Durability of thoracoabdominal aortic aneurysm repair in patients with connective tissue disorders

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Objective: **Thoracoabdominal aortic aneurysm (TAAA) repair is a durable procedure performed with reasonable perioperative mortality and morbidity in patients with atherosclerotic aortic disease. However, the long-term outcome and durability of TAAA repair performed in patients with a connective tissue disorder (CTD) is not well known.**

Methods: **The records of 257 patients who underwent TAAA repair at the Johns Hopkins Hospital between January 1992 and December 2001 were reviewed. Survival analysis was performed with Kaplan-Meier analysis, and subgroups were compared with the log-rank test. Multivariable analysis was performed with the Cox proportional hazards model and logistic regression.**

Results: **Patients with CTD (n = 31) were seen earlier (mean age, 48.6 ± 2.9 years) than patients without CTD (mean age, 69.1 ± 0.6 years; $P < .0001$, Mann-Whitney *U* test) and had a greater incidence rate of aortic dissection (52% versus 19%; $P < .0001$, χ^2 test) and extent I or II aneurysm (77% versus 64%; $P = .04$). The perioperative (30-day) mortality rate was 6.5% in patients with CTD, which was similar to the rest of the cohort ($P = .39$, Fisher exact test). The incidence rate of paraparesis/paraplegia was 12.9%/6.5% in patients with CTD, and CTD was the only factor predictive of paraparesis ($P = .03$; odds ratio, 9.3; logistic regression). The cumulative survival rate among the entire cohort was 53.4% ± 4.4% at 5 years (Kaplan-Meier), and no difference was seen among patients with or without CTD ($P = .16$, log-rank test) or among different Crawford extents ($P = .29$). Of the two late (>6 months) deaths in patients with CTD, none were from aortic rupture or dissection, compared with two of 31 late deaths in patients without CTD. Multivariable analysis confirmed that postoperative renal failure ($P = .03$) predicted mortality but neither CTD ($P = .93$), nor Crawford extent ($P = .21$, Cox regression) predicted mortality. Among survivors, no mean difference was found in largest aortic diameter on follow-up imaging in patients with or without CTD (4.7 ± 0.3 cm versus 4.4 ± 0.3 cm; $P = .47$, Mann-Whitney *U* test). The cumulative graft patency rate, representing long-term graft stability and with death, rupture, dissection, or recurrent aneurysm as endpoints, was 47.5% ± 4.6% at 5 years (Kaplan-Meier) and was similar in patients with or without CTD ($P = .10$, log-rank test).**

Conclusion: **TAAA repair appears to be a durable operation, with a reasonable 5-year patient survival rate and a low risk of postoperative paraplegia or additional aortic events. Patients with CTD can expect their outcome, including long-term survival and aortic stability, to be similar to patients without CTD. (J Vasc Surg 2002;36:696-703.)**

Thoracoabdominal aortic aneurysm (TAAA) repair is a durable procedure performed with acceptable perioperative mortality and morbidity in patients with atherosclerotic aortic disease.[1] The long-term outcome of TAAA repair likewise is good, with approximately 50% to 60% cumulative 5-year survival rates in several series.[2-7]

The connective tissue disorders (CTDs) are a diverse group of syndromes with defects in connective tissue structural integrity; vascular wall weakness is common to all these syndromes. The most common CTD is Marfan syndrome, an autosomal dominant disorder characterized by defective fibrillin, a structural component of microfibrils, and manifested clinically by characteristic skeletal, ocular, and cardiovascular defects.[8-12] Mortality in untreated patients with CTD such as Marfan syndrome is usually the result of cardiovascular disease, most commonly rupture of the aorta.[13-15] Replacement of a dilated aortic annulus and ascending aorta has been shown to significantly prolong the life expectancy of patients with Marfan syndrome.[12,16-20] In addition, the need for ascending aortic replacement in patients with Marfan syndrome predicts the development of a large TAAA necessitating repair.[17,19-21]

However, the long-term outcome and durability of TAAA repair performed in patients with CTD is not well known. This study reports the outcome after TAAA repair in patients with CTD compared with those patients without CTD.

