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In patients born with CHD, dilatation of the aorta is a frequent feature at presentation and during follow up after surgical intervention.





Primary aortic dilatation is mainly associated with

- Coarctation of the aorta
- Bicuspid aortic valve
- Conotruncal abnormalities such as TOF,
 PA/VSD or Truncus Arteriosus
- Genetic syndromes with connective tissue disorders such as Marfan, Loeys-Dietz, Ehler-Danlos, Turner Syndromes.

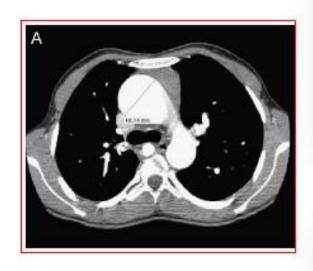


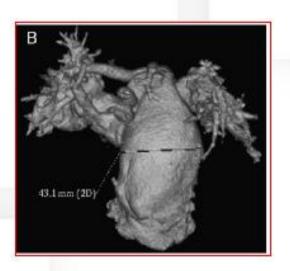


Aortopathy in Conotruncal Anomalies

14-year old boy with PA/VSD

6-year old girl with Truncus Arteriosus









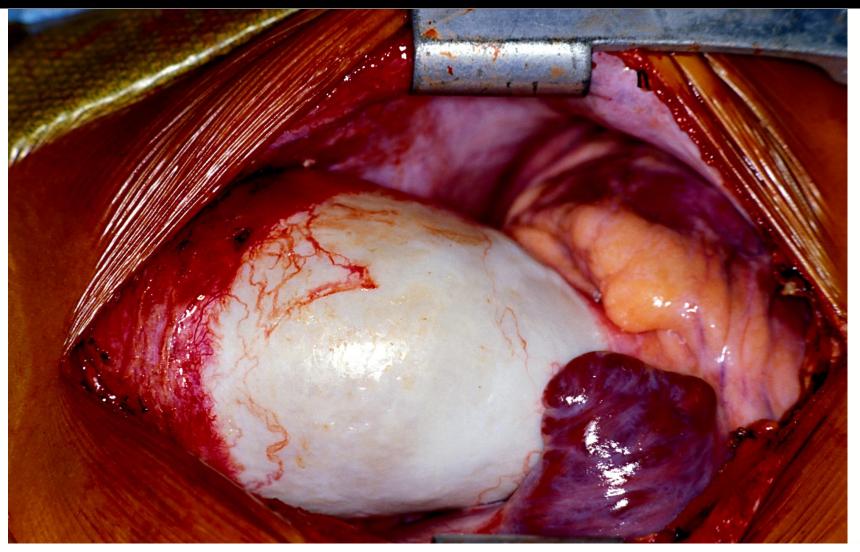


Other Rare Pathologies:

- Familial aneurysms
- Sporadic aneurysms including young patients without family history of aneurysms







Sporadic ascending aortic aneurysm in a 6-year old boy



The aortic root may be involved and actually in patients with Marfan Syndrome, the aneurysms usually begins in the aortic sinuses.







Secondary dilatation of the aortic root and ascending aorta can be seen after the congenital cardiac surgery when the original aortic root is replaced by the pulmonary root (e.g Ross Procedure, Arterial Switch, Damus-Kaye-Stansel)

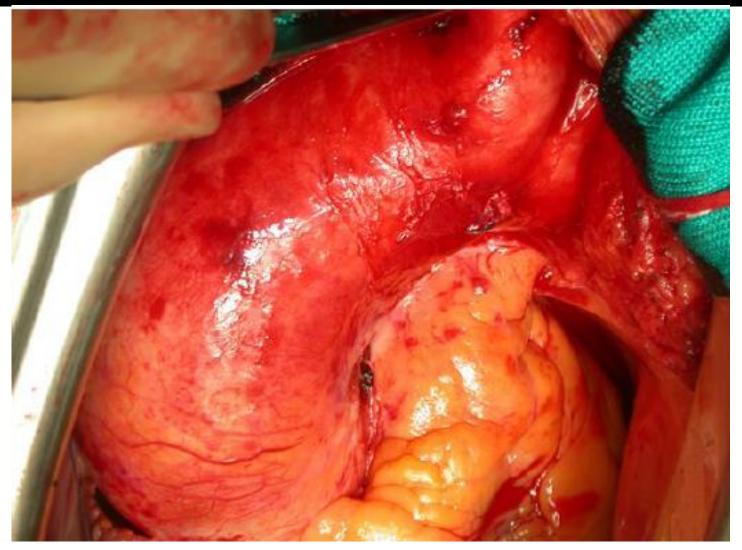




The magnitude of this problem continues to grow; as large cohort of surgically treated patients survive into adulthood.

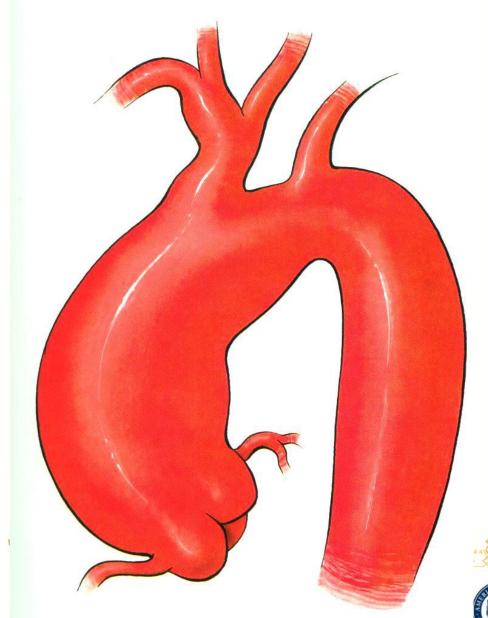






Dilated aorta post TOF repair





Post stenotic dilatation in patients with aortic stenosis





Aortopathy in CHD.

 In 88 CHD patients with dilated aorta with age of 3 weeks to 81 years (32 +/- 6 years) (48 males, 40 females), surgical biopsy aortic specimens was obtained, and cystic medial necrosis in the aortic media was observed in all of these patients.

