

Appendix 1 Supplemental methods

Surgical techniques

All The median sternotomy and cardiopulmonary bypass (CPB) were consistently performed on all patients. During the cooling period following CPB, aortic root procedures were performed using individualized strategies determined by the severity of pathological changes. When root involvement was minimal (partial cusp damage without dilatation or significant aortic regurgitation), we employed direct running suture reinforcement at the sinotubular junction without using felt or bioglu. In cases of extensive root pathology (root diameter exceeding 45 mm, presence of Marfan syndrome, root tear, or substantial aortic root destruction), complete root replacement was the preferred intervention. For intermediate scenarios falling between minimal and extensive involvement, various root preservation techniques were utilized, including the adventitial inversion method, selective replacement of affected aortic sinuses, and valve resuspension procedures. The adventitial inversion technique involved trimming the intimal layer approximately 1.0 cm below the adventitial margin. The excess adventitial tissue was subsequently folded inward into the aortic cavity and reattached to the intima using continuous 5-0 prolene sutures. This technique established a reconstructed aortic wall with a tri-layered "outer-inner-outer" configuration, providing a suitable foundation for Dacron graft anastomosis. It should be emphasized that the selection of specific surgical approaches was primarily influenced by individual surgeon expertise and clinical judgment.

From 2010 to 2018, right axillary artery cannulation and unilateral antegrade cerebral perfusion, combined with hypothermia circulatory arrest, were the preferred circulatory management approach for patients at our institution. Cerebral perfusion commenced upon reaching the target core temperature (18 °C during the first five years and 25 °C over the following years), maintaining a flow rate of 5-10 mL/kg/min. Total arch replacement with with frozen elephant trunk (TAR with FET) was then performed. The diameter of the prosthetic graft and FET (MicroPort Medical Company Limited, Shanghai, China) was determined based on preoperative measurements of the distal aortic diameter, typically ranging from 26-28 mm, with matching sizes chosen for both the graft and stent. The length of the FET (100/120/150 mm) was selected based on the patient's height and the distance from the left common carotid artery to the T8 level (considering safe length to

avoid spinal cord injury), positioning the distal anchoring zone at the T6-T7 level.

In brief, the left subclavian artery was circumferentially transected 0.5-1.0 cm distal to its origin, and the proximal arterial segment was sutured. After transecting the distal aorta between the origins of the left common carotid and left subclavian arteries, a FET stent graft was implanted into the true lumen of the descending aorta, followed by distal anastomosis with the tetrafurcate graft. Lower body perfusion was then restored through the perfusion limb of the tetrafurcate graft. The left common carotid artery, left subclavian artery, and innominate artery were sequentially reconstructed, after which rewarming began. Finally, an end-to-end anastomosis was performed between the proximal end of the artificial blood vessel and either the ascending aorta or the repaired aortic root.

Statistical analysis

Categorical variables were described as frequencies with percentages and analyzed using the Chi-square or Fisher's exact tests, as appropriate. Continuous variables were assessed for normal distribution using the Kolmogorov-Smirnov test and expressed as mean and standard deviation for normally distributed data or median and interquartile range for non-normally distributed data. Student's *t*-test was applied to normally distributed variables, while the Mann-Whitney U test was used for non-normally distributed variables. Survival rates were estimated using the Kaplan-Meier method and compared using the Log-rank test. The cumulative incidence of reoperation and incomplete self-care (ADL \leq 2) was analyzed using the Gray-Fine regression model, with death considered as a competing risk.

A total of 44 baseline and intraoperative variables were incorporated into univariable logistic, Cox, and competing risk Cox regression analyses for early mortality, long-term mortality, and reoperation, respectively. Three different regression models were constructed to identify independent risk factors for the corresponding outcomes. Model 1 included Marfan syndrome, age, and sex; Model 2 added baseline variables with $P < 0.05$ from univariable analyses to Model 1; Model 3 further incorporated intraoperative variables with $P < 0.05$ from univariable analyses to Model 2.

Statistical analysis and data visualization were performed using R version 4.2.1 and Microsoft Excel version 2402. A two-tailed P value < 0.05 was considered statistically significant.

