Long-Term Results of Frozen Elephant Trunk and Total Arch Replacement for Type A Aortic Dissection in Marfan Syndrome

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No Conflict of Interest

Surgery for Marfan Patients With Acute Type A Dissection Using a Stented Elephant Trunk Procedure

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Background. The purpose of t
efficacy of total arch replacemen
elephant trunk implantation f
acute Stanford type A aortic
aortic arch.
Methods. Between January 20
secutive Marfan patients (4 fema
A aortic dissection involving t
total arch replacement combine
stented elephant trunk. Aortic d
iliac artery was seen in 10 patie
aorta in 3 patients. Ages ranged
39 ± 13). Computed tomography
residual false lumen in the desc
Results. All patients survive
from hospital. One patient wi
nominate artery suffered cereb

Repair of acute type A aortic
Issues regarding aortic enla
tion after repair of acute type A d
particularly for patients with M
replacement of the arch combin
trunk for Stanford type A aort
thrombosis of the false lumen,
remaining dissected aorta, and
for late reoperation [2]. In this
reviewed our experience of tota
combined with implantation of a
patients with Marfan syndrome
dissection involving the aortic ar

Patients and Methods

A consecutive series of 13 Marf
9 male) aged 17 to 65 years (me

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*Drs Sun and Qi contributed equally to

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going total arch replacement combined with elephant trunk implantation are listed in Table 1.

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Long-term outcomes of frozen elephant trunk for type A aortic dissection in patients with Marfan syndrome

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Incidences of reoperation, death, and event-free survival were 15%, 25% and 60%, respectively, at 10 years.

Central Message

In 106 Marfan syndrome patients with type A aortic dissection, the frozen elephant trunk and total arch replacement technique has achieved low operative mortality, favorable long-term survival and freedom from reoperation.

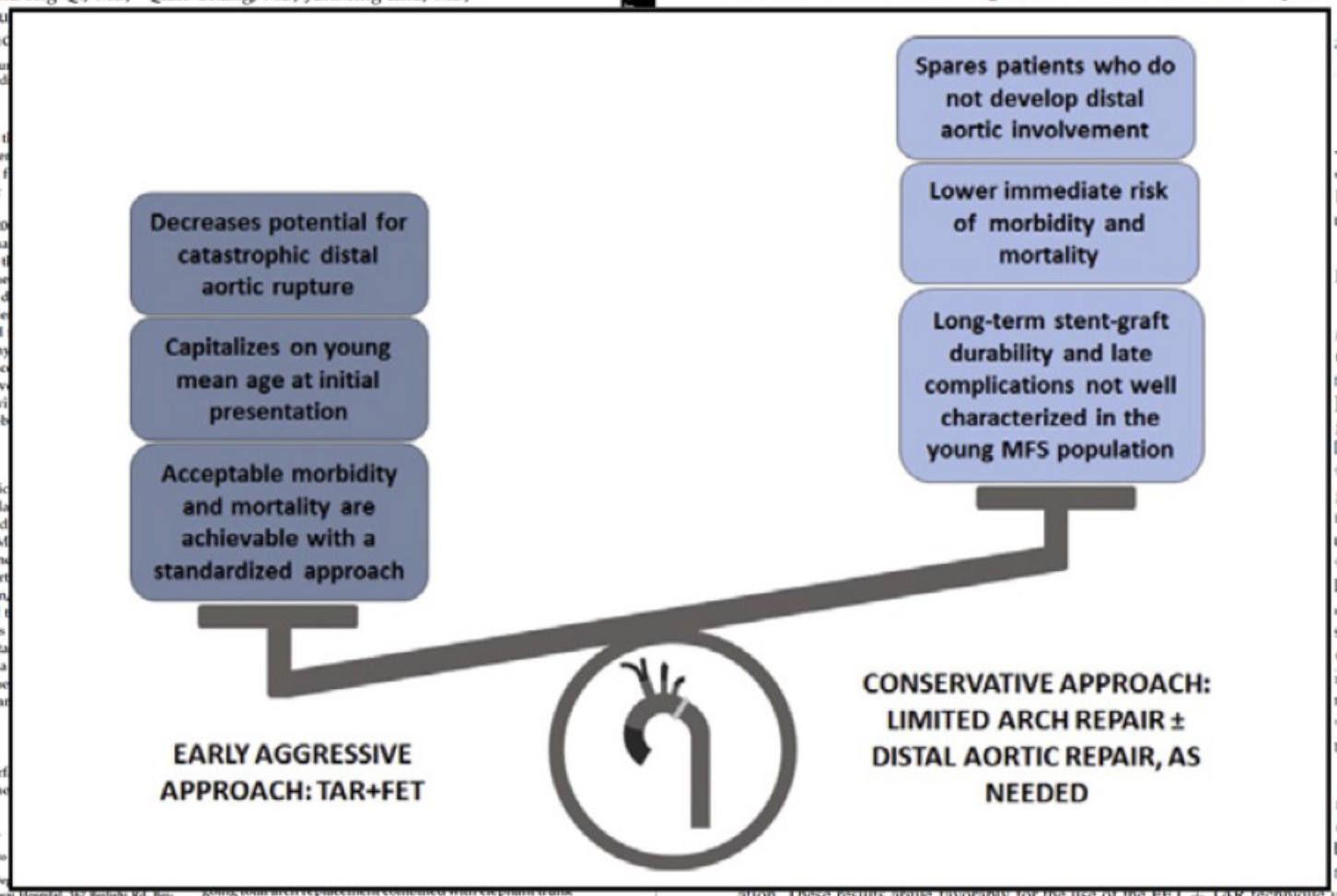
Perspective

The extent of surgical repair and the use of the frozen elephant trunk (FET) technique are controversial for type A dissection in Marfan syndrome. This study found favorable early and long-term results via total arch replacement with FET. A Bentall procedure during FET was predictive of better late survival and increased risk for reoperation. This extensive surgical approach is recommended in such a setting.

See Editorial Commentary page 1190.