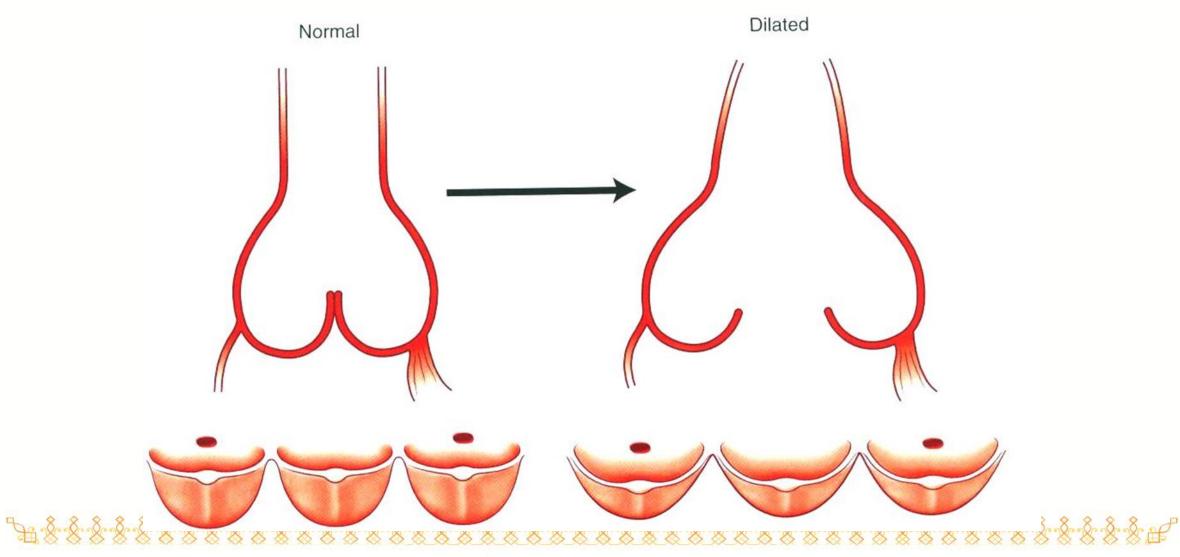
(Niwa K, Perloff JK, Bhuta SM, et al. Structural abnormalities of great arterial walls in congenital heart disease: light and electron microscopic analyses.

Circulation 2001;103:393-400)

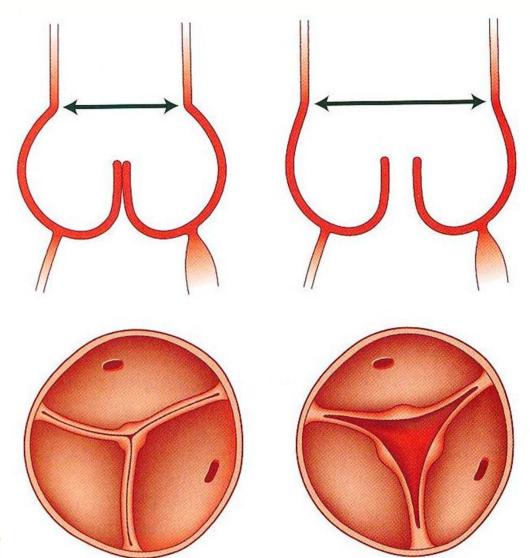








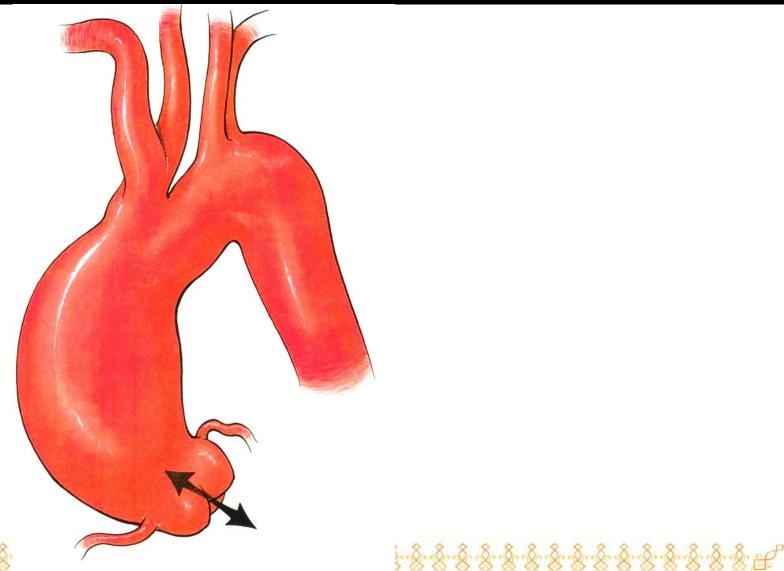














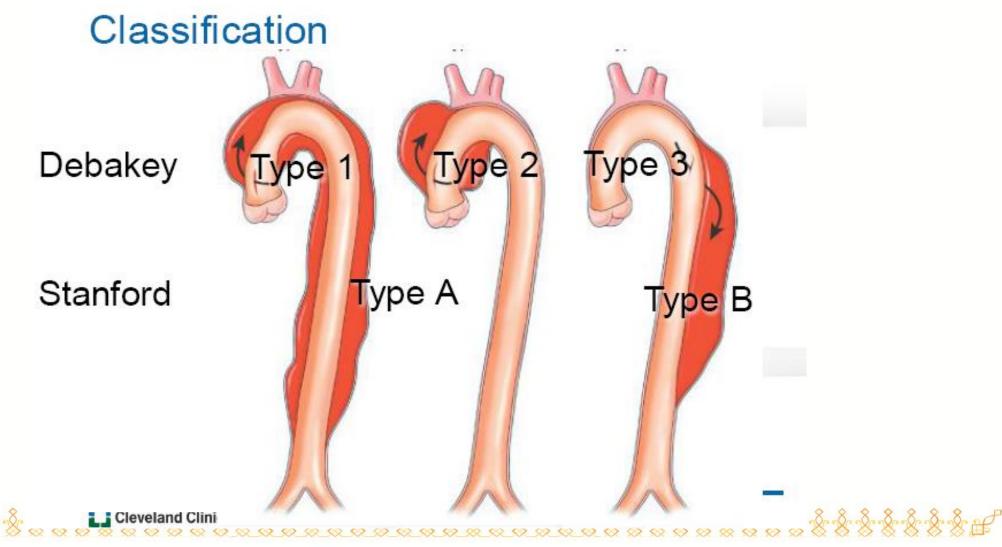


Ascending aortic aneurysm tend to increase in size and eventually rupture or cause aortic dissection.

