Oklahoma Heart Institute

Recent advances in Surgery of the Thoracic Aorta

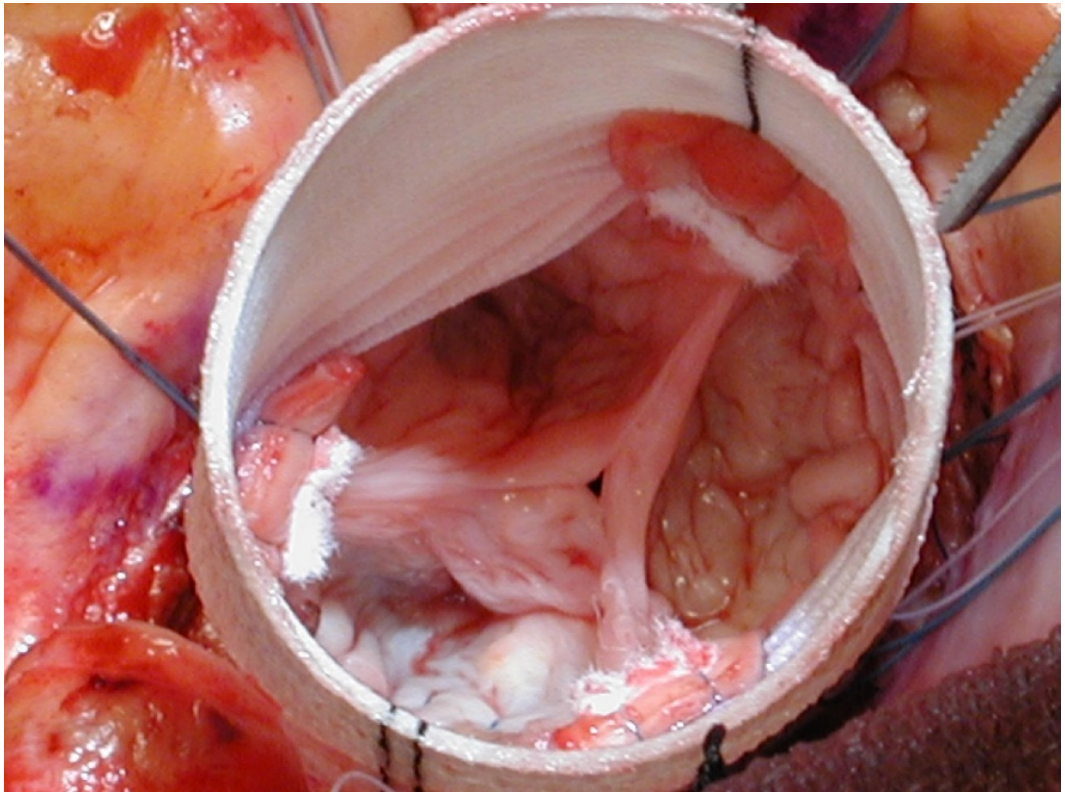
Ajit K. Tharakan, MD

Cardiothoracic Surgery

No conflict of interest

Recent advances

- Surgery of the aortic root
- Surgery of the aortic arch
- Surgery for aortic dissections
- Cerebral protection during arch surgery
- Genetic aortopathies



SURGERY OF THE AORTIC ROOT

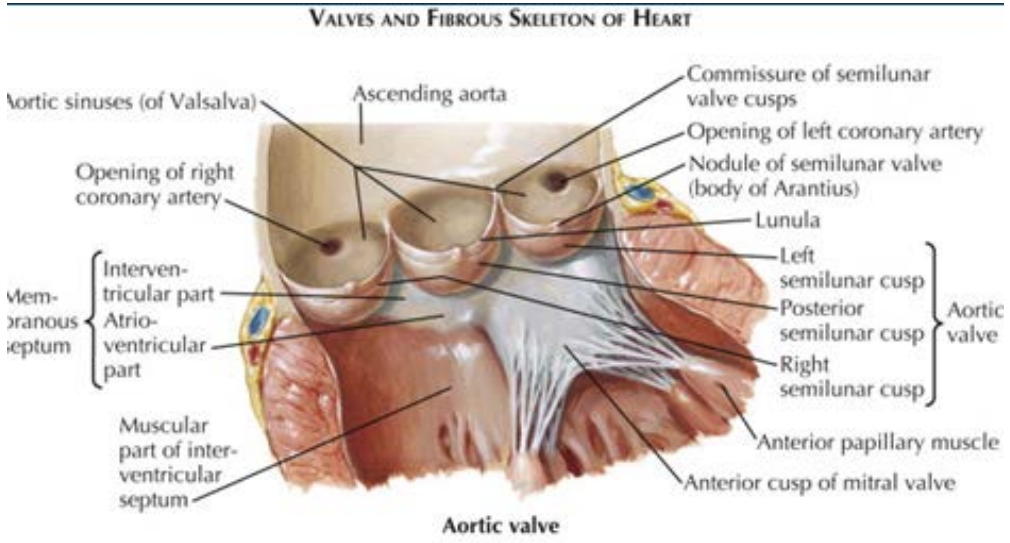
Components of the aortic root

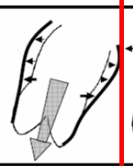
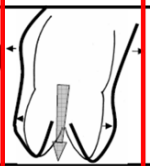
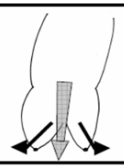
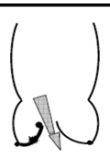
Aortic valve annulus

Valve leaflets

Sinuses of Valsalva

Sinotubular junction



AI Class	Type I Normal cusp motion with FAA dilatation or cusp perforation				Type II Cusp Prolapse	Type III Cusp Restriction
	Ia	Ib	Ic	Id		
Mechanism						
Repair Techniques (Primary)	STJ remodeling *Ascending aortic graft*	Aortic Valve sparing: *Reimplantation or Remodeling with SCA*	SCA	Patch Repair *Autologous or bovine pericardium*	Prolapse Repair *Plication* *Triangular resection* *Free margin Resuspension* *Patch*	Leaflet Repair *Shaving* *Decalcification* *Patch*
(Secondary)	SCA		STJ Annuloplasty	SCA	SCA	SCA

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Pathology

Degenerative aortic root aneurysm

Connective tissue disorders

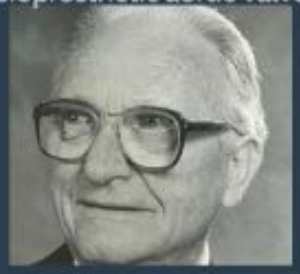
Marfans/ Loeys Dietz/ Ehlers Danlos IV

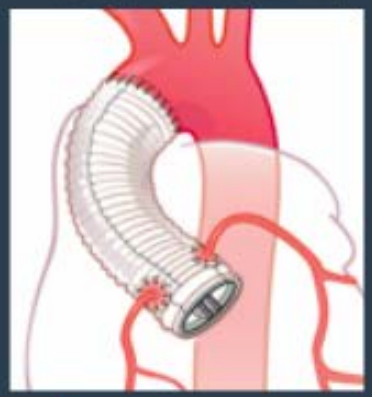
Arteritis

Type A aortic dissection

1968

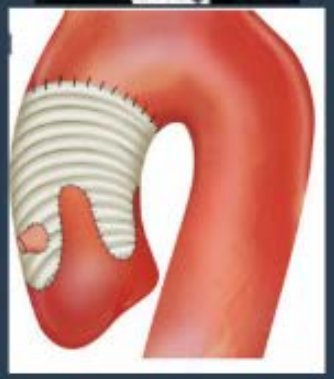
Hugh Bentall & Anthony de Bono
composite graft with mechanical / bioprosthetic aortic valve





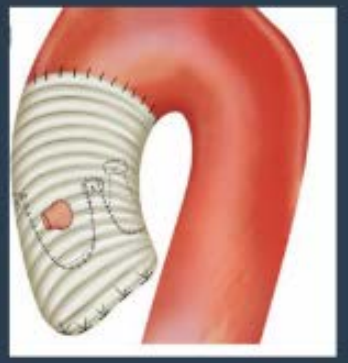
1979

Sir Magdi Yacoub
remodelling procedure



198?

Tirone David
reimplantation procedure



Prezi

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Thorax (1968), **23**, 338.

A technique for complete replacement of the ascending aorta

HUGH BENTALL AND ANTONY DE BONO

From the Royal Postgraduate Medical School, London, and Hammersmith Hospital

A technique for complete replacement of the aortic valve and ascending aorta in cases of aneurysm of the ascending aorta with aortic valve ectasia is described. The proximal aortic root was too attenuated to afford anchorage to the aortic prosthesis, so this was sutured to the ring of a Starr valve and the prostheses were inserted *en bloc*. The ostia of the coronary arteries were anastomosed to the side of the aortic prosthesis.

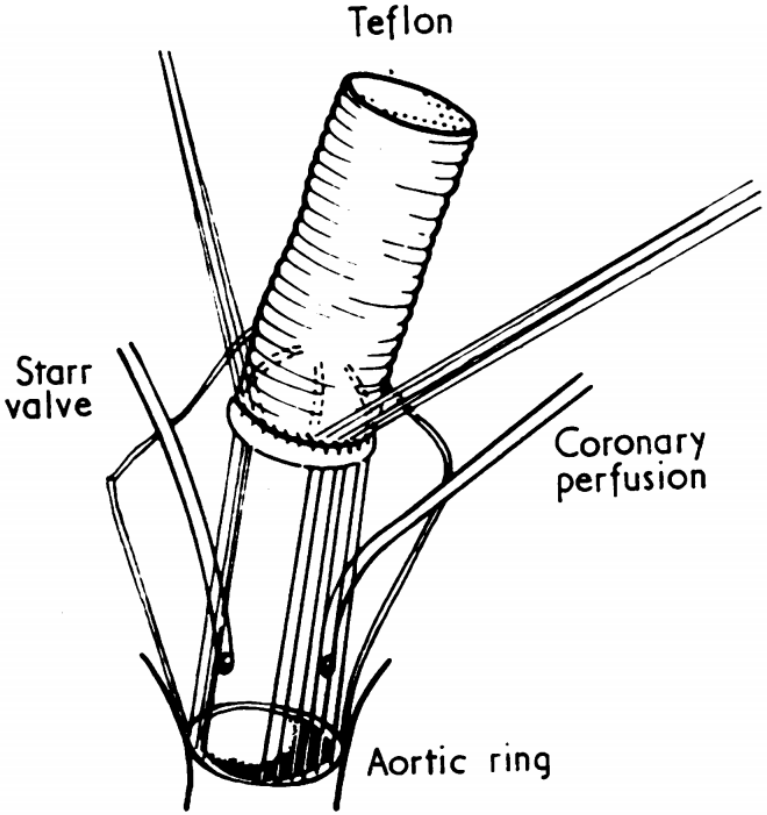


FIG. 2. *Starr valve has been sutured to aortic prosthesis: sutures have been placed in aortic ring before fixing the combined prostheses.*

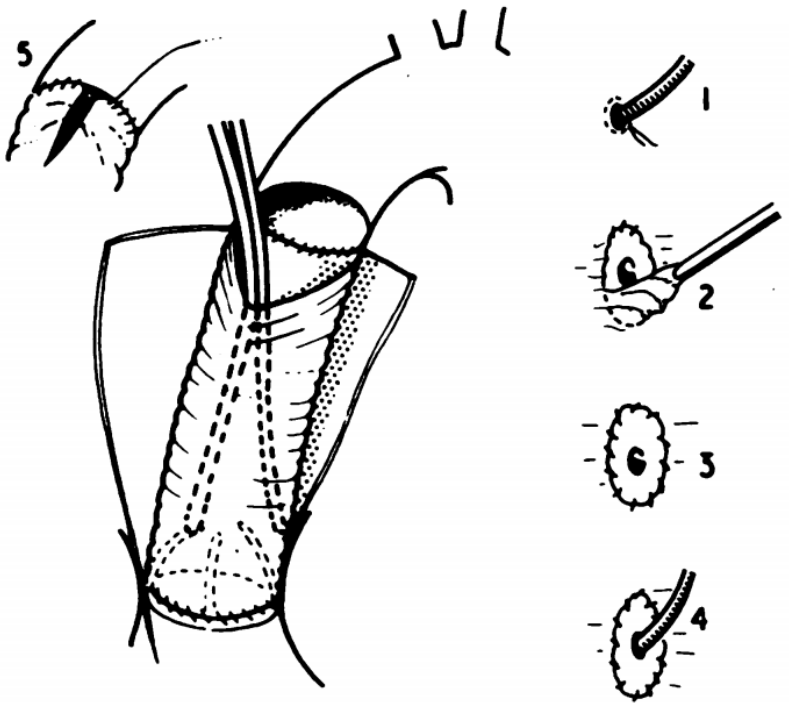
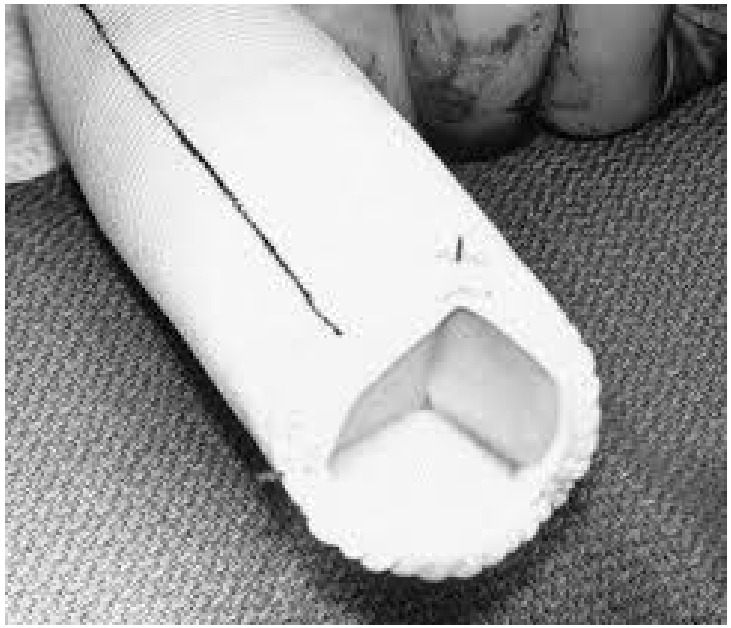


FIG. 3. *Combined prostheses in situ. Insets* 1 *to* 4 *show details of holes fashioned in the side wall of the Teflon tube to reincorporate the coronary ostia within the lumen of the new ascending aorta. Inset* 5 *shows the vertical slit in the prosthesis.*



Surgery for Aneurysms of the Aortic Root

A 30-Year Experience

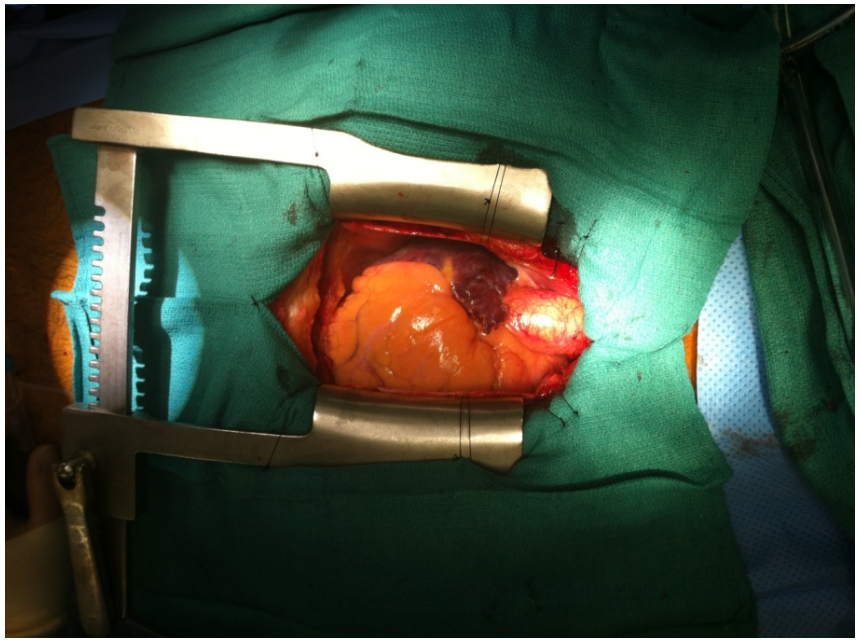
Kenton J. Zehr, MD; Thomas A. Orszulak, MD; Charles J. Mullany, MD; Alireza Matloobi, MD; Richard C. Daly, MD; Joseph A. Dearani, MD; Thoralf M. Sundt III, MD; Francisco J. Puga, MD; Gordon K. Danielson, MD; Hartzell V. Schaff, MD

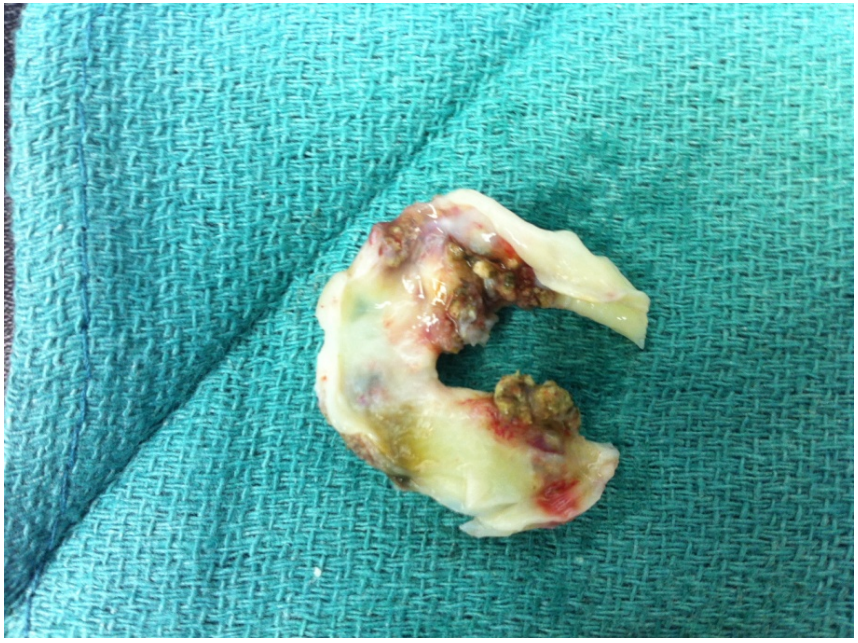
Background—This study evaluated long-term results of aortic root replacement and valve-preserving aortic root reconstruction for patients with aneurysms involving the aortic root.

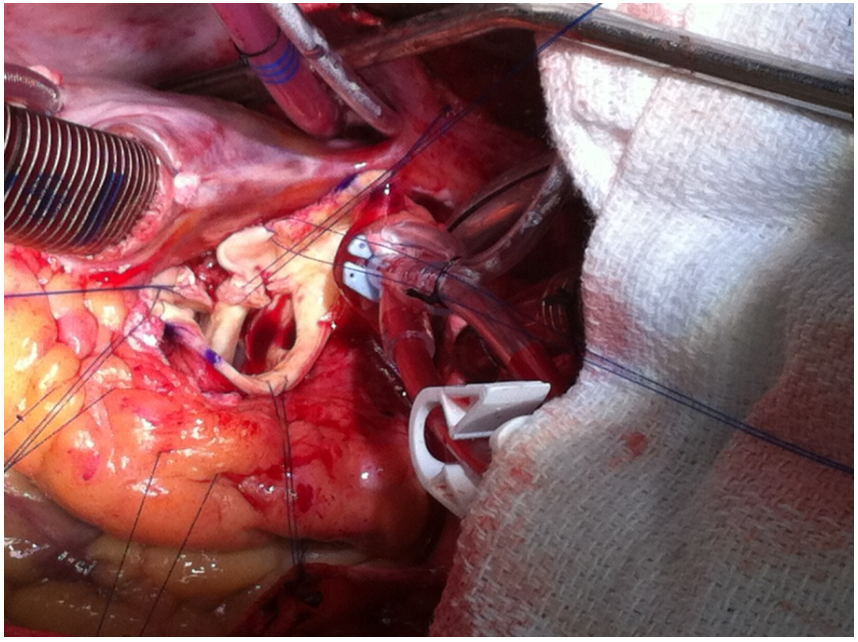
Methods and Results—Two-hundred three patients aged 53±16 years (mean±SD; 153 male, 50 female) underwent elective or urgent aortic root surgery from 1971 to 2000 for an aortic root aneurysm: 149 patients underwent a composite valve conduit reconstruction, and 54 patients underwent valve-preserving aortic root reconstruction. Fifty patients had Marfan syndrome. In-hospital and 30-day mortality was 4.0% (8/203) overall: for a composite valve conduit procedure, the corresponding value was 4.0% (6/149) and for valve-preserving procedure, 3.7% (2/54) (P=NS). Morbidity included 3 strokes (1%), 10 perioperative myocardial infarctions (5%), and 8 reoperations for bleeding (4%). Actuarial survival at 5, 10, 15, and 20 years was 93% (95% confidence interval [CI] = 88% to 97%), 79% (95% CI = 71% to 87%), 67% (95% CI = 57% to 79%), and 52% (95% CI = 36% to 69%), respectively. Freedom from reoperation was 72% (95% CI = 54% to 86%) at 20 years. Complications with anticoagulation occurred in 29 patients; with valve thrombosis, in 2; and with hemorrhage, in 27 (4 life threatening and 23 minor). Freedom from thromboembolism was 91% (95% CI = 77% to 98%) at 20 years. Freedom from endocarditis was 99% (95% CI = 92% to 100%) at 20 years. Multivariate analysis revealed preoperative mitral valve regurgitation (+3 to 4) and older age to be significant predictors of late death ($P \leq 0.005$), and Marfan syndrome, initial valve-preserving aortic root reconstruction, and need for a concomitant procedure at initial operation to be significant predictors of the need for reoperation ($P \leq 0.01$).

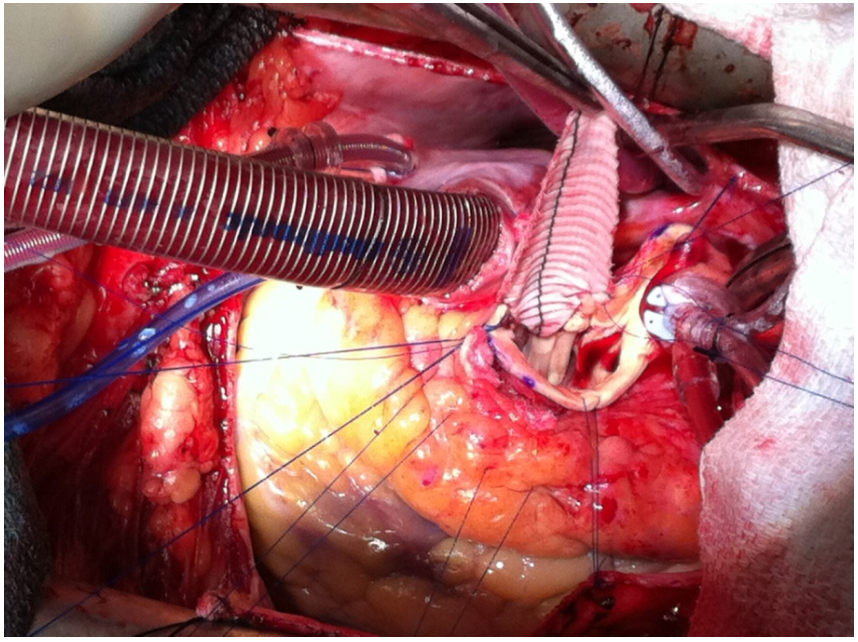
Conclusions—Aortic root replacement for aortic root aneurysms can be done with low morbidity and mortality. Composite valve conduit reconstruction resulted in a durable result. There were few serious complications related to the need for long-term anticoagulation or a prosthetic valve. Reoperation was most commonly required because of failure of the aortic valve when a valve-preserving aortic root reconstruction was performed or for other cardiac or aortic disease elsewhere. **(*Circulation*. 2004;110:1364-1371.)**

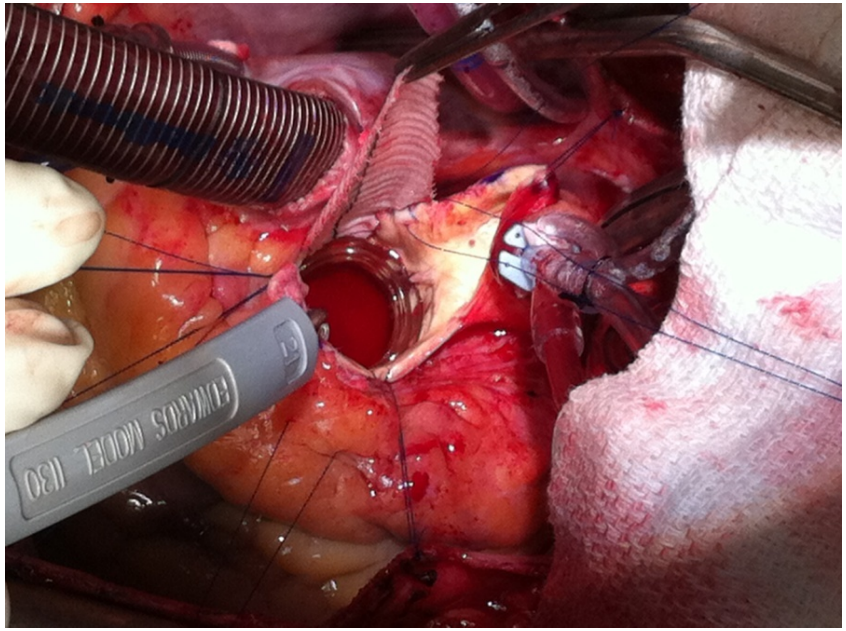
Key Words: surgery ■ aneurysm ■ aorta

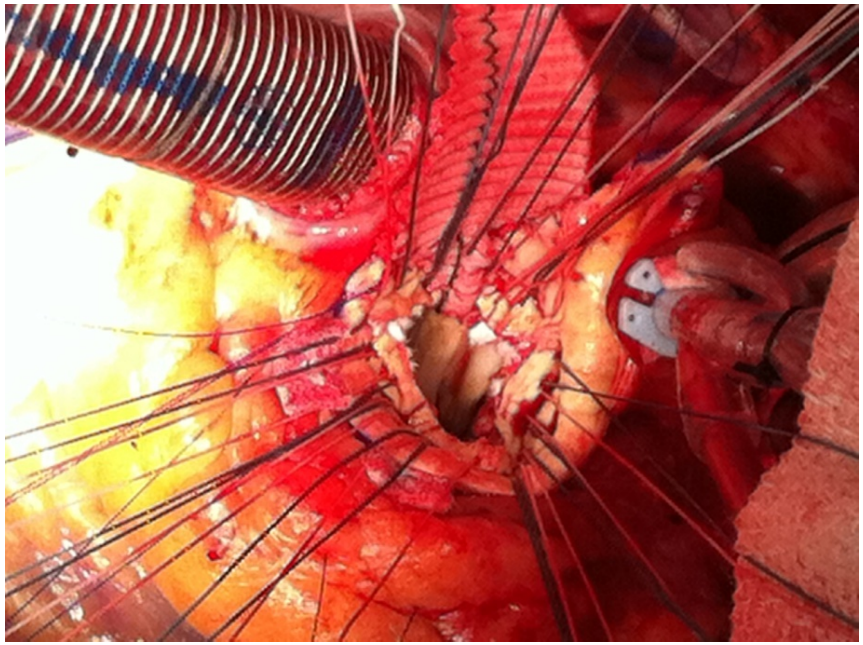


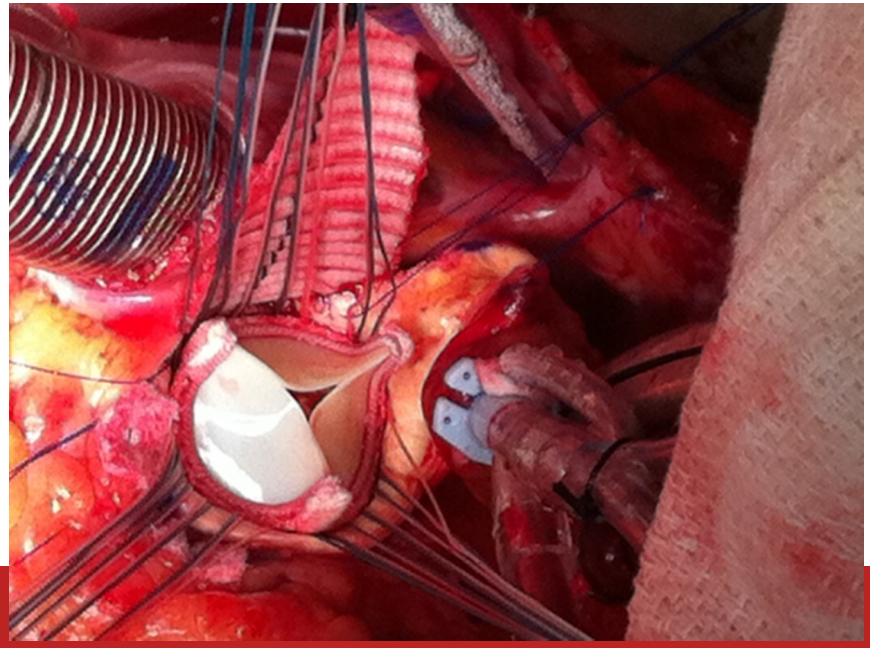


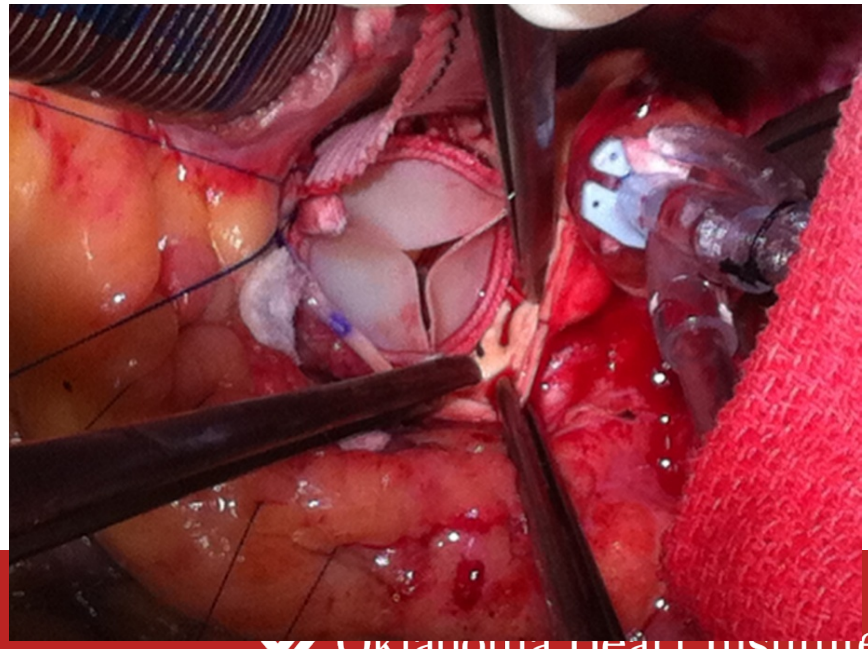





EDWARDS MODEL 1130







Why valve sparing operation

- Mechanical valve have need for lifelong anticoagulation with anticoagulation related complication 0.5-1%/ year
- Tissue valve replacement in young patients have an early failure rate

Indications

Aortic root aneurysms without valve pathology

Aortic root aneurysms

- With trileaflet valve: > 55 mm
- Marfan’s / Bicuspid valve: > 45 mm
- Loeys-Dietz’s syndrome: > 40 mm

Growth of > 0.5 cm/ year

Valve sparing aortic root replacement

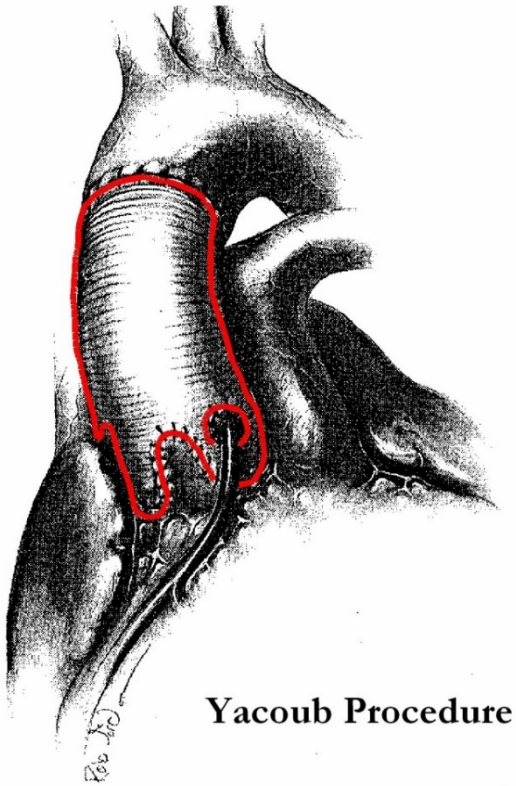


FIGURE 31–8 (A) A tubular Dacron graft is tailored to create three neo-aortic sinuses. (B) The complete repair. *(Reproduced from David TE: Remodeling of the aortic root with preservation of the native aortic valve. Op Tech Cardiac Thorac Surg 1996; 1:44, with permission from WB Saunders.)*

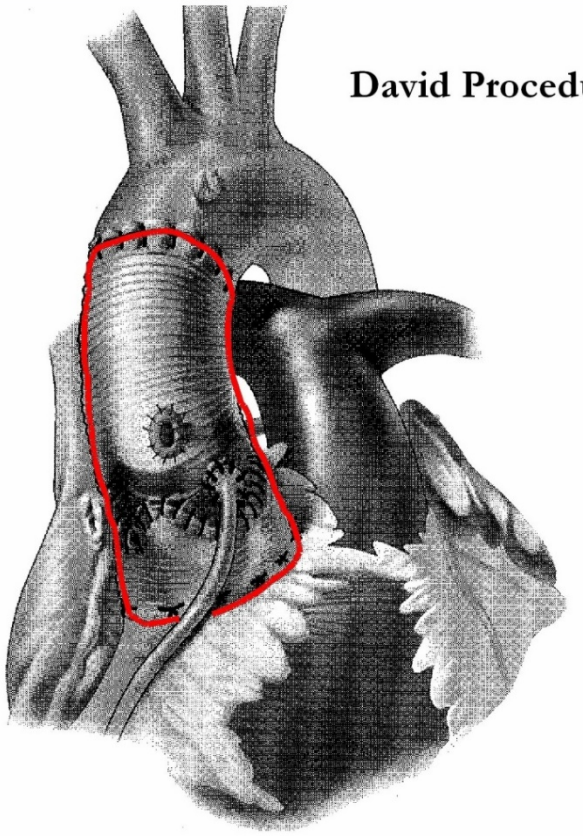


FIGURE 31–13 Reimplantation of the aortic valve. The three commissures are resuspended inside the graft and the remnants of the aortic sinuses are secured to the Dacron graft. The coronary arteries are reimplanted.