See Editorial page 1169.

ation. These results argue favorably for the use of the FET + TAR technique in the management of TAAD in patients with MFS. (J Thorac Cardiovasc Surg 2017;154:1175-89)



Experience in Beijing

- 1993-2018, > 600 patients with Marfan syndrome
- 1996-2017, 223 type A dissections
  - Acute, 141
  - Chronic, 82
- Since 2003, TAR + FET
- One-stage vs two stage repair for acute type I dissection
  - Similar early and late survival
  - Two-stage repair: ↑ distal aortic dilation
  - One-stage repair: ↓ distal aortic rupture, dilation and reintervention

Objectives

- To evaluate the long-term outcomes in terms of survival and reoperation in 172 patients with Marfan syndrome
- To analyze the temporal changes of the distal aorta after FET with respect to the false lumen, true lumen and maximum aortic size, growth rates, dilation and remodeling
- To identify risk factors for late adverse events, including distal aortic dilation, reoperation and death

Profile of Patients

Variable	Total (n = 172, %)	Acute (n = 94)	Chronic (n = 78)	P value
Age (year)	34.6 ± 9.3	34.2 ± 9.6	35.0 ± 9.1	.602
Male	121 (70.3%)	72 (76.6%)	49 (62.8%)	.049
Hypertension	59 (34.3%)	32 (34.0%)	27 (34.6%)	.937
Family history of aortic dissection	71 (41.3%)	37 (39.4%)	34 (43.6%)	.575
History of proximal aortic surgery	29 (16.9%)	8 (8.5%)	21 (26.9%)	.001
Malperfusion syndrome	14 (8.1%)	11 (11.7%)	3 (3.8%)	.061
Preoperative aortic diameter (mm)				
Aortic sinus	63.4 ± 13.4	59.3 ± 11.7	69.2 ± 13.7	<.001
Aortic arch	35.8 ± 11.6	33.8 ± 10.5	38.2 ± 12.4	.020
Proximal descending aorta	37.3 ± 11.6	35.1 ± 10.7	40.2 ± 12.0	.006
Mid-descending aorta	31.0 ± 9.3	29.3 ± 8.3	33.0 ± 10.2	.017
Diaphragm	30.2 ± 9.6	28.2 ± 7.8	32.8 ± 11.1	.005
Renal arteries	26.5 ± 8.9	24.8 ± 7.4	28.7 ± 10.1	.007
Arch vessel involvement	165 (95.9%)	91 (96.8%)	74 (94.9%)	.522

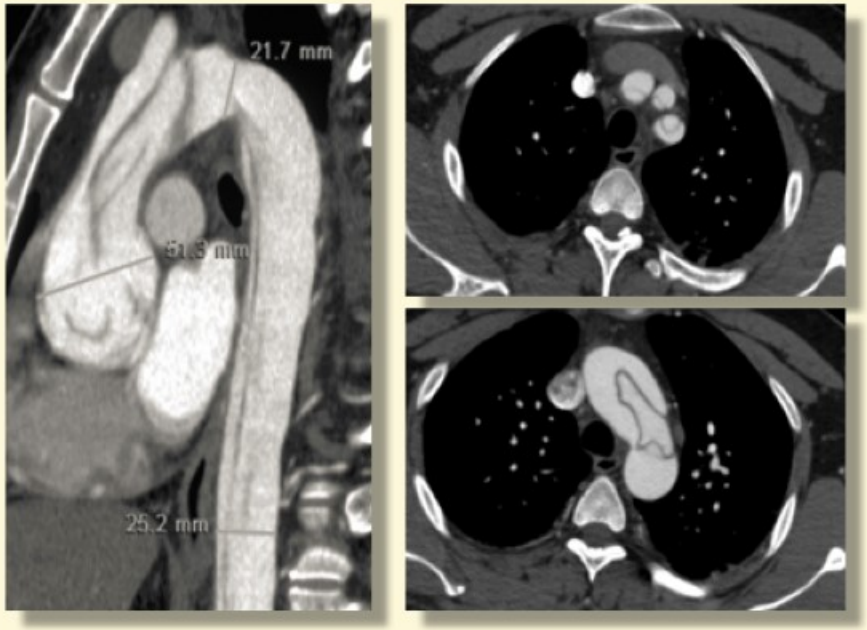
Surgical Indications and Techniques

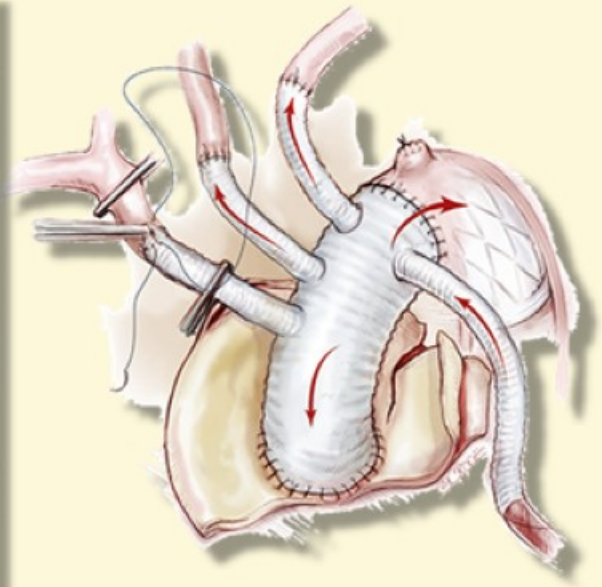
Indications

- Intimal tear located in arch or descending aorta
- Aneurysm of the arch or proximal descending aorta (> 40 mm in diameter)
- dissection, aneurysm, or occlusion of arch vessels

Technical Details

- Right axillary cannulation
- Unilateral antegrade cerebral perfusion
- Hypothermic circulatory arrest at 20-25°C
- Arch transected between LCA and LSCA
- Distal first strategy