The transverse diameter of the aneurysm is the most important predictor of rupture or dissection.

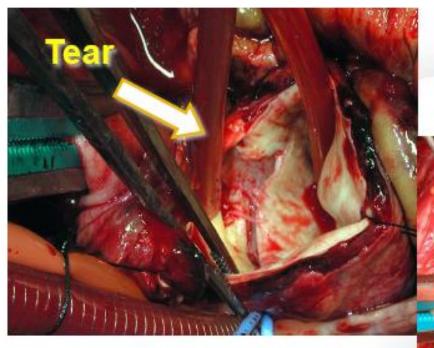


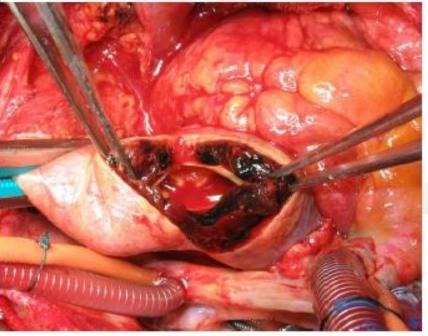






Acute Aortic Dissection

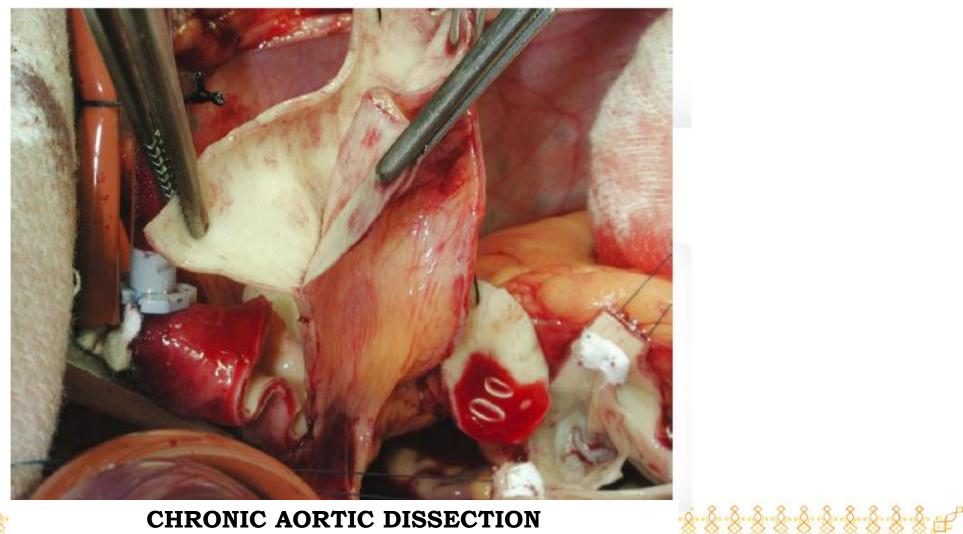














CHRONIC AORTIC DISSECTION



Complications / Causes of Death

- Rupture exsanguination
- Pericardial effusion tamponade
- Aortic insufficiency heart failure
- Branch vessel obstruction ischemia





Diagnostic Modalities



Recommendations on imaging of the aorta

Recommendations	Classa	Level ^b	Ref
It is recommended that diameters be measured at pre-specified anatomical landmarks, perpendicular to the longitudinal axis.	1	U	
In the case of repetitive imaging of the aorta over time, to assess change in diameter, it is recommended that the imaging modality with the lowest iatrogenic risk be used.	1	C	
In the case of repetitive imaging of the aorta over time to assess change in diameter, it is recommended that the same imaging modality be used, with a similar method of measurement.	1	U	
It is recommended that all relevant aortic diameters and abnormalities be reported according to the aortic segmentation.	1	U	
It is recommended that renal function, pregnancy, and history of allergy to contrast media be assessed, in order to select the optimal imaging modality of the aorta with minimal radiation exposure, except for emergency cases.		U	
The risk of radiation exposure should be assessed, especially in younger adults and in those undergoing repetitive imaging.	lla	В	72
Aortic diameters may be indexed to the body surface area, especially for the outliers in body size.	ПР	В	19,20, 46





The corner stone investigation is transthoracic echocardiography. It delineates:

* exact aortic valve anatomy

* aortic valve function and severity of stenosis and/or

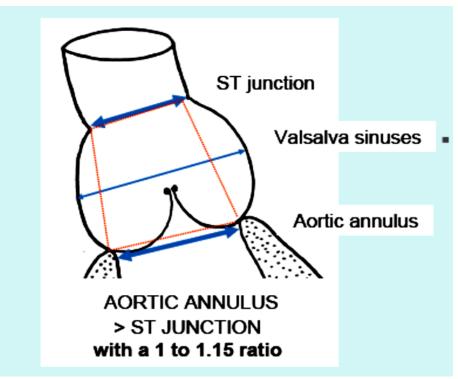
regurgitation

ventricular function

* measurement of the aortic root

(normally performed at 4 levels during enddiastole: aortic annulus, sinus, sinotubular junction, and proximal ascending aorta).





It may be difficult to assess the distal ascending aorta by TTE. Better image quality can be obtained with TEE.