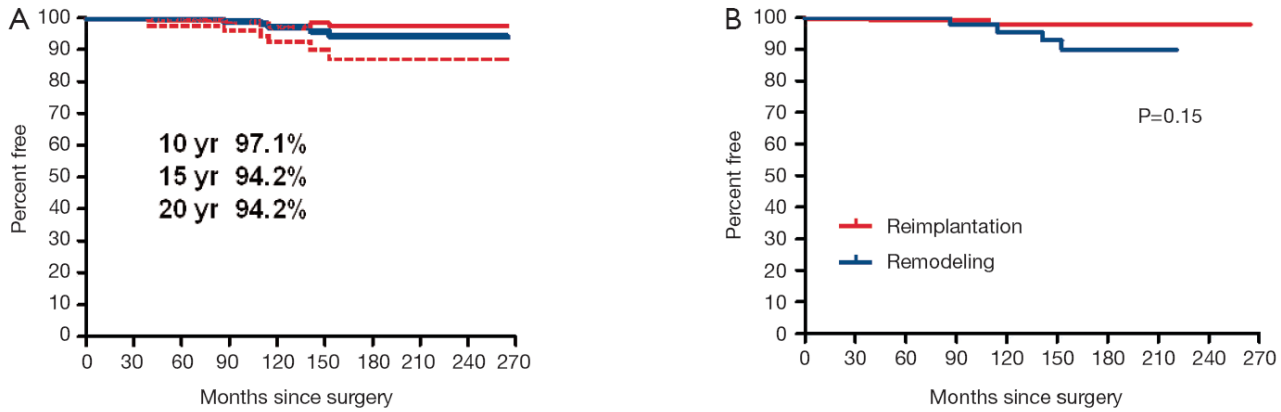


Figure 2 A. Freedom from reoperation on the aortic valve after aortic valve sparing operations; B. Freedom from reoperation after reimplantation and remodeling procedures

Table 3 Freedom from moderate or severe aortic insufficiency

	All		Reimplantation		Remodeling	
	N	Freedom	N	Freedom	N	Freedom
1 year	334	100.0%	265	100.0%	69	100.0%
5 years	206	98.2%	161	98.3%	45	98.0%
10 years	97	93.0%	59	92.9%	38	93.2%
15 years	28	78.1%	12	89.4%	16	70.7%
20 years	6	74.4%	5	89.4%	1	63.6%

Abbreviation: N = number of patients at risk

Annals of Cardiothoracic Surgery : Tirone David, January 2013

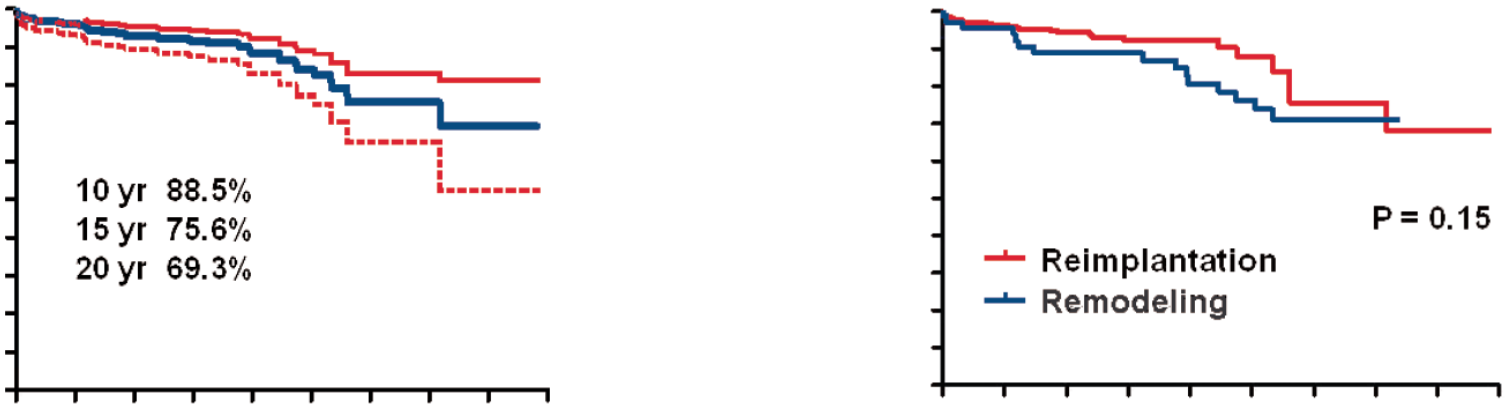
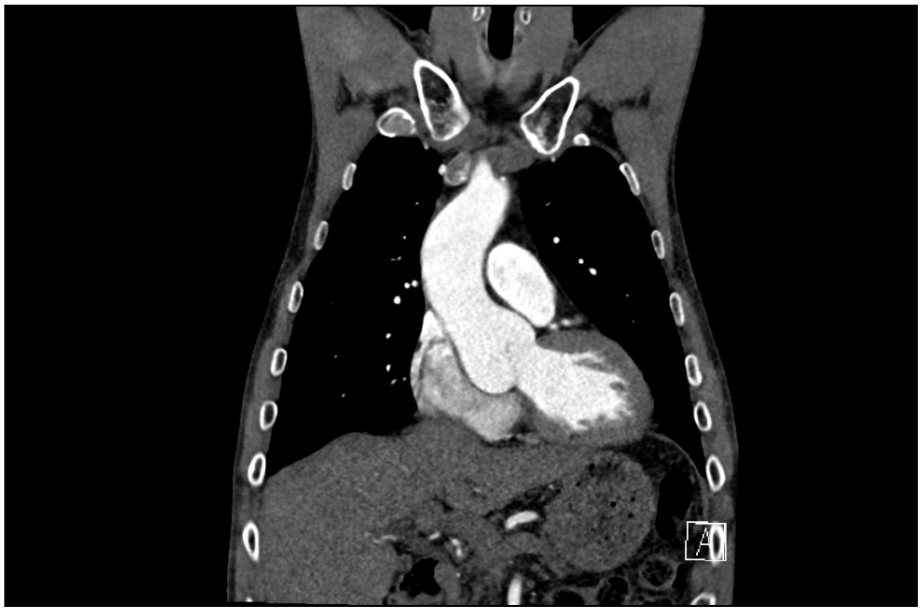
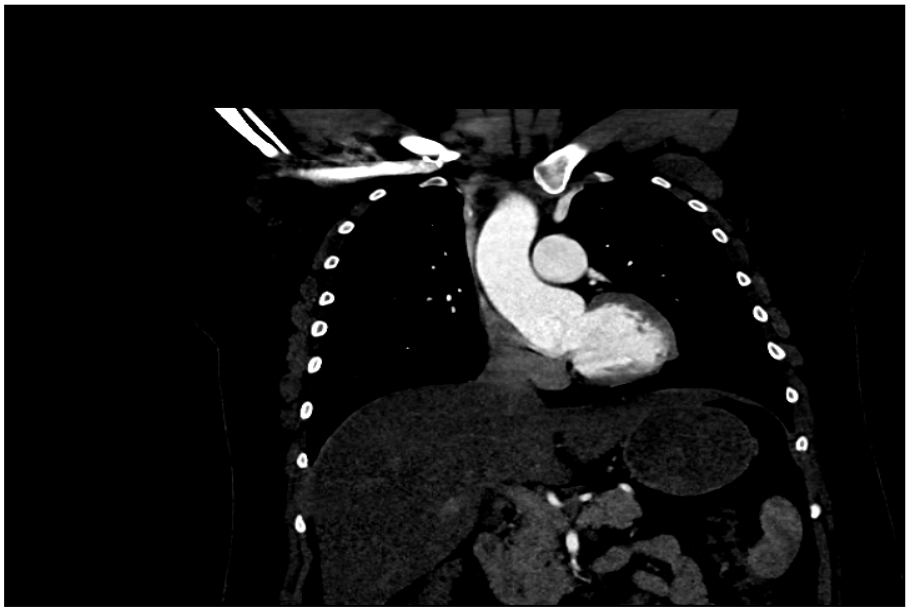
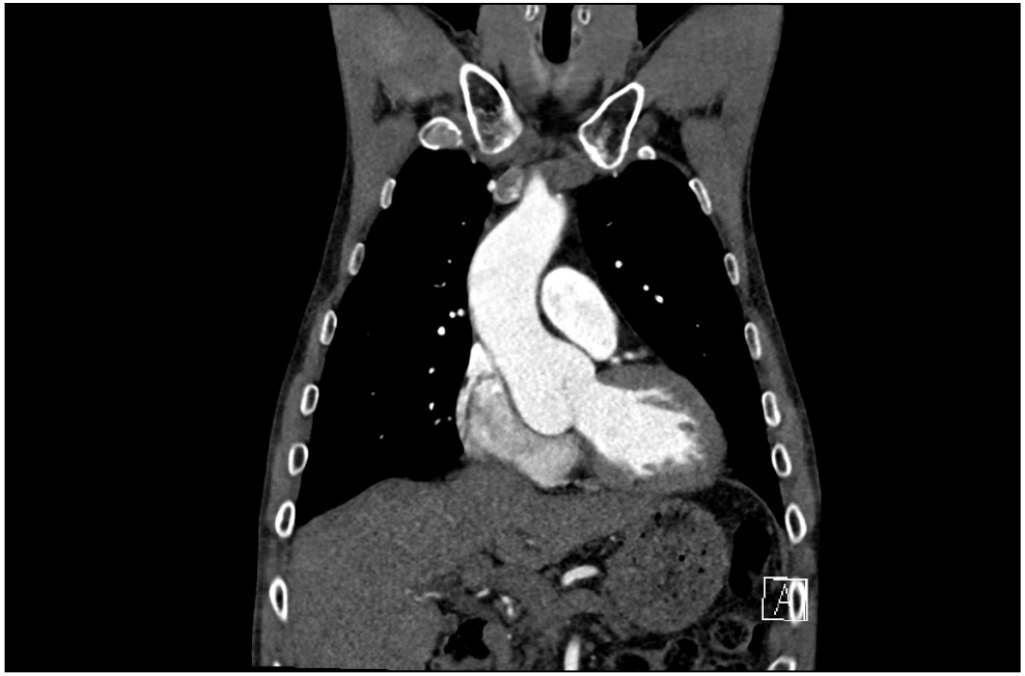


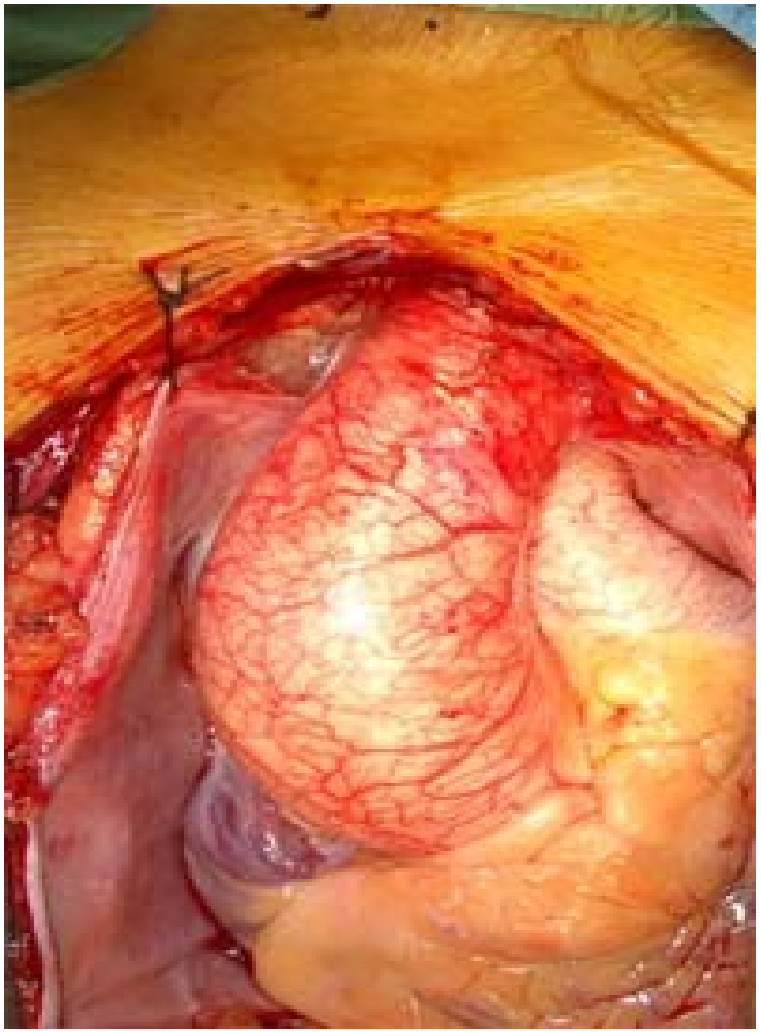
Figure 1 A. Patients' survival after aortic valve sparing operations; B. Survival after reimplantation and remodeling procedures

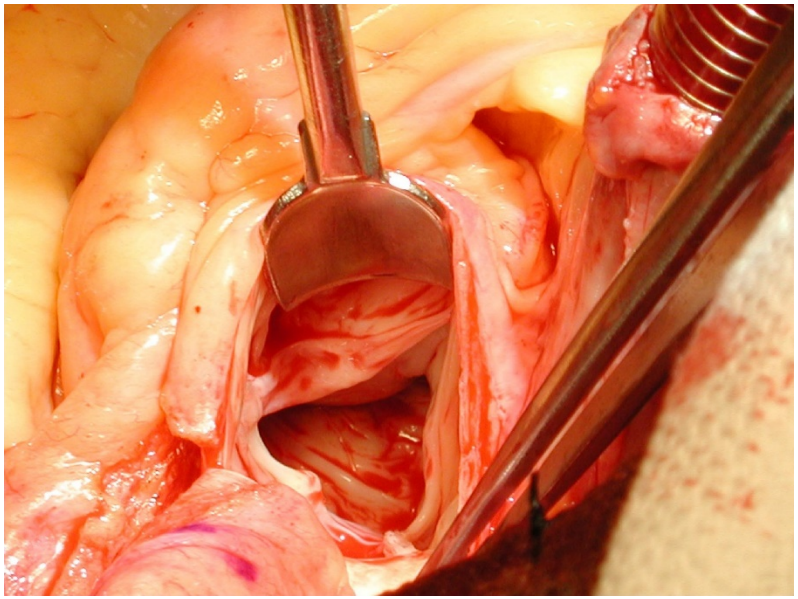
- 30 yr old female
- Strong family history of aortic complications from Marfan’s syndrome
- Serial CT scan showed enlargement to 5 cm at the aortic root
- Active, no symptoms
- One child with no peripartum complications