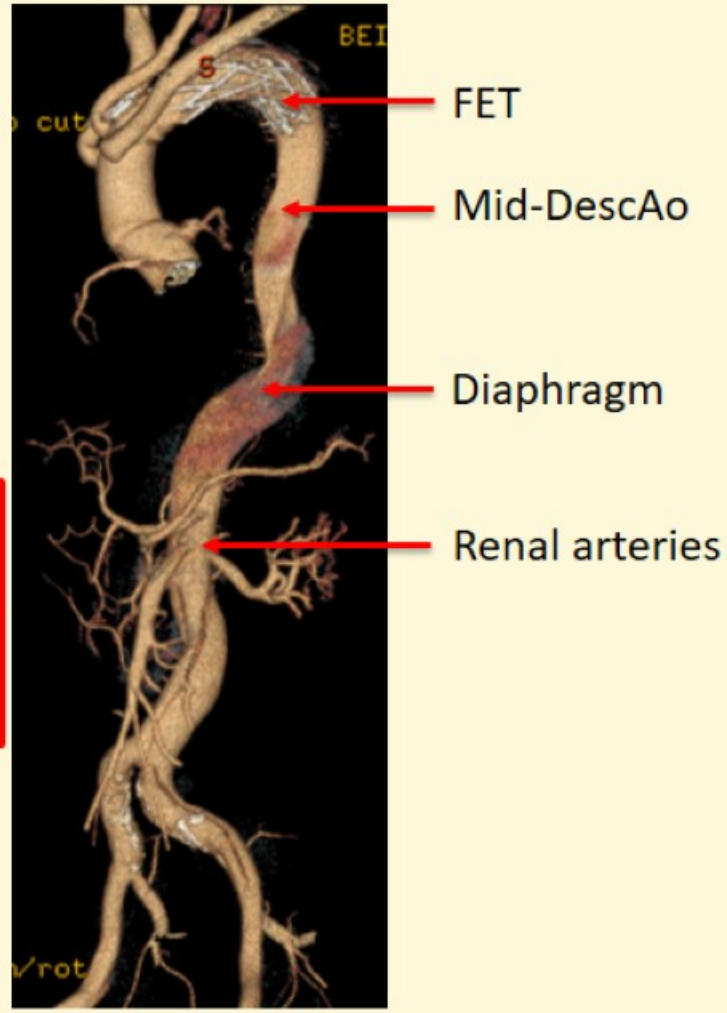


Early Mortality and Morbidity

Mortality *and* Morbidity	Total (n = 172, %)	Acute (n=94)	Chronic (n=78)	*P* value
Operative mortality	14 (8.1%)	7 (7.4%)	7 (9.0%)	.715
Operative complications	39 (22.7%)	22 (23.4%)	17 (21.8%)	.802
Spinal cord injury	2 (1.2%)	2 (2.1%)	0	.195
Stroke	5 (2.9%)	3 (3.2%)	2 (2.6%)	.807
Low cardiac output	7 (4.1%)	4 (4.3%)	3 (3.8%)	.892
Lower limb ischemia	10 (5.8%)	3 (3.2%)	7 (9.0%)	.107
Acute renal failure	6 (3.5%)	3 (3.2%)	3 (3.8%)	.816
Distal aortic rupture	2 (1.2%)	2 (2.1%)	0	.195
Re-exploration for bleeding	10 (5.8%)	5 (5.3%)	5 (6.4%)	.761

Follow-Up and Endpoints

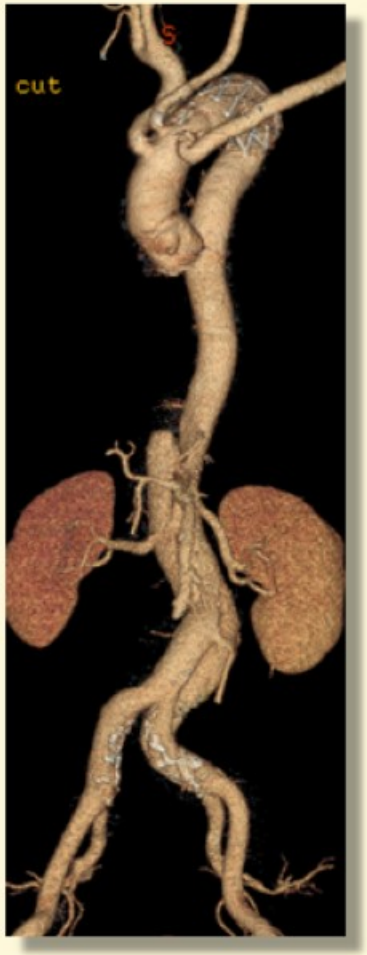
- **Follow-up:** 98.7% (156/158) for 6.2 ± 3.3 years
- **Clinical endpoints** (Cox regression)
  - Late death
  - Distal aortic reoperations
- **Imaging follow-up**
  - Aortic dilatation
    1) Maximal aortic diameter (DMax) of > 50 mm
       (45 mm for family history of aortic surgery or rupture)
    2) An average growth rate of > 5 mm/year
  - Trends of changes in TL, FL and maximal aortic size (mixed linear model)
  - False lumen obliteration



False Lumen Obliteration and Remodeling

False lumen status before discharge

Aortic segments	Complete (%)	Partial (%)	Patent (%)
Frozen elephant trunk	86.1	12.0	1.9
Mid-descending aorta	39.8	22.2	38.0
Diaphragmatic hiatus	25.9	14.6	59.5
Renal arteries	20.9	12.0	67.1