Other modalities: CT Scan MRI Angiography





Comparison of methods for imaging the aorta

Advantages/disadvantages	TTE	TOE	СТ	MRI	Aortography
Ease of use	+++	++	+++	++	+
Diagnostic reliability	+	+++	+++	+++	++
Bedside/interventional use ^a	++	++	-	-	++
Serial examinations	++	+	++(+) ^b	+++	-
Aortic wall visualization ^c	+	+++	+++	+++	-
Cost	-	-			
Radiation	0	0		-	
Nephrotoxicity	0	0			

⁺ means a positive remark and—means a negative remark. The number of signs indicates the estimated potential value

CT = computed tomography; MRI = magnetic resonance imaging; TOE = transoesophageal echocardiography; TTE = transthoracic echocardiography.





^aIVUS can be used to guide interventions (see web addenda)

b+++ only for follow-up after aortic stenting (metallic struts), otherwise limit radiation

^cPET can be used to visualize suspected aortic inflammatory disease

- * MRI is preferable to CT scan because of no radiation since patients with dilated aorta will need multiple studies during their lifetime.
- ❖ When following patients with aortic dilatation not only the absolute diameter of the aortic segment is important but also the diameter indexed to body surface area. This allows serial comparison in growing children − can be related to Z-values.
 - ✓ Upper limit of normal 21 mm/m²
 - ✓ Significant dilatation >25 mm/m²
 - ✓ Significant risk of rupture >27.5 mm/m²





Recommendations on interventions on ascending aortic aneurysms

Recommendations	Class	Level⁵				
Surgery is indicated in patients who have aortic root aneurysm, with maximal aortic diameter ^e ≥50 mm for patients with Marfan syndrome.	-	U				
Surgery should be considered in patients who have aortic root aneurysm, with maximal ascending aortic diameters: • ≥45 mm for patients with Marfan syndrome with risk factors. ^d • ≥50 mm for patients with bicuspid valve with risk factors. ^{ef} • ≥55 mm for other patients with no elastopathy. ^{gh}	lla	U				
Lower thresholds for intervention may be considered according to body surface area in patients of small stature or in the case of rapid progression, aortic valve regurgitation, planned pregnancy, and patient's preference.	ШЬ	U				
Interventions on aortic arch aneurysn	15					
Surgery should be considered in patients who have isolated aortic arch aneurysm with maximal diameter ≥55 mm.	lla	C				
Aortic arch repair may be considered in patients with aortic arch aneurysm who already have an indication for surgery of an adjacent aneurysm located in the ascending or descending aorta.	Шь	U				
Interventions on descending aortic aneurysms						
TEVAR should be considered, rather than surgery, when anatomy is suitable.	lla	U				
TEVAR should be considered in patients who have descending aortic aneurysm with maximal diameter ≥55 mm.	lla	U				
When TEVAR is not technically possible, surgery should be considered in patients who have descending aortic aneurysm with maximal diameter ≥60 mm.	lla	C				
When intervention is indicated, in cases of Marfan syndrome or other elastopathies, surgery should be indicated rather than TEVAR.	IIa	U				

^{*}Class of recommendation.

All guidelines are based on diameters,
Aortic size index is not used up to now!

bLevel of evidence.

^eDecision should also take into account the shape of the different parts of the aorta. Lower thresholds can be used for combining surgery on the ascending aorta for patients who have an indication for surgery on the aortic valve.

❖ All patients with an aortic diameter of >40mm should be followed up regularly.

❖ When diameter is > 45mm and the patient has a family history of dissection or show rapid progression more frequent follow up is mandatory.





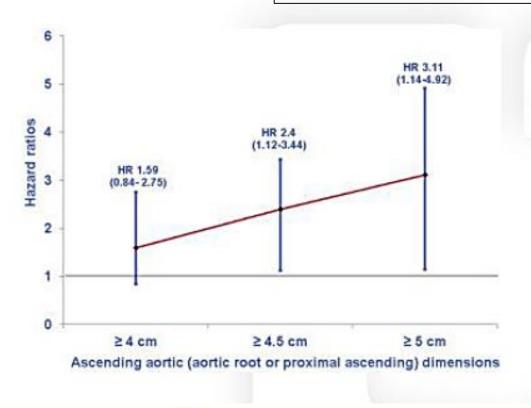
AORTIC SURGERY FOR BICUSPID AORTIC VALVES

- 1. Surgical intervention is recommended in asymptomatic patients with a bicuspid aortic valve and an aortic root or aortic ascending aorta ≥ 5.5 cm in size (Level B)
- 2. Surgical intervention is reasonable at 5 5.5 cm in diameter. If an additional risk factor is present for a ortic dissection (a ortic growth ≥ 0.5 cm/year or family history of a ortic dissection) (Level B) ? Also in Low surgical risk patients.
- 3. In patients with bicuspid aortic valve planned for surgery on the aortic valve, it is reasonable to intervene on the ascending aorta if size > 4.5 cm (consensus of expert opinion).

Risk of Death over Time by Size for

BAV

Still dissection can occur at smaller diameters [in a series of BAV patients 12.5% with dissection had aortic sizes <5 cm estimated at the time of surgery]

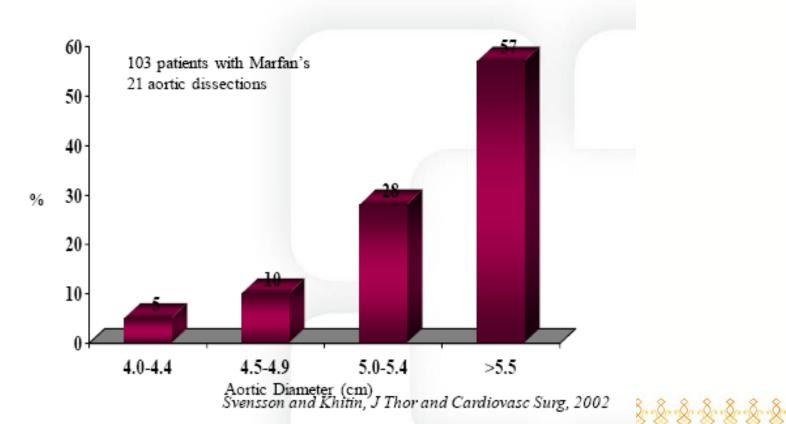








Aneurysm size vs dissection IN MARFAN SYNDROME







Unfortunately no guidelines exist for aortopathy in CHD. In ESC guidelines for ACHD only the following are mentioned:

•Surgical treatment for ascending aortic dilatation in coarctation may be considered when the diameter is >55mm (>27.5 mm/m²) or if there is rapid progression.