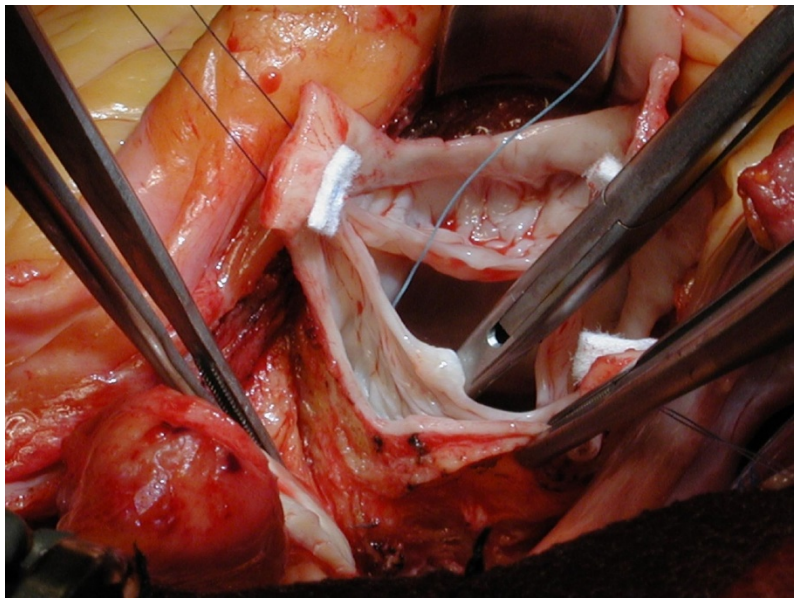


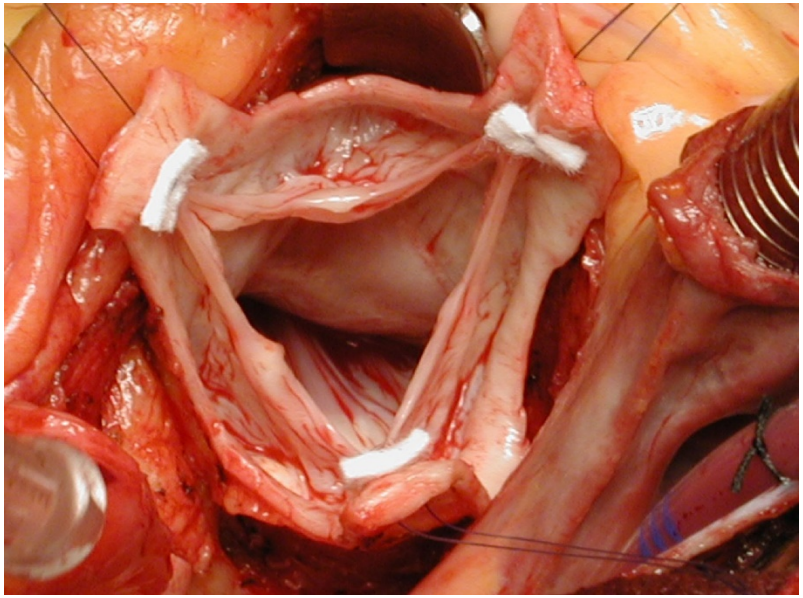


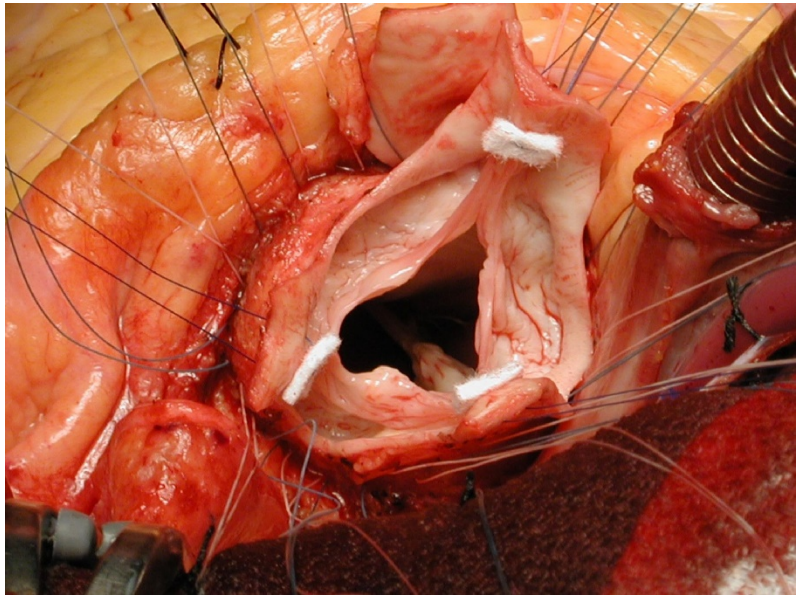

A

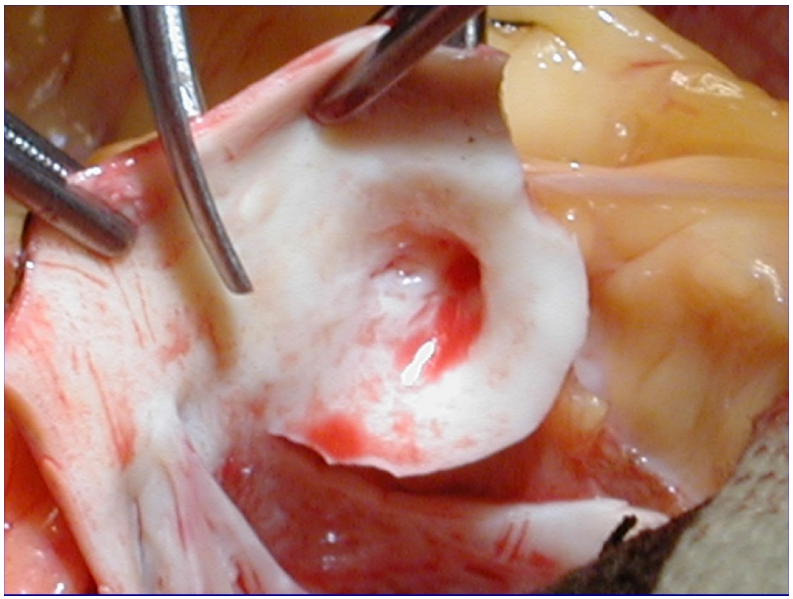


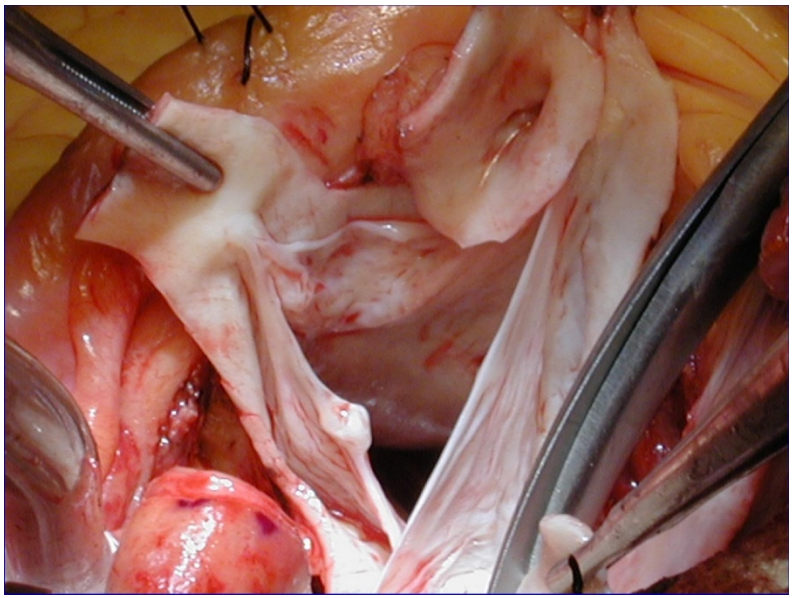


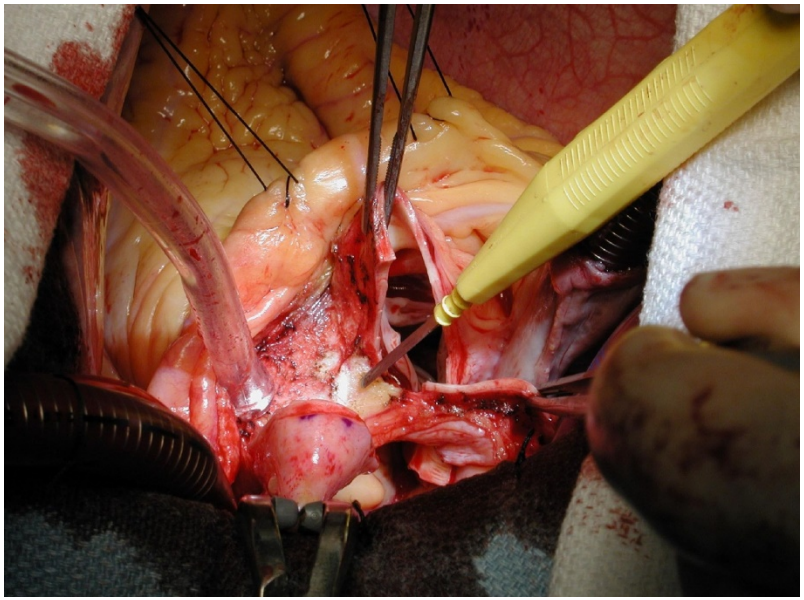


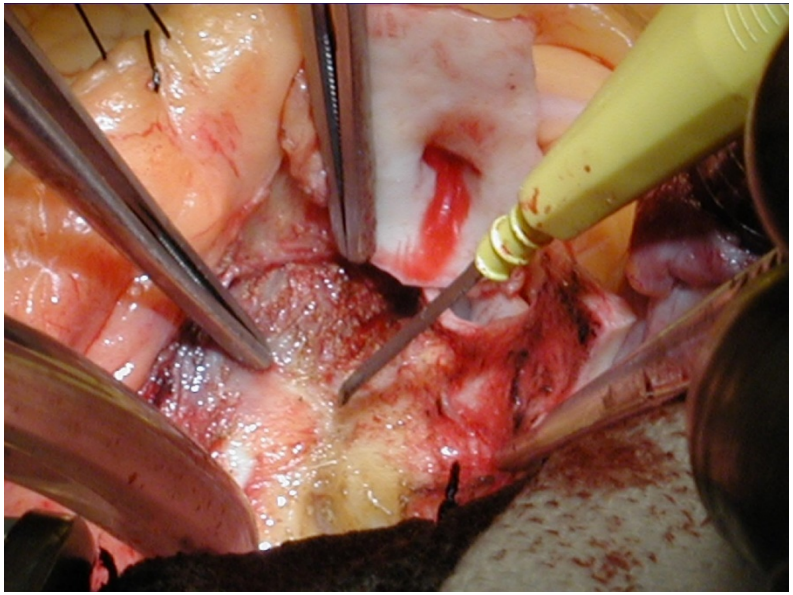


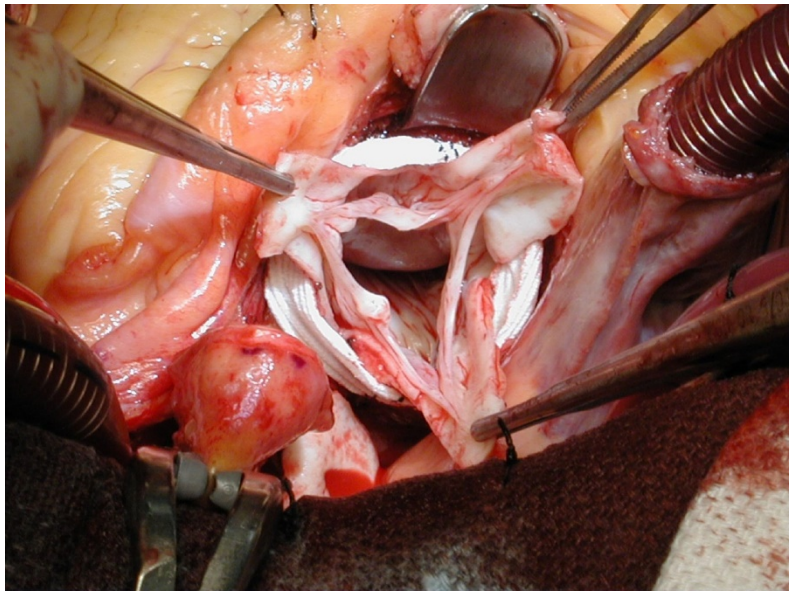


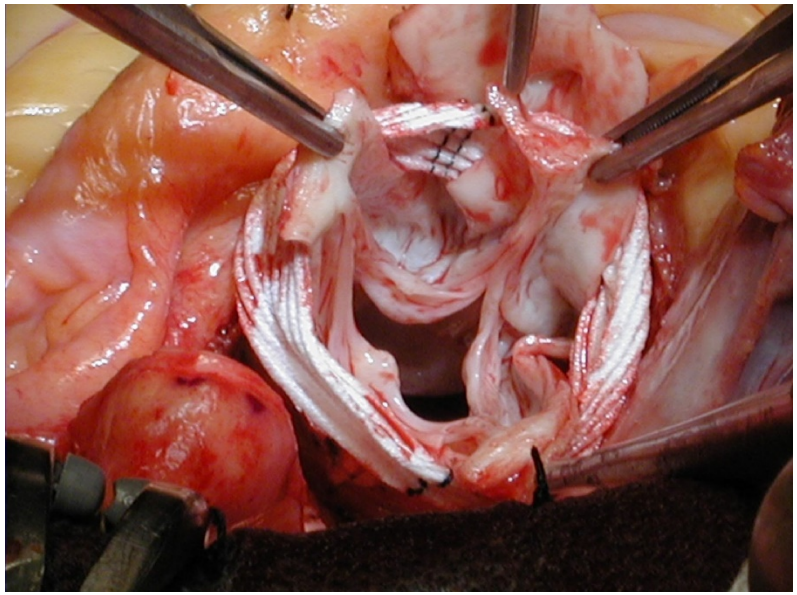


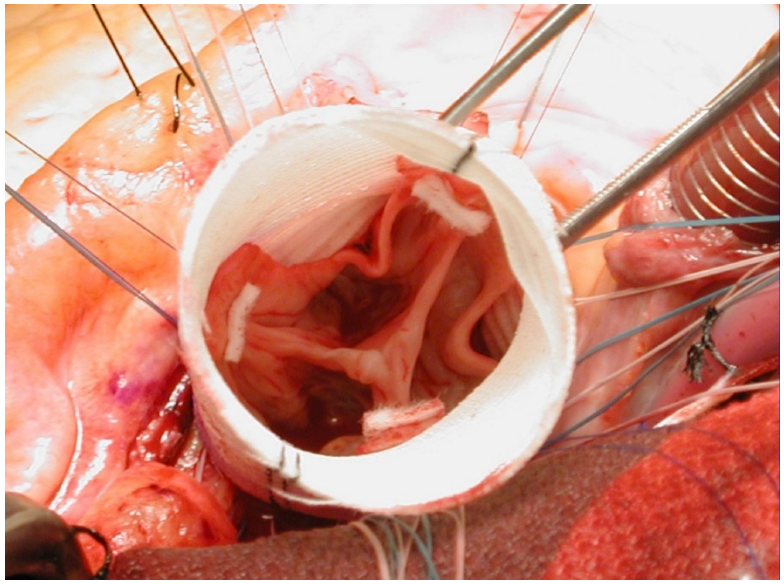


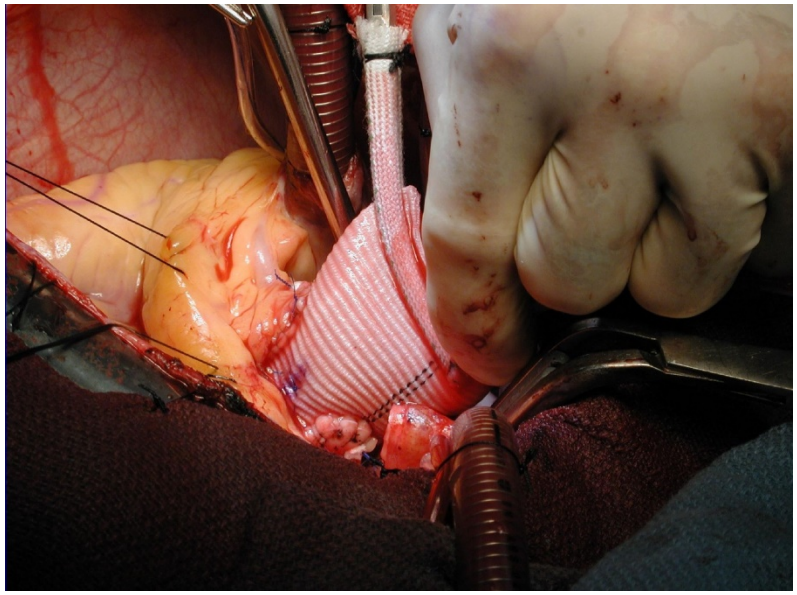


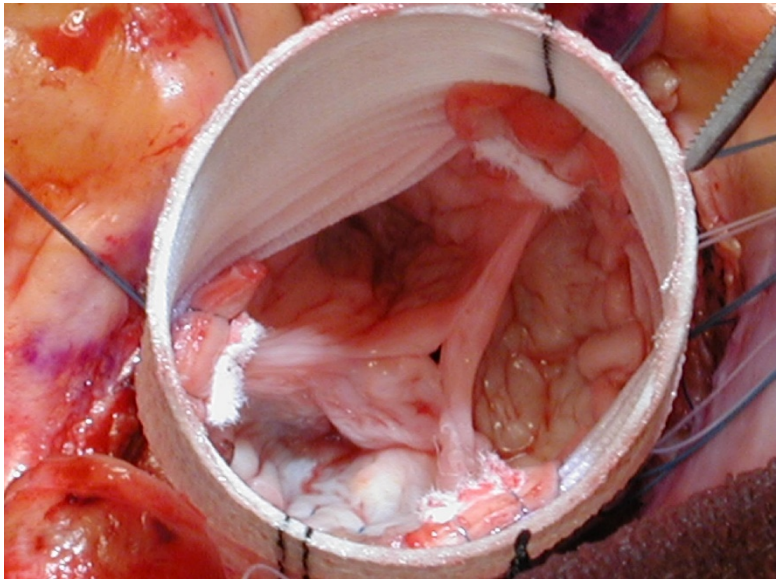


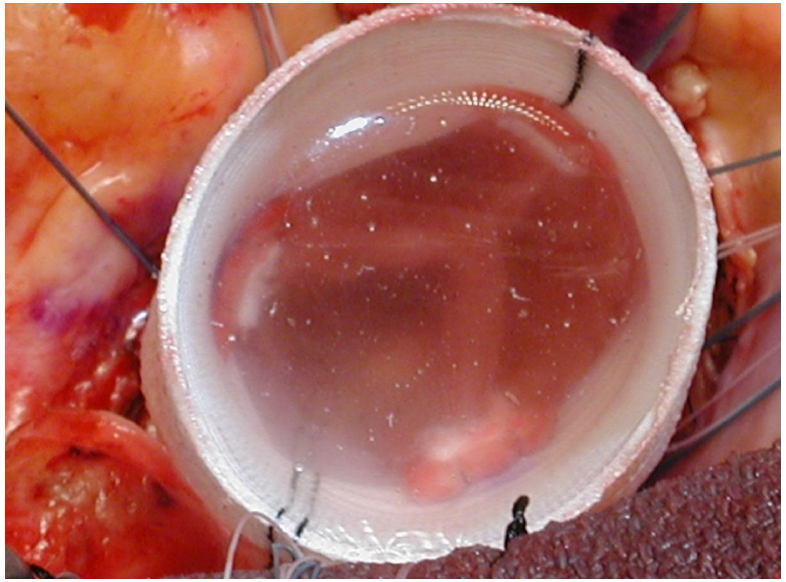


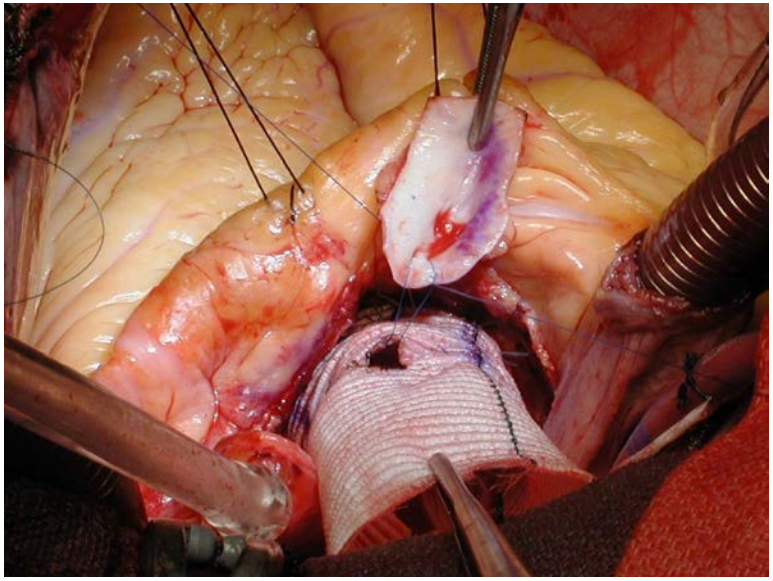


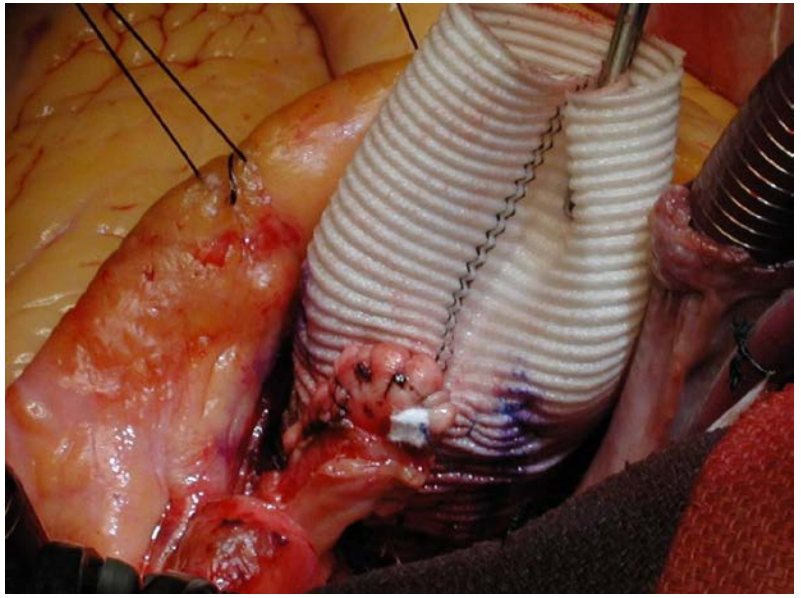


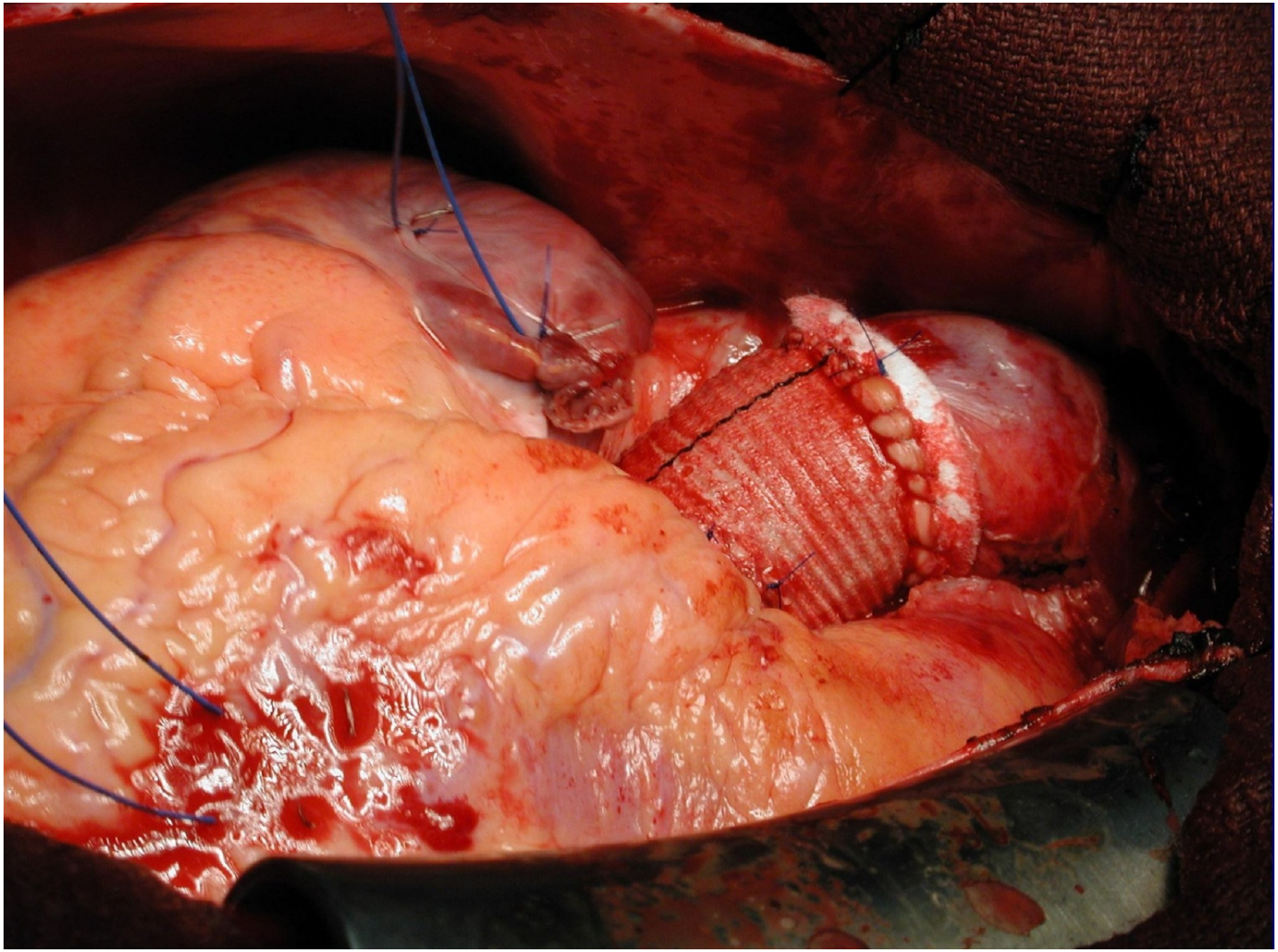


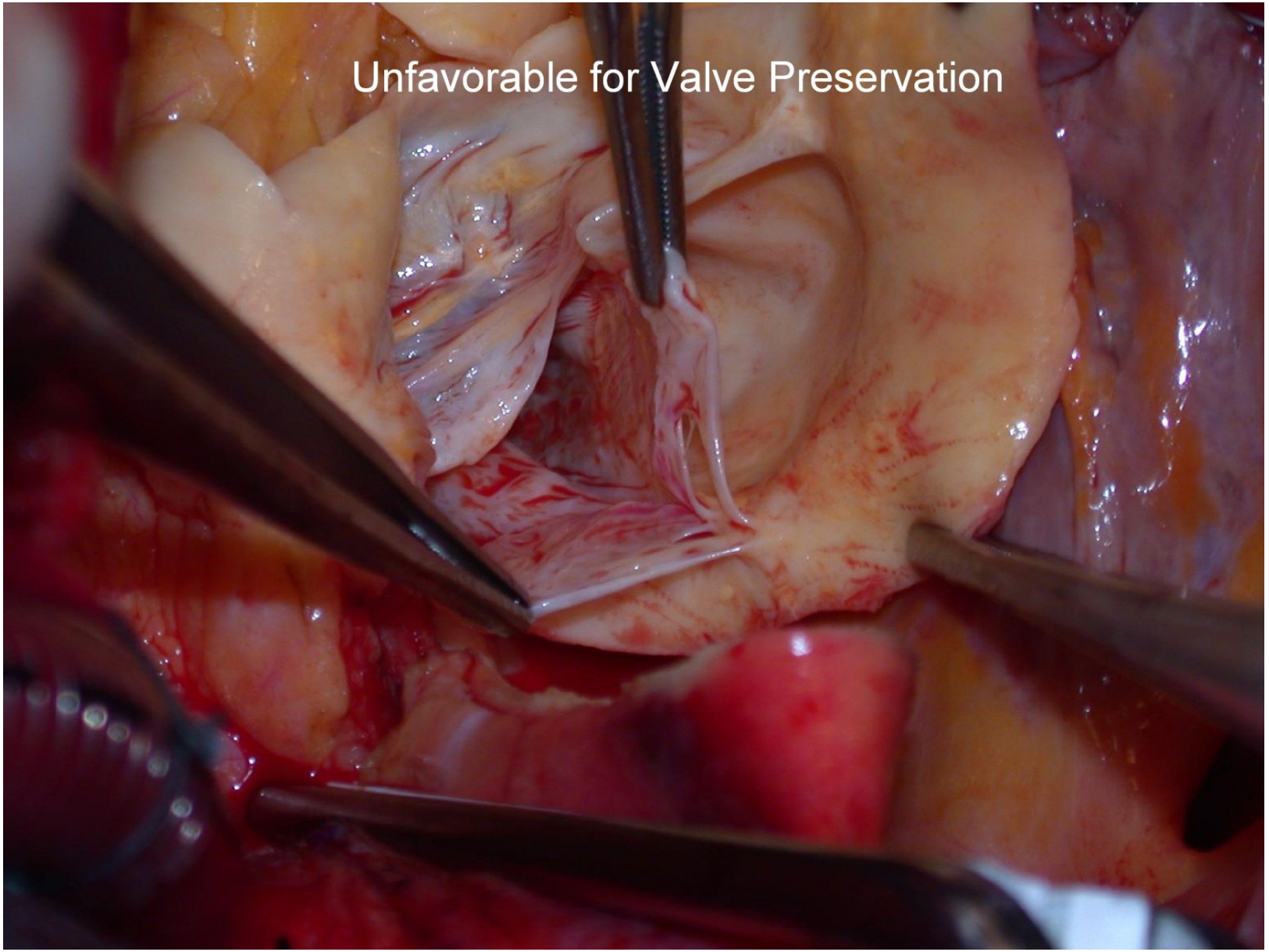


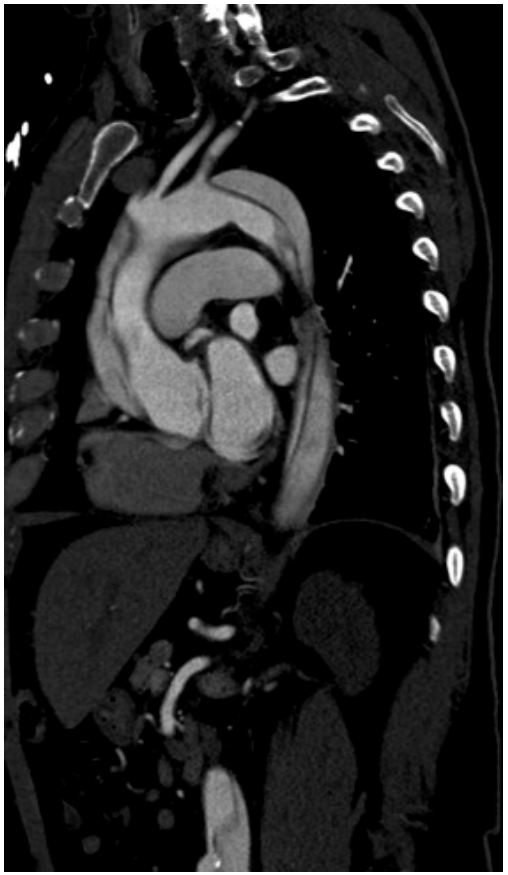


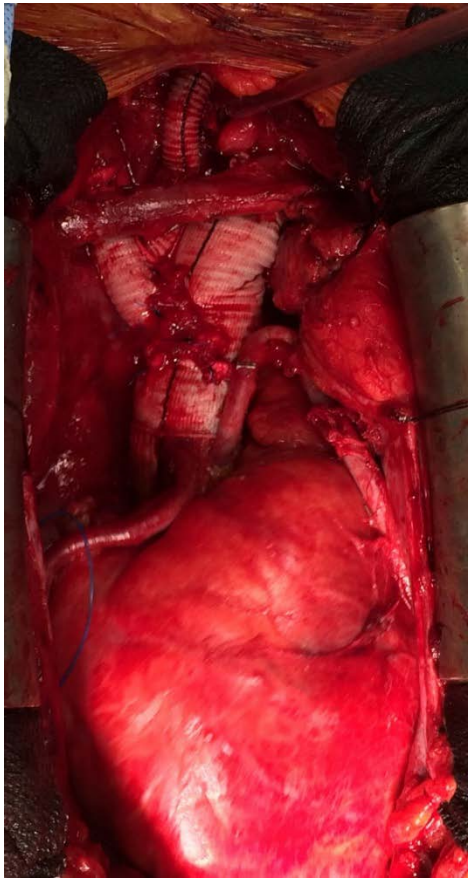







Unfavorable for Valve Preservation

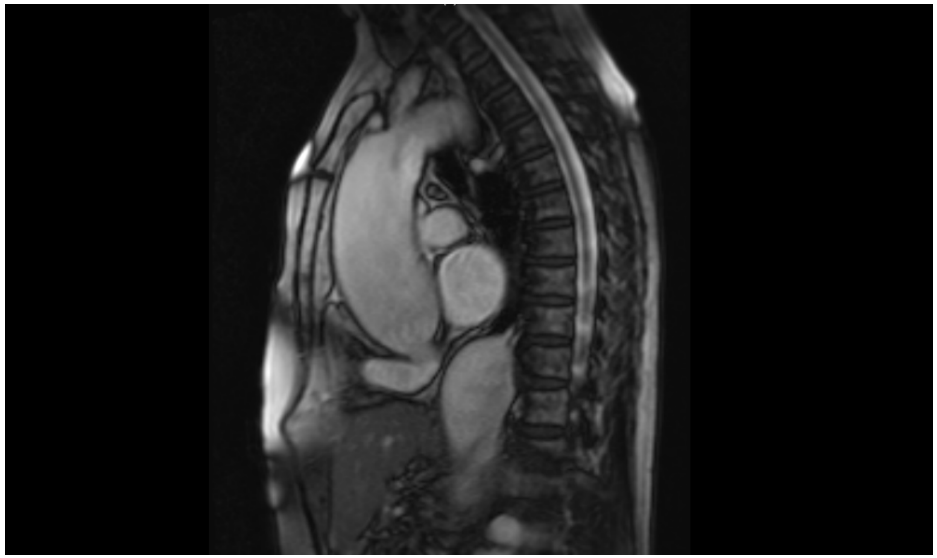


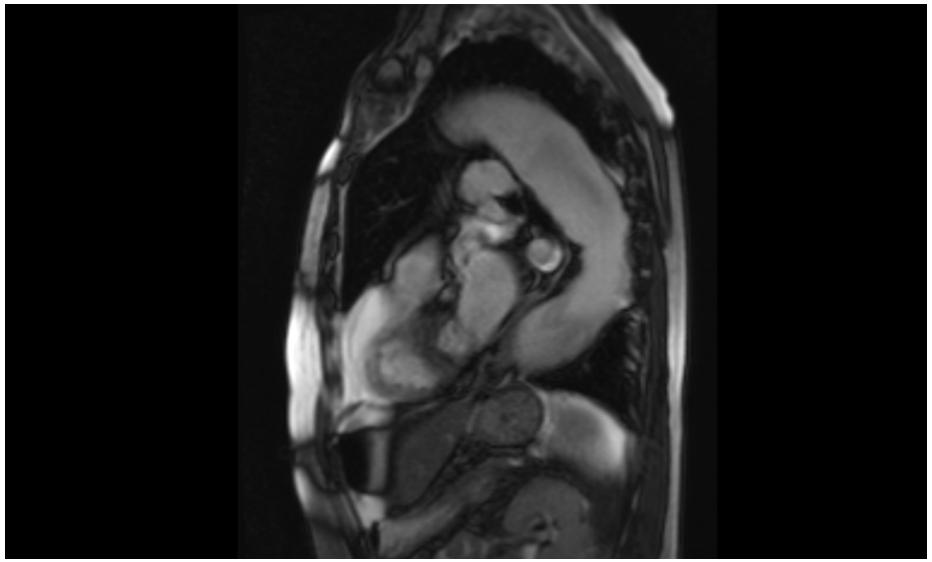


SURGERY OF THE AORTIC ARCH

Aortic arch aneurysm

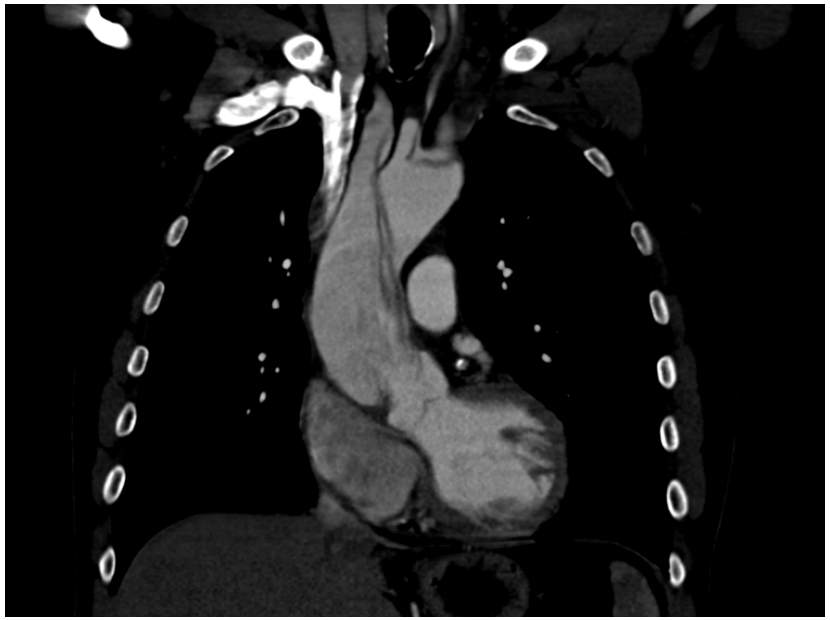
- 65 yr old male with upper chest discomfort
- No history of aortic syndromes
- 6.5 cm aortic arch at the level of left carotid
- Active