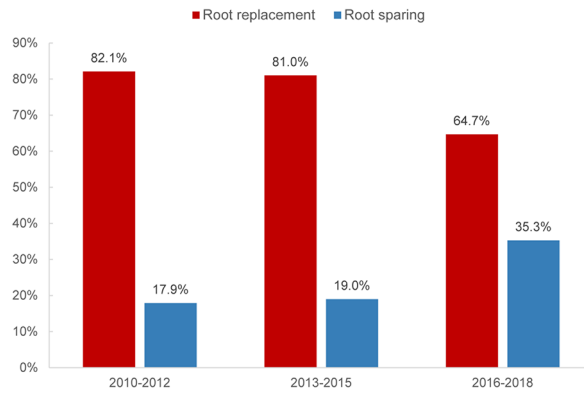


Figure S1 Histogram showing the proportion of root-sparing procedures in Marfan patients across different time periods..

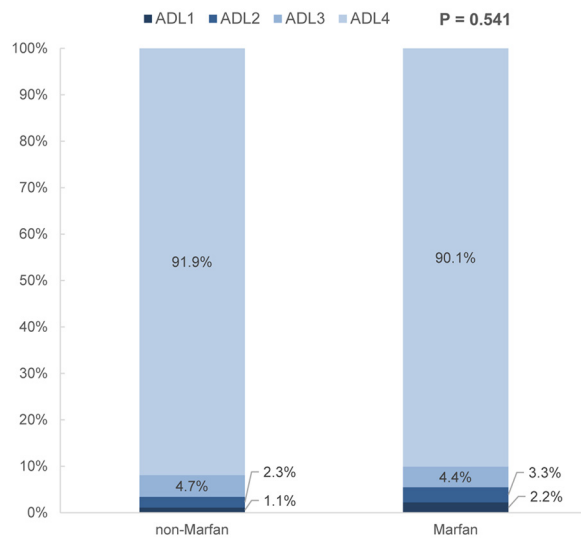


Figure S2 Histogram showing long-term activity of daily living grade based on the presence of Marfan syndrome.

Table S1 Late outcomes including causes of death, false lumen status, and reoperation in hospital survivors

Variables	Non-Marfan, N=906*	Marfan, N=100*	P value
Causes of late death			0.781
Survival	902 (91.2)	95 (91.0)	
Distal aortic rupture	8 (0.9)	1 (1.0)	
Cardiac causes	7 (0.8)	0 (0.0)	
Reoperation	4 (0.4)	1 (1.0)	
Non-cardiovascular causes	58 (6.4)	7 (7.0)	
Accident	3 (0.3)	0 (0.0)	
Aortic remodeling			
Complete false lumen thrombosis at the distal end of the stent graft	772 (85.2)	61 (61.0)	< 0.001
Causes of reoperation			< 0.001
No reoperation	862 (95.1)	75 (75.0)	
Aortic endovascular repair	22 (2.4)	2 (2.0)	
Aortic surgery	22 (2.4)	21 (21.0)	
Cardiac surgery	0 (0.0)	2 (2.0)	

Data are presented as n (%). *, Analysis restricted to hospital survivors.