• Aortic surgery should be considered after ASO when the neo-aortic root in >55 mm.





OTHER PATHOLOGIES

There has been no case reports of aortic rupture in aortas <55mm in adult size patients. This diameter can be used as a cut off!

• A diameter of >60mm is considered a threshold at which one should consider surgical intervention particularly if there is evidence of rapid progression.





Aortic Dissection in Turner Syndrome.

- 20 individuals from the International Turner Syndrome Aortic Dissection Registry.
- Type A dissections occurred in 17 of 20 (85%) cases, and type B occurred in 3 cases.
- Of those with spontaneous aortic dissections, 18 of 19 (95%) had an associated cardiac malformation that included a bicuspid aortic valve.
- For those with type A dissections, the mean ascending aortic size index was 2.7 + 0.6 cm/m2.
- Recommendation: Individuals with Turner syndrome who are >18 years of age with an ascending aortic size index 2.5 cm/m2 should be considered for an aortic operation to prevent aortic dissection.





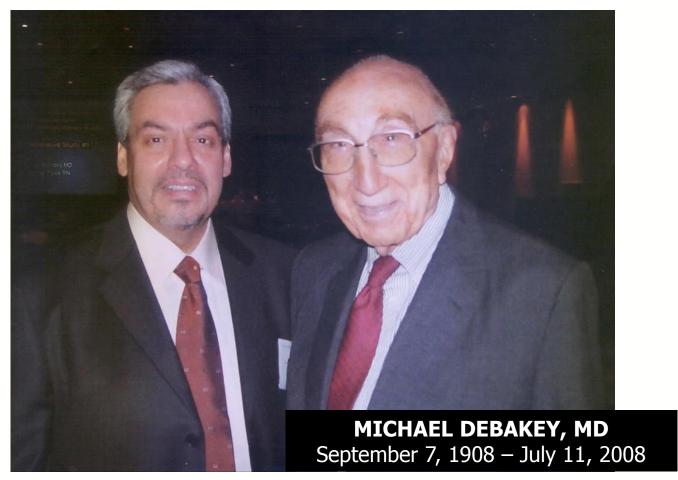


MEDICAL THERAPY

- Co-existent arterial hypertension in patient with CHD must be effectively treated.
- General principles of cardiovascular risk adjustment should be followed.
- The usefulness of medical therapy to prevent aortic expansion in normotensive patients with dilated aortas related to CHD is controversial.
- The goal of medical therapy have been to reduce shear stress on the dilated aortic segment by reducing BP and contractility (dp/dt)
 - **√** β Blockers
 - ✓ Angiotensin II receptor blocker/Losartan
 - ✓ Angiotensin converting enzyme inhibitors (ACE)/Lisinopril



SURGICAL THERAPY







- Valve sparing procedures are preferred in growing children and young adults whenever possible as the aortic valve is often normal.
- However usually a 'good size' prosthesis can be implanted as the aortic annulus is usually large.
- Surgery should be performed in centers with experience in this type of surgery.





SURGERY

- Cardiopulmonary bypass
- Arterial cannulation transverse arch or right axillary artery or femoral artery
- Venous Cannulation right atrium
- Temperature depends on the distal end anastomosis. This may require circulatory arrest.
- Cardioplegic cardiac arrest

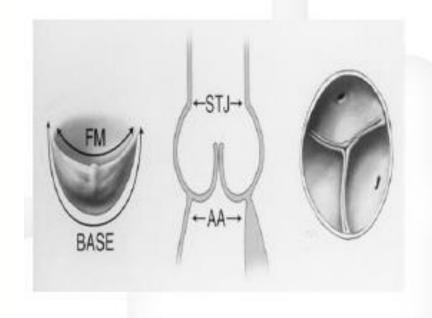




'CLASS' EVALUATION

Functional anatomy of the aortic valve complex

- C = Commissures
- L = Leaflets
- A = Annulus
- S = Sinuses
- S = Sinotubular junction







Aortic Valve Repair: Is it relevant?

- Safe, durable, and effective?
- Precludes the need for anticoagulation
- Eliminates the risk of prosthetic valve complications



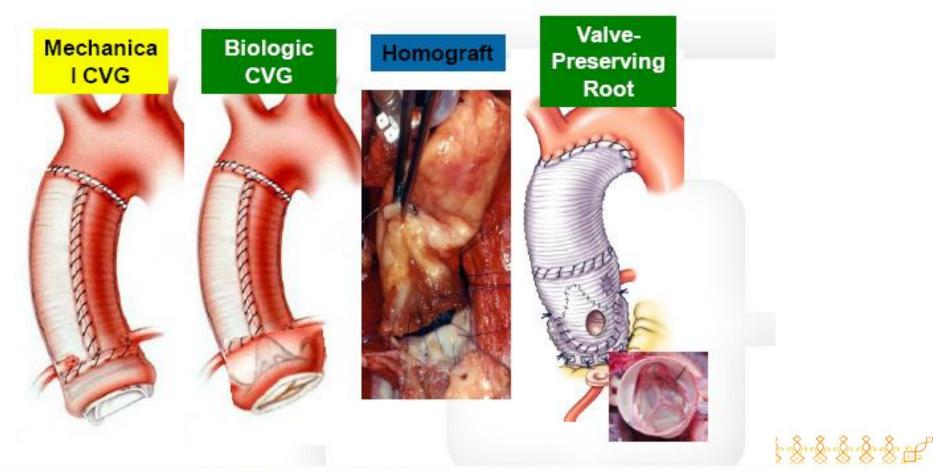


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Four Root Procedures



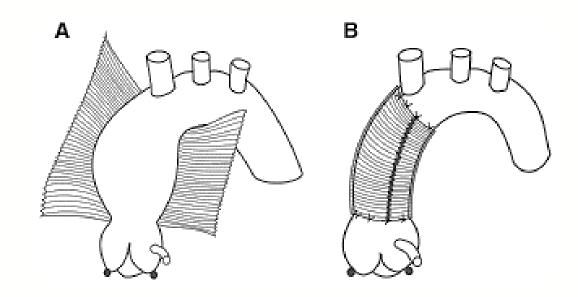




SURGERY

• For mild to moderate ascending aortic dilatation particularly with other cardiac surgery aortic wrapping has been performed.

