), we recommended TAR + FET be performed during initial surgery if dissection involves the aortic arch in MFS patients. This approach may at least eliminate the need for an additional operation on the arch and proximal DA via a repeat median sternotomy in some patients. In addition, the Cronus FET has an extra centimeter of Dacron sewing cuff at the distal end, which facilitates manipulations during second-stage repair or reoperations should this be required (37).

The present study identified hypertension as the sole risk factor for distal aortic dilatation and one of two predictors of late adverse events. Therefore, antihypertensive therapy and close monitoring of the entire aorta with CT are essential for the optimal management of all patients with MFS (38). This is of particular importance to those who underwent a Bentall procedure for initial TAAD with unresected intimal tears in the arch and DA.

Study limitations

The limitations of this study include a small sample size and the lack of a control group. Additionally, this is a retrospective analysis of a single-center experience with a mixture of different acuity and etiology conditions. The patients are extremely young even in view of the fact that they are Marfan, and therefore, an average follow-up period of 5.1 years is not long enough. Due to the large case volume at our center, the recommendations we made may not be directly transferable to other centers.

Conclusions

The TAR and FET technique was feasible and efficacious for TAAD following previous Bentall procedure in patients with MFS. Early and late survival did not differ with acute and chronic dissections, while freedom from late rupture and reoperation is significantly higher in patients with acute TAAD. Patients with hypertension and aged <35 years are at higher risk for late distal aortic dilation, reoperation and death.

Acknowledgements

Funding: This study was supported by the Special Research Fund for Public Health and Welfare (No. 201402009).

Footnote

Conflicts of Interest: Abstract of this work was presented at the American Association for Thoracic Surgery Aortic Symposium 2018, New York, NY, USA, April 26–27, 2018.

Ethical Statement: This study was approved by the Ethic Committees of Beijing Anzhen Hospital of Capital Medical University and Fu Wai Hospital Chinese Academy of Medical Sciences (No. 2013013X).

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Cite this article as: Chen Y, Ma WG, Zheng J, Liu YM, Zhu JM, Sun LZ. Total arch replacement and frozen elephant trunk for type A aortic dissection after Bentall procedure in Marfan syndrome. J Thorac Dis 2018;10(4):2377-2387. doi: 10.21037/jtd.2018.03.79