METHODS

The records of all patients who underwent repair of a TAAA at the Johns Hopkins Hospital between January 1992 and December 2001 were reviewed retrospectively. Patients without an aneurysm but with a similar thoracoabdominal operative procedure, such as those patients with aortic dissection, and patients with purely thoracic aneurysms were excluded from this analysis.

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Table I. Demographics of patients who underwent TAAA repair at Johns Hopkins Hospital from 1992 to 2001

	Whole group	*CTD*	*No CTD*	P *value*
No.	257	31 (12%)	226 (88%)	
Age (mean ± SEM; y)	66.6 ± 0.8	48.6 ± 2.9	69.1 ± 0.6	<.0001
Male	158 (61%)	22 (71%)	136 (60%)	.25
White	210 (82%)	29 (94%)	181 (80%)	.08
Smoking history	193 (75%)	18 (58%)	175 (77%)	.02
Hypertension	227 (88%)	27 (87%)	200 (88%)	.77
Diabetes	18 (7%)	2 (6%)	16 (7%)	.99
Coronary artery disease	119 (46%)	12 (39%)	107 (47%)	.37
Peripheral vascular disease	87 (34%)	12 (39%)	75 (33%)	.58
COPD	57 (22%)	3 (10%)	54 (24%)	.10
Preoperative renal insufficiency				
Chronic renal insufficiency (1.8 ≤ Cr ≤ 2.4)	20 (8%)	1 (3%)	19 (8%)	.70
Chronic renal failure (Cr ≥ 2.5)	10 (4%)	1 (3%)	9 (4%)	.99
CTD	31 (12%)	31	0	
Marfan syndrome	28 (90%)	28 (90%)	0	
Ehlers-Danlos syndrome	3 (10%)	3 (10%)	0	
Crawford extent				
I	91 (35%)	8 (26%)	83 (37%)	
II	77 (30%)	16 (52%)	61 (27%)	
III	71 (28%)	5 (16%)	66 (29%)	
IV	18 (7%)	2 (6%)	16 (7%)	
Previous aortic dissection	60 (23%)	16 (52%)	44 (19%)	<.0001
Emergent presentation/rupture	17 (7%)	2 (6%)	15 (7%)	.99

SEM, Standard error of mean; *COPD,* chronic obstructive pulmonary disease.

Patients with CTD were previously known and identified either with genetic testing for Marfan (n = 22) or Ehlers-Danlos (n = 3) syndromes or with a strong family history of Marfan syndrome and a Marfanoid appearance (n = 6). Routine preoperative screening for CTD was not performed.

Patients were considered before surgery to have renal insufficiency if the serum creatinine level (Cr) was 1.8 or more and 2.4 or less, and renal failure if the Cr was 2.5 or more. Patients were considered to have postoperative renal failure if the Cr either doubled from its baseline value or was 3.0 or more at discharge from the hospital.[5]

TAAA repair was performed with distal aortic perfusion and sequential aortic clamping.[1] Spinal arteries identified on preoperative arteriography were routinely reimplanted intraoperatively. Lumbar drains were routinely placed before elective surgery and cerebrospinal fluid (CSF) drainage began in the operating room and continued for 3 days after surgery.[22] Spinal cooling or instillation of pharmacologic agents was not performed in any patient. Patients with TAAA and concomitant aortic dissection had adjunctive aortic fenestration or tailoring procedures performed only in small sections of the aortas not needing graft repair of an aneurysm.[23]

Routine follow-up imaging for patients after TAAA repair included yearly computed tomographic (CT) scans with three-dimensional reconstruction. Development of additional aneurysms in areas of the aorta that were not initially repaired prompted close observation and possibly surgical repair, on the basis of standard criteria.[1,2]

Comparison between groups was performed with the Mann-Whitney *U* test for nonparametric variables and the χ^2 test or Fisher exact test for categorical variables. Survival analysis was performed with Kaplan-Meier analysis, and subgroups were compared with the log-rank or Breslow-Gehan-Wilcoxon tests. Freedom from additional aortic events was calculated as cumulative graft patency with Kaplan-Meier analysis and with death, aortic or graft rupture, aortic dissection, or recurrent aneurysm as endpoints. Multivariable analysis was performed with the Cox proportional hazards model and logistic regression. *P* values were considered significant at .05 or less.