Table S2 Details of reoperation in Marfan patients

Patients	Age	Sex	Primary operation*	Reoperation	Cause of reoperation	Time interval (years) [†]
1	34	Male	Ascending aorta replacement	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	2.16
2	34	Male	Ascending aorta replacement	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm + type B aortic dissection	4.28
3	42	Male	Ascending aorta replacement	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	7.57
4	23	Male	Bentall	Root pseudoaneurysm repair	Root pseudoaneurysm	1.19
5	26	Male	Bentall + aorta-femoral artery bypass	Mitral valve replacement	Severe mitral valve insufficiency	4.99
6	36	Male	Bentall	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	5.02
7	22	Male	Bentall	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm + type B aortic dissection	8.00
8	52	Male	Ascending aorta replacement	Transcatheter closure	Root anastomotic leakage	0.76
9	49	Female	Ascending aorta replacement	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	8.11
10	24	Female	Bentall	Reconstruction of the aortic arch branches	Severe stenosis of the innominate artery + occlusion of the left common carotid and left subclavian arteries	5.28
11	31	Female	Ascending aorta replacement	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	3.02
12	23	Male	Bentall	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	0.83
13	24	Male	Ascending aorta replacement	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	0.48
14	30	Male	Bentall	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm + type B aortic dissection	2.38
15	31	Male	Bentall	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	5.79
16	26	Male	Bentall	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	1.30
17	33	Male	Bentall	Mitral valve replacement	Severe mitral valve insufficiency	4.30
18	28	Female	Bentall	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	2.97
19	24	Male	Bentall	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm + type B aortic dissection	5.65
20	30	Male	Bentall	Root pseudoaneurysm repair	Root pseudoaneurysm	1.15
21	29	Male	Bentall	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	0.74
22	34	Male	Bentall	Root pseudoaneurysm repair	Root pseudoaneurysm	2.63
23	42	Male	Ascending aorta replacement	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	0.96
24	36	Female	Bentall + mitral valve replacement	Thoracoabdominal aorta replacement	Thoracoabdominal aortic aneurysm	5.24
25	29	Male	Bentall	Thoracic endovascular aortic repair	Type B aortic dissection	2.01

* Concomitant procedures in addition to total arch replacement with frozen elephant trunk. † Time interval between primary operation and reoperation.

Table S3 Univariable logistic regression analysis for operative mortality

Variables	Odds ratio	95% confidence interval	P value
Marfan syndrome	0.477	0.143-1.179	0.157
Age	1.030	1.007-1.054	0.010
Body mass index	0.988	0.935-1.038	0.665
Male sex	0.509	0.312-0.850	0.008
Time interval between onset and admission	1.012	0.940-1.080	0.737
Smoking	0.864	0.540-1.367	0.535
Alcohol addiction	1.428	0.696-2.684	0.296
Hypertension	1.013	0.594-1.819	0.963
Diabetes mellitus	0.426	0.024-2.035	0.405
Hyperlipemia	0.771	0.352-1.500	0.477
Coronary artery disease	15.878	6.481-39.382	< 0.001
NYHA \geq 3	3.763	1.575-8.992	0.003
Chronic kidney disease	4.212	1.902-8.617	< 0.001
Chronic obstructive pulmonary disease	2.110	0.111-12.560	0.492
Cerebrovascular disease	3.062	1.005-9.327	0.049
Family history of aortic disease	0.000	0-Inf	0.979
History of cardiovascular surgery	1.712	0.499-4.480	0.324
History of TEVAR	0.836	0.046-4.208	0.863
History of PCI	2.110	0.111-12.560	0.492
Aortic insufficiency	1.104	0.690-1.748	0.674
Pericardial effusion	1.873	1.048-3.201	0.027
Cardiac wall motion abnormality	2.124	0.327-7.969	0.330
Preoperative ejection fraction	0.972	0.932-1.017	0.203
Preoperative LVEDD	0.966	0.931-1.001	0.060
Preoperative white blood cell	1.062	1.010-1.115	0.017
Preoperative platelet	0.997	0.993-1.001	0.122
Preoperative hemoglobin	0.990	0.979-1.002	0.101
TEM-E*			
E1	1.178	0.458-4.007	0.761
E2	1.341	0.499-4.667	0.598
E3	2.889	0.856-11.361	0.099
TEM-M0	1.015	0.512-1.854	0.964
TEM-M1 †			
M1 without symptoms	1.815	1.010-3.261	0.046
M1 with symptoms	1.787	0.760-3.712	0.146
TEM-M2 †			
M2 without symptoms	0.918	0.566-1.512	0.731
M2 with symptoms	4.262	1.569-10.506	0.002
TEM-M3†			
M3 without symptoms	1.271	0.707-2.232	0.410
M3 with symptoms	2.336	1.355-3.986	0.002
Root Surgery [§]			
Root repair	1.400	0.705-2.613	0.310
Root replacement	1.283	0.757-2.132	0.343
Coronary artery bypass grafting	3.254	1.898-5.425	< 0.001
Aorta-femoral artery bypass	3.849	1.747-7.813	< 0.001
Operation time	1.408	1.282-1.553	< 0.001
Cardiopulmonary bypass time	1.013	1.010-1.017	< 0.001
Cross-clamp time	1.015	1.010-1.021	< 0.001
Hypothermic circulatory arrest time	1.060	1.028-1.093	< 0.001
Lowest core temperature	0.856	0.796-0.918	< 0.001
Blood loss	1.000	1.000-1.001	0.001
Red blood cell transfusion	1.050	0.957-1.139	0.262
Plasma transfusion	1.001	1.000-1.001	0.001
Platelets transfusion	1.118	0.892-1.376	0.311