Complete aortic remodeling on latest CT

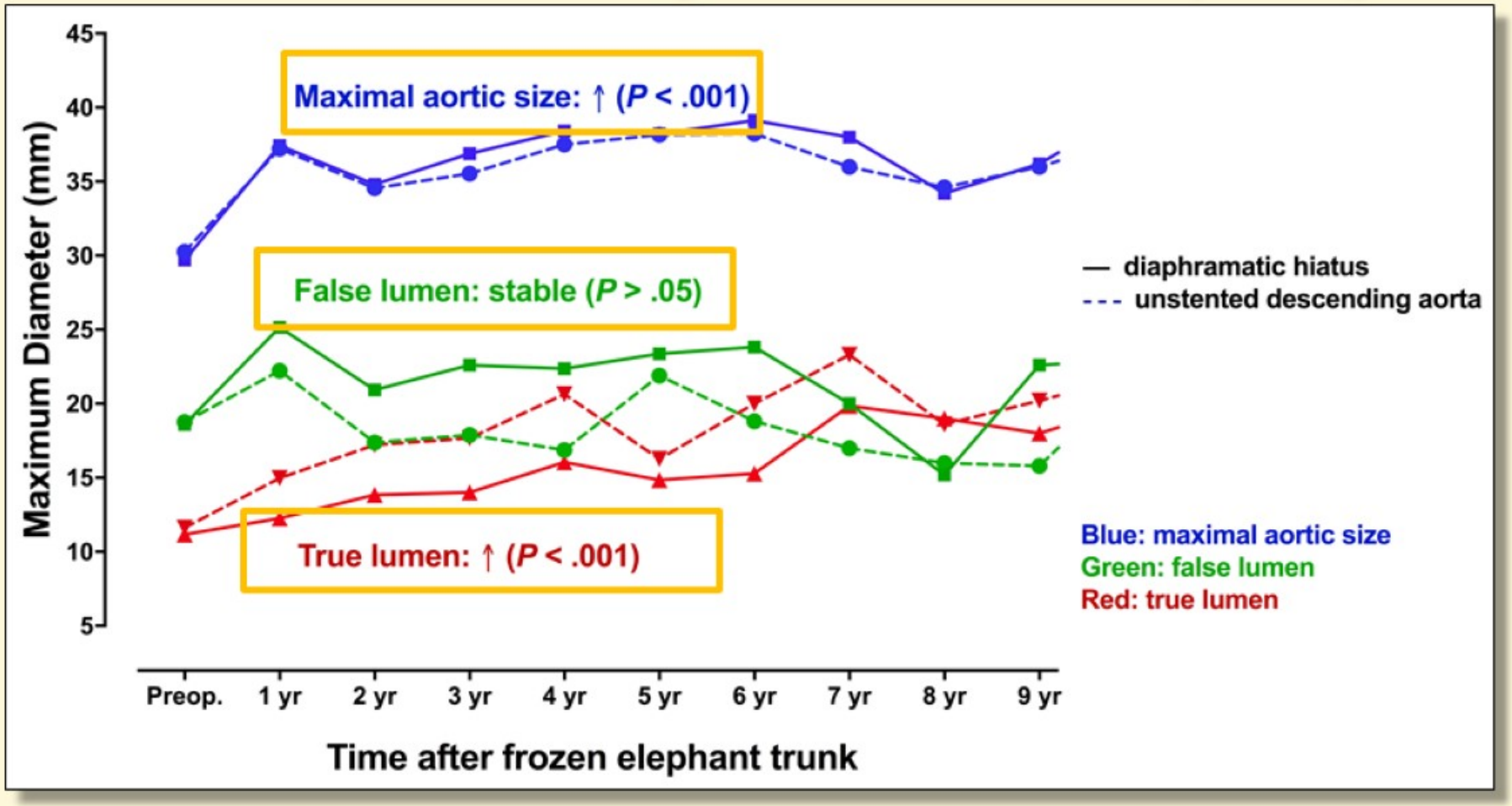
- FET: 56.4% (88/156)
- Mid-descending aorta: 28.8% (45/156)

Trend of Changes in True Lumen, False Lumen and Maximal Aortic Size over Time

Aortic segments	True lumen			False lumen			Maximal aortic size		
	Trend	β	*P* value	Trend	β	*P* value	Trend	β	*P* value
Frozen elephant trunk	↑	2.102	<.001	↓	-2.959	<.001	S	-0.043	.924
Mid-descending aorta	↑	1.304	<.001	S	-0.072	.890	↑	1.308	.001
Diaphragmatic hiatus	↑	0.725	<.001	S	0.910	.076	↑	1.698	<.001
Renal arteries	↑	0.684	<.001	S	0.706	.104	S	1.249	.752

In linear mixed modeling, ↑, expansion ($P < 0.05$); ↓, shrinkage ($P < 0.05$); S, stable ($P > 0.05$); β, relative effect of time

Trends of Changes in Aortic Size, True and False Lumen at Mid-Descending Aorta and Diaphragm



Growth Rates and Dilation

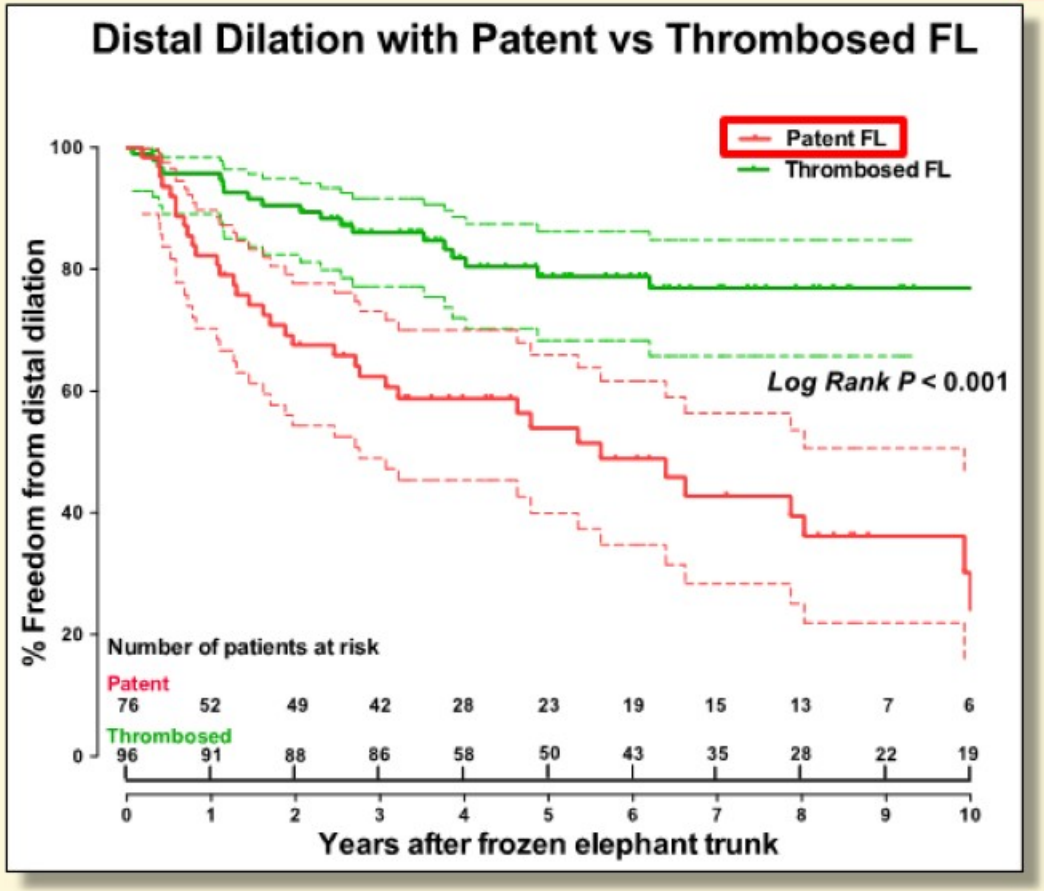
Aortic segment	Whole cohort (n = 120)	Acute (n = 69)	Chronic (n = 51)	P value
Frozen elephant trunk (FET)	0.4	-0.6	1.8	.002
Unstented descending aorta (DA)	2.8	3.5	2.0	.145
Diaphragm hiatus (DH)	3.6	4.5	2.2	.015
Renal arteries (RA)	2.6	3.3	1.7	.031