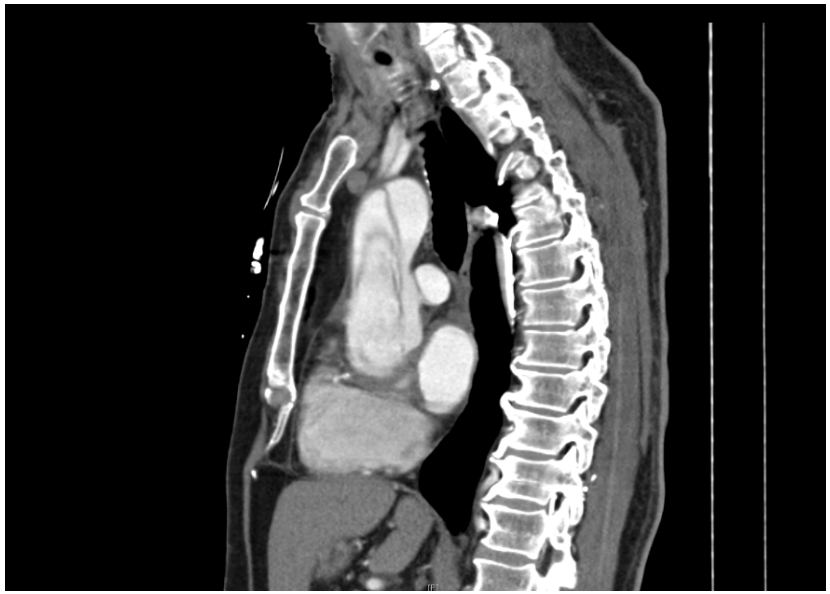


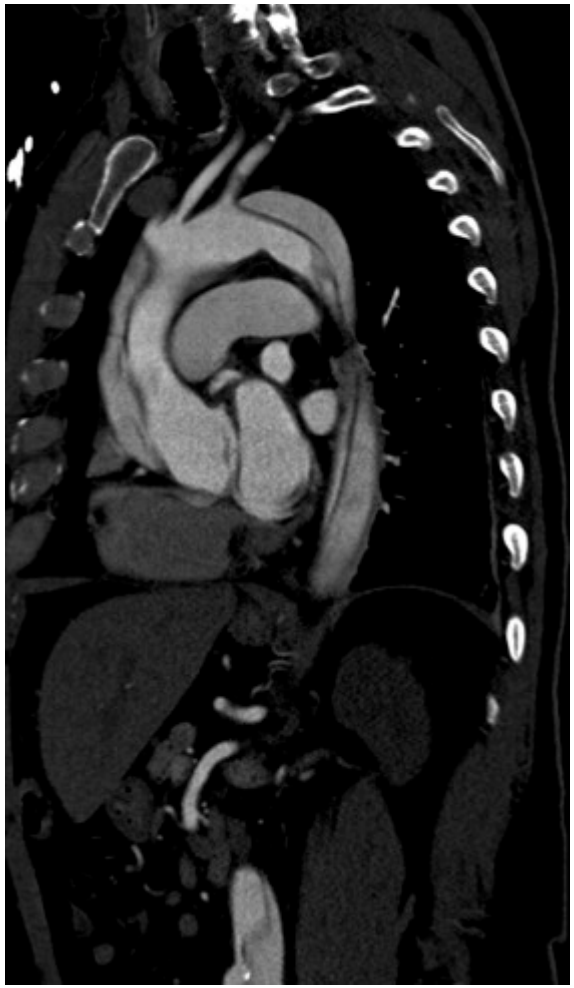


Patient 2

- 51 yr old
- Severe chest pain while moving furniture
- BP 170/90

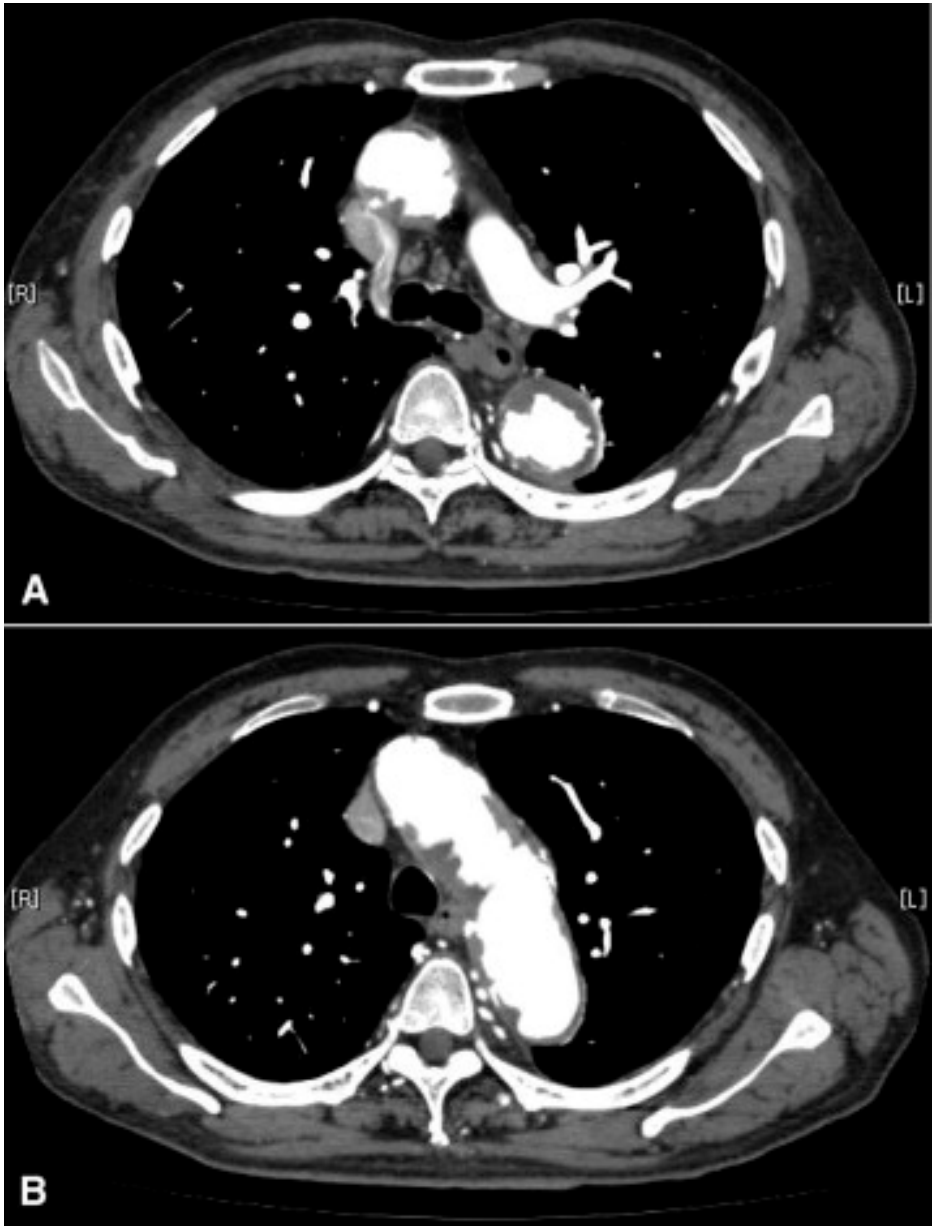


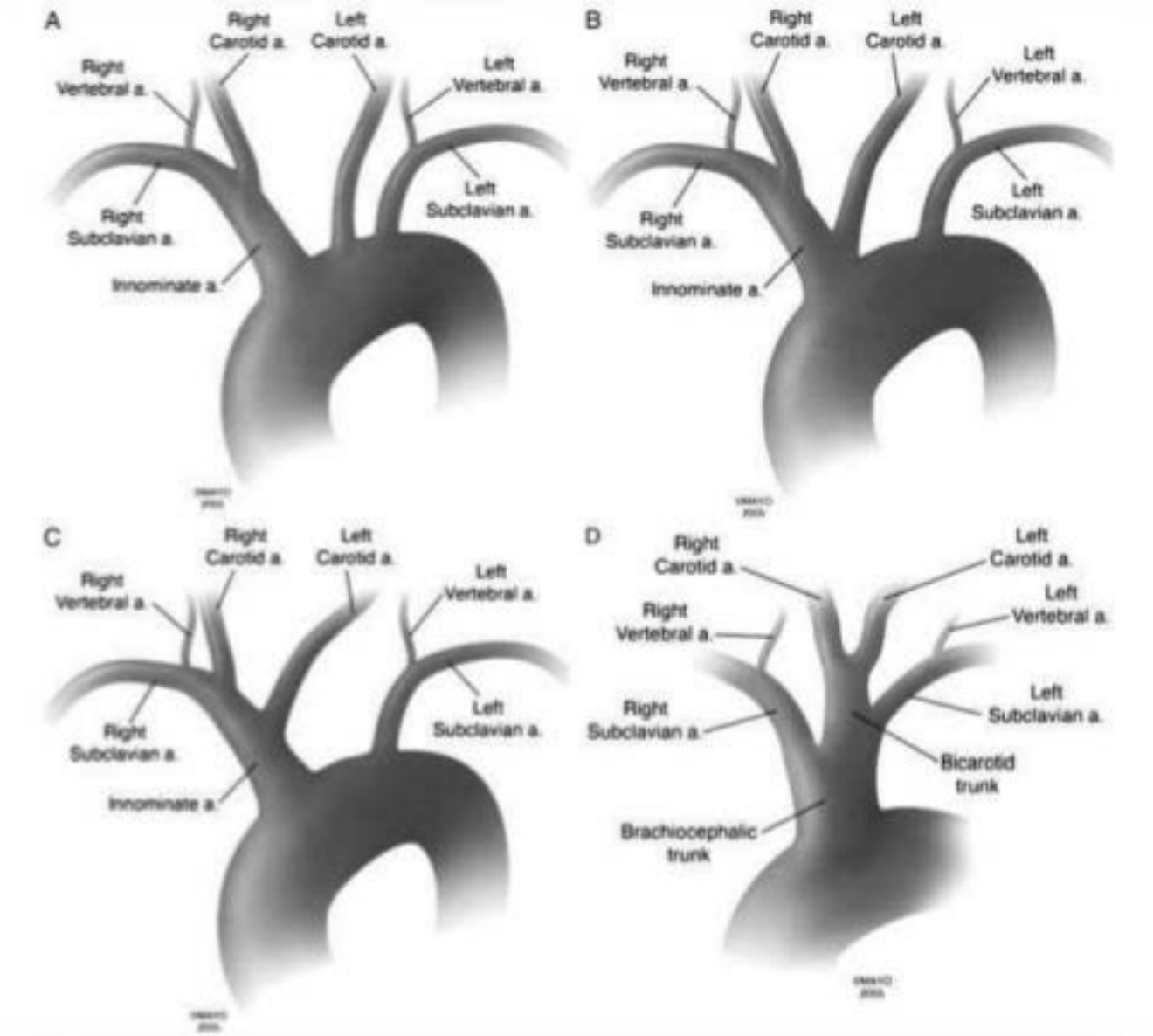




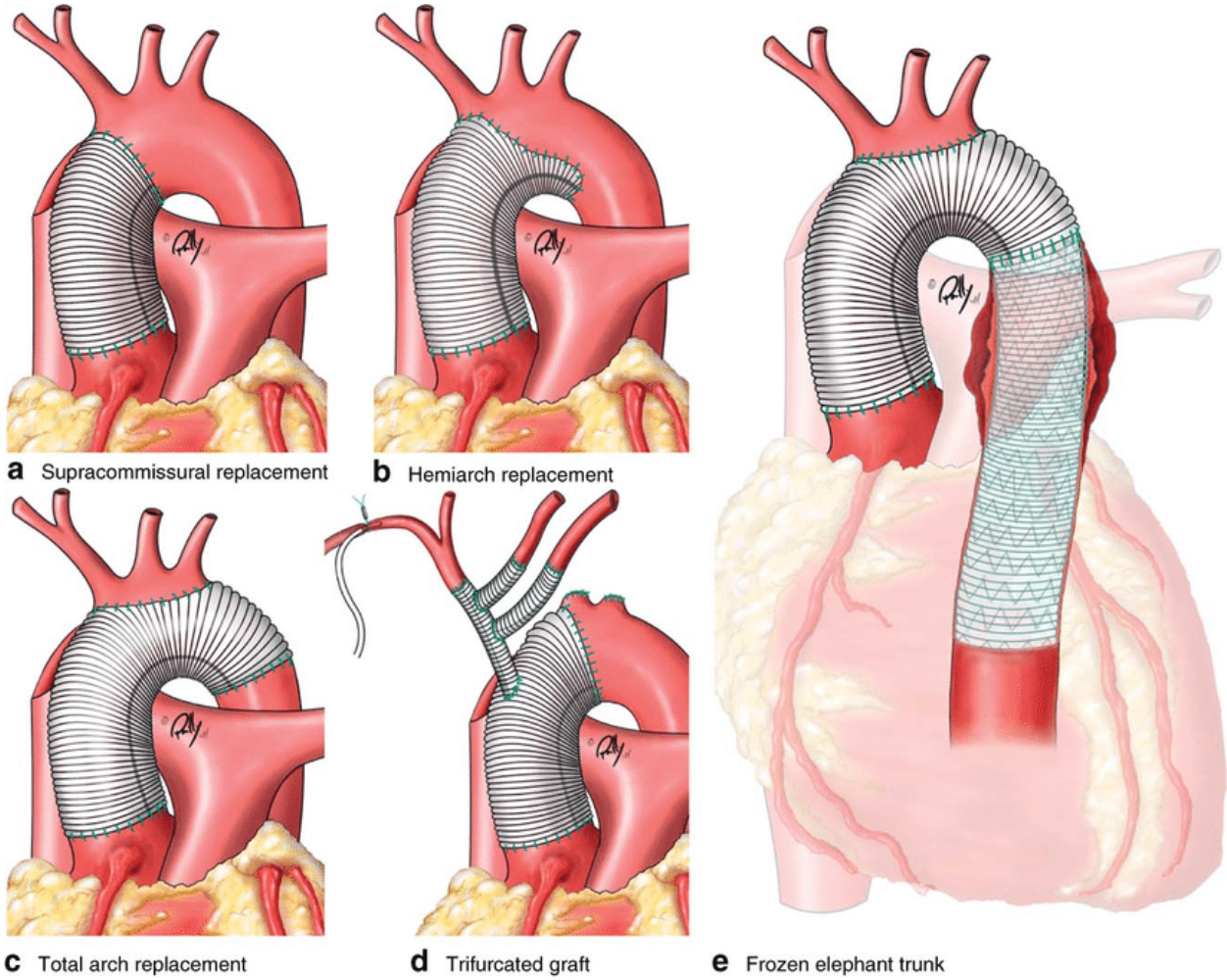
Patient 3

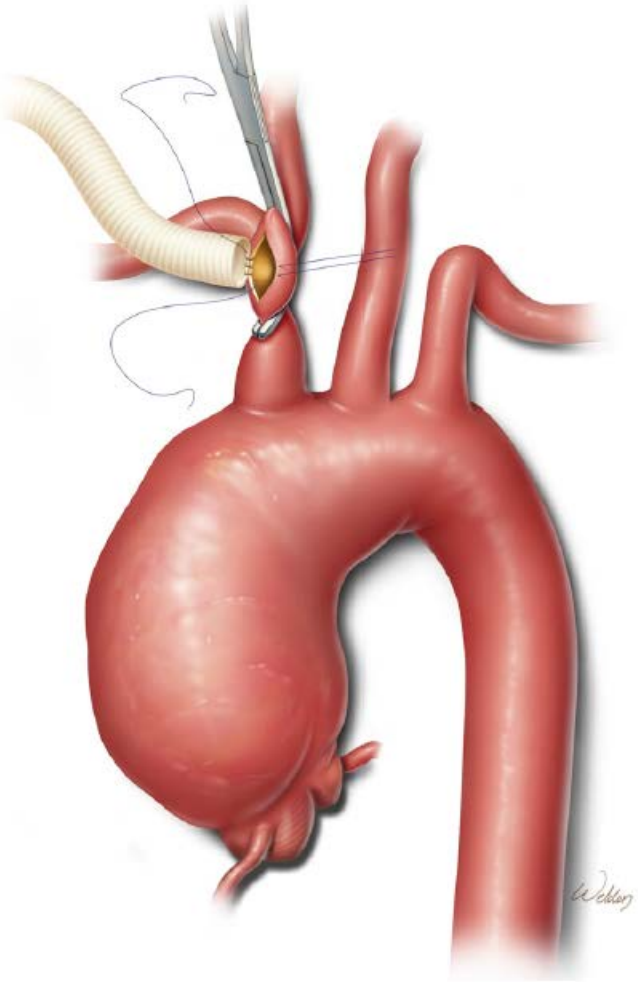
- 82 yr old gentleman
- Active
- Syncopal spell
- Stable hemodynamics

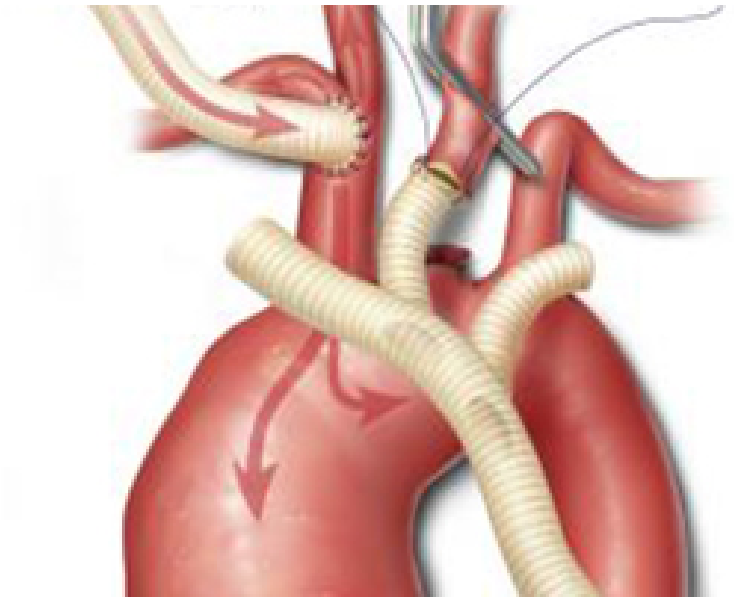

[R]
[L]
A
[R]
[L]
B

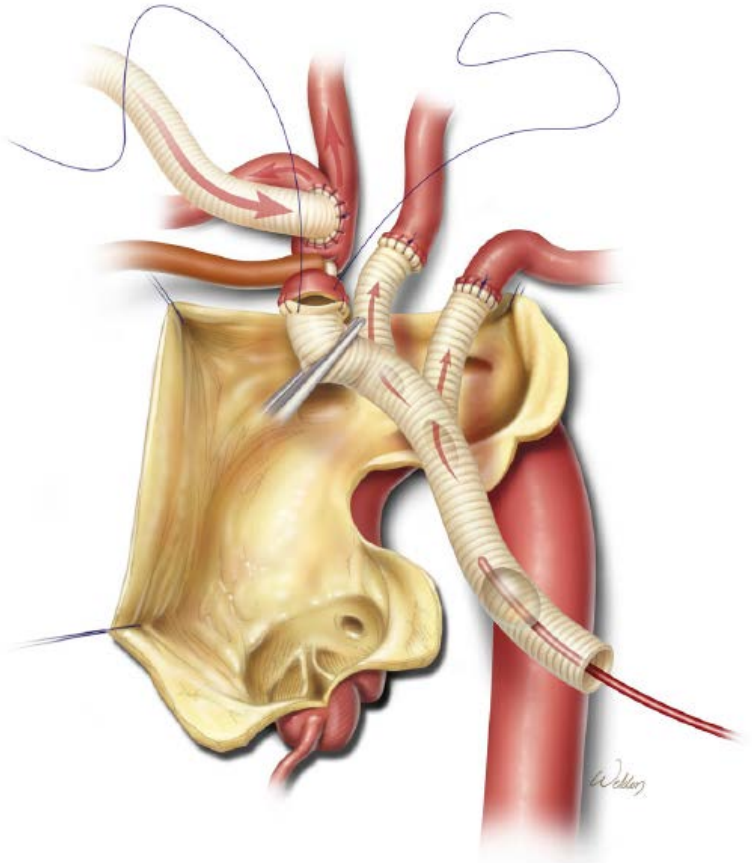

A
Right Carotid a.
Left Carotid a.
Right Vertebral a.
Left Vertebral a.
Right Subclavian a.
Left Subclavian a.
Innominate a.
B
Right Carotid a.
Left Carotid a.
Right Vertebral a.
Left Vertebral a.
Right Subclavian a.
Left Subclavian a.
Innominate a.
C
Right Carotid a.
Left Carotid a.
Right Vertebral a.
Left Vertebral a.
Right Subclavian a.
Left Subclavian a.
Innominate a.
D
Right Carotid a.
Left Carotid a.
Right Vertebral a.
Left Vertebral a.
Right Subclavian a.
Left Subclavian a.
Bicarotid trunk
Brachiocephalic trunk

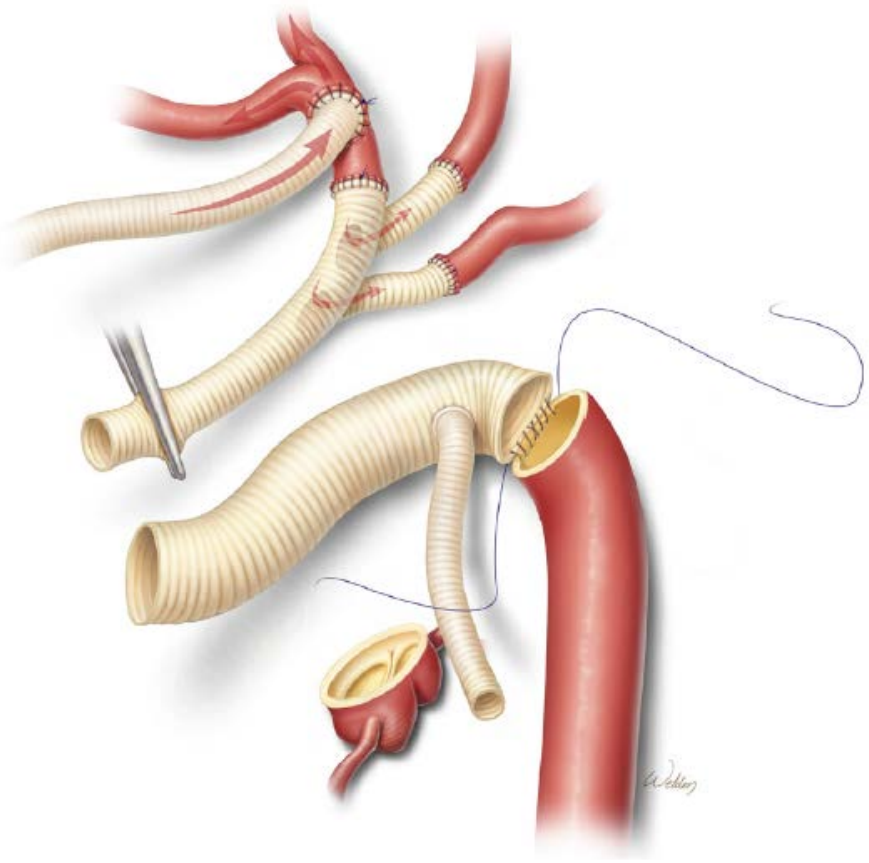
Options for Arch replacement

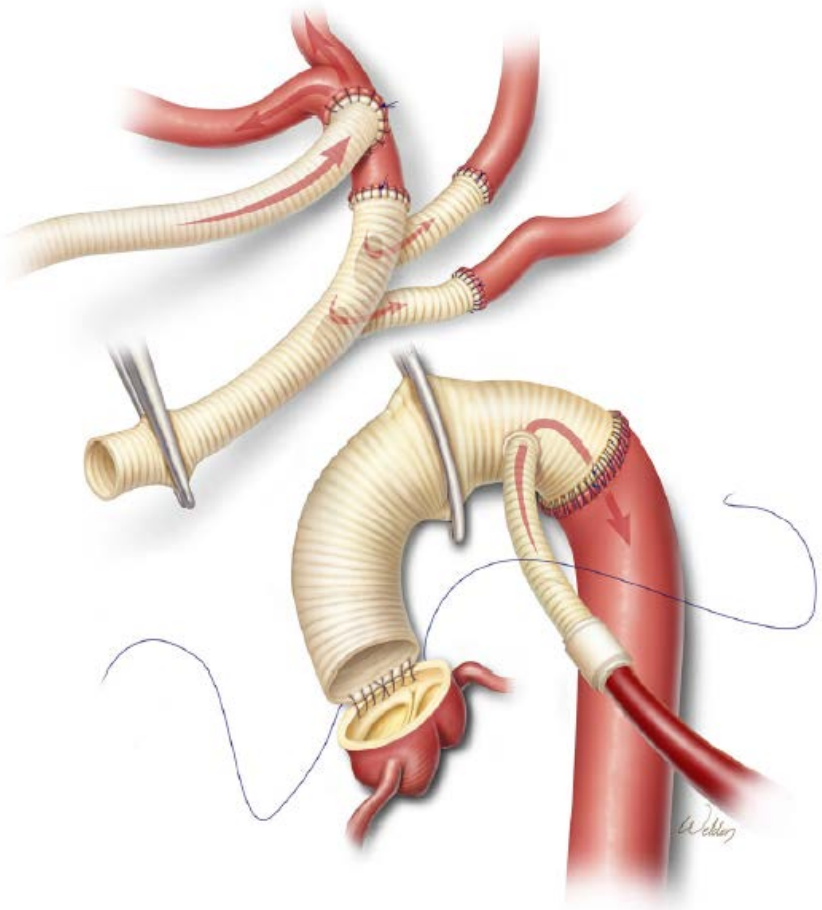


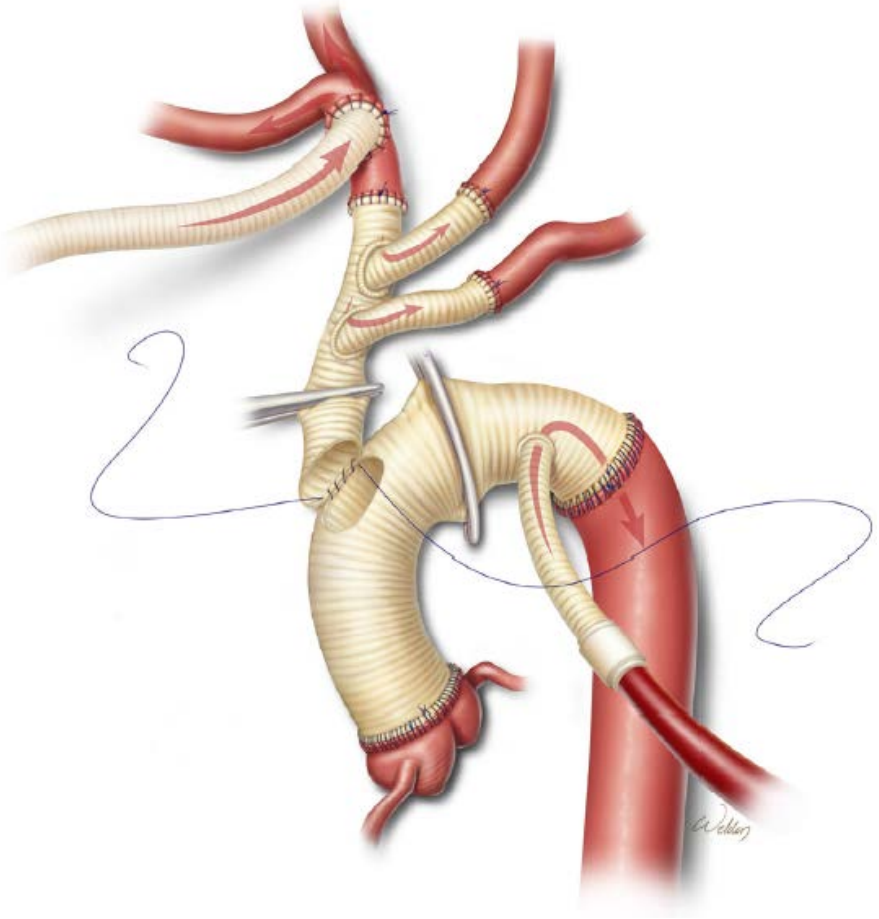


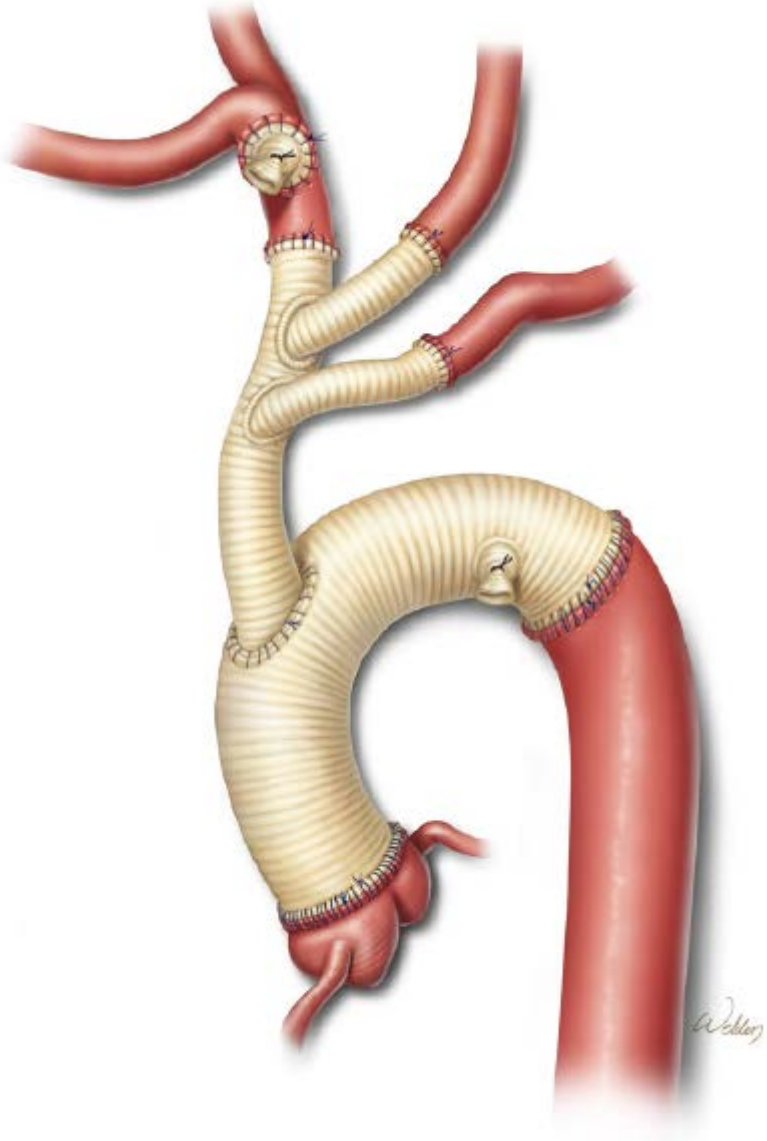


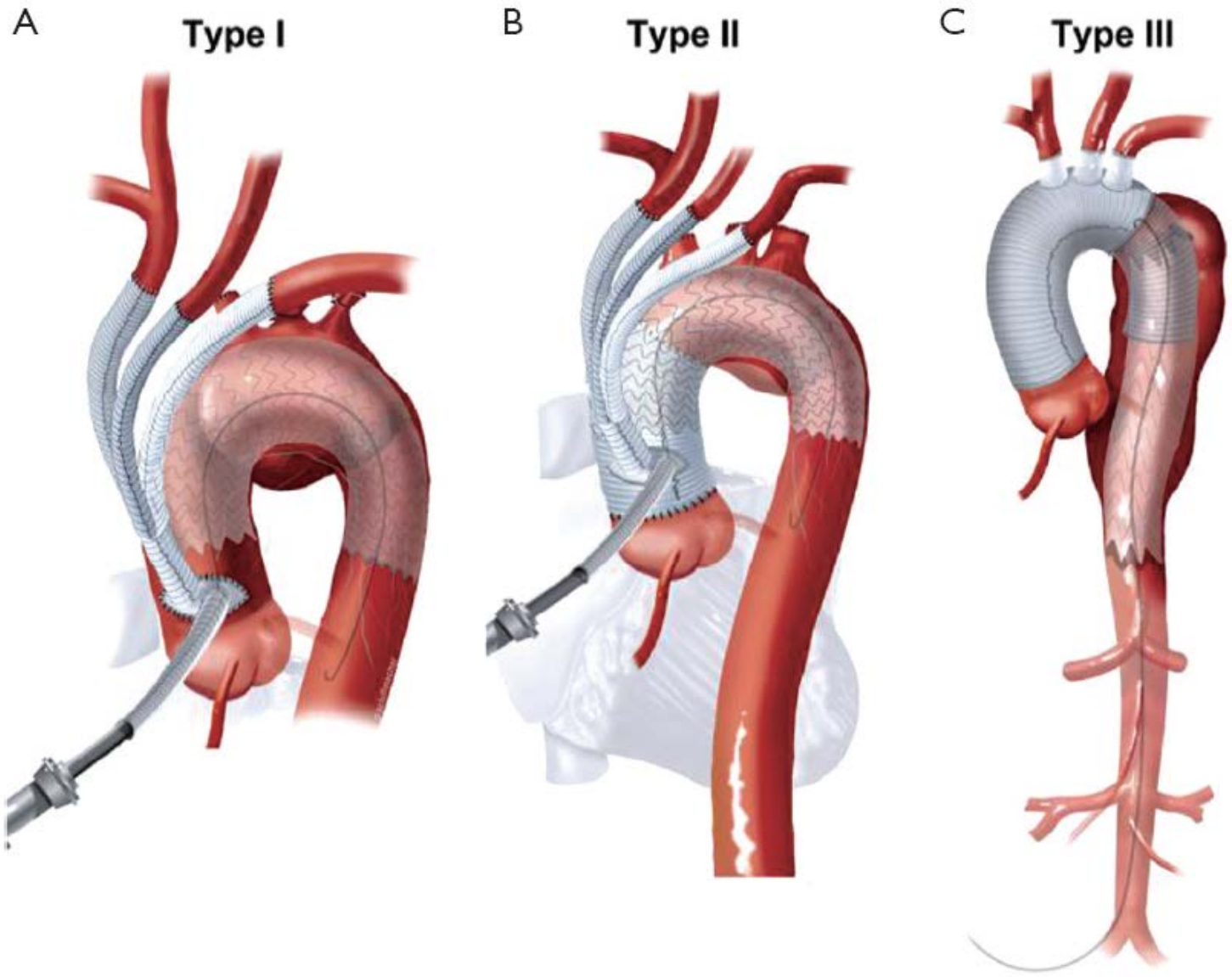











A
Type I
B
Type II
C
Type III

Evolution of Simplified Frozen Elephant Trunk Repair for Acute DeBakey Type I Dissection: Midterm Outcomes

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Background. A modified technique for frozen elephant trunk (FET) repair of acute DeBakey type I dissection has evolved. Procedural modifications are described and midterm outcomes evaluated.

Methods. From 2009 to 2016, 72 patients with DeBakey type I dissection underwent emergency simplified FET. Mean age was 59 ± 15 years. Presentation included malperfusion (n = 22, 31%), rupture (n = 12, 16%), and aortic insufficiency (n = 42, 58%). Concomitant procedures included valve replacement (n = 9), root replacement (n = 11; valve sparing n = 6), cusp repair (n = 11), and valve resuspension (n = 21). The first 39 were treated by modifying an early generation stent graft. The next 16 received newer modified stent grafts, and the latest 17 underwent branched single anastomosis technique with left subclavian stent grafting.

Results. Operative mortality was 4.2% (n = 3 of 72). Two presented comatose without recovering, the other died from coagulopathy complications. Morbidity included stroke (n = 3, 4.2%), spinal injury (n = 3, 4.2%; 1 permanent), tracheostomy (n = 7, 9.7%), and renal failure (n = 2, 2.8%). Median follow-up was 28 ± 25 months. Survival was 92% at 6 months, 92% at 1 year, 89% at 3 years, and 80% at 5 years. Among 69 survivors, follow-up imaging was available in 63 (91%). Of these, 58 (92%) patients thrombosed the treated false lumen, with shrinkage in 37(54%) patients from 42 ± 8 mm to 37 ± 7 mm. Ten patients underwent 14 late reinterventions for growth and incomplete thrombosis (7 endo extension, 4 left subclavian embolization, 1 bypass, 2 false lumen embolization). Freedom from reintervention was 93% at 6 months, 87% at 1 year, 77% at 3 years, and 72% at 5 years.

Conclusions. Simplified FET for treating acute DeBakey type I dissection has evolved and remained safe. It promotes aortic remodeling, and simplifies management of chronic aortic complications.