*Compared with TEM-E0. †Compared with non-TEM-M1, M2, or M3. §Compared with no root surgery. NYHA, New York Heart Association; TEVAR, thoracic endovascular aortic repair; PCI, percutaneous coronary intervention; LVEDD, left ventricular end-diastolic diameter; TEM, type, entry, malperfusion.

Table S4 Detailed logistic regression analysis models for the association of Marfan syndrome with early mortality

Variables	Unadjusted		Model 1		Model 2		Model 3	
	OR (95% CI)	P value	OR (95% CI)	P value	OR (95% CI)	P value	OR (95% CI)	P value
Marfan syndrome	0.477 (0.143-1.179)	0.157	0.583 (0.200-1.702)	0.324	0.705 (0.233-2.138)	0.537	0.550 (0.141-2.146)	0.389
Age			1.020 (0.995-1.045)	0.114	1.017 (0.990-1.045)	0.220	1.030 (0.999-1.062)	0.061
Male sex			0.562 (0.334-0.946)	0.030	0.467 (0.264-0.829)	0.009	0.368 (0.195-0.697)	0.002
Coronary artery disease					12.886 (4.780-34.735)	< 0.001	13.641 (4.074-45.672)	< 0.001
NYHA 3					2.901 (1.071-7.855)	0.036	2.778 (0.859-8.984)	0.088
Chronic kidney disease					2.583 (0.975-6.841)	0.056	3.924 (1.355-11.360)	0.012
Cerebrovascular disease					2.824 (0.773-10.317)	0.116	2.961 (0.739-11.863)	0.125
Pericardial effusion					1.321 (0.705-2.477)	0.385	1.740 (0.865-3.504)	0.121
Preoperative white blood cell					1.089 (1.030-1.151)	0.003	1.077 (1.015-1.144)	0.015
TEM-M1*								
M1 without symptoms					1.875 (1.003-3.506)	0.049	0.903 (0.206-3.954)	0.892
M1 with symptoms					1.648 (0.681-3.990)	0.268	1.606 (0.581-4.438)	0.361
TEM-M2*								
M2 without symptoms					1.014 (0.596-1.726)	0.958	0.734 (0.403-1.334)	0.310
M2 with symptoms					3.760 (1.327-10.650)	0.013	4.168 (1.265-13.738)	0.019
TEM-M3*								
M3 without symptoms					1.320 (0.710-2.454)	0.380	1.090 (0.504-2.357)	0.828
M3 with symptoms					1.403 (0.697-2.823)	0.342	0.923 (0.397-2.150)	0.853
Coronary artery bypass grafting							0.732 (0.175-3.063)	0.669
Aorta-femoral artery bypass							7.716 (2.958-20.128)	< 0.001
Operation time							1.043 (0.886-1.226)	0.615
Cardiopulmonary bypass time							1.016 (1.009-1.024)	< 0.001
Cross-clamp time							0.990 (0.980-1.001)	0.062
Hypothermic circulatory arrest time							1.023 (0.980-1.067)	0.295
Lowest core temperature							0.943 (0.849-1.047)	0.270
Blood loss							1.000 (1.000-1.001)	0.127
Plasma transfusion							1.000 (1.000-1.001)	0.663

*Compared with non-TEM-M1, M2, or M3. OR, odds ratio; CI, confidence interval; NYHA, New York Heart Association; TEM, type, entry, malperfusion.