RESULTS

During this study, 257 patients underwent TAAA repair at the Johns Hopkins Hospital. Of these 257 patients, 158 were men and 99 were women; demographics are presented in Table I. The mean size of the thoracic aorta was 6.7 ± 0.1 cm, and the mean size of the abdominal aorta was 5.5 ± 0.2 cm. Twenty-two patients (9%) had previous thoracic segment repairs, and 53 (21%) had prior abdominal repairs.

Patients with CTD (n = 31) most commonly had Marfan syndrome (Table I). Fourteen patients (45%) had prior surgical repair of the ascending aorta, including eight (26%) with elephant trunk procedures.[24] These patients were seen earlier (mean age, 48.6 ± 2.9 years) than patients without CTD (mean age, 69.1 ± 0.6 years; $P < .0001$, Mann-Whitney *U* test) and had a greater incidence rate of aortic dissection (52% versus 19%; $P < .0001$, χ^2 test). Eight extent I (26%), 16 extent II (52%), five extent III (16%), and two (6%) extent IV aneurysms were seen, which was a more heavily weighted distribution of extent I and II aneurysms (77% versus 64%) than in the rest of the study

Table II. Comparison of operative and postoperative parameters and perioperative complications between patients with and without CTD

	CTD	*No CTD*	P *value*
No. of Crawford patches	2.1 ± 0.2	1.7 ± 0.1	.10
No. of bypass grafts	0.7 ± 0.2	0.4 ± 0.1	.01
Intercostal revascularization	15 (48%)	90 (40%)	.44
Estimated blood loss (L)	18 ± 2	11 ± 1	.01
PRBC transfusion (U)	18 ± 3	14 ± 1	.16
Cell-saver transfusion (L)	5 ± 1	3 ± 0.2	.003
FFP transfusion (U)	23 ± 4	18 ± 1	.37
Platelet transfusion (U)	10 ± 2	11 ± 1	.19
ICU length of stay (d)	6.4 ± 0.8	10.1 ± 0.8	.22
Intermediate care length of stay (d)	2.0 ± 0.3	4.5 ± 0.5	.12
Hospital length of stay (d)	14 ± 1	23 ± 2	.22
Paraparesis	4 (12.9%)	22 (9.7%)	.58
Permanent paraplegia	2 (6.5%)	12 (5.3%)	.68
Myocardial infarction	2 (6%)	17 (8%)	.99
Renal failure	4 (13%)	31 (14%)	.99
Pulmonary embolism	0	10 (4%)	.61
Stroke	0	4 (2%)	.99
Vocal cord paralysis	1 (3%)	21 (10%)	.49
Prolonged ventilation	3 (10%)	45 (20%)	.22
Tracheostomy	1 (3%)	35 (15%)	.09
Subdural hematoma	2 (6%)	6 (3%)	.25

PRBC, Packed red blood cells; *FFP,* fresh frozen plasma; *ICU,* intensive care unit.

Table III. Multivariable analysis of risk factors that predict paraparesis after TAAA repair (logistic regression)

Factor	P *value*	*Odds ratio*	*95% CI*
Age	.70	1.01	0.96-1.07
Race	.38	2.22	0.37-13.47
Gender	.91	0.94	0.27-3.22
Hypertension	.99	1.00	0.01-99.99
COPD	.23	2.30	0.59-8.96
CTD	.03	9.32	1.24-70.03
Previous aortic dissection	.81	1.20	0.29-5.03
Crawford extent (I, II)	.23	2.27	0.59-8.75
Preoperative ruptured aneurysm	.84	1.29	0.11-15.42
Preoperative renal insufficiency	.39	2.37	0.33-17.07
Preoperative renal failure	.30	4.17	0.28-62.58
Preoperative stroke	.27	2.42	0.50-11.63
Spinal artery identified before surgery	.73	0.79	0.21-3.02
Intercostal artery revascularized	.15	2.44	0.72-8.29
Lumbar drain	.008	0.16	0.04-0.62
Postoperative renal failure	.60	0.65	0.13-3.22

COPD, Chronic obstructive pulmonary disease.

group ($P = .04$, χ^2 test). No difference was found in mean aneurysm size in patients with CTD compared with patients without CTD (thoracic size, 6.7 ± 0.3 cm versus 6.7 ± 0.1 cm; $P = .65$, Mann-Whitney *U* test). Patients with CTD had a similar rate of ruptured TAAA as patients without CTD (6.5% versus 6.6%; $P = .99$, Fisher exact test).