(Ann Thorac Surg 2018;105:749–56)

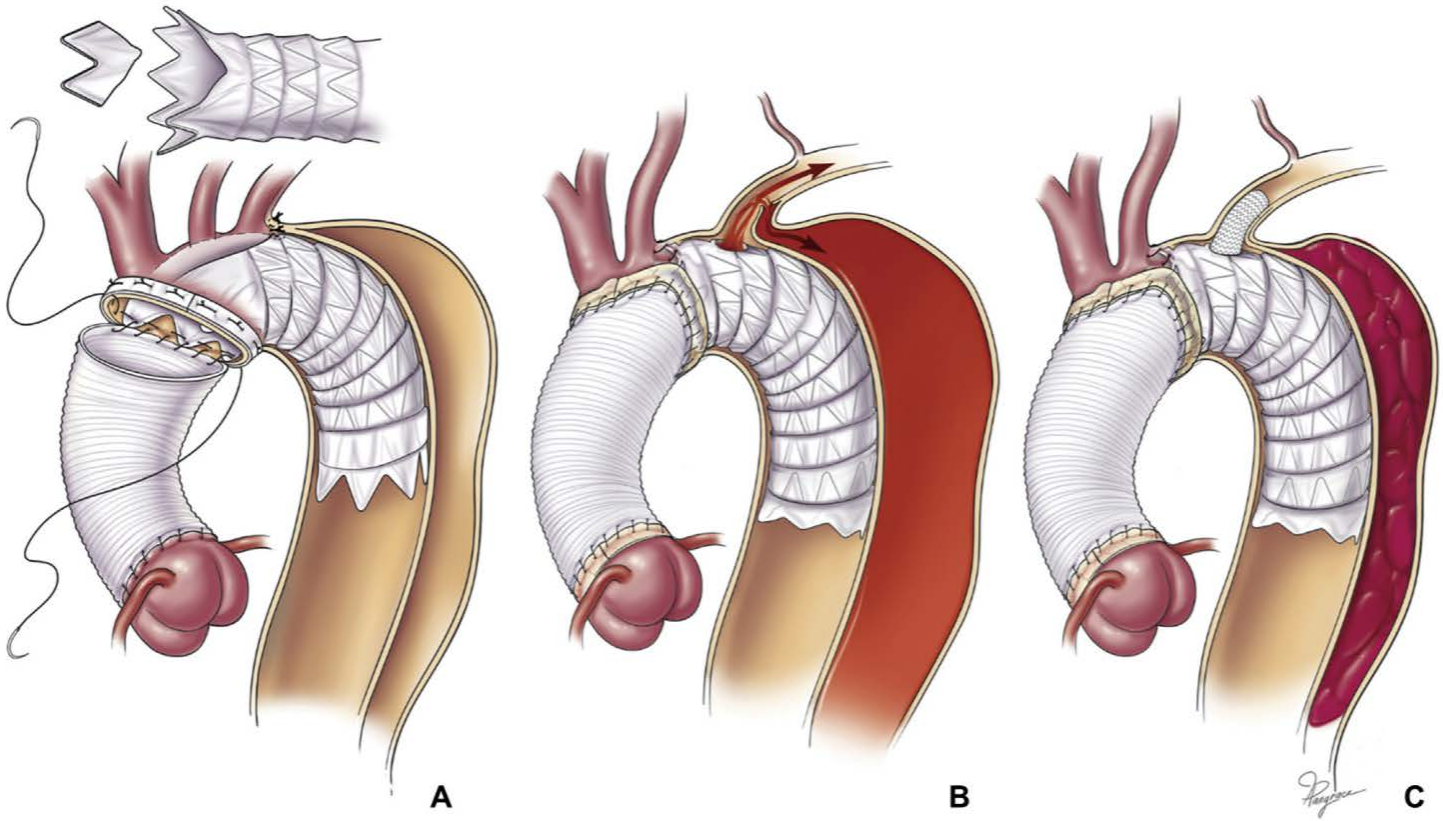

Fig 1. Evolution of the simplified frozen elephant trunk technique. (A) Intraoperative creation of fenestration by resection of a portion of proximal stent graft (2009). (B) Fenestration below the left subclavian artery (2012). (C) Direct branch vessel stent grafting (2015).

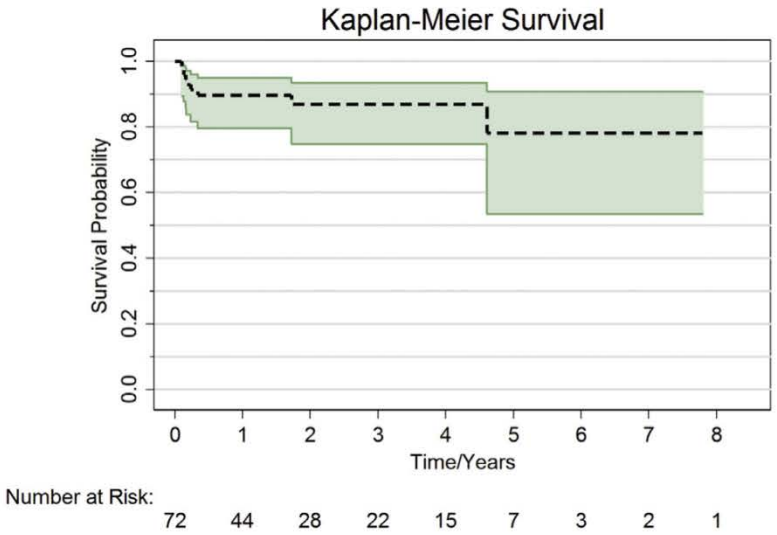

Fig 3. Survival was 92% at 6 months, 92% at 1 year, 89% at 3 years, and 80% at 5 years.

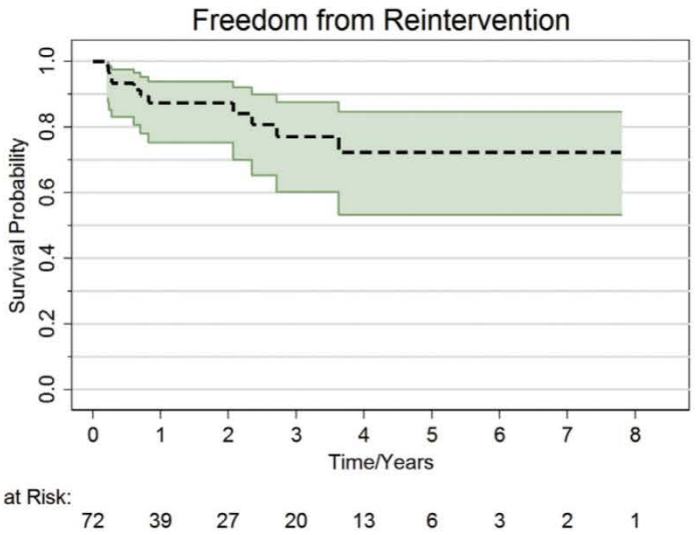

Fig 4. Freedom from reintervention was 93% at 6 months, 87% at 1 year, 77% at 3 years, and 72% at 5 years.

Masters of Cardiothoracic Surgery

Total endograft replacement of aortic arch

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Total endovascular replacement of the aortic arch is a complex procedure that is often favoured when the pathology anatomy precludes a standard median sternotomy. Here we present the case of endograft repair in a 79 year old male with 6.5 cm arch aneurysm and 5.4 cm descending thoracoabdominal aneurysm. Following bilateral carotid-subclavian bypasses, a long 7 Fr sheath was advanced into the descending aorta through the common iliac artery purse string. A double curved long Lunderquist wire was guided to deep within the left ventricle, and the endograft carefully advanced over the wire. The graft was radiologically orientated, and deployed under asystolic conditions. Retrograde cannulation of the branches were accomplished, with carotid sheath placed into the branches followed by bridging stents. The graft delivery system was then removed. This approach obviates the need for a sternotomy, cumbersome extra-anatomic debranching, and hypothermic circulatory arrest.

Keywords: TEVAR; total arch replacement; endograft; case report

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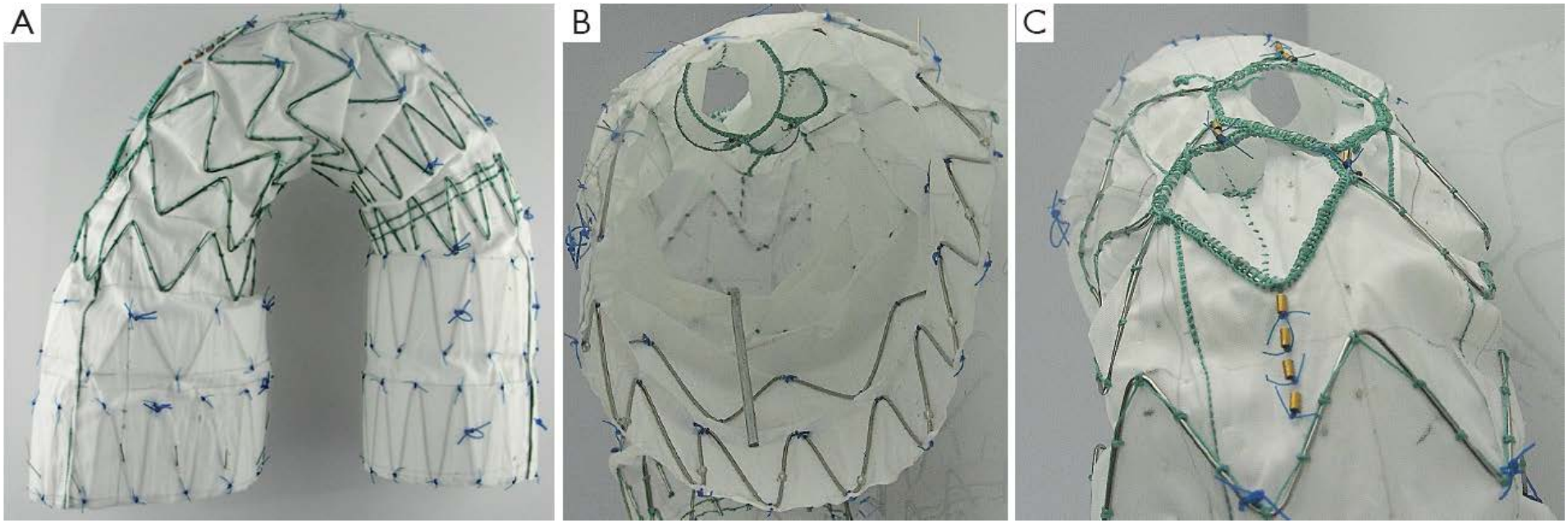
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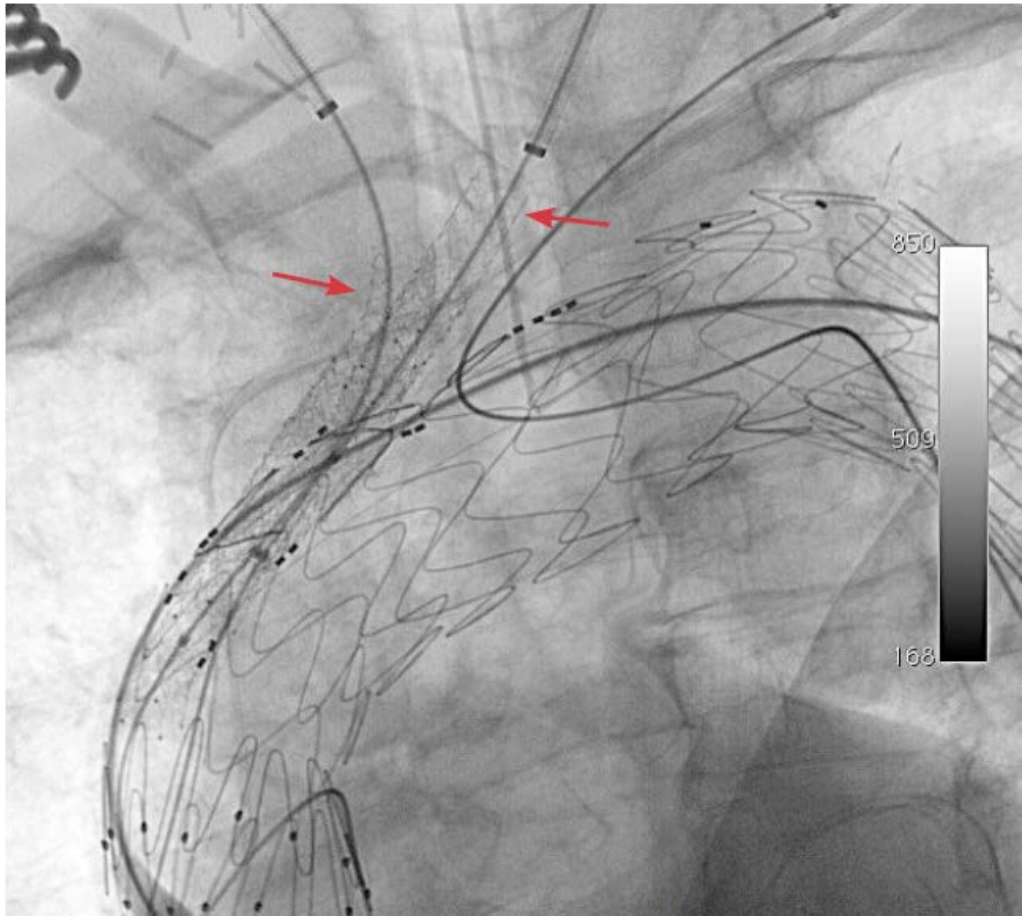
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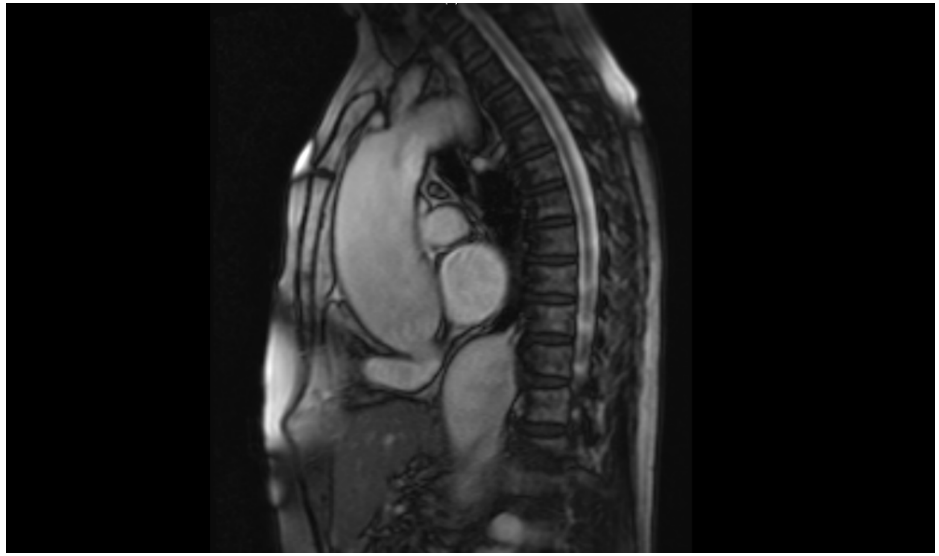
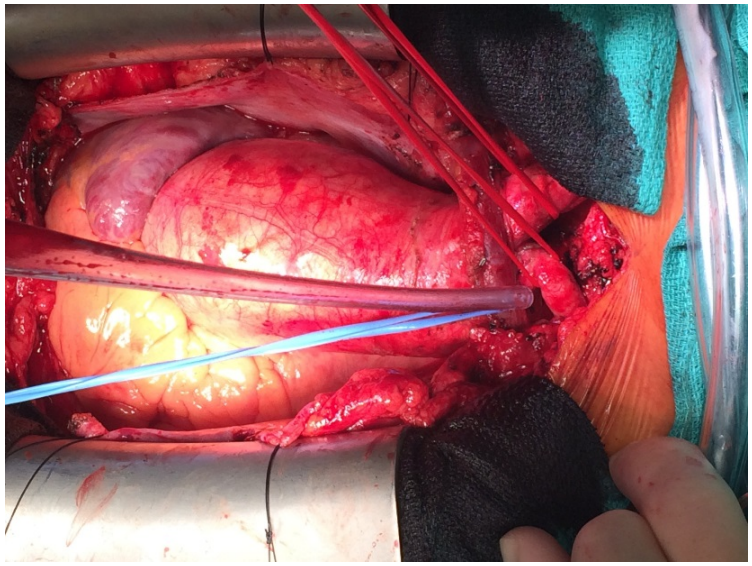
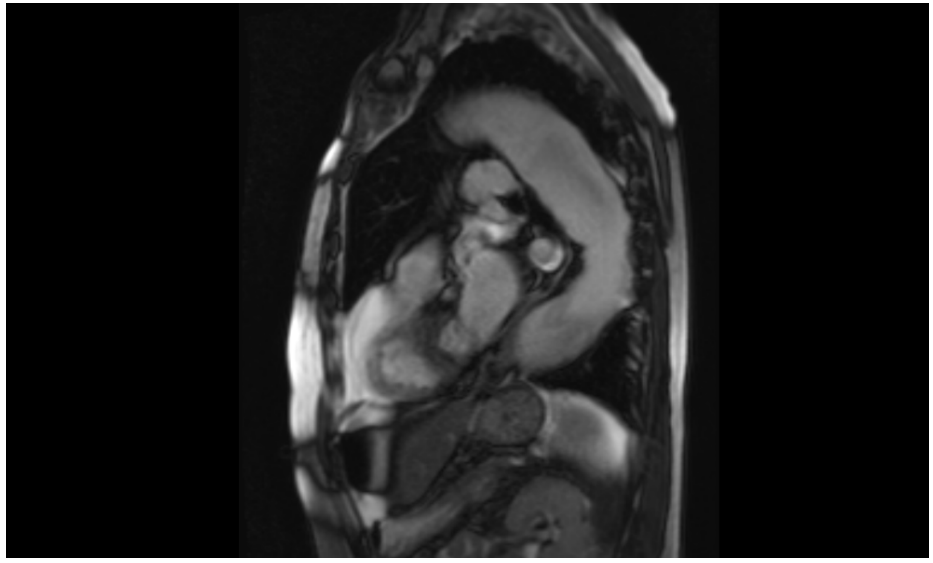
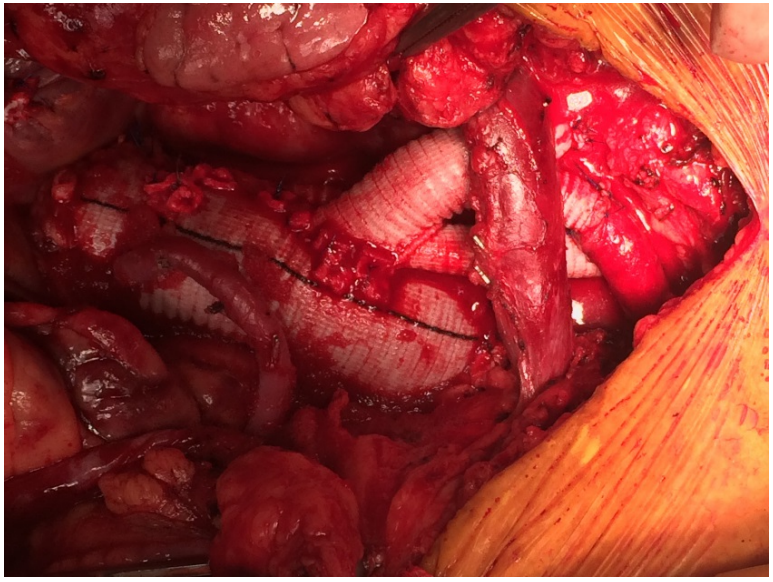
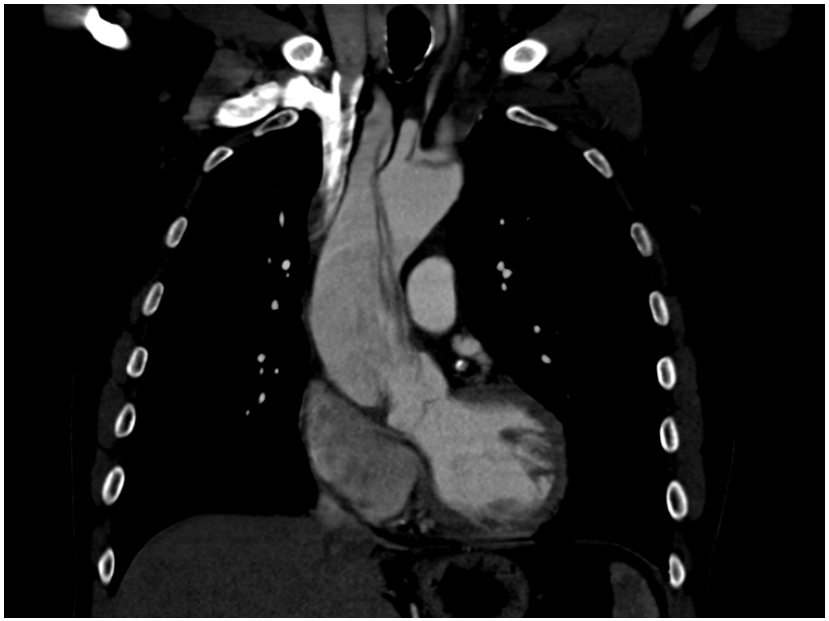
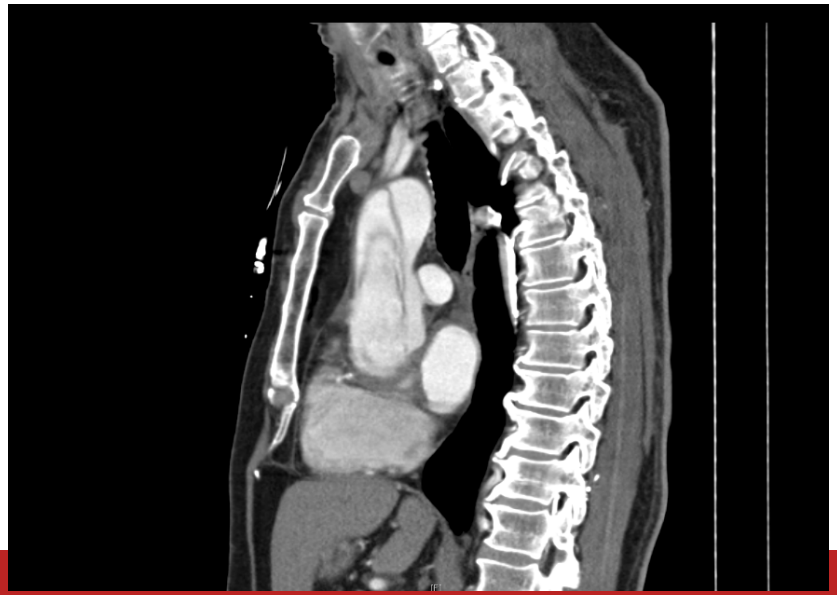