Table S5 Univariable Cox regression analysis for long-term mortality

Variables	Hazard ratio	95% confidence interval	P value
Marfan syndrome	0.705	0.400-1.242	0.226
Age	1.039	1.024-1.055	< 0.001
Body mass index	0.983	0.948-1.019	0.343
Male sex	0.674	0.478-0.951	0.025
Time interval between onset and admission	1.049	1.008-1.093	0.020
Smoking	1.182	0.873-1.599	0.280
Alcohol addiction	1.294	0.819-2.046	0.269
Hypertension	1.131	0.779-1.643	0.516
Diabetes mellitus	1.353	0.599-3.058	0.467
Hyperlipemia	1.008	0.654-1.553	0.972
Coronary artery disease	8.266	4.760-14.353	< 0.001
NYHA \geq 3	2.906	1.529-5.526	0.001
Chronic kidney disease	2.066	1.147-3.719	0.016
Chronic obstructive pulmonary disease	1.933	0.479-7.797	0.354
Cerebrovascular disease	2.867	1.408-5.836	0.004
Family history of aortic disease	0.808	0.258-2.531	0.714
History of cardiovascular surgery	2.207	1.198-4.067	0.011
History of TEVAR	1.367	0.436-4.285	0.592
History of PCI	3.284	1.047-10.299	0.041
Aortic insufficiency	1.209	0.891-1.642	0.223
Pericardial effusion	1.824	1.261-2.637	0.001
Cardiac wall motion abnormality	1.181	0.293-4.770	0.815
Preoperative ejection fraction	0.960	0.933-0.988	0.006
Preoperative LVEDD	0.985	0.962-1.009	0.211
Preoperative white blood cell	0.999	0.963-1.037	0.960
Preoperative platelet	1.000	0.997-1.002	0.885
Preoperative hemoglobin	0.990	0.983-0.998	0.013
TEM-E *			
E1	0.703	0.408-1.211	0.204
E2	0.745	0.416-1.334	0.321
E3	1.188	0.532-2.654	0.674
TEM-M0	1.131	0.754-1.699	0.552
TEM-M1 †			
M1 without symptoms	1.570	1.041-2.368	0.031
M1 with symptoms	1.941	1.182-3.189	0.009
TEM-M2 †			
M2 without symptoms	0.870	0.628-1.206	0.403
M2 with symptoms	3.734	2.002-6.965	< 0.001
TEM-M3 †			
M3 without symptoms	1.149	0.797-1.655	0.457
M3 with symptoms	1.504	1.035-2.186	0.032
Root Surgery [§]			
Root repair	1.137	0.722-1.792	0.579
Root replacement	0.978	0.688-1.390	0.900
Coronary artery bypass grafting	2.422	1.687-3.478	< 0.001
Aorta-femoral artery bypass	2.351	1.360-4.064	0.002
Operation time	1.169	1.115-1.226	< 0.001
Cardiopulmonary bypass time	1.009	1.007-1.011	< 0.001
Cross-clamp time	1.011	1.008-1.015	< 0.001
Hypothermic circulatory arrest time	1.047	1.027-1.069	< 0.001
Lowest core temperature	0.900	0.858-0.944	< 0.001
Blood loss	1.000	1.000-1.001	< 0.001
Red blood cell transfusion	1.066	1.007-1.128	0.027
Plasma transfusion	1.000	1.000-1.000	0.095
Platelets transfusion	1.194	1.046-1.363	0.009

*Compared with TEM-E0. †Compared with non-TEM-M1, M2, or M3. §Compared with no root surgery. NYHA, New York Heart Association; TEVAR, thoracic endovascular aortic repair; PCI, percutaneous coronary intervention; LVEDD, left ventricular end-diastolic diameter; TEM, type, entry, malperfusion.