Among the entire patient group, preoperative spinal arteriography was successfully performed in 191 patients (74%), and a spinal artery was identified in 86 of these cases (45%). Twenty of 31 patients (65%) with CTD successfully underwent arteriography, which was statistically similar to the rate of arteriography in patients without CTD ($P = .38$, χ^2 test). Spinal artery identification was successful in seven of these 20 patients (35%), which was not statistically different than the rate of success in patients without CTD ($P = .25$, χ^2 test).

Lumbar drains were used in 168 patients (65%), including 22 of the 31 patients with CTD (71%), which was not different than the usage in patients without CTD ($P = .48$, χ^2 test). Drains were usually removed on postoperative day 3 (mean, 2.4 ± 0.1 days) and removed a mean of 367 ± 22 mL of CSF. No difference was seen in the amount of CSF removed in patients with (408 ± 82 mL) or without (361 ± 23 mL) CTD ($P = .98$, Mann-Whitney *U* test).

Aortic fenestration was used as an adjunctive procedure in 13 patients, 10 without CTD (4%) and three with CTD (10%; $P = .20$, Fisher exact Test). Aortic tailoring was also performed in 17 patients, more frequently in patients with CTD (n = 5; 16%) than in patients without CTD (n = 12; 5%; $P = .02$, χ^2 test).

Operative results. The mean estimated blood loss for all cases was 12 ± 1 L, with intraoperative transfusion of 14 ± 1 units of packed red blood cells, 3 ± 0.2 L cell saver, 19 ± 1 units of fresh frozen plasma, and 11 ± 1 units of platelets. The mean length of stay in the intensive care unit was 9.7 ± 0.7 days, and 4.0 ± 0.4 days were spent in the intermediate care unit. The total hospital length of stay was 22 ± 2 days. Patients with CTD had greater intraoperative blood loss than patients without CTD (Table II).

Among the entire cohort of 257 patients, four intraoperative deaths (1.6%) and 32 deaths (12.5%) occurred within 30 days of operation. No intraoperative deaths and only two (6.5%) of the perioperative deaths occurred in patients with CTD ($P = .39$, Fisher exact test). In patients with a preoperatively ruptured aneurysm, six deaths (35%) occurred within 30 days, for a rate greater than the 30-day mortality rate for elective repairs (11%; $P = .003$, χ^2 test).

Among the entire cohort, 19 perioperative myocardial infarctions, four perioperative strokes, and 10 pulmonary emboli were seen. Twenty-two patients had vocal cord paralysis, 48 patients had delayed (>1 week) ventilator weaning, with 36 needing tracheostomy, and eight patients had subdural hematomas. Thirty-five patients (14%) had postoperative renal failure, including nine patients with preoperative renal insufficiency and six patients with preoperative renal failure. No difference was seen in the rate of these complications among patients with and without CTD (Table II).

Among all 257 patients, neuromuscular deficits (any paraparesis) occurred in 31 patients (12.1%). Although the incidence rate was higher in patients with CTD (12.9%) compared with those without CTD (9.7%), this increase was not statistically significant with univariate analysis ($P = .58$, χ^2 test). Permanent paraplegia occurred in only 14

patients among the entire group (5.4%) and also did not vary with or without the presence of CTD (6.5% versus 5.3%; $P = .68$, Fisher exact test). However, multiple logistic regression revealed that the presence of CTD was the only factor predictive of any paraparesis ($P = .03$; odds ratio, 9.3), whereas the usage of a lumbar drain was the only factor predictive of decreased incidence of paraparesis ($P = .008$; odds ratio, 6.1 fold decrease; Table III). Similar analysis of factors predictive of permanent paraplegia was too small to achieve statistical significance (data not shown).