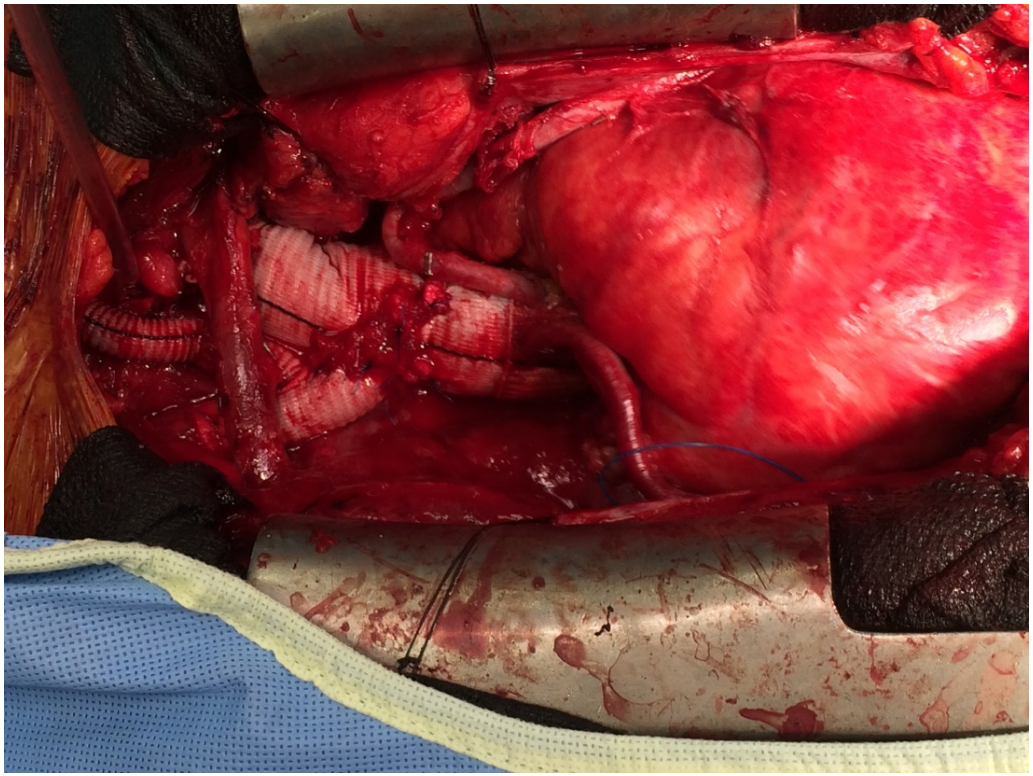


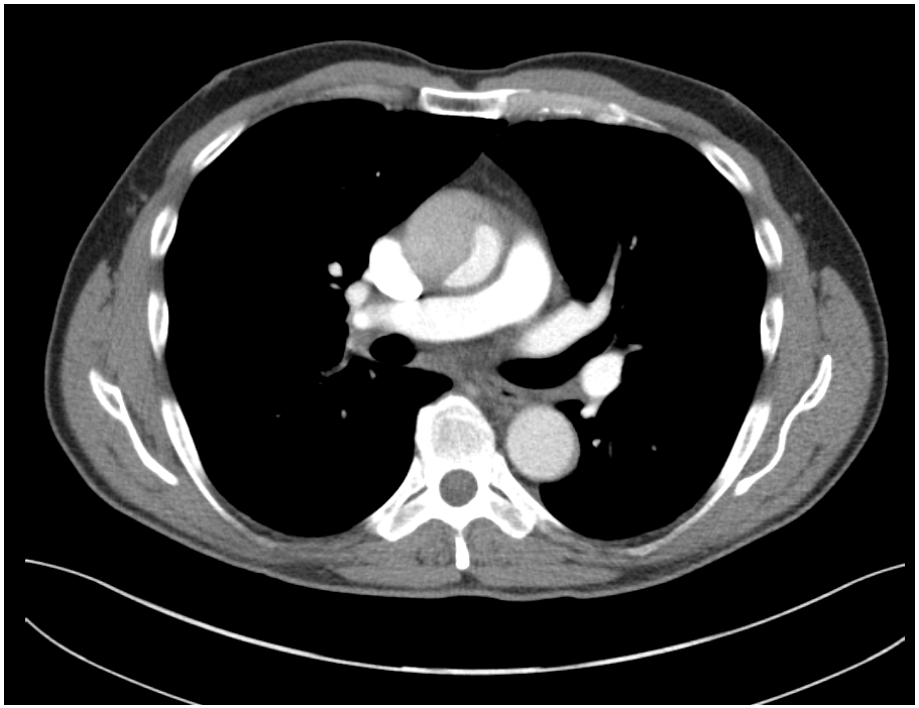


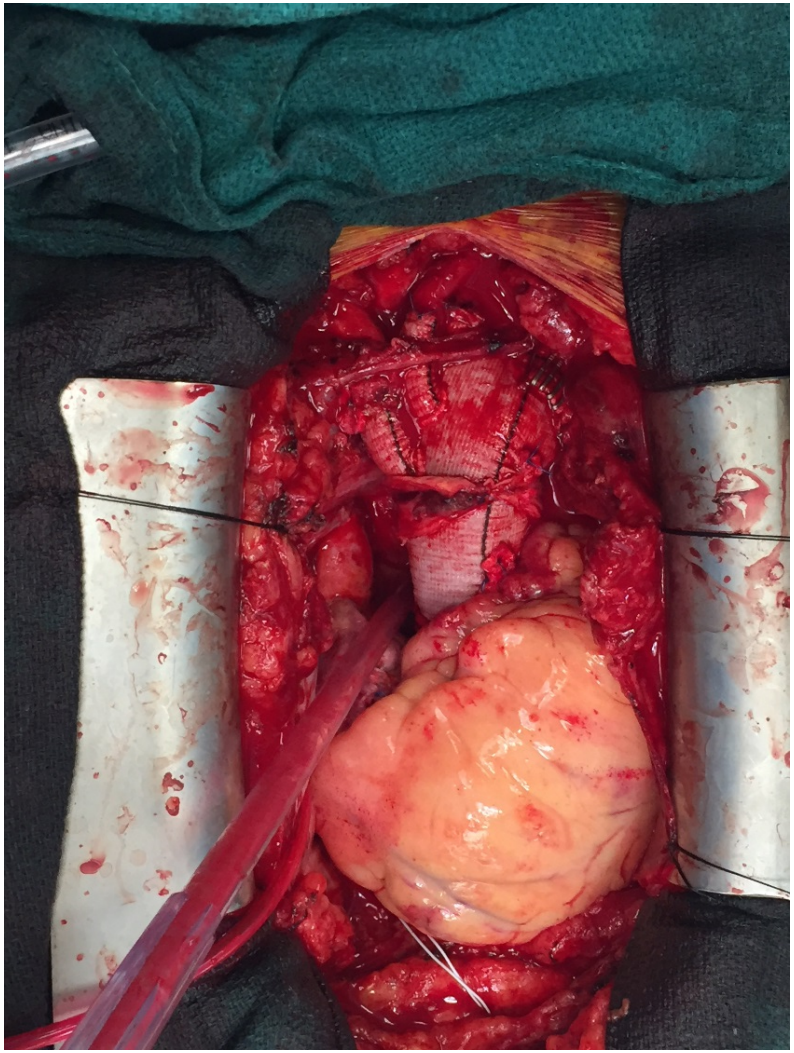


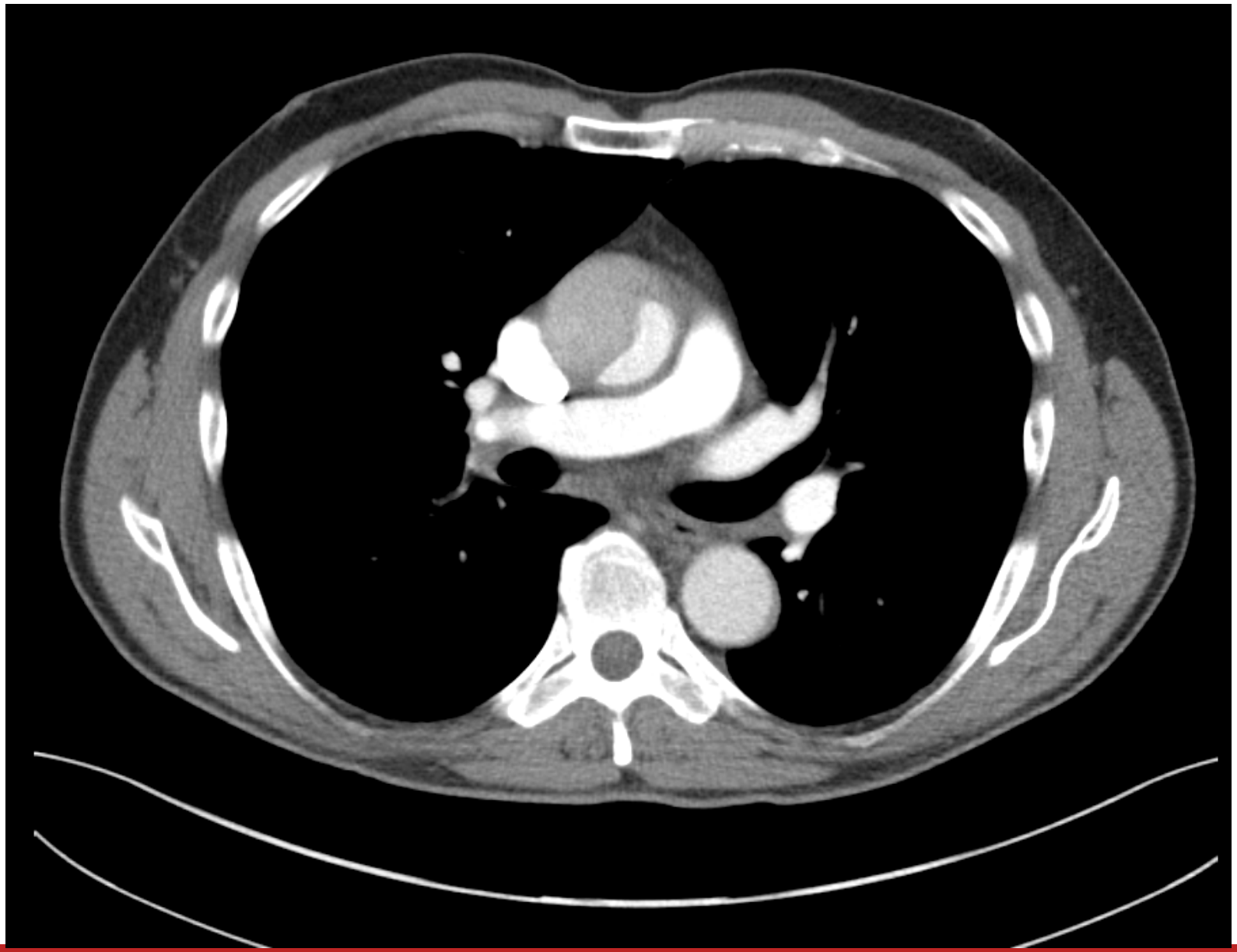












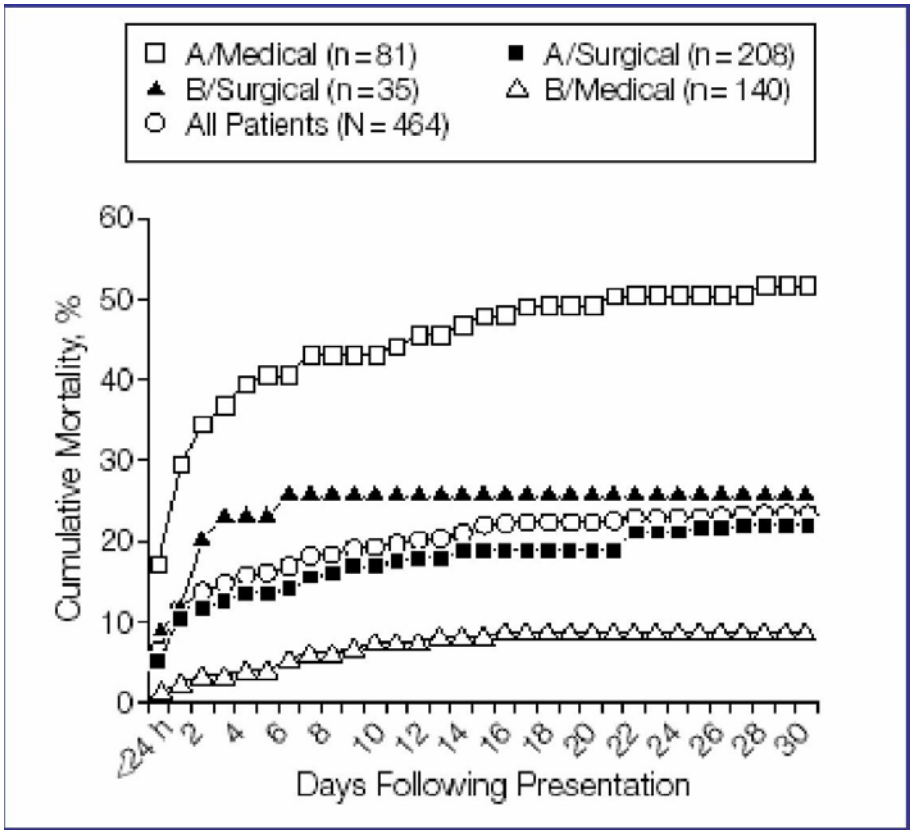
Surgical Background

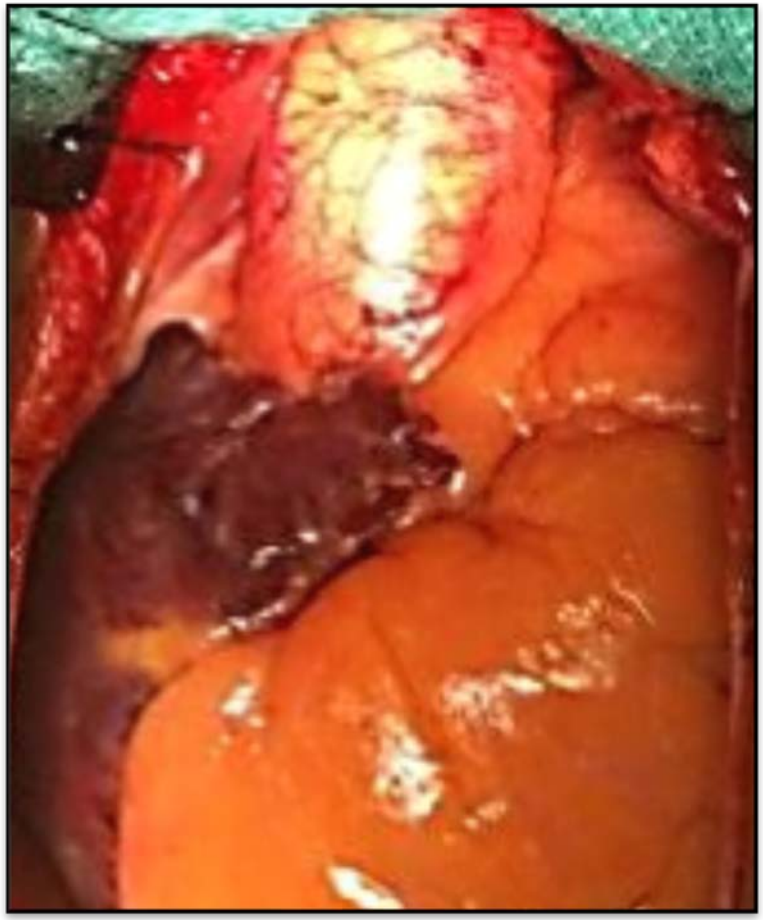
In 1992, Dr Crawford projecting trends from 1975, that we could achieve a 3% mortality for uncomplicated ascending aortic replacement

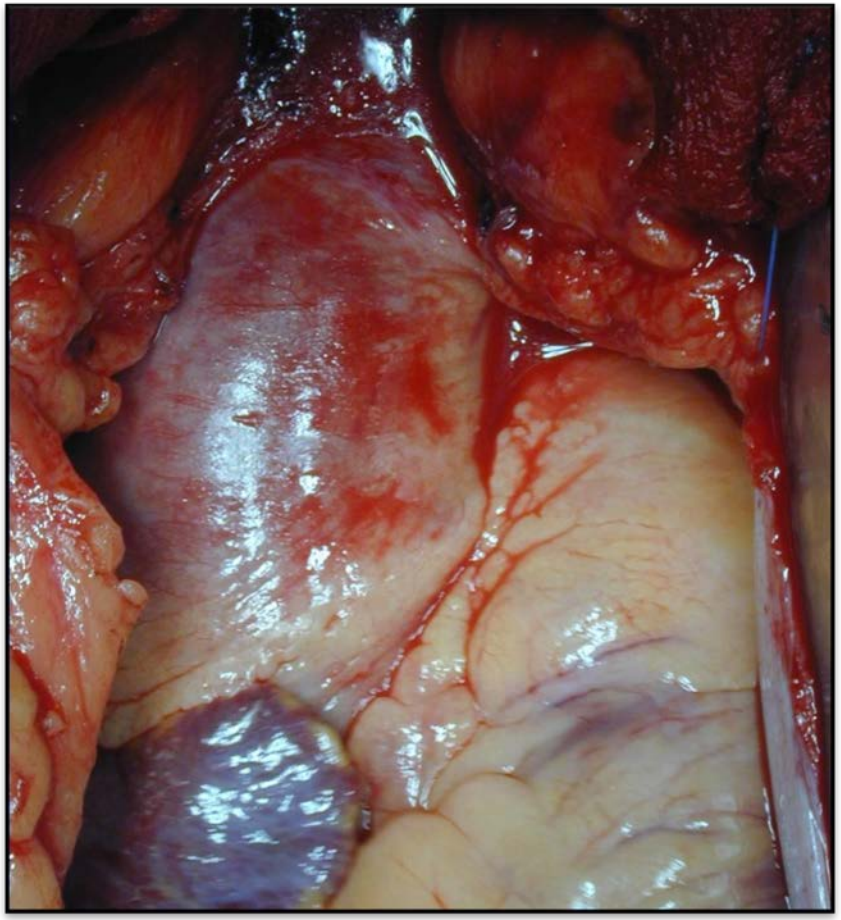
- Crawford et al, JTCVS 1992

IRAD data from 12 specialized aortic centers, note a 26 ± 3 % acute mortality

- Hagan et al, JAMA 2000



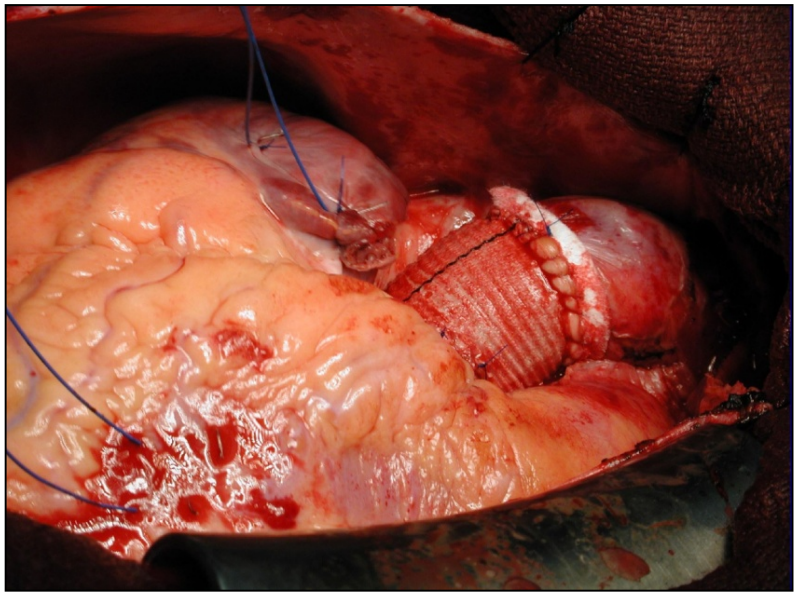




Acute Aortic Dissection

Priorities of Surgical management

- Live Patient !
- Stable aortic root
- Competent aortic valve
- Normal coronary perfusion
- Resection of arch tear
- Single lumen descending thoracic aorta



Cannulation Techniques

Femoral artery

Rt subclavian artery

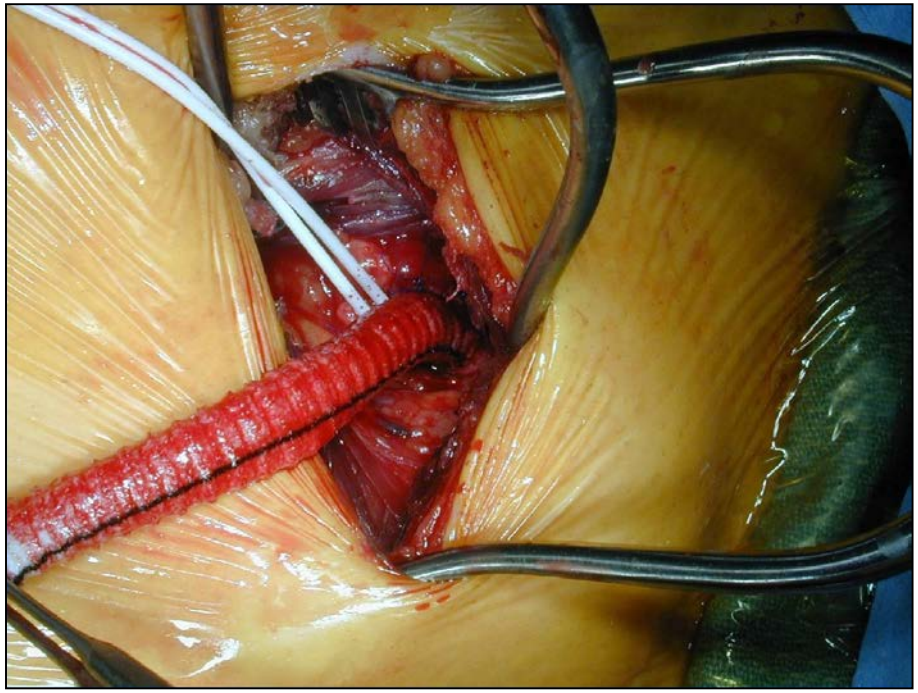
Innominate artery

Lt carotid artery

Direct aortic cannulation

(Seldinger technique)

Transapical LV

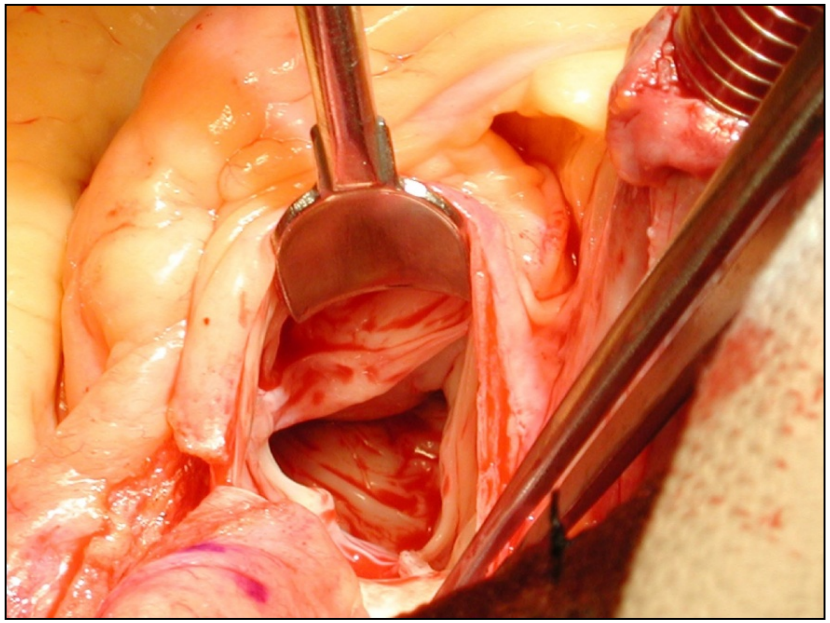


Acute Type A Aortic Dissection

Management of the Proximal anastomosis

Assess:

- Condition of the patient
- Proximal extent
- Size of the aortic sinuses
- Integrity of the aortic valve
- Location and integrity of the coronary ostia



Acute Type A Aortic Dissection

Options for proximal reconstruction

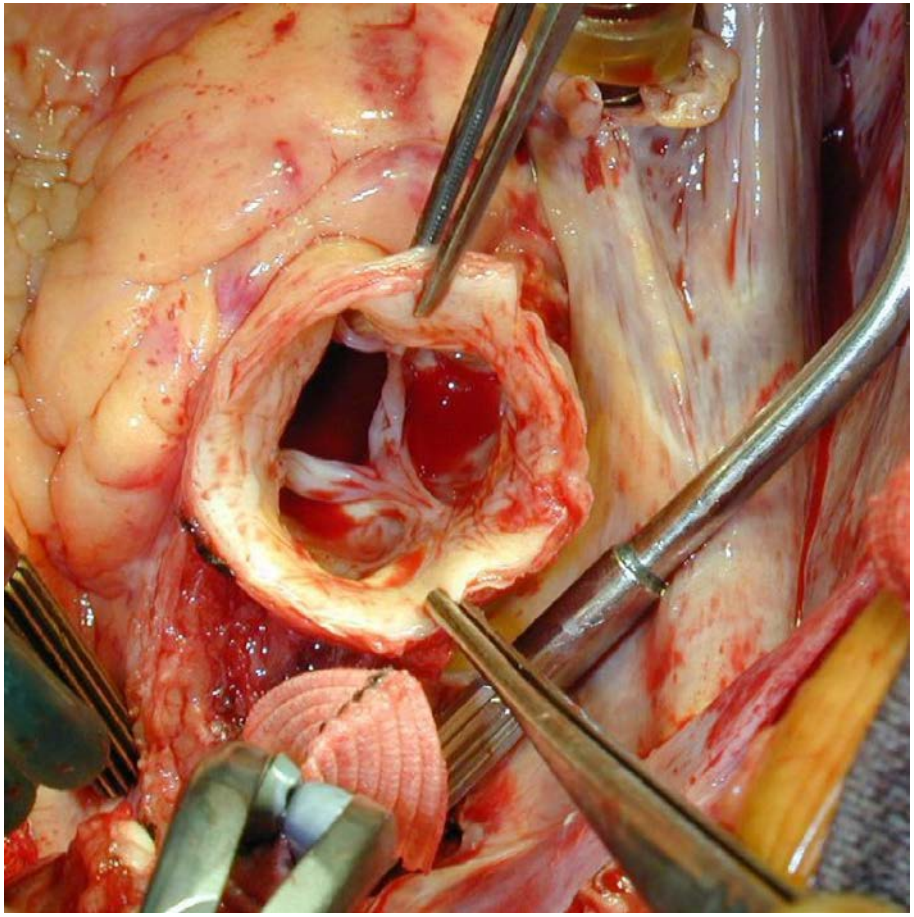
- Reconstruct the proximal root
  (felt/ glue the affected layers)
- Resuspend or replace aortic valve
- Composite valve graft
  - *Yacoub* aortic reconstruction
  - *David* aortic reconstruction

TABLE 2. Intraoperative Data for Patients With AADA Treated With Different Surgical Methods

	SCR	Comp	AVS	Overall	*P*
Operation time, min	242±68	301±121	305±75	267±90	<0.001
ECC time, min	141±54	189±100	209±57	166±74	<0.001
X-clamp time, min	74±29	108±35	156±39	98±45	<0.001
Circulatory arrest, min	24±14	24±16	29±18	25±15	0.20
Aortic cannulation (%)	31 (22)	11 (19)	28 (58)	28	<0.001
Proximal arch replacement (%)	74 (51)	32 (50)	22 (46)	128 (50)	0.18
Sub/total arch replacement (%)	28 (19)	12 (19)	18 (38)	58 (23)	0.023
Elephant trunk (%)	5 (3)	2 (3)	7 (15)	14 (5)	0.008
CABG (%)	13 (9)	8 (13)	3 (6)	24 (9)	0.52
Fenestration (%)	4 (3)	1 (2)	2 (4)	7 (3)	0.70

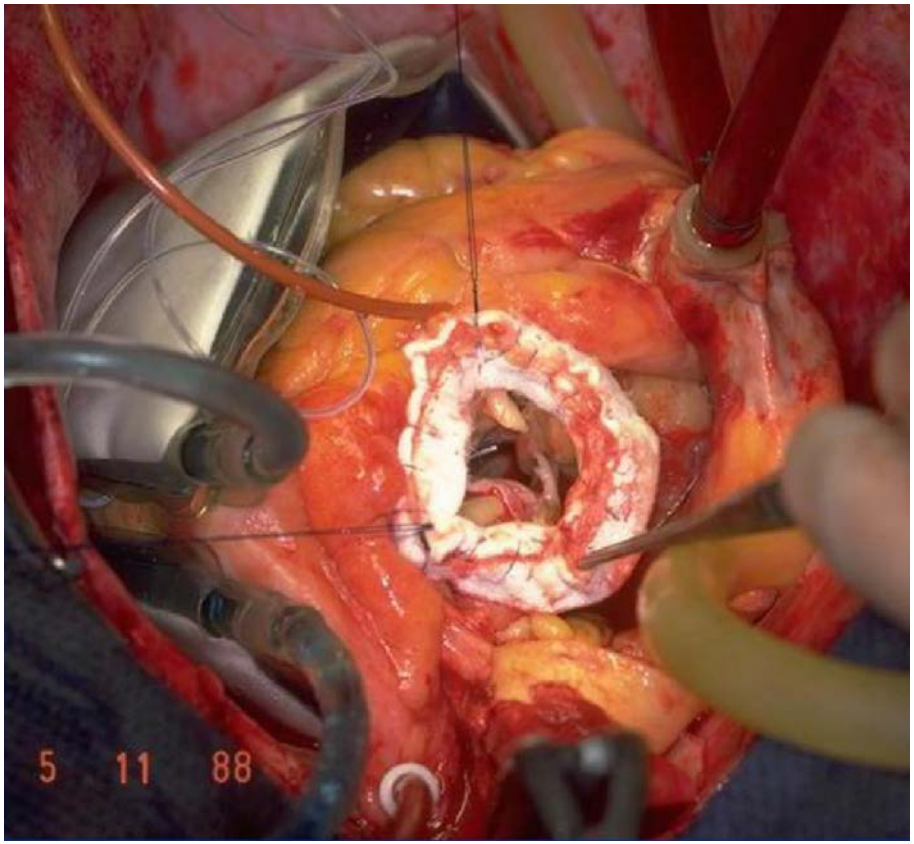
Acute Type A Aortic Dissection

Supracommisural tube placement



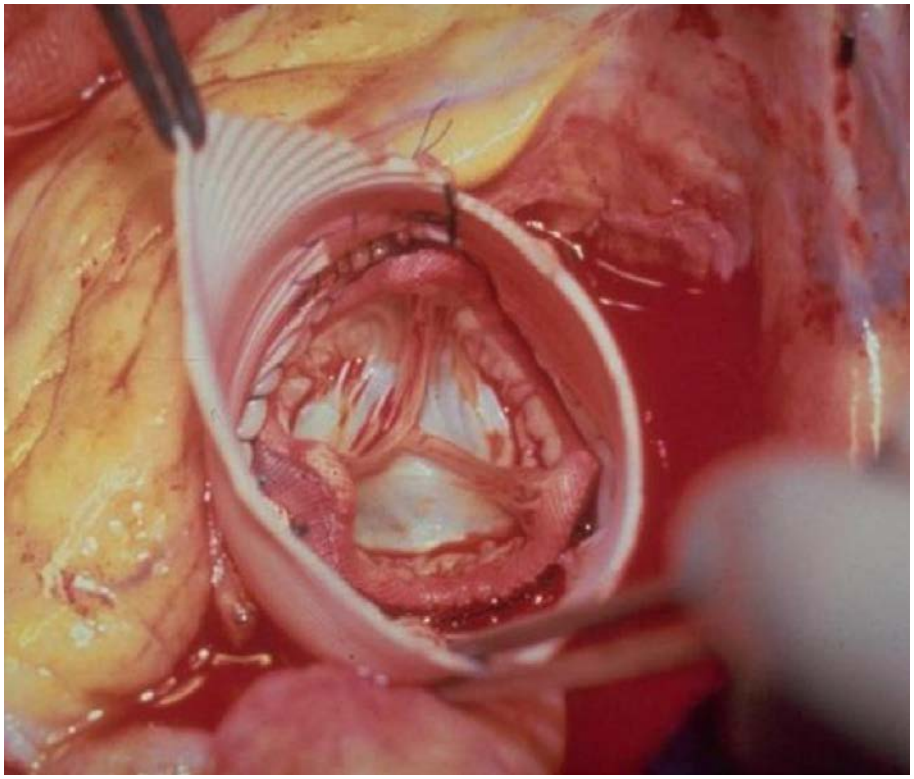
Acute Type A Aortic Dissection

- Root reconstruction w/ felt/ pericardium sandwich ± bioglue



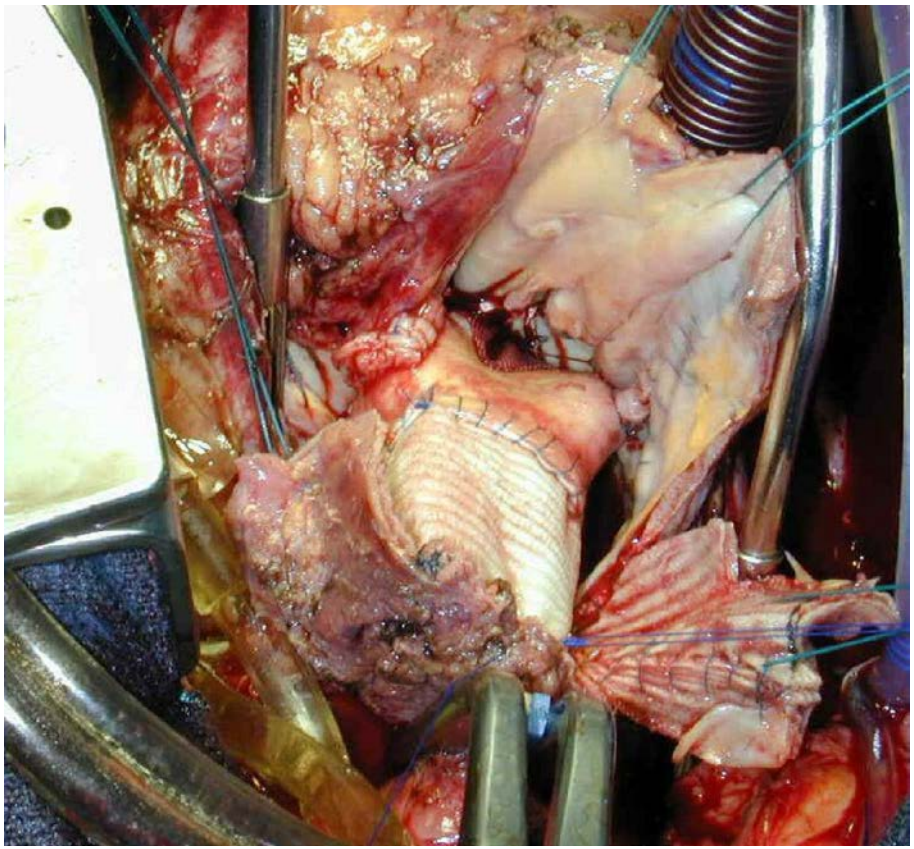
Acute Type A Aortic Dissection

- Valve replacement with supracommisural tube
- Composite valve graft



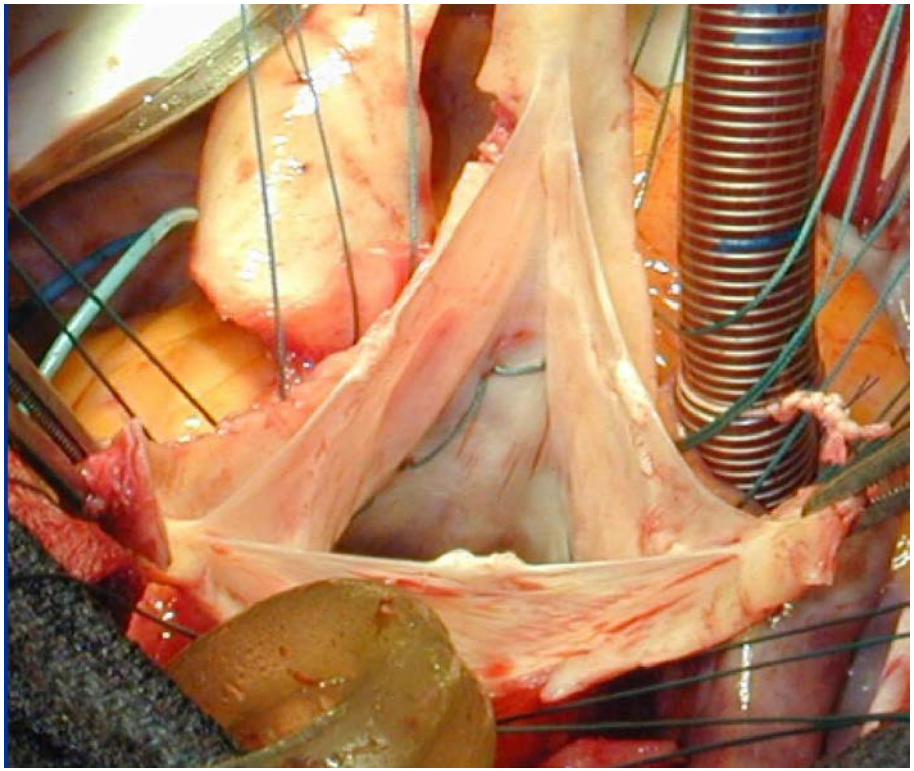
Acute Type A Aortic Dissection

Composite valve graft



Acute Type A Aortic Dissection

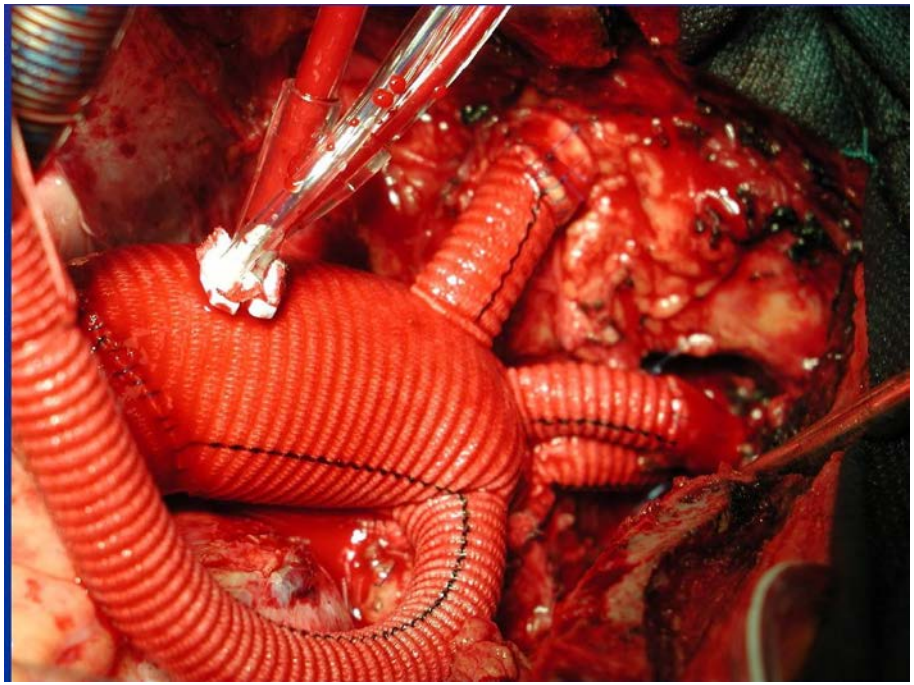
Valve sparing root replacement



Acute Type A Aortic Dissection

Does a more distal radical reconstruction improve survival?

- Distal clamp
- Open distal
- Open hemiarch
- Total arch
- Total arch with elephant trunk



Systematic Review

Hemiarch versus total aortic arch replacement in acute type A dissection: a systematic review and meta-analysis

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Background: Despite recent advances in aortic surgery, acute type A aortic dissection remains a surgical emergency associated with high mortality and morbidity. Appropriate management is crucial to achieve satisfactory outcomes but the optimal surgical approach is controversial. The present systematic review and meta-analysis sought to access cumulative data from comparative studies between hemiarch and total aortic arch replacement in patients with acute type A aortic dissection.

Methods: A systematic review of the literature using six databases. Eligible studies include comparative studies on hemiarch versus total arch replacement reporting short, medium and long term outcomes. A meta-analysis was performed on eligible studies reporting outcome of interest to quantify the effects of hemiarch replacement on mortality and morbidity risk compared to total arch replacement.