• This can offer good lasting outcomes in selected patients preventing progression of aortic dilatation.



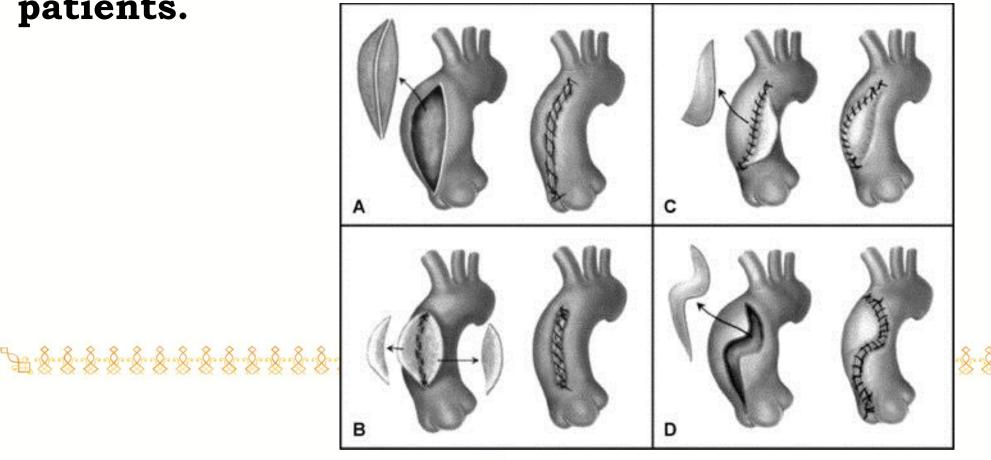




SURGERY

Reduction aortoplasty of ascending aortic aneurysms can be a possible alternative to graft replacement in selected

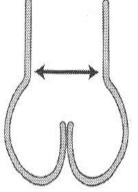
patients.

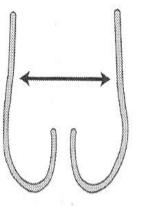


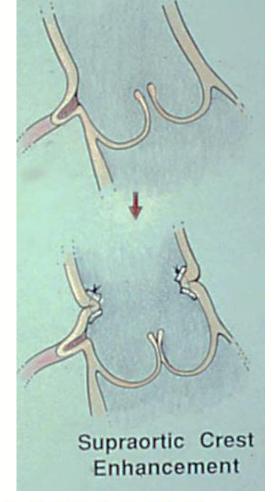
SURGERY

- When sinotubular junction is dilated (or effaced) the aortic valve can become incompetent in the presence of normal leaflets.
- All that is needed to re-establish valve competence is to restore the normal diameter of the sinotubular junction to allow leaflets to coapt again.





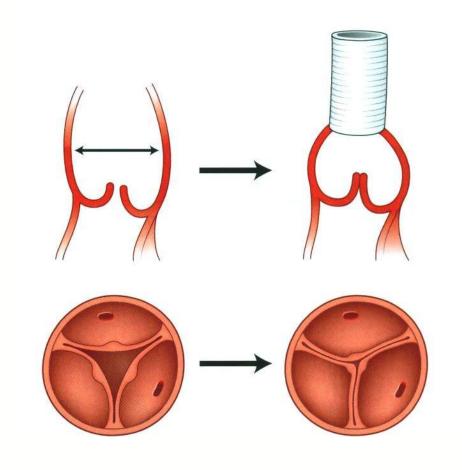






SURGERY

Replacement of the aneurysmal ascending aorta with a Dacron tube graft has been the traditional method of dealing with aortic aneurysms with normal sinuses. That can also adjust the sinotubular junction and restore aortic valve competence.







SURGERY

When the ascending aorta expands to develop an aneurysm it also becomes elongated.

Thus during its replacement the graft should be much shorter than the aneurysm (too long a graft can easily kink).





SURGERY

AORTIC ROOT ANEURYSMS

The aortic valve leaflets are often normal or minimally stretched or diseased and an aortic valve sparing operation should be performed.

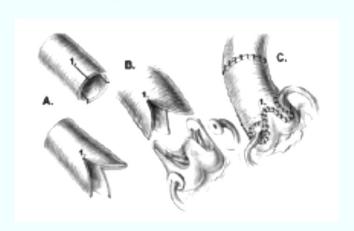




SURGERY

The 2 basic types of aortic valve – sparing operations are:

Remodeling or Yacoub technique

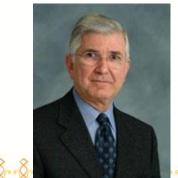




Reimplantation or David technique



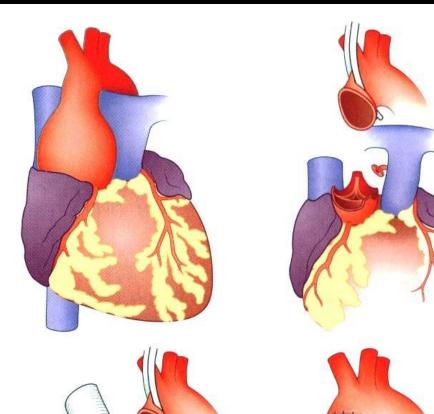








SURGERY

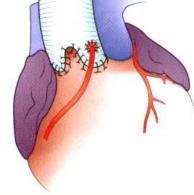


Aprtic Valve
Sparing
(remodeling)

Robicsak
Urbanski
The tailor of Gloucester
The Florida Sleeve
Hopkins
Lansac



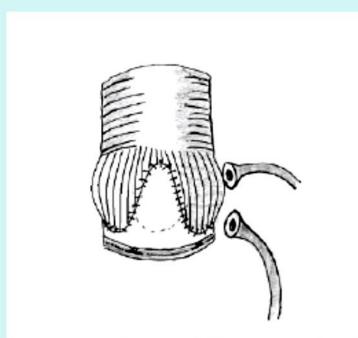






SURGERY

Lansac E (2005)