Long-term results. The mean length of follow-up was 27 months (range, 0 to 106 months), and follow-up was complete in 244 patients (95%). The cumulative survival rate was 53.4% ± 4.4% at 5 years (Kaplan-Meier; Fig 1, *A*), and no difference was seen among patients with or without CTD ($P = .16$, log-rank test; Fig 1, *B*) or among different Crawford extents ($P = .29$, log-rank test; Fig 1, *C*). Patients with either an extent I or II aneurysm had similar 5-year cumulative survival rates compared with patients with an extent III or IV aneurysm (51.5% ± 5.1% versus 58.2% ± 8.5%; $P = .50$, log-rank test). Postoperative renal failure was associated with a decreased 5-year survival rate (39.8% ± 17.2% versus 55.4% ± 4.6%; $P = .004$, log-rank test; Fig 1, *D*). Presence of a preoperatively ruptured TAAA diminished early (30 days) (58.8% ± 11.9% versus 86.0% ± 2.3%; $P = .03$, Breslow-Gehan-Wilcoxon test) but not late (5 years) (50.4% ± 12.9% versus 53.3% ± 4.7%; $P = .34$, log-rank test) survival rates (Fig 1, *E*).

Of the two late (>6 months) deaths in patients with CTD, none were from aortic aneurysm rupture or dissection, compared with two of 31 late deaths in patients without CTD; one death in a patient with CTD was from rupture of a small unrepaired iliac aneurysm. Multivariable analysis confirmed that age ($P = .04$) and postoperative renal failure ($P = .03$) predicted late outcome, but neither preoperative rupture ($P = .17$), CTD ($P = .93$), nor Crawford classification ($P = .21$) predicted late mortality (Table IV).

Among late survivors, no mean difference was seen in largest aortic diameter in patients with or without CTD (4.7 ± 0.3 cm versus 4.4 ± 0.3 cm; $P = .47$, Mann-Whitney *U* test). Also, no difference was seen in the mean size of the unrepaired proximal thoracic segment in patients who originally had an extent III or IV TAAA (4.8 ± 1.3 cm with CTD versus 5.0 ± 0.4 cm without CTD; $P = .75$). Long-term freedom from recurrent aortic events was calculated with death, aortic rupture, aortic dissection, or recurrent aneurysm as endpoints and was 47.5% ± 4.6% at 5 years (Kaplan-Meier; Fig 2, *A*). This long-term graft stability was similar in patients with or without CTD ($P = .10$, log-rank test; Fig 2, *B*).

Table IV. Multivariable analysis of risk factors that predict late survival after TAAA repair (Cox regression)

Factor	P *value*	*Risk ratio*	*95% CI*
Age	.04	1.03	1.00-1.07
Gender	.24	0.69	0.37-1.29
Race	.47	0.70	0.27-1.83
Hypertension	.43	1.48	0.56-3.92
Coronary artery disease	.34	0.74	0.40-1.37
Peripheral vascular disease	.83	1.08	0.55-2.12
COPD	.28	0.70	0.36-1.35
CTD	.93	0.94	0.25-3.59
Crawford extent (I, II)	.21	0.64	0.32-1.28
Preoperative ruptured aneurysm	.17	0.50	0.18-1.35
Preoperative renal insufficiency	.24	0.54	0.19-1.53
Preoperative renal failure	.44	1.85	0.39-8.75
Lumbar drain	.81	1.09	0.55-2.19
Postoperative renal failure	.03	0.40	0.17-0.90
Paraparesis	.58	1.58	0.32-7.84
Permanent paraplegia	.11	0.23	0.04-1.38

COPD, Chronic obstructive pulmonary disease.

DISCUSSION

We show that patients with CTD have similar perioperative and long-term survival rates after TAAA repair compared with patients without CTD. However, patients with CTD had a greater incidence rate of extent I and II aneurysms and a nine-fold increased risk of paraparesis. Use of a lumbar drain was six-fold protective against the occurrence of paraparesis. The early survival rate was diminished in patients with a ruptured TAAA, and the late survival rate was diminished in patients with postoperative renal failure. Finally, long-term CT scan surveillance showed no relative increase in aortic size, and persistent freedom from recurrent aortic events, in patients with CTD.