Table S6 Detailed Cox regression analysis models for the association of Marfan syndrome with long-term mortality

Variables	Unadjusted		Model 1		Model 2		Model3	
	HR (95% CI)	P value	HR (95% CI)	P value	HR (95% CI)	P value	HR (95% CI)	P value
Marfan syndrome	0.705 (0.400-1.242)	0.226	1.080 (0.594-1.965)	0.800	0.836 (0.447-1.561)	0.574	0.984 (0.519-1.865)	0.961
Age			1.038 (1.021-1.055)	< 0.001	1.027 (1.009-1.045)	0.003	1.033 (1.014-1.051)	0.001
Male sex			0.819 (0.574-1.168)	0.271	0.820 (0.558-1.203)	0.310	0.744 (0.503-1.100)	0.138
Time interval between onset and admission					1.039 (0.993-1.086)	0.095	1.023 (0.974-1.074)	0.372
Coronary artery disease					7.284 (3.764-14.095)	< 0.001	5.121 (2.189-11.979)	< 0.001
NYHA 3					1.728 (0.865-3.451)	0.121	1.731 (0.833-3.600)	0.142
Chronic kidney disease					1.457 (0.724-2.931)	0.291	2.110 (1.049-4.245)	0.036
Cerebrovascular disease					2.384 (1.140-4.984)	0.021	2.252 (1.075-4.714)	0.031
History of cardiovascular surgery					2.843 (1.501-5.385)	0.001	2.551 (1.334-4.878)	0.005
History of PCI					0.278 (0.074-1.044)	0.058	0.369 (0.095-1.435)	0.150
Pericardial effusion					1.307 (0.883-1.936)	0.181	1.564 (1.046-2.339)	0.029
Preoperative ejection fraction					0.971 (0.941-1.002)	0.067	0.970 (0.941-1.001)	0.057
Preoperative hemoglobin					0.994 (0.986-1.003)	0.210	0.997 (0.988-1.006)	0.528
TEM-M1 *								
M1 without symptoms					1.608 (1.055-2.450)	0.027	1.197 (0.548-2.615)	0.653
M1 with symptoms					1.954 (1.174-3.251)	0.010	1.671 (0.977-2.860)	0.061
TEM-M2 *								
M2 without symptoms					0.907 (0.646-1.271)	0.570	0.719 (0.507-1.021)	0.065
M2 with symptoms					2.838 (1.481-5.441)	0.002	3.128 (1.573-6.219)	0.001
TEM-M3 *								
M3 without symptoms					1.429 (0.979-2.087)	0.064	1.223 (0.798-1.876)	0.355
M3 with symptoms					1.505 (0.973-2.327)	0.066	1.072 (0.672-1.711)	0.771
Coronary artery bypass grafting							0.881 (0.404-1.921)	0.749
Aorta-femoral artery bypass							2.707 (1.442-5.085)	0.002
Operation time							1.010 (0.914-1.117)	0.838
Cardiopulmonary bypass time							1.008 (1.005-1.011)	< 0.001
Cross-clamp time							0.998 (0.992-1.004)	0.485
Hypothermic circulatory arrest time							1.017 (0.993-1.042)	0.157
Lowest core temperature							0.981 (0.922-1.044)	0.551
Blood loss							1.000 (1.000-1.000)	0.112
Red blood cell transfusion							1.028 (0.966-1.094)	0.385
Platelets transfusion							1.085 (0.936-1.258)	0.279

* Compared with non-TEM-M1, M2, or M3. HR, hazard ratio; CI, confidence interval; NYHA, New York Heart Association; PCI, percutaneous coronary intervention; TEM, type, entry, malperfusion.