■To combine the advantages of remodeling and reimplantation procedures

by adding an external subvalvular prosthetic ring annuloplasty



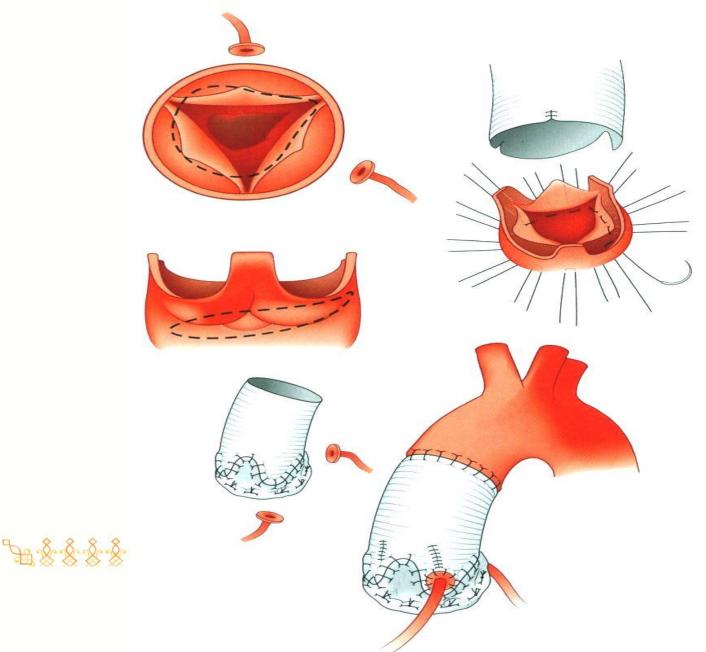


Characteristics of Remodeling

- Very anatomic and physiologic (sinuses, cusp motion)
- Increased risk of bleeding (long external sutures)
- Incresed risk of valve distortion (longe tongue, short valve remnants)



stability of results



Aortic Valve sparing (reimplantation)

- Cochran sinus reconstruction
- The David's number (I to V)
- The Stanford medication (DavidV)
- Gleason
- The Valsalva graft
 - ✓ Cameron
 - ✓ Mazzola
 - ✓ Maselli



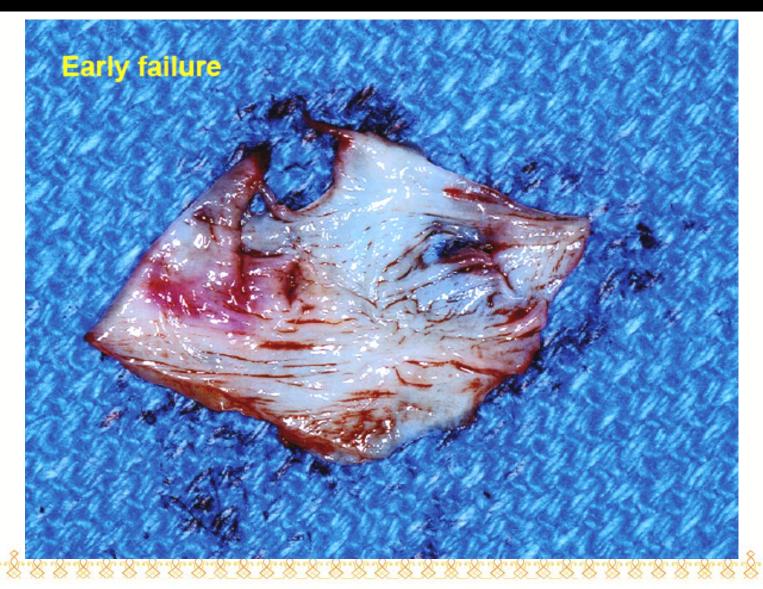
Reimplantation

- •Main differences over the remodeling procedure:
- 1. Annulus stabilization

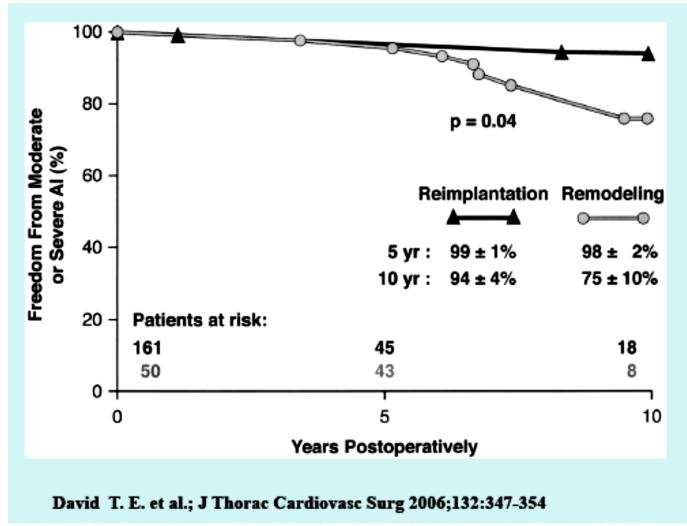
- Support of the aortic wall
- Less risk of suture bleeding















Aortic valve sparing operations in aortic root aneurysms: remodeling or reimplantation?