The long-term results of TAAA repair continue to reaffirm the utility of surgical repair, and the Crawford technique in particular.[25] In our series, the cumulative 5-year survival rate was 53% and no difference was seen among patients with the more extensive Crawford extent I and II aneurysms. Our inability to detect a difference in survival among patients with greater aneurysm extent is similar to the observations of some series[5] but not others.[2,7,26] Our small sample size, and especially our relatively few number of extent IV aneurysms, with their superior survival rate[4] may be insufficiently powered to detect a difference from aneurysm extent. However, we were able to detect a significant decrease in perioperative survival rate among patients with a ruptured TAAA (Fig 1, *E*) or with postoperative renal failure (Fig 1, *D*), as noted by others.[2,7,26] Our data also suggest that the long-term survival rate after TAAA repair performed for aneurysm rupture is similar to that of the rest of the electively performed cohort among survivors beyond the perioperative period, similar to that suggested by some series examining survivors of ruptured infrarenal aortic aneurysms.[27-29]

Patients with CTD had similar long-term survival rates after TAAA repair in this series compared with patients without CTD (Fig 1, *B*). Although this finding may have been anticipated in this group of younger patients with fewer associated comorbid diseases, we believe that this information is unexpected. Life expectancy in patients with

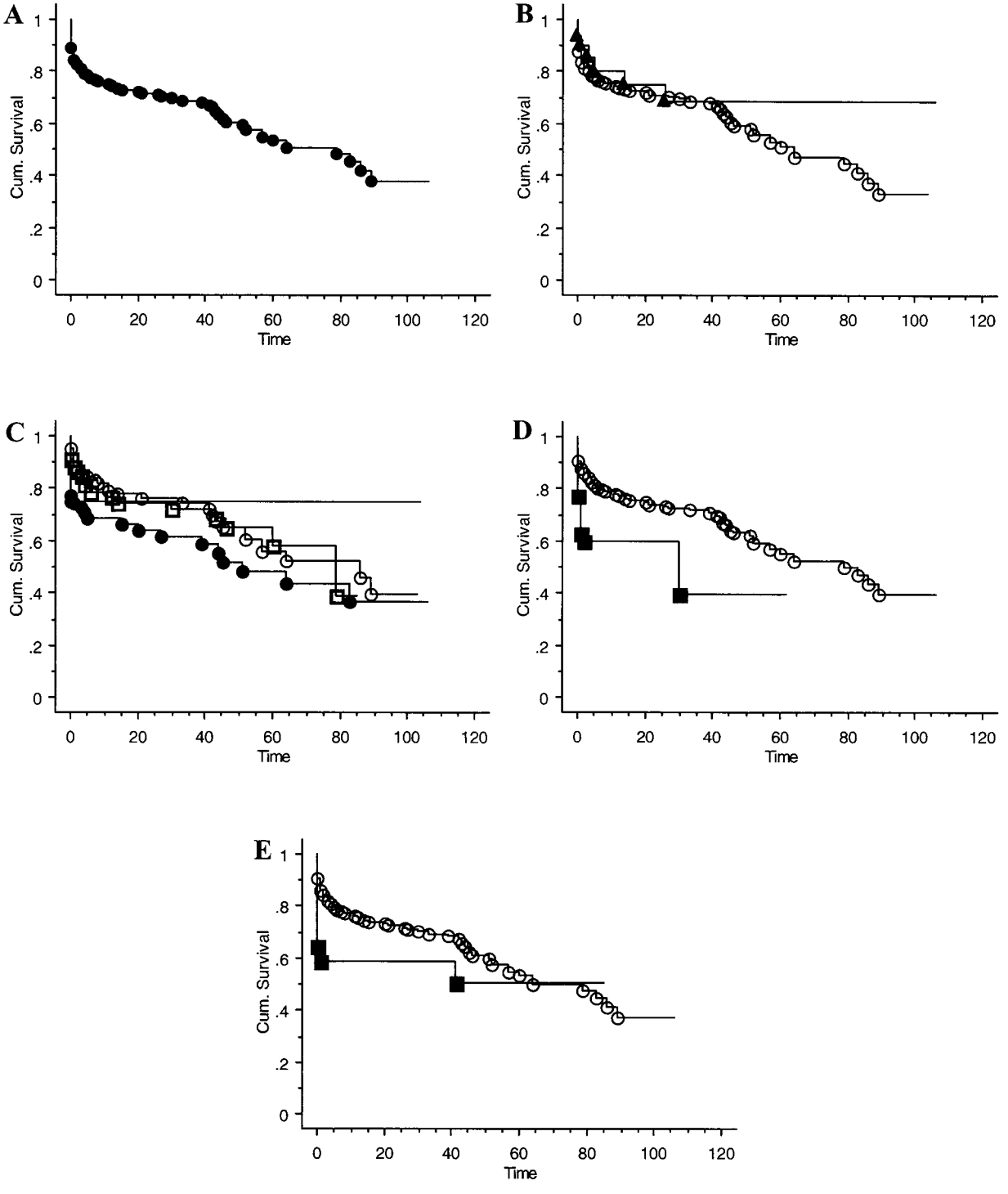

Fig 1. A, Kaplan-Meier plot of cumulative survival rate of 257 patients followed after TAAA repair. Time is in months. **B,** Kaplan-Meier plots of cumulative survival rate after TAAA repair in patients with *(▲)* and without *(○)* CTD. **C,** Kaplan-Meier plots of cumulative survival rate after TAAA repair stratified by Crawford extent (I, ○; II, ●; III, □; IV, −). **D,** Kaplan-Meier plots of cumulative survival rate after TAAA repair in patients with *(■)* and without *(○)* postoperative renal failure. **E,** Kaplan-Meier plots of cumulative survival rate after TAAA repair in patients with *(■)* and without *(○)* preoperative ruptured aneurysm.

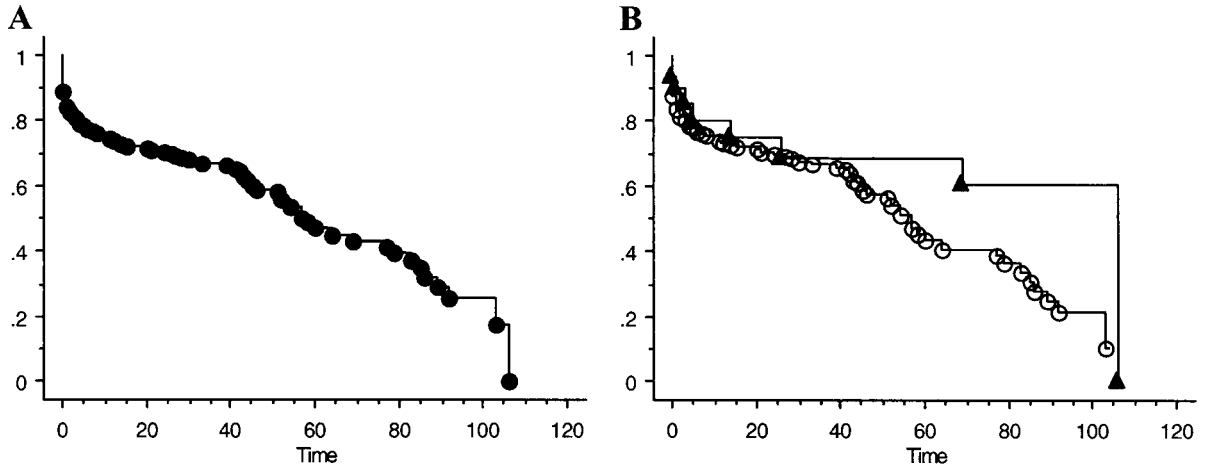

Fig 2. A, Kaplan-Meier plot of cumulative graft freedom from additional degeneration in 257 patients followed after TAAA repair. Time is in months. **B,** Kaplan-Meier plots of cumulative graft freedom from additional degeneration after TAAA repair in patients with *(▲)* and without *(○)* CTD.