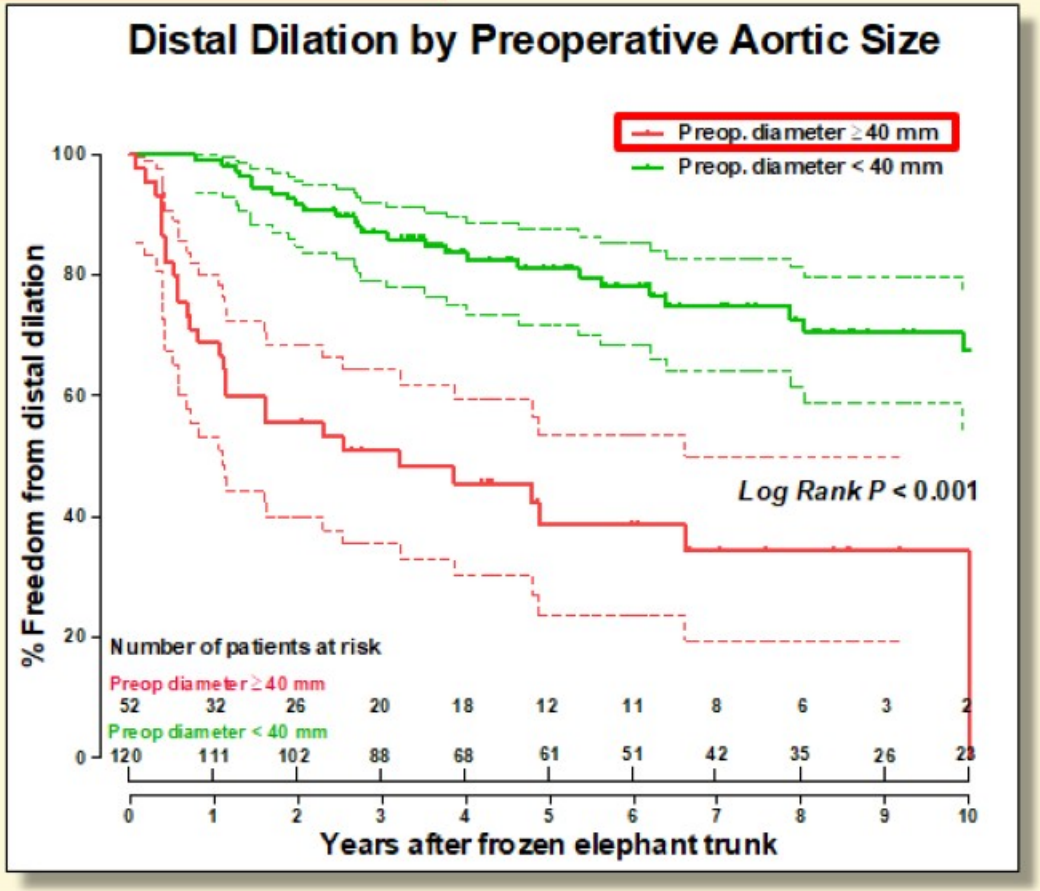
Maximal size of distal aorta

- Non-dilated: 63.5% (99/156)
- Dilated: 36.5% (57/156)
- Complete remodeling: 33 TAADS confined to mid-descending aorta

Freedom from Aortic Dilation

At 5 years: 69% (95% CI 60.6-76.0%); 57.6% at 10 years (95% CI 46.9-66.8%)

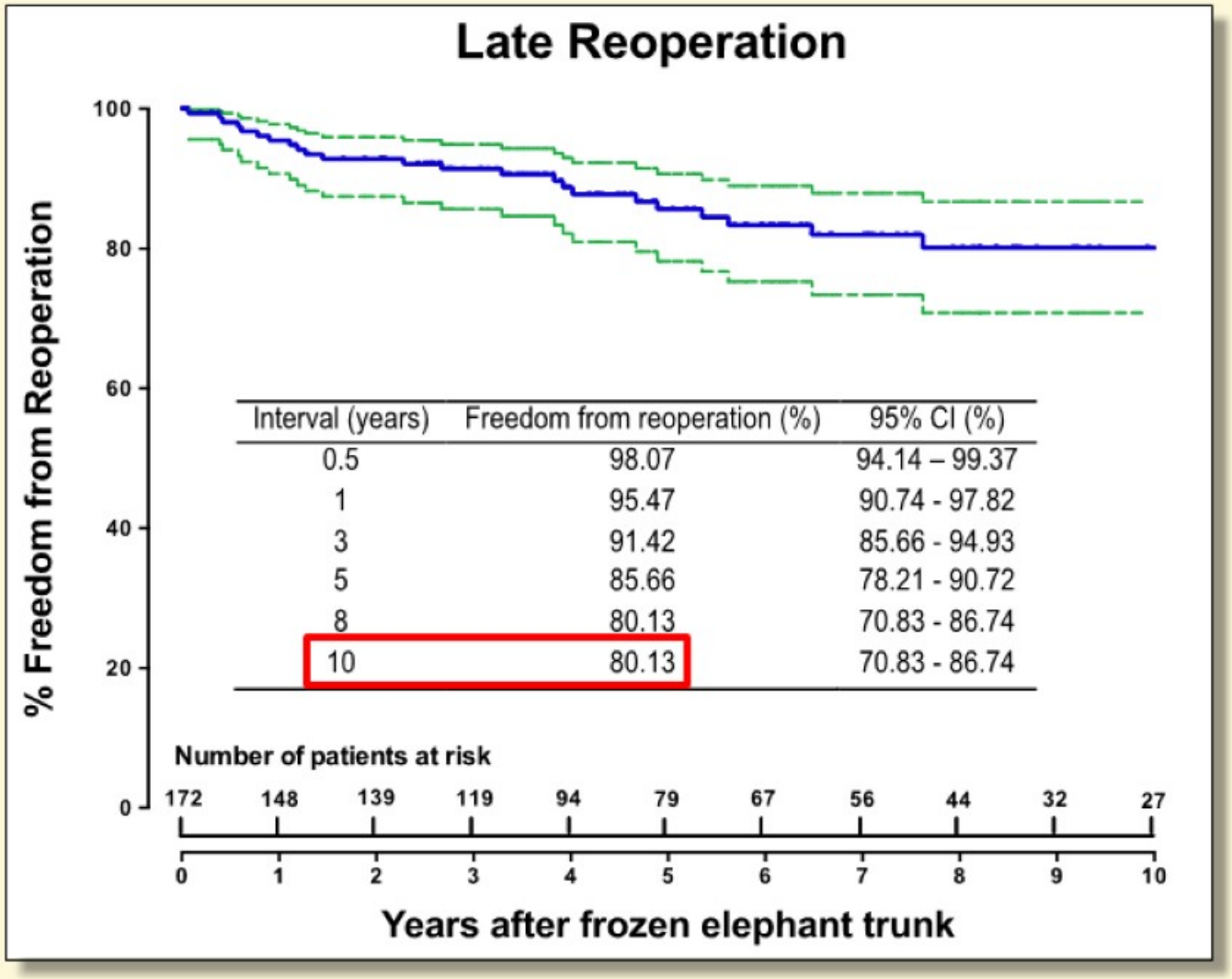




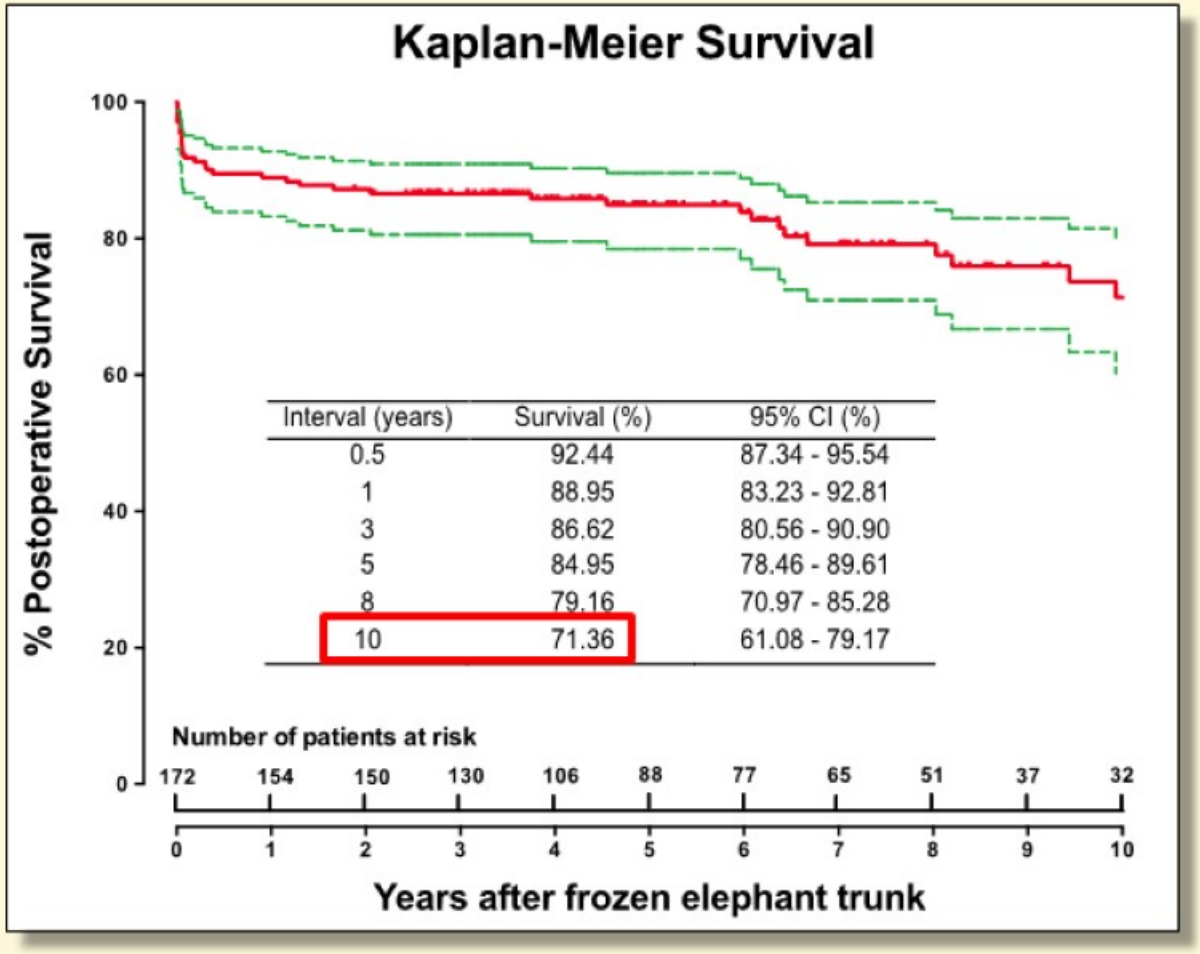
Long-Term Outcomes

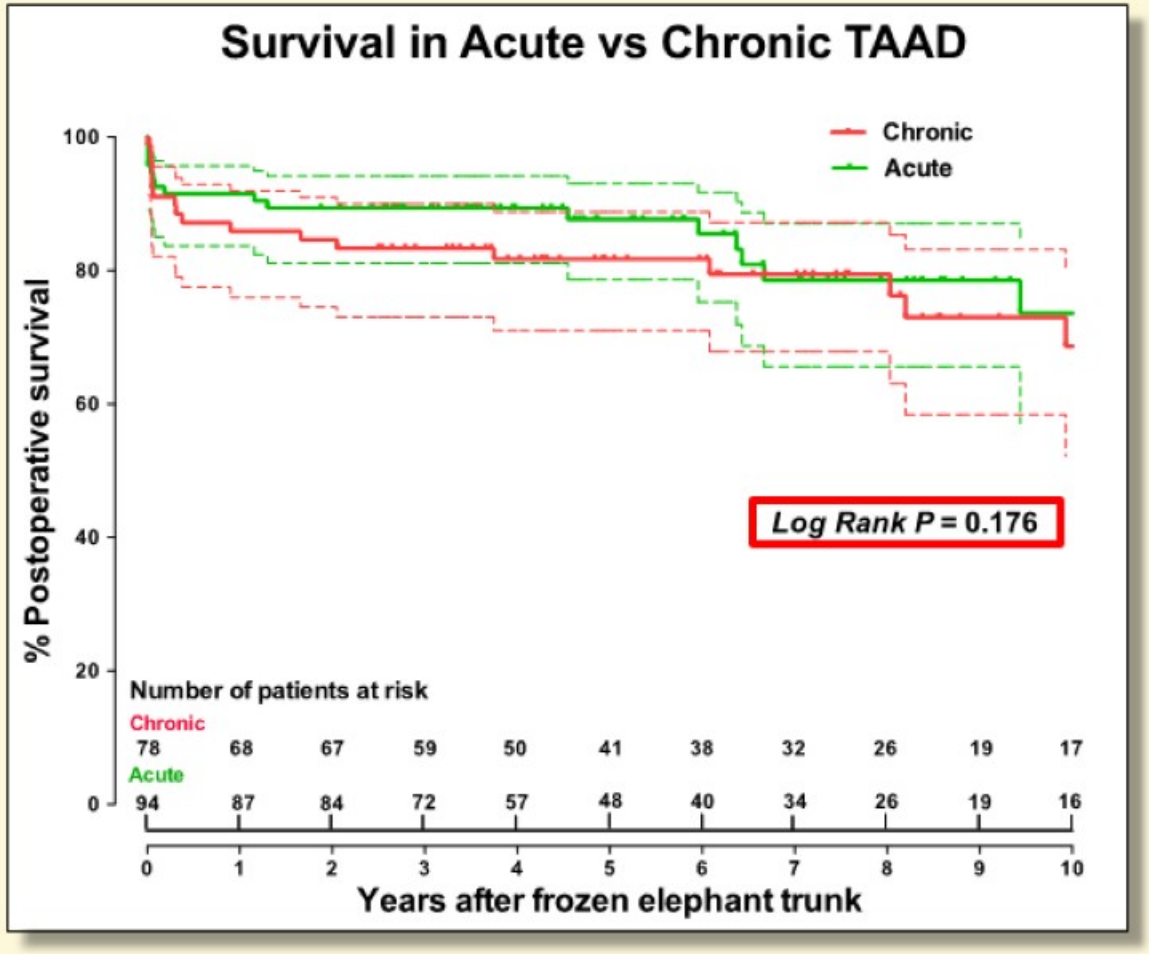
Variable	Total (n = 172, %)	Acute (n=94)	Chronic (n=78)	*P* value
Late death	22 (12.8%)	11 (11.7%)	11 (14.1%)	.639
Distal aortic rupture	9 (5.2%)	4 (4.3%)	5 (6.4%)	.528
Heart failure and arrhythmia	3 (1.7%)	1 (1.1%)	2 (2.6%)	.454
Non-cardiac cause	10 (5.8%)	6 (6.4%)	4 (5.1%)	.726
Late complications	8 (4.7%)	8 (14.0%)	8 (4.7%)	.635
Proximal stent leakage	2 (1.2%)	1 (1.1%)	1 (1.3%)	.894
Distal end of FET entering false lumen	4 (2.3%)	2 (2.1%)	2 (2.6%)	.816
Distal reintervention	23 (13.4%)	14 (14.9%)	9 (11.5%)	.520
TAAAR	19 (11.0%)	12 (12.8%)	7 (9.0%)	.430
TEVAR	4 (2.3%)	2 (2.1%)	2 (2.6%)	.450