David Tian¹, Mohammad Rahnavardi^{1,2,3}, Tristan D. Yan^{1,2,3}

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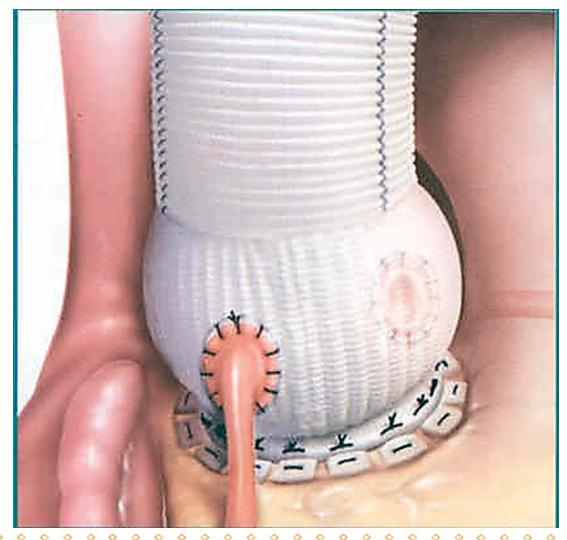
In conclusion, current evidence is in favour of the David technique over the Yacoub technique in pathologies such as MFS, acute type A aortic dissection, and excessive annular dilatation, which may impair aortic root integrity. Careful selection of patients for each technique and successful restoration of normal cusp geometry are the keys to success in AVS operations.







Annals of Cardiothoracic Surgery, Vol 2, No 1 January 2013



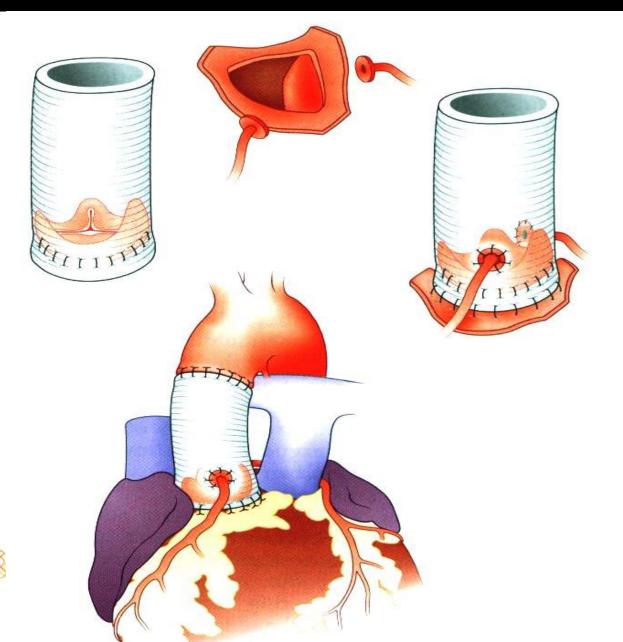




When the aortic valve is deemed unrepairable then a valve replacement becomes necessary.



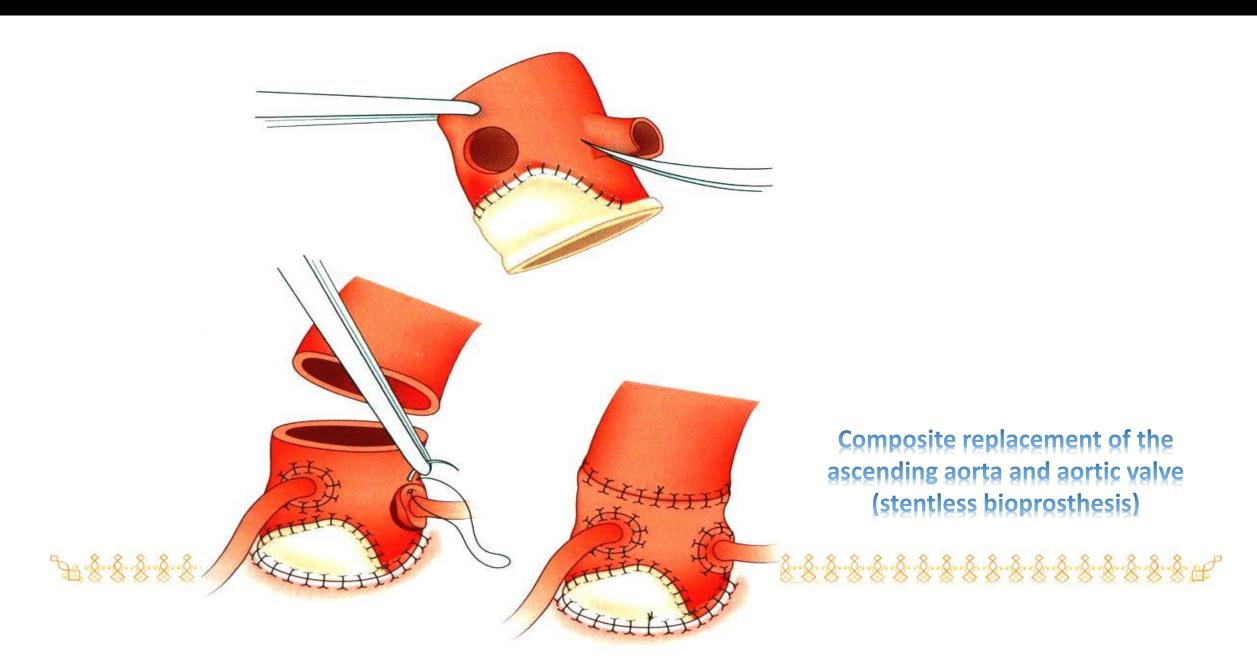




Composite replacement of the ascending aorta and aortic valve (stented bioprosthesis)

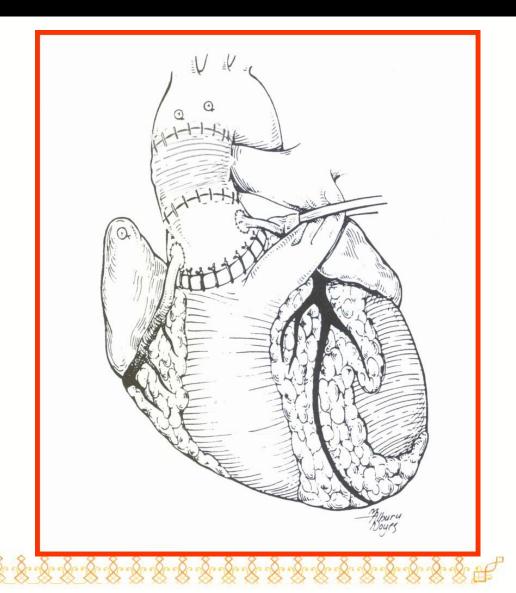