CTD is reduced compared with the population.[13] Although several series have reported excellent survival rates in patients with CTD after aortic repair, it is unclear with what this rate is compared.[17,19,20] In addition, these series included cardiac and ascending aortic repairs that may result in improved outcome. We note, however, that the small number of patients with CTD, especially in longer term follow-up, may not allow us to exclude a type II error (ie, that patients with CTD have reduced survival rates after TAAA repair).

Furthermore, our finding that patients with CTD have long-term aortic stability after repair, as reflected by lack of additional aortic dilation, aneurysm formation, dissection, or rupture (Fig 2, *B*), was not anticipated; leaving behind any area of unrepaired aorta places the patient at continued risk of aneurysmal expansion and rupture.[30] This observation explains our higher frequency of placement of a bypass graft to the left renal artery in patients with CTD, minimizing visceral patch size (Table II).[30] Our high percentage of extent I and II aneurysms, however, suggests that our patients have their entire aorta, previously at risk, repaired and thus are unlikely to have a subsequent event. In addition, it is unclear whether or not patients with progression to aneurysmal stage have a different subsequent event rate compared with patients with chronic dissection alone; patients with aortic tailoring or fenestration procedures for dissection alone, without aneurysm, were excluded from this report.

A previous report from our institution suggested that 35% of patients with Marfan syndrome had evidence on magnetic resonance imaging of progressive dilation of the thoracoabdominal aorta within a mean of 5 years after composite repair of the aortic root.[31] This evidence of aortic instability in a large percentage of patients with CTD underscores the need to image the thoracoabdominal aorta and potentially repair it; lack of additional events after TAAA repair, as reported here, confirms the efficacy of the repair. On the other hand, our sample size may be too small to detect the 35% of long-term survivors who do have progressive aortic dilation. In addition, our follow-up with CT scan may not be as sensitive for events as is magnetic resonance imaging. However, we believe that the previously reported large incidence rate of thoracoabdominal aortic dilation is present only in patients with only the ascending aorta replaced and, therefore, at an earlier stage of the cardiovascular disease.[17,19-20] Because after repair of an extent I or II aneurysm all native proximal thoracoabdominal aorta other than the visceral patches is obliterated, no aorta is left that can undergo progressive dilation (Fig 2, *A*).

In our series, paraparesis and permanent paraplegia occurred in 12.9% and 6.5%, respectively, of patients with CTD, a rate similar to that previously reported.[2,17] Our inclusion of patients with CTD other than Marfan syndrome may possibly slightly elevate our neurologic event rate above other reports,[32] although the differences in these small groups are unlikely to be significant. However, patients with CTD were more likely to have paraparesis than patients without CTD (Table III). This increase may be due to the increased prevalence of extent I and II aneurysms in patients with CTD.[2,7,32-35] Also unknown is whether some of these patients have their spinal arteries obliterated with dissection as a consequence of the operative procedure, but the significance of this possibility is unclear. Nonetheless, the protective effect of lumbar drainage[36-39] was reaffirmed even in this high-risk group of patients (Table III). This suggests that paraparesis in patients with CTD is likely to be the result of a multifactorial process, reflecting a relative balance of factors maintaining spinal cord perfusion,[40] rather than simple extent of the aneurysm and interruption of a single spinal artery.[7,35]

The durability of TAAA repair in patients with CTD may reflect surgical replacement of defective structural elements, such as fibrillin or collagen, limited to the aorta.[12,23] Whether a different distribution of fibrillin exists in the aortic branches compared with the aorta and whether this may be the case only in subsets of patients are not known. Continued postprocedural surveillance is, therefore, vital to detect potential events in the extreme (>10 year) long-term follow-up.

We reaffirm that outcome after TAAA repair is durable, with reasonable cumulative 5-year survival, low risk of postoperative paraplegia, and long-term aortic stability expected for most patients. Patients with CTD can expect their outcome after TAAA repair, including long-term survival and aortic stability, to be similar to patients without CTD who undergo TAAA repair. We advocate early elective TAAA repair in patients with CTD because of the safety, efficacy, and durability of the procedure. Early elective repair also allows placement of a lumbar CSF drain that helps protect against the higher incidence of paraparesis in these patients.

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