Result: Fourteen retrospective studies met the inclusion criteria and 2,221 patients were included in the final analysis. Pooled analysis showed that hemiarch replacement was associated with a lower risk of post-operative renal dialysis [risk ratio (RR) =0.72; 95% confidence interval (CI): 0.56–0.94; P=0.02; I^2=0%]. There was no significant difference in terms of in-hospital mortality between the two groups (RR =0.84; 95% CI: 0.65–1.09; P=0.20; I^2=0%). Cardiopulmonary bypass, aortic cross clamp and circulatory arrest times were significantly longer in total arch replacement. During follow up, no significant difference was reported from current studies between the two operative approaches in terms of aortic re-intervention and freedom from aortic reoperation.

Conclusions: Within the context of publication bias by high volume aortic centres and non-randomized data sets, there was no difference in mortality outcomes between the two groups. This analysis serves to demonstrate that for those centers doing sufficient total aortic arch activity to allow for publication, excellent and equivalent outcomes are achievable. Conclusions on differences in longer term outcome data are required. We do not, however, advocate total arch as a primary approach by all centers and surgeons irrespective of patient characteristics, but rather, a tailored approach based on surgeon and center experience and patient presentation.

Keywords: Hemiarch replacement; total arch replacement; acute type A dissection; meta-analysis

Table 1 An overview of publication from selected studies

First author	Year	Study period	Centers	No. of hemiarch	No. of total arch	Total sample size, n	Mean follow up time	Newcastle Ottawa Score
Shi (10)	2014	2006–2011	Shenyang, China	71	84	155	42.7±17.8 months (3.6 years)	8
Ohtsubo (11)	2002	1989–2001	Saga, Japan	23	24	47	42.0±36 months (0–147 months) (3.5 years)	7
Tan (12)	2003	1986–2001	Nieuwegein, The Netherlands	53	17	70	2.6 years (0–14.5 years)	8
Uchida (13)	2009	1997–2008	Hiroshima, Japan	55	65	120	67 months (3–124 months) (5.6 years)	7
Rylski (14)	2014	2001–2013	Freiburg, Germany	37	14	51	4.9 years 45% >5 years	9
Kim (15)	2010	1999–2009	Seoul, South Korea	144	44	188	47.5 months (0–130.4 months) (4.0 years)	9
Shiono (16)	2006	1995–2005	Tokyo, Japan	105	29	134	FU up to 10 years	8
Zhang (17)	2014	2002–2010	Shanghai, China	74	88	162	55.7±33.1 months (4.6 years)	7
Sun (4)	2014	2003–2008	Beijing, China	66	148	214	42–49 months (3.5–4.1 years)	8
Di Eusanio (18)	2015	1997–2012	Bologna, Italy	187	53	240	4.8±3.9 years (0.1–15.5 years)	8
Rice (19)	2015	NS	Texas, USA	440	49	489	49 months	9
Omura (20)	2016	1999–2014	Kobe, Japan	109	88	197	60±48 months	9
Vallabhajosyula (21)	2015	2006–2013	Philadelphia, USA	30	31	61	60±41 months	7
Dai (22)	2015	2008–2010	Fujian, China	41	52	93	64±5.3 months	7

Table 4 Neuroprotection strategies in hemiarch and total arch replacement

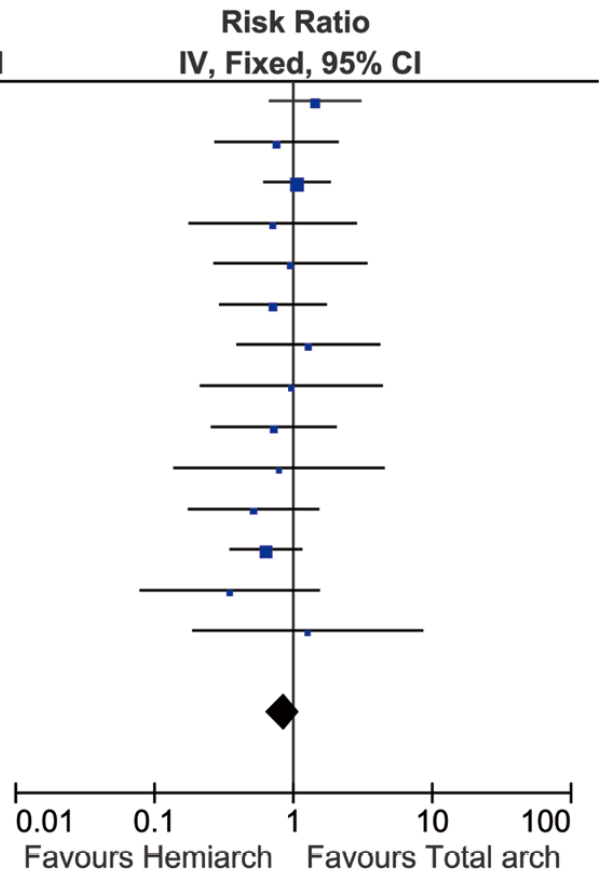
Study	Degree of hypothermia, HA/TA	Temperature measurement/ temperature range, HA/TA	Use of cerebral adjunct (antegrade or retrograde)		Duration of antegrade/ retrograde cerebral perfusion (min)		Comments
			HA	TA	HA	TA	
Shi	MHCA with SACP	Hemiarch: rectal, lowest, 23.3±1.60 °C; total arch: rectal, lowest, 23.7±1.10 °C	SACP	SACP	30.6±4.9	55.2±6.2	Longer duration of antegrade perfusion in total arch group; P<0.001
Ohstubo	PHCA, 3 arch vessels perfusion	NS	SACP in 21.7% of cases	SACP in 100% of cases	29.0±12.7	106.0±6.0	SACP is more commonly used in total arch replacement
Tan	MHCA with bilateral SACP	NP, 250 °C	Bilateral SACP	Bilateral SACP	NS	NS	NS
Uchida	MHCA with SACP	NS	NS	NS	21.0±12.0	70.0±18.0	Longer duration of cerebral perfusion in total arch replacement
Rylski	MHCA with SACP	NS	SACP	SACP	32.0±9.0	71.0±25.0	Longer duration of cerebral perfusion in total arch replacement; P<0.001
Kim	Hemiarch: DHCA (11.0–19.90 C) in 76.4%; MHCA (20.0–26.50 C) in 23.6%. Total arch: DHCA (11.0–19.90 C) in 70.5%; MHCA (20.0–26.50 C) in 29.5%	Hemiarch: OP, lowest, 16.3±40 °C; total arch: OP, lowest, 17.2±4.50 °C	SACP in 29.2%, RCP in 68.8%	SACP in 61.4%, RCP in 38.6%	NS	NS	SACP is more commonly utilised in total arch replacement, retrograde is preferred in hemiarch group. Moderate hypothermia is more likely to be used in SACP
Shiono	DHCA with SACP	Below 200 °C	NS	NS	NS	NS	NS
Zhang	MHCA with SACP	Core temperature 26.0–28.00 °C	SACP in 89.2%; RCP in 10.8%	SACP in 92%; RCP in 8.0%	NS	NS	Moderate hypothermia is more commonly used in recent practices. Similar use of antegrade and retrograde in both groups. P=0.53
Sun	MHCA with unilateral SACP	NP, 18.0–22.00 °C	SACP	SACP	18.0±7.0	24.0±9.0	Similar use and duration of hypothermia and cerebral adjunct in both groups. P=0.097
Di Eusanio	MHCA with SACP	NP, 260 °C	SACP	SACP	45.1±13.7	86.9±33.3	Significantly longer duration of SACP in total arch replacement; P<0.001
Omura	MHCA with SACP	Tympanic <23.00 °C, rectal <30.00 °C	SACP in 56.0% of cases	SACP in 97.7% of cases	48.1±26.6	124.0±42.5	SACP is more commonly utilized in total arch replacement; the duration of perfusion if significantly longer; P≤0.01
Vallabhajosyula	MHCA with SACP/RCP	NS	RCP only in 80%, SACP only 0%; both methods in 20%	RCP only 0%, SACP only 32%; both methods in 68%	NS	NS	RCP is used more frequently in hemiarch replacement
Dai	DHCA with SACP	NP, <20.00 °C	NS	NS	NS	NS	NS
Pooled mean (range)	NA	NA	NA	NA	Mean difference of pooled cerebral time (min): 22.71; (95% CI: 21.50–23.92), P<0.00001		The duration of cerebral perfusion time is significantly longer in total arch replacement

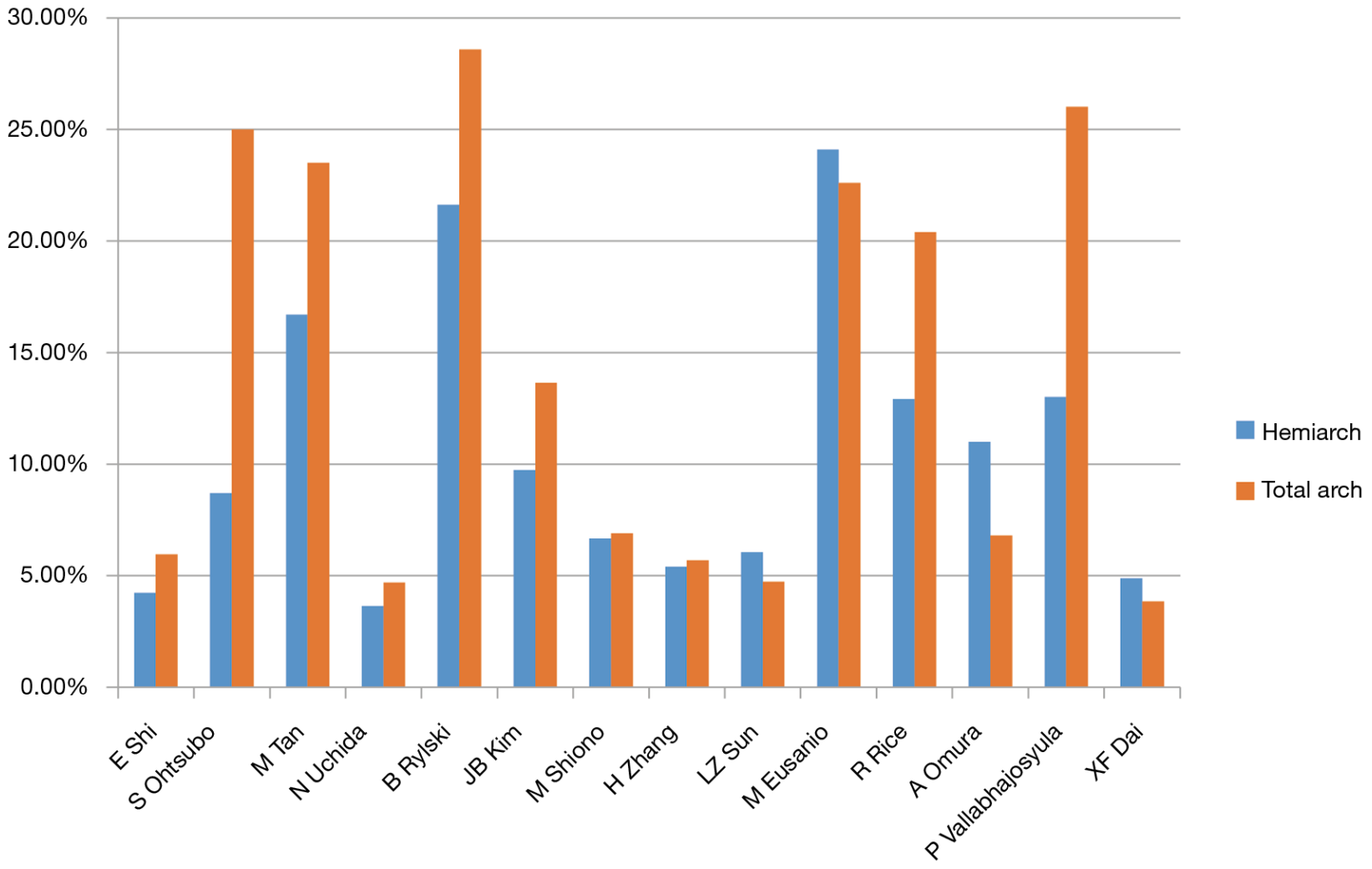
SACP, selective antegrade cerebral perfusion; RCP, retrograde cerebral perfusion; MHCA, moderate hypothermic circulatory arrest; DHCA, deep hypothermic circulatory arrest; PHCA, profound hypothermic circulatory arrest; NP, nasopharyngeal; OP, oropharyngeal; NS, not specified; RR, risk ratio; CI, confidence interval; HA, hemiarch; TA, total arch.

Study or Subgroup	Hemiarch Events	Hemiarch Total	Total arch Events	Total arch Total	Weight	Risk Ratio IV, Fixed, 95% CI
A Omura 2016	16	109	9	88	11.4%	1.44 [0.67, 3.09]
B Rylski 2014	8	37	4	14	6.3%	0.76 [0.27, 2.12]
D Eusanio 2015	45	187	12	53	21.5%	1.06 [0.61, 1.86]
Enyi Shi 2014	3	71	5	84	3.4%	0.71 [0.18, 2.87]
H Zhang 2014	4	74	5	88	4.1%	0.95 [0.27, 3.41]
JB Kim 2011	14	144	6	44	8.4%	0.71 [0.29, 1.74]
LZ Sun 2014	4	66	7	148	4.7%	1.28 [0.39, 4.23]
M Shiono 2006	7	105	2	29	2.9%	0.97 [0.21, 4.41]
MH Tan 2003	9	53	4	17	6.2%	0.72 [0.25, 2.05]
N Uchida 2009	2	55	3	65	2.2%	0.79 [0.14, 4.55]
P Vallabhajosyula 2015	4	30	8	31	5.6%	0.52 [0.17, 1.54]
R Rice 2015	57	440	10	49	18.4%	0.63 [0.35, 1.16]
S Ohtsubo 2002	2	23	6	24	3.0%	0.35 [0.08, 1.55]
XF Dai 2015	2	41	2	52	1.8%	1.27 [0.19, 8.62]
Total (95% CI)		**1435**		**786**	**100.0%**	**0.84 [0.65, 1.09]**
Total events	177		83			

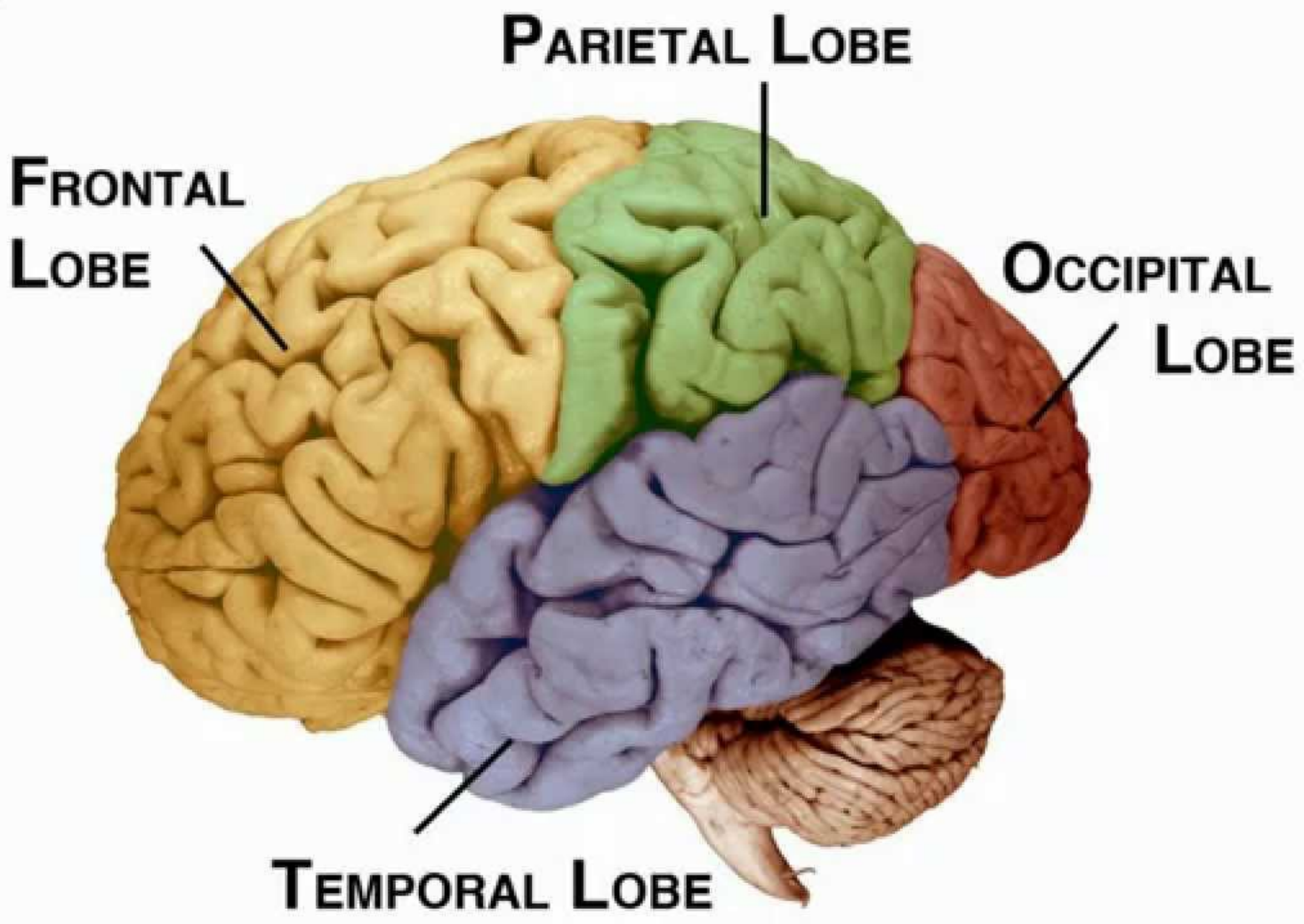
Heterogeneity: Chi² = 6.52, df = 13 (P = 0.93); I² = 0%

Test for overall effect: Z = 1.27 (P = 0.20)




30.00%
25.00%
20.00%
15.00%
10.00%
5.00%
0.00%
E Shi
S Ohtsubo
M Tan
N Uchida
B Rylski
JB Kim
M Shiono
H Zhang
LZ Sun
M Eusanio
R Rice
A Omura
P Vallabhajosyula
XF Dai
Hemiarch
Total arch

Conclusions: Within the context of publication bias by high volume aortic centres and non-randomized data sets, there was no difference in mortality outcomes between the two groups. This analysis serves to demonstrate that for those centers doing sufficient total aortic arch activity to allow for publication, excellent and equivalent outcomes are achievable. Conclusions on differences in longer term outcome data are required. We do not, however, advocate total arch as a primary approach by all centers and surgeons irrespective of patient characteristics, but rather, a tailored approach based on surgeon and center experience and patient presentation.

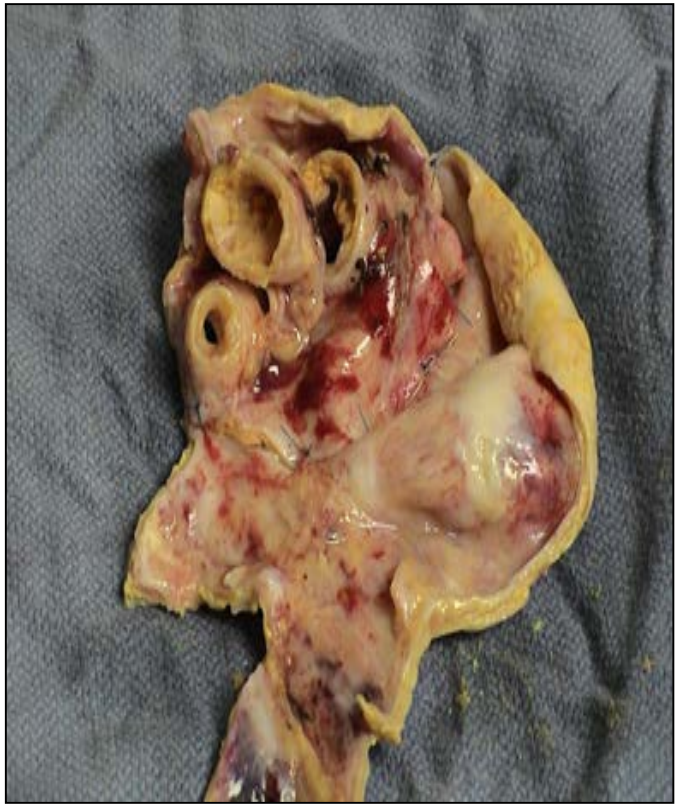

PARIETAL LOBE
FRONTAL LOBE
OCCIPITAL LOBE
TEMPORAL LOBE

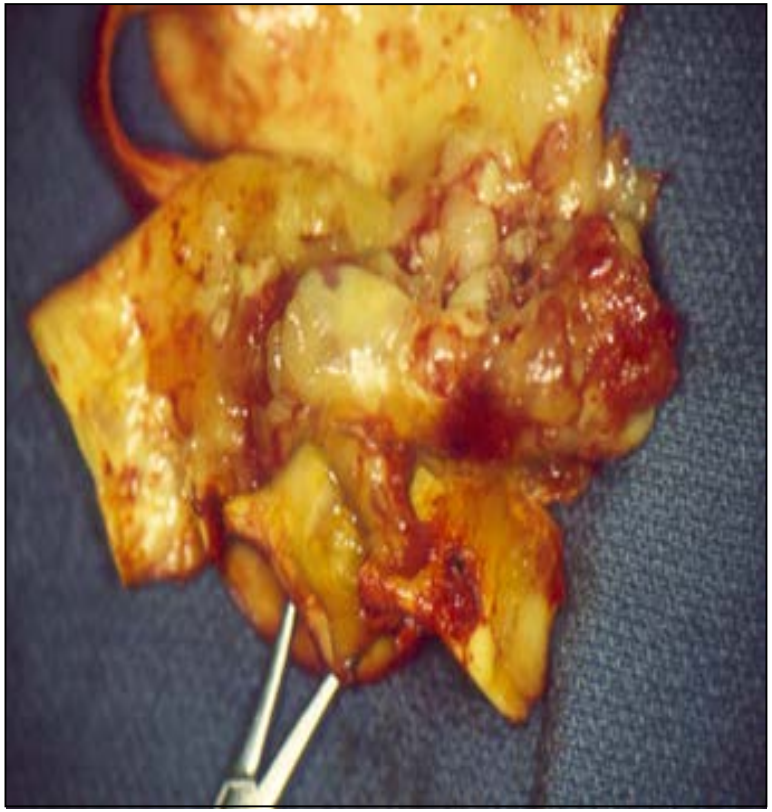
Brain

- 1400g (5 % body weight)
- 20% total body oxygen consumption
  - »40% Cellular
  - »60% Transmission Nerve Impulses

Neuro-cognitive Events

- Focal Cerebral Vascular Events (Stroke)
- Embolic Events





Global cerebral necrosis

- Minimum O2 vs Cerebral Metabolic Requirements
- Ischemia depletes ATP
    - Failure basic cellular Ionic pumps
    - Influx Na Cl
    - Calcium accumulation

Excessive neuronal stimulation (excitotoxicity)

- Excitatory amino acids (glutamate) – Accelerates neuronal death

Selective vulnerability

- Hippocampus
- Cerebellum
- Striatum
- Amygdala
- Lateral Thalamic Nucleus
- 3^{rd}-5^{th} Layer Neocortex
- Clinically Improve, but Neuronal Death Continues Up to 1 Week.

Mechanisms of Cerebral Protection

- Deep Hypothermic Circulatory Arrest (DHCA)
- Retrograde Cerebral Perfusion (RCP)
- Selective Antegrade Cerebral Perfusion (SACP)

Antegrade selective cerebral perfusion: historical notes

Year	Surgeons	Techniques	Outcome
1957	De Bakey	Normothermic CPB and bilateral perfusion of subclavian and carotid arteries by means of several pumps.	Hospital mortality: 75%
1968	Dubost and Pearce	Single pump with a Y-connection of the arterial line.	
1986	Frist	Moderate hypothermia (26°–28 °C) and Y-connection on arterial line to perfuse femoral and brachiocephalic arteries.	Hospital mortality: 20% No stroke
1999	Bachet	Cold cerebroplegia: innominate and left carotid arteries perfusion with blood between 6° and 12 °C through separate pumps and heat exchangers.	Hospital mortality: 13% One stroke
1992	Kazui	Moderate hypothermia (25 °C) and separate arterial pump heads for cerebral and systemic circulations. Endoluminal cannulation of the brachiocephalic and left common carotid arteries and occlusion of the left subclavian artery.	Hospital mortality: 4.3% No stroke

CPB, cardiopulmonary bypass.

Roberto Di Bartolomeo et al. MMCTS 2011;2011:mmcts.2010.004457

Define – hypothermia

- Mild - 30˚ - 34˚ C
- Moderate - 23˚ - 29˚ C
- Deep - 13˚ - 22˚ C

PH Management

ALPHA -STAT

(Normal PH and $PaCO_2$)

PH of 7.4 and $PaCO_2$ of 40 mm Hg in 37°

blood alkaline and hypocapnic

PH -STAT

Maintain a PH of 7.40 and $PaCO_2$ of 40 mm Hg under hypothermic conditions

when warmed, blood becomes acidotic and hypercapnic

Glycemic control

- Hyperglycemia results in anaerobic conversion glucose to lactate resulting in acidosis

- Hyperglycemia releases excitatory amino acids with negative neurologic effects

Hematocrit monitoring

- Hemodilution decreases the oxygen carrying capacity of the perfusate

- Optimal Hematocrit level of 30%

Pharmacologic adjustments

- Barbituates
  - Thiopentone Studies
  - Thiopental 15- 30 mg/kg
  - Negative Inotropes
  - Benefit Unknown

Volatile anesthetics

Halothane – Isoflurane

- Provide protection via inhibition of excitotoxicity
- Depression of metabolism
- Improvement of cerebral oxygenation
- Inhibits glutamate release
- Can provide an isoelectric EEG

Corticosteroids

- High dose reduce cytokines and TNF – alpha and seems beneficial to
- Improve cerebral oxygen metabolism ?
- Recovery of cerebral blood flow after DHCA

Patient Monitoring

- EEG – Electroencephalographic
- BIS – Bispectral Index Monitor – 0 (flat line) to 100 awake
- SSEP – Somatosensory evoked potentials – Median Nerve
  - Not significantly Influenced by Anesthetics
  - Can be nearly 100% in determining early neurological events
- Jugular Venous bulb Saturation
  - SvO2 > 95%
  - Associated with lower central temperature but not with better outcome

Rewarming

- Begin slowly
- “Cold blood, low -pressure reperfusion”
- Vulnerable period 6-8 hours after initiation of rewarming

Advantages of DHCA

- Dry and motionless field
- Avoidance of clamping and manipulation of the aorta
- Simplicity and no need for additional perfusion equipment

Disadvantages of DHCA

- Limited time of circulatory arrest
- Prolonged CPB time to cool down and rewarm increases pulmonary renal cardiac and endothelial dysfunction
- Reperfusion Injury
- Coagulopathy

Retrograde cerebral perfusion (RCP)

- Once circulatory arrest initiated then retrograde cold, oxygenated blood into superior vena cave
- above or below azygous?
- Flow 100-500 ml/min
- Adjust to keep central venous pressure 15-25 mm/Hg

RCP Neuro-protective mechanisms

- Maintain cerebral hypothermia
- Washout of embolic air and debris
- Cerebral Perfusion and Metabolic Support?

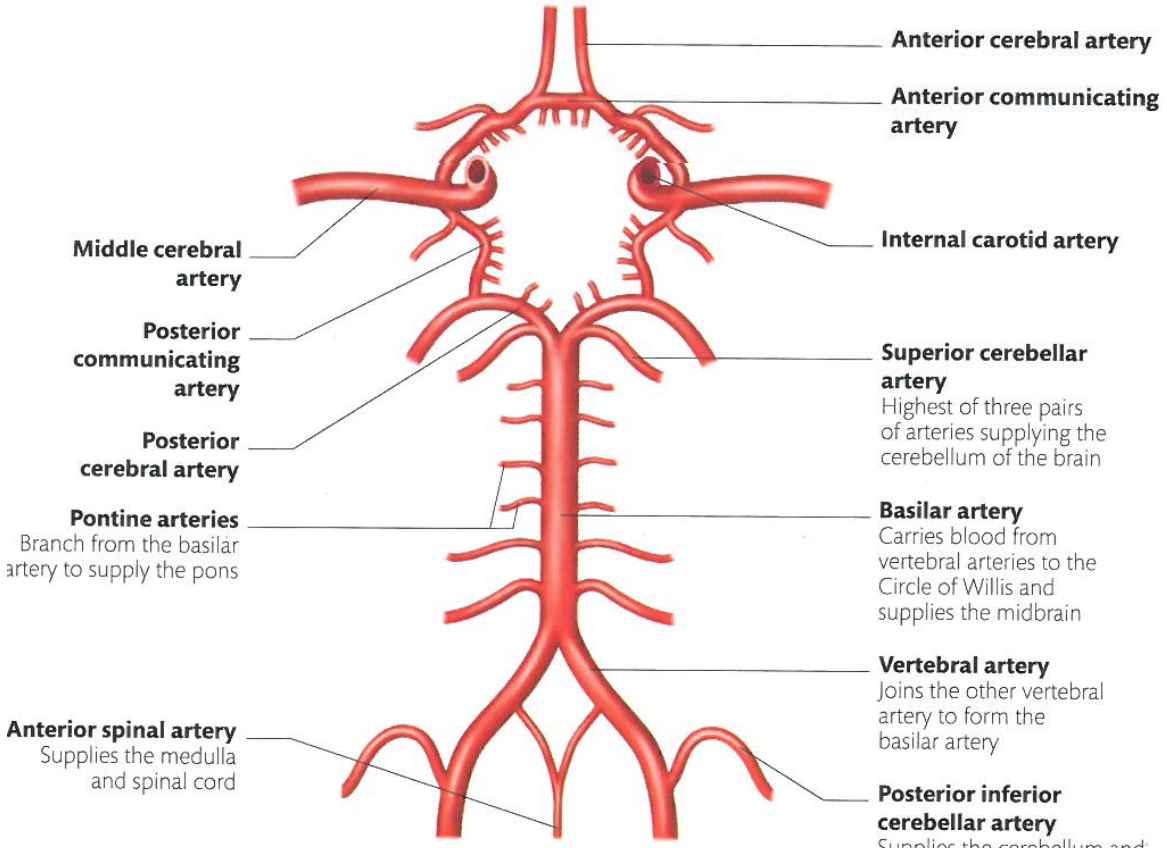
However….

- Provides only 20-60 % of overall intracranial flow as compared to hypothermic bypass

- Relationship of use of RCP and neurologic outcome is unclear

Selective cerebral perfusion (SCP)

- Kazui – 1989 described perfusion through both innominate and left carotid arteries
- Has become popular for cerebral protection for complex arch reconstructions
- Most often the right axillary artery is directly cannulated or 8 mm graft
- Intraclavicular or deltopectoral groove incision



CIRCLE OF WILLIS

SACP

- Right axillary flow 10-20 ml kg/min
- Pressure adjusted to 40 – 50 mm/Hg
- Temperature perfusate, 18ºC
- To cannulate left carotid artery or not ?