Table S7 Univariable competing risk Cox regression analysis for reoperation

Variables	Hazard ratio	95% confidence interval	P value
Marfan syndrome	3.863	2.425-6.155	< 0.001
Age	0.953	0.932-0.974	< 0.001
Body mass index	0.984	0.937-1.033	0.506
Male sex	1.078	0.640-1.816	0.778
Time interval between onset and admission	1.004	0.953-1.058	0.888
Smoking	0.708	0.459-1.093	0.120
Alcohol addiction	0.309	0.098-0.973	0.045
Hypertension	0.776	0.486-1.240	0.289
Diabetes mellitus	0.401	0.055-2.918	0.367
Hyperlipemia	0.948	0.516-1.743	0.864
Coronary artery disease	1.319	0.332-5.236	0.694
NYHA \geq 3	0.000	0.000-Inf	0.995
Chronic kidney disease	1.064	0.404-2.807	0.900
Chronic obstructive pulmonary disease	1.759	0.229-13.488	0.587
Cerebrovascular disease	0.511	0.072-3.646	0.503
Family history of aortic disease	4.359	1.993-9.534	< 0.001
History of cardiovascular surgery	2.199	1.008-4.798	0.048
History of TEVAR	4.393	1.581-12.208	0.005
History of PCI	2.299	0.309-17.117	0.416
Aortic insufficiency	1.344	0.886-2.041	0.165
Pericardial effusion	1.030	0.557-1.904	0.925
Cardiac wall motion abnormality	0.000	0.000-Inf	0.994
Preoperative ejection fraction	0.980	0.939-1.022	0.340
Preoperative LVEDD	1.020	0.989-1.053	0.214
Preoperative white blood cell	0.993	0.949-1.038	0.745
Preoperative platelet	1.001	0.998-1.004	0.676
Preoperative hemoglobin	1.006	0.995-1.017	0.282
TEM-E *			
E1	2.449	0.775-7.741	0.127
E2	1.634	0.487-5.484	0.426
E3	1.246	0.207-7.515	0.810
TEM-M0	1.169	0.674-2.027	0.578
TEM-M1 †			
M1 without symptoms	0.431	0.174-1.068	0.069
M1 with symptoms	1.808	0.931-3.515	0.081
TEM-M2 †			
M2 without symptoms	0.778	0.504-1.200	0.256
M2 with symptoms	0.899	0.220-3.676	0.882
TEM-M3 †			
M3 without symptoms	1.097	0.686-1.753	0.700
M3 with symptoms	0.598	0.313-1.141	0.119
Root Surgery [§]			
Root repair	0.912	0.449-1.850	0.798
Root replacement	1.241	0.787-1.956	0.352
Coronary artery bypass grafting	0.420	0.170-1.036	0.060
Aorta-femoral artery bypass	1.310	0.555-3.094	0.537
Operation time	0.949	0.853-1.055	0.333
Cardiopulmonary bypass time	0.999	0.996-1.002	0.506
Cross-clamp time	0.999	0.993-1.006	0.868
Hypothermic circulatory arrest time	1.044	1.015-1.073	0.002
Lowest core temperature	1.016	0.952-1.085	0.624
Blood loss	1.000	1.000-1.000	0.163
Red blood cell transfusion	0.961	0.876-1.054	0.395
Plasma transfusion	1.000	0.999-1.000	0.181
Platelets transfusion	1.032	0.855-1.246	0.741

*Compared with TEM-E0. †Compared with non-TEM-M1, M2, or M3. §Compared with no root surgery. NYHA, New York Heart Association; TEVAR, thoracic endovascular aortic repair; PCI, percutaneous coronary intervention; LVEDD, left ventricular end-diastolic diameter; TEM, type, entry, malperfusion.

Table S8 Detailed competing risk Cox regression analysis models for the association of Marfan syndrome with reoperation

Variables	Unadjusted		Model 1		Model 2		Model 3	
	HR (95% CI)	P value	HR (95% CI)	P value	HR (95% CI)	P value	HR (95% CI)	P value
Marfan syndrome	3.863 (2.425-6.155)	< 0.001	2.744 (1.643-4.582)	< 0.001	2.122 (1.220-3.692)	0.008	2.060 (1.183-3.586)	0.011
Age			0.971 (0.950-0.992)	0.008	0.967 (0.947-0.988)	0.002	0.967 (0.946-0.988)	0.002
Male sex			1.011 (0.595-1.718)	0.968	1.021 (0.598-1.743)	0.939	1.007 (0.597-1.699)	0.980
Alcohol addiction					0.378 (0.121-1.182)	0.094	0.405 (0.130-1.263)	0.119
Family history of aortic disease					1.804 (0.759-4.287)	0.182	1.722 (0.730-4.064)	0.215
History of cardiovascular surgery					1.567 (0.693-3.546)	0.281	1.347 (0.616-2.944)	0.455
History of TEVAR					5.472 (2.095-14.294)	0.001	5.824 (2.331-14.549)	< 0.001
Hypothermic circulatory arrest time							1.041 (1.009-1.074)	0.011

HR, hazard ratio; CI, confidence interval; TEVAR, thoracic endovascular aortic repair.