Freedom from Reintervention

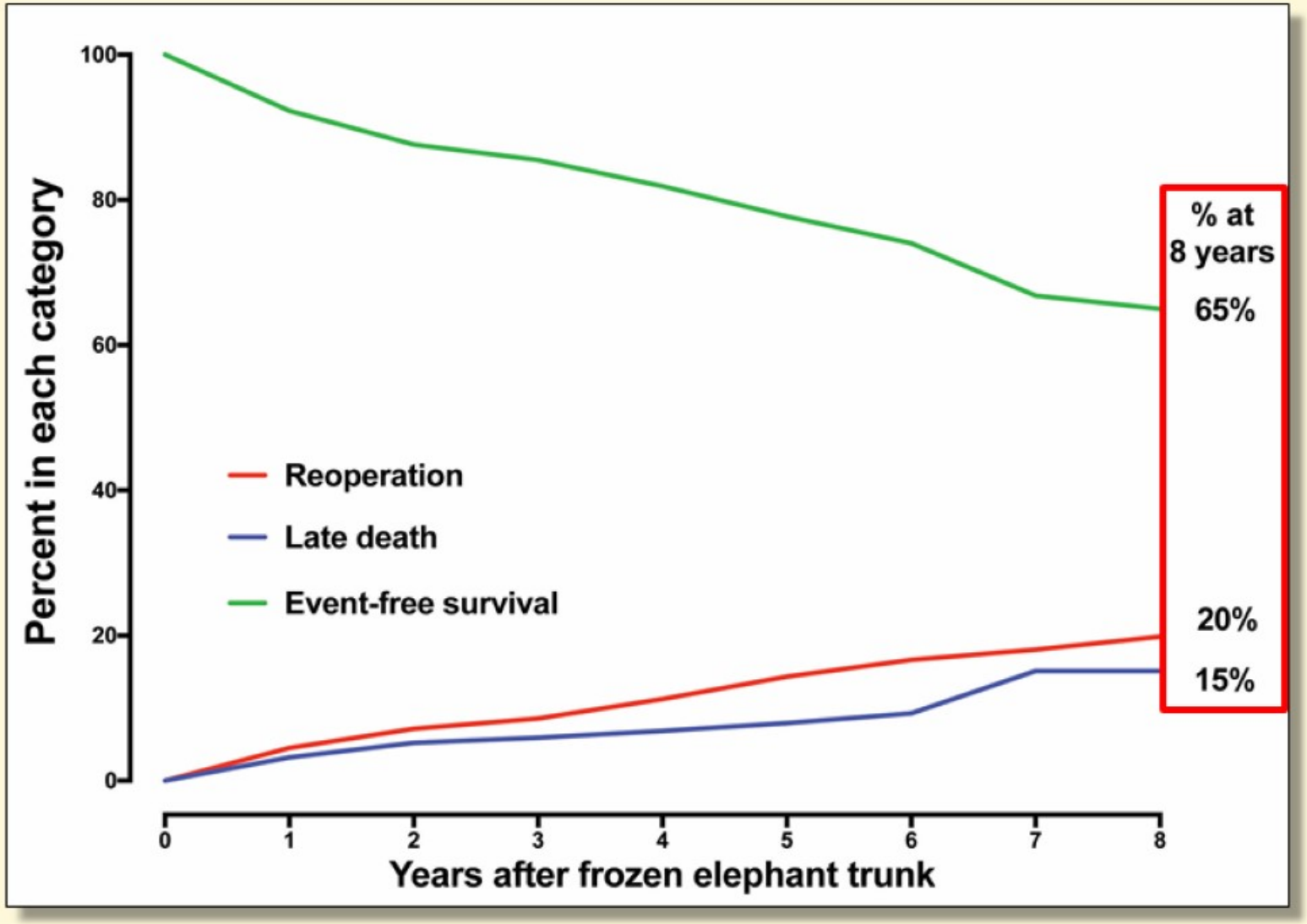


Kaplan-Meier Survival





Competing Risks of Death and Reoperation



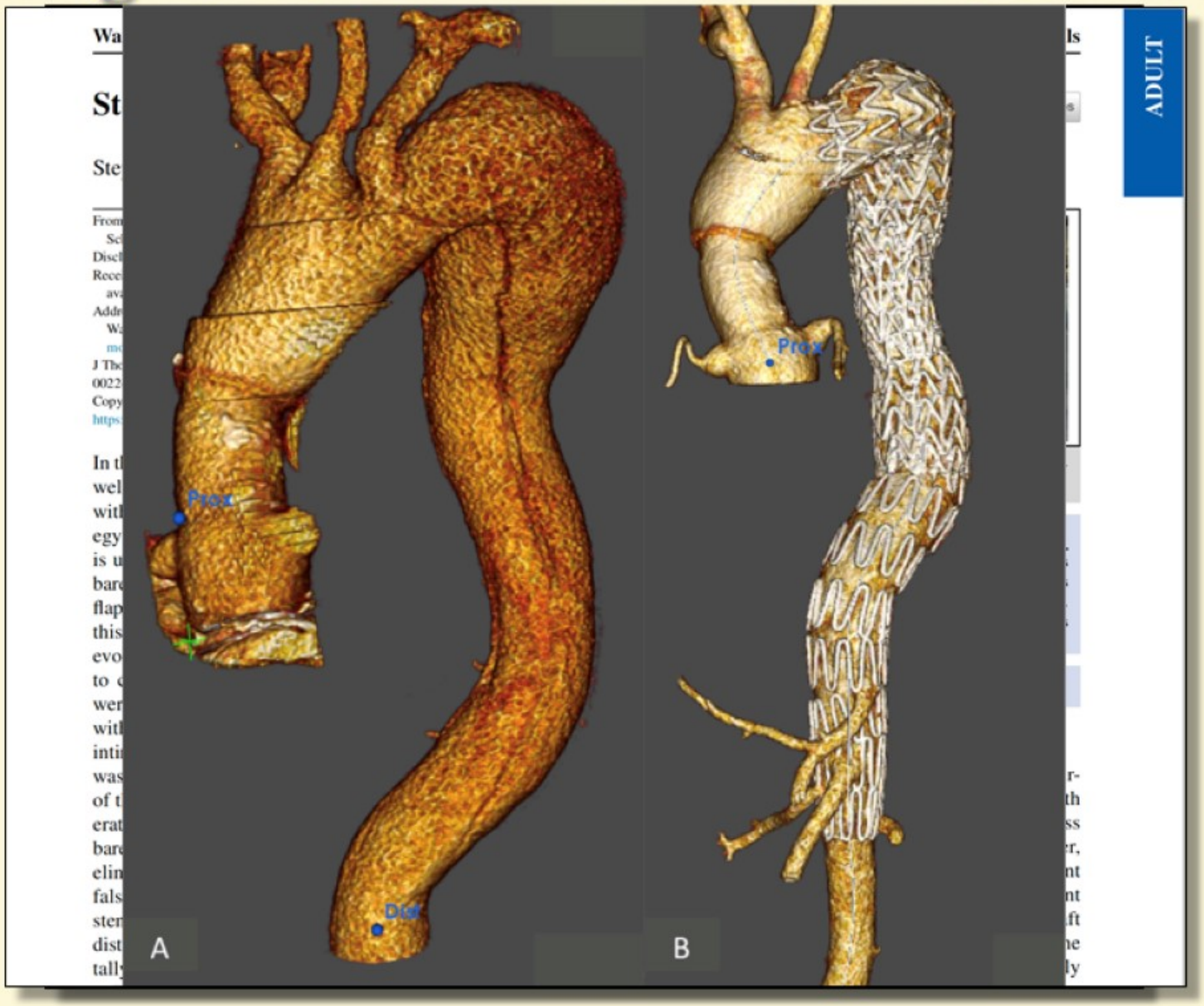
Risk Factors for Dilation, Reoperation and Death

Endpoint/Risk factors	Hazard Ratio	95% Confidence Interval	*P* value
Distal aortic dilatation			
Patent false lumen in descending aorta	3.88	1.99 - 7.57	<.001
Preoperative distal DMax (mm)	1.11	1.08 - 1.14	<.001
FET diameter < 26 mm	3.98	1.90 - 8.33	<.001
Male gender	3.35	1.52 - 7.37	.003
Distal aortic reoperation			
Patent false lumen in descending aorta	3.36	1.28 - 8.85	.014
Preoperative distal DMax (mm)	1.07	1.03 - 1.10	<.001
Late death			
Patent false lumen in descending aorta	3.31	1.03 - 10.67	.045
Preoperative distal DMax ≥ 45 mm	3.29	1.14 - 9.46	.027

Conclusions

- In Marfan patients with type A dissection, FET could induce favorable aortic remodeling by expanding the true lumen, and decreasing or stabilizing the false lumen, which led to satisfactory survival and low reoperation rates in the long term
- Our experience adds clinical and imaging evidence supporting the use of the frozen elephant trunk technique for type A dissection in patients with Marfan syndrome
- Future efforts should be aimed at reducing false lumen patency to improve long-term outcomes

Looking Forward: Metalize the Distal Aorta?



Thank you !