Advantages of SACP

- Circulatory arrest times can extend to 90 minutes ?
- Moderate (nasopharyngeal 25˚ C) temperatures with reduced coagulopathy and systems complications (renal failure)

Disadvantages of SACP

- Technical Complexity
- Reduced Surgical Visibility
- Increased manipulation of the aortic and arch vessels

What to do…

- Lots of experimental data but few prospective , randomized trials

- Very difficult to compare trials because of variations of techniques between and within various trials

There are known, knowns .

These are things that we know.

There are known unknowns.

That is to say, there are things we know we don’t know.

But there are also unknown unknowns.

There are things we don’t know we don’t know.

Donald Rumsfeld

Genetic aortic syndromes

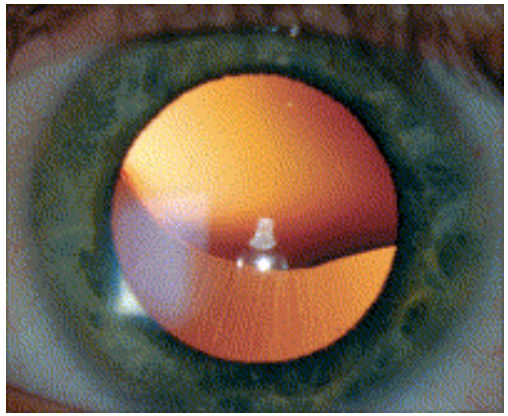
- Marfan syndrome(MFS)
- Loeys – Dietz syndrome (LDS)
- Shprintzen – Goldberg syndrome (SGS)
- Aneurysms-osteoarthritis syndrome (AOS)
- Syndromic TAA (sTAA)

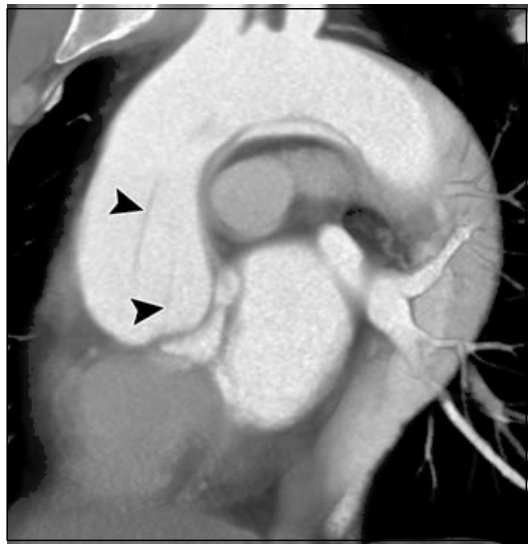
Marfan Syndrome(MFS)

- Marfan syndrome
  - relatively common connective tissue
  - involves eyes, cardiovascular and musculoskeletal systems
- Mutations in fibrillin-1
- Autosomal dominant
- Professor Antoine Bernard-Jean Marfan
  - described a 5 year old girl with disproportionately long limbs and digits

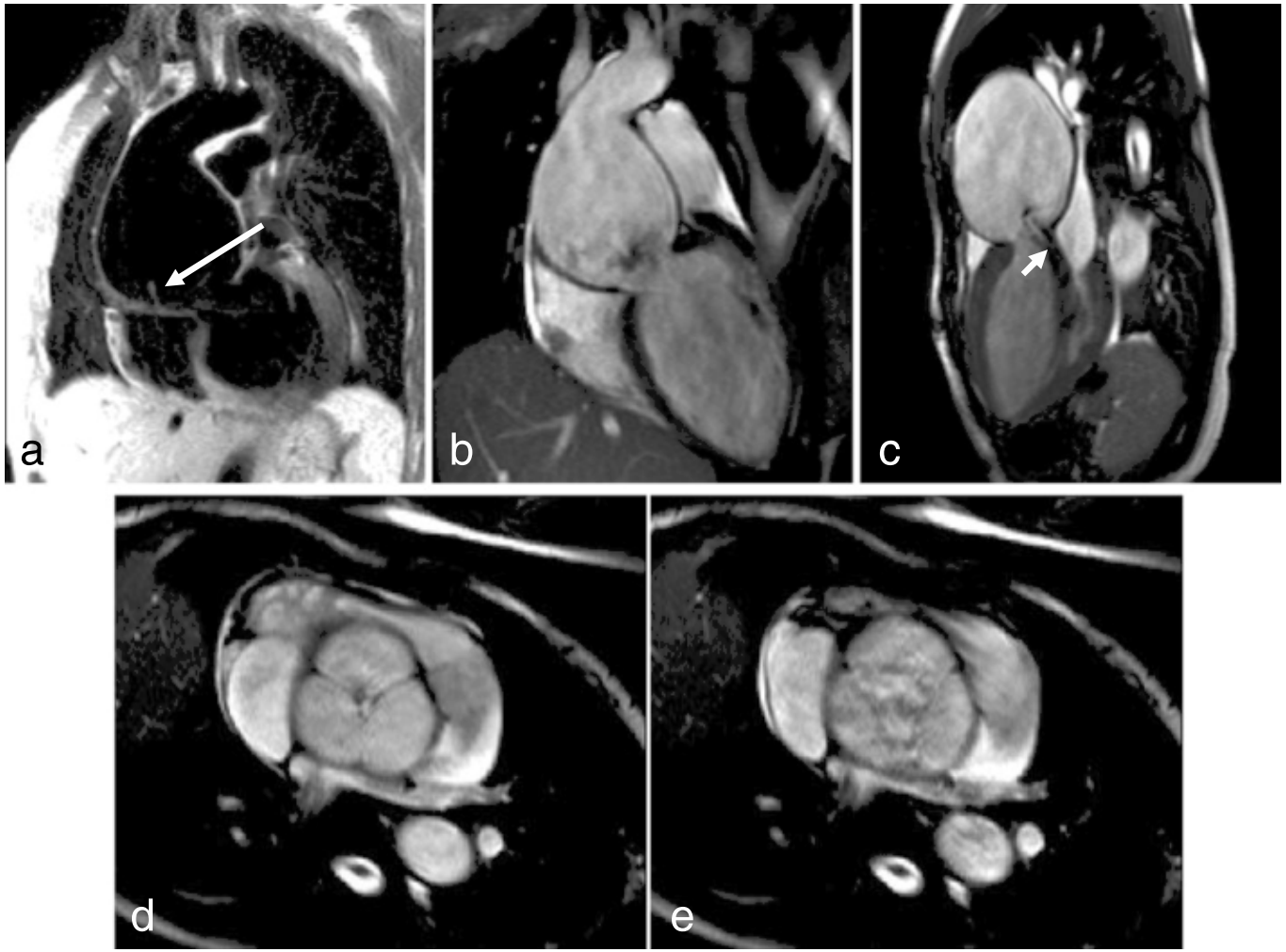
Marfan syndrome

- Cardinal manifestations
  - aortic root diameter Z-score >2
  - ectopia lentis
  - inherited Fbn1 mutation



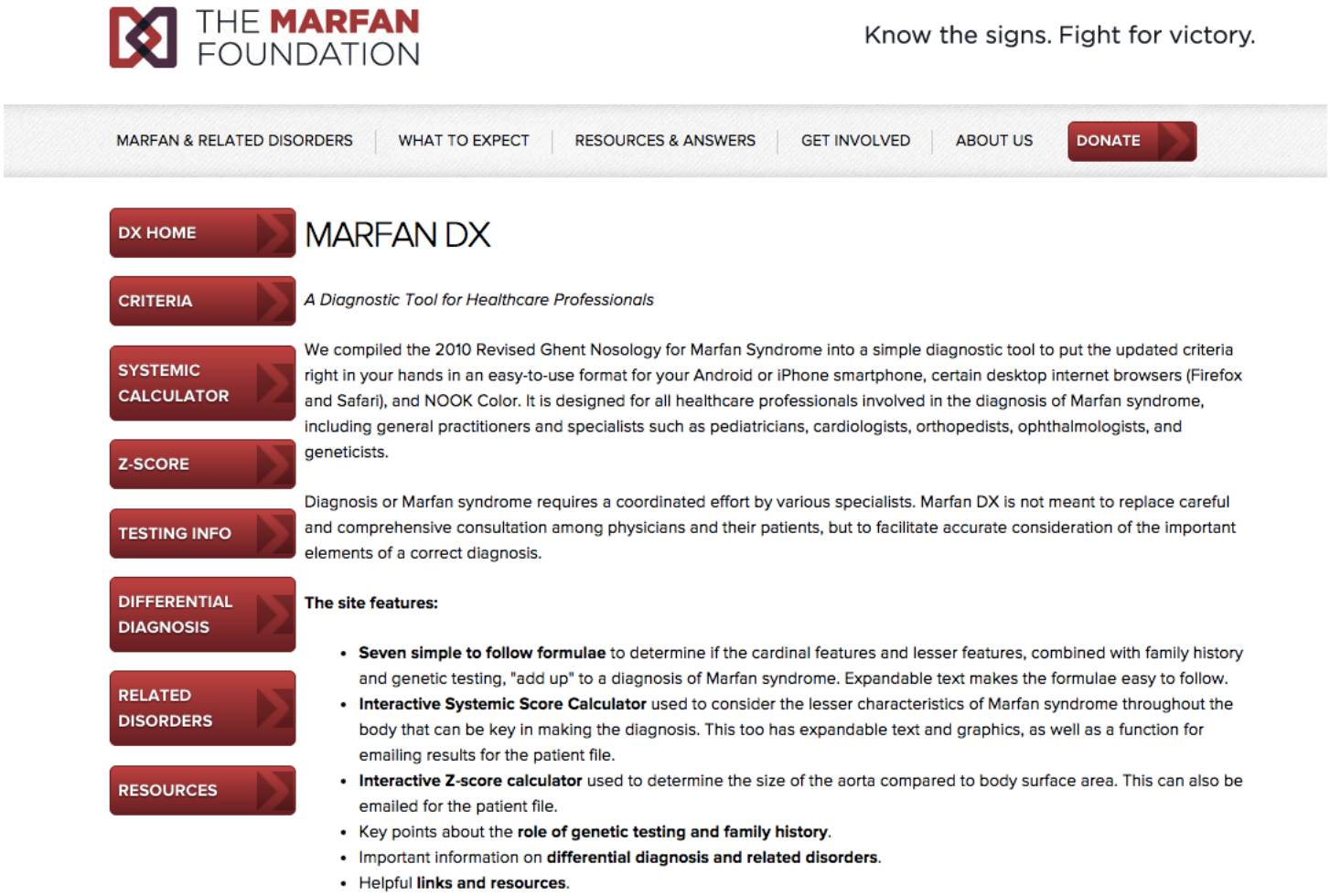


MFS – aortic aneurysm



Dormand and Mohiaddin Journal of Cardiovascular Magnetic Resonance 2013, 15:33 http://www.jcmr-

Marfan diagnosis



Z score

- “z score”
  - the number of standard deviations from the mean of “aortic” measurement when compared with normal individuals with the same body surface area,

Marfan diagnosis

CALCULATION OF SYSTEMIC SCORE

Clinical manifestations of MFS in other organ systems were critically evaluated for their specificity and diagnostic utility based on expert opinion and the available literature. Several of the "minor" criteria from the old Ghent nosology were eliminated, but the most selective systemic features were included in the "systemic score".

Feature		Value	Click to include
Wrist AND thumb sign	+	3	☐
Wrist OR thumb sign	+	1	☐
Pectus Carinatum Deformity	+	2	☐
Pectus Excavatum or Chest Asymmetry	+	1	☐
Hindfoot Deformity	+	2	☐
Plain Flat Foot	+	1	☐
Spontaneous Pneumothorax	+	2	☐
Dural Ectasia	+	2	☐
Protucio Acetabulae	+	2	☐
Scoliosis or Thoracolumbar Kyphosis	+	1	☐
Reduced Elbow Extension	+	1	☐
3 of 5 Facial Features	+	1	☐
Skin Striae	+	1	☐
Severe Myopia	+	1	☐
Mitral Valve Prolapse	+	1	☐
Reduced Upper Segment / Lower Segment & Increased Arm span / Height	+	0	Open to calculate

*A score of ≥ 7 is considered a positive systemic score.

Cardiovascular system in Marfan syndrome

- Mitral valve prolapse and regurgitation
- Left ventricular dilatation and CHF
- Pulmonary artery dilatation
- Aortic root dilatation
  - the most common cause of morbidity and mortality

Aorta in Marfan syndrome

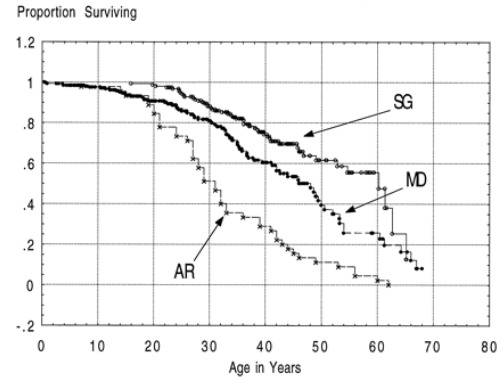
- Aortic aneurysm
- Aortic dissection
- Aortic regurgitation is secondary to above

Aorta in Marfan syndrome

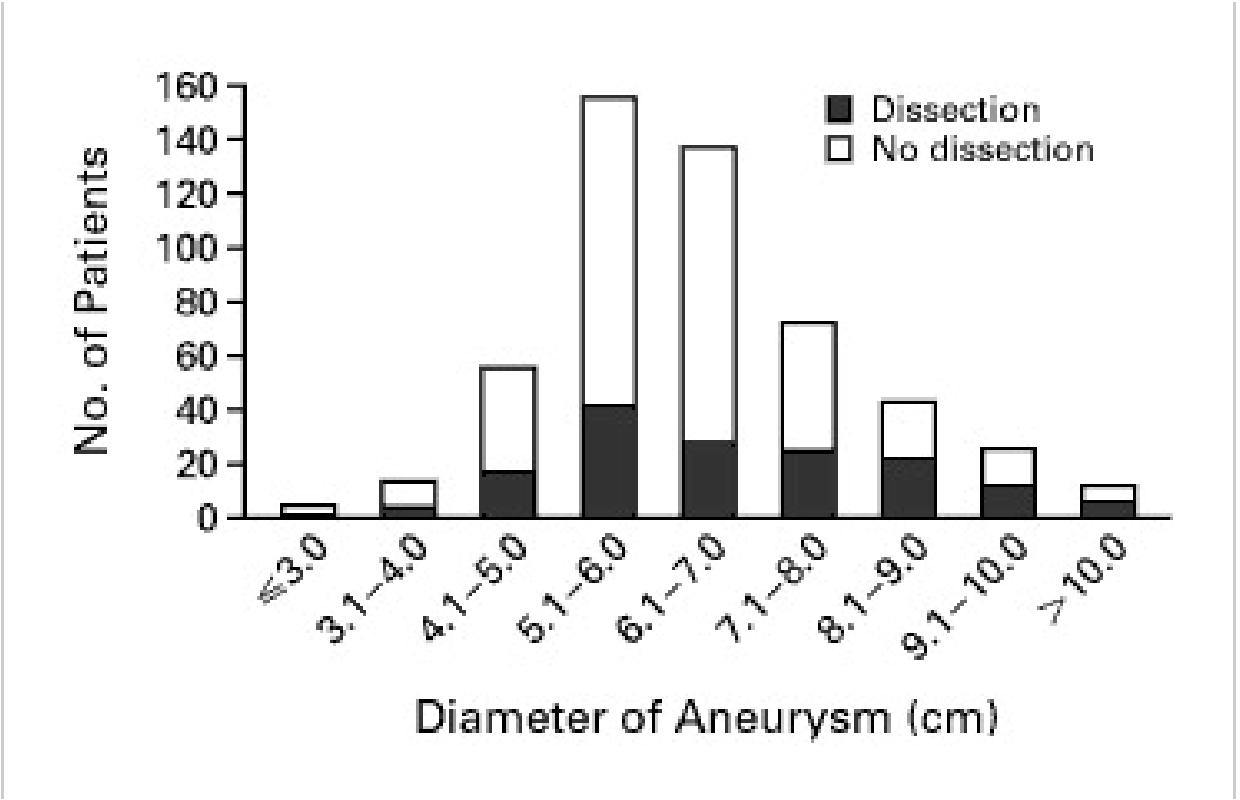
- Risk of aortic dissection
  - diameter at the sinus of Valsalva exceeds 5 cm
  - rate of dilatation exceeds 0.5 mm per year
  - family history of aortic dissection.

Natural history of Marfan

- Mortality from aortic complications has decreased
  - 70% in 1972, 48% in 1995
- Life expectancy has increased
  - mean age at death 32 years in 1972 versus 45 years in 1998



Natural history of Marfan



N Engl J Med 1999; 340:1307-1313April 29, 1999

Medical management

- If the aorta is dilated
  - Beta-Blocker therapy
- Prophylactic treatment is recommended
  - in those with an aortic diameter of less than 4 cm.

Surveillance

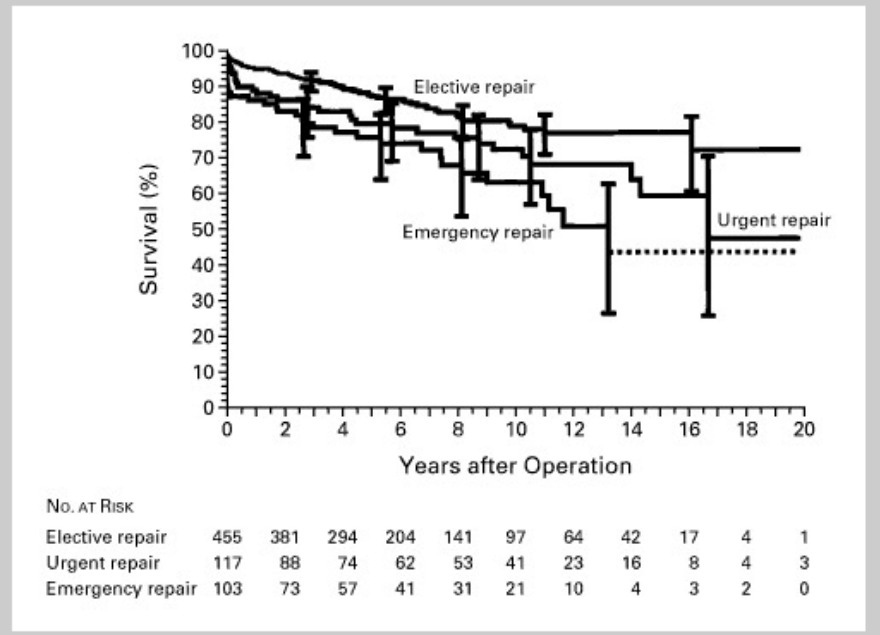
- Annual evaluation
  - clinical history, examination
  - echocardiography

Imaging in Marfan

- Echocardiography at diagnosis to assess the aortic root and ascending aorta
- Follow-up echocardiogram after 6 months.
- If aortic root diameter is stable,
  - annual imaging until the diameter reaches 4.5 cm or
  - changes significantly, when more frequent imaging is advised (Class I, Level of Evidence: C).

Surgical management

- Prophylactic aortic root surgery
  - when the aortic diameter at the Sinus of Valsalva exceeds 5 cm.



N Engl J Med 1999; 340:1307-1313April 29, 1999

Prophylactic surgery vs emergent

- Performing prophylactic surgery
  - <2% mortality rate
- Performing emergent repair during acute dissection
  - >10% mortality

Post surgical followup

- Repair site is protected from further dilation
- However, continued risk to the arch and descending aorta
  - development of aneurysm formation and dissection later in life
- Careful monitoring is still required proximal and distal to the repair site (1,3,6,12 months and yearly)
- Routine lifelong imaging of the entire aorta is recommended(every 5 years)

MFS and Exercise

- Avoid high intensity static exercise
  - (most competitive athletics, heavy weight lifting)
- Encouraged to participate in lower intensity dynamic exercise (such as golf and bowling)
- Contact sports are not advised

Marfan syndrome and pregnancy

- Increased risk of aortic complications
  - hormonal and hemodynamic alterations during pregnancy
  - pregnancy is a hypervolemic and hyperdynamic state.

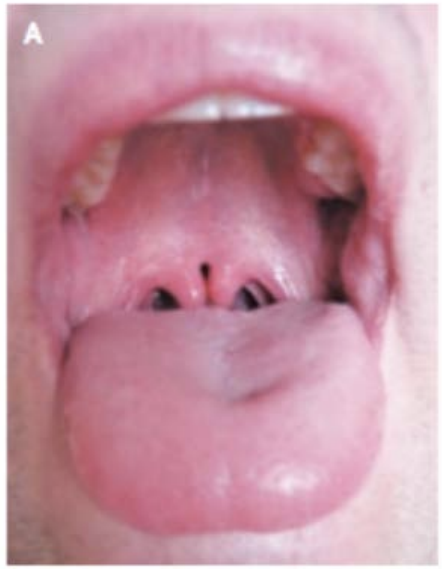
Loeys-Dietz syndrome (LDS)

- Autosomal dominant disorder
  - connective tissue with widespread systemic involvement
- Mutations in the genes encoding transforming growth factor beta receptor 1 or 2

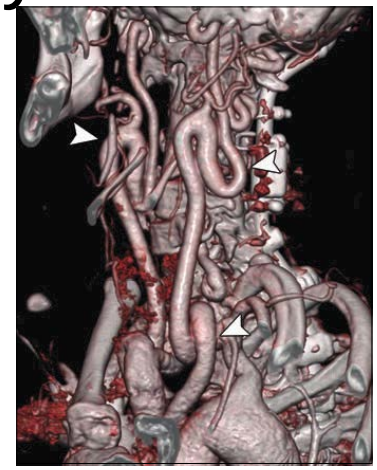
Loeys-Dietz syndrome (LDS)

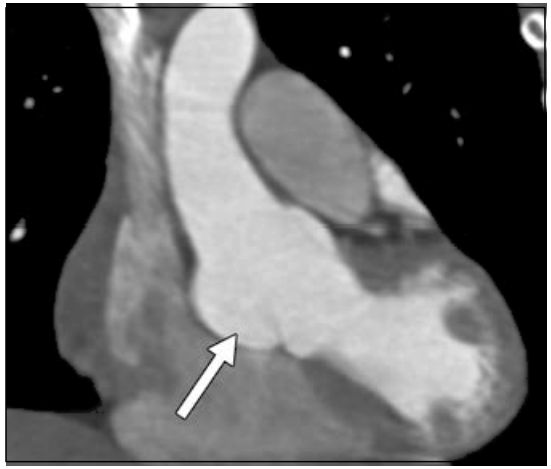
- In contrast to other aortopathies
  - catastrophic in children
- Aortic rupture and dissection
  - Can occur at very young ages (6 months of age) and at very small (nearly normal) aortic diameters.

Loeys-Dietz syndrome (LDS)



- Cardinal triad
  - hypertelorism
  - cleft palate or bifid uvula
  - arterial/aortic aneurysms and arterial tortuosity





LDS – Aortic pathology

- Progressive dilatation of the aorta
  - maximal at the level of the sinuses of Valsalva
- Risk of aortic dissection and rupture
  - major source of early mortality
  - mean age at death 26.1 years in the initially reported series

LDS – natural history

- At the time of diagnosis,
  - about two-thirds already have an aneurysm of the aortic root
  - one-fifth already have an aortic dissection
  - all LDS patients will eventually develop dilatation of the aortic root

LDS – arterial pathology

- Arterial involvement is widespread
- Arterial tortuosity, specifically of the head and neck vessels

LDS – other cardiac manifestations

- Associated anomalies
  - bicuspid aortic valve
  - atrial septal defect (ASD)
  - patent ductus arteriosus (PDA)
  - mitral valve prolapse

LDS - diagnosis

- Diagnosis is considered if typical clinical characteristics are present in the patient
- Family history is important
  - although only 25% of the newly diagnosed patients have an affected parent

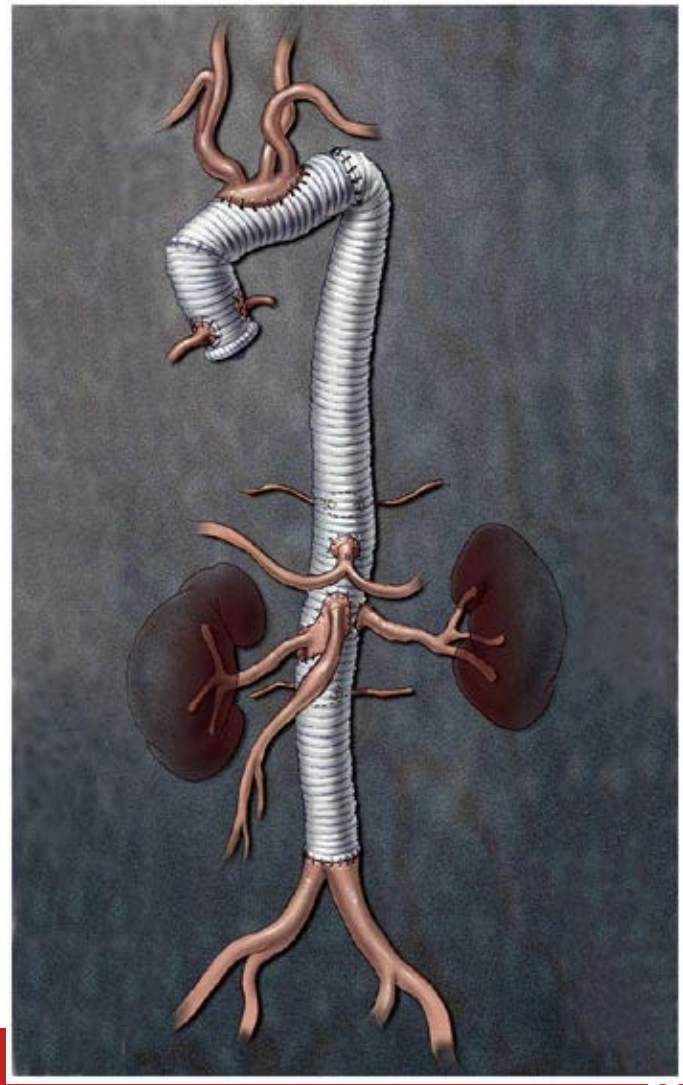
LDS-natural history

- In the largest study of Loeys-Dietz patients (n=90)
  - 98% had aortic root aneurysms
  - The mean age at death was 26 years (range 0.5 to 47 years)
    - thoracic aortic dissection (67%),
    - abdominal aortic dissection (22%)
    - intracerebral bleeding (7%)

LDS- medical management

- Patients should be treated to control hypertension to the lowest tolerated blood pressure
- **β**-blocker (atenolol, nadolol or propranolol)
  - Reduces hypertension and pulsatile wall stress.
  - Studied in patients with Marfan syndrome
- Angiotensin-II receptor blockers (ARBs) (losartan or irbesartan)
  - inhibits levels of TGF-β.
  - Selective blockade of AT1.
  - Studied in patients with Marfan syndrome

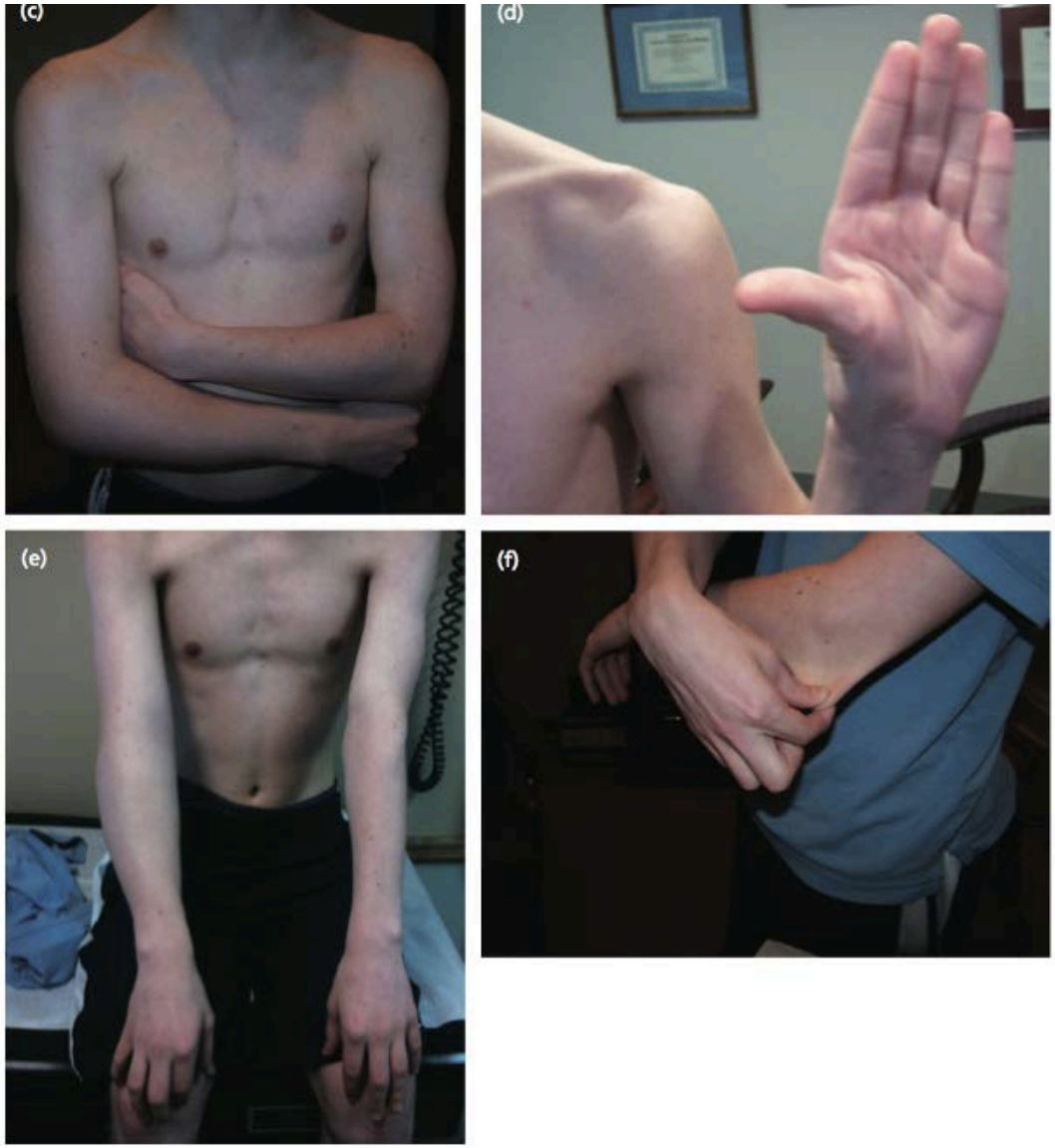
LDS - surgical management



LDS- exercise

- Circumstances to be avoided
  - Contact sports, isometric exercises, competitive sports or activities

EDS



When to operate?

- Marfan syndrome
  - ≥5.0 cm unless family history of dissection at < 5.0 cm, or growth >0.5 cm/ year
  - ≥4.0 cm if contemplating pregnancy
- Loeys-Dietz
  - ≥4.2 cm for dissection (family history of aortic dissection or growth >0.5 cm/ year),
- Ehlers Danlos, vascular form
  - Surgery is high risk because of tissue fragility

Conclusions

- The aortic valve should be preserved whenever possible
- The David operation has long term freedom from aortic reoperations
- Endovascular operations of the aortic arch are largely evolving and open proximal aortic surgery is still the gold standard.
- **Need for a team who are familiar with complex aortic pathology including surgical team, perfusion, CV anesthesia, blood bank, access to imaging- hybrid room, postop ICU care**

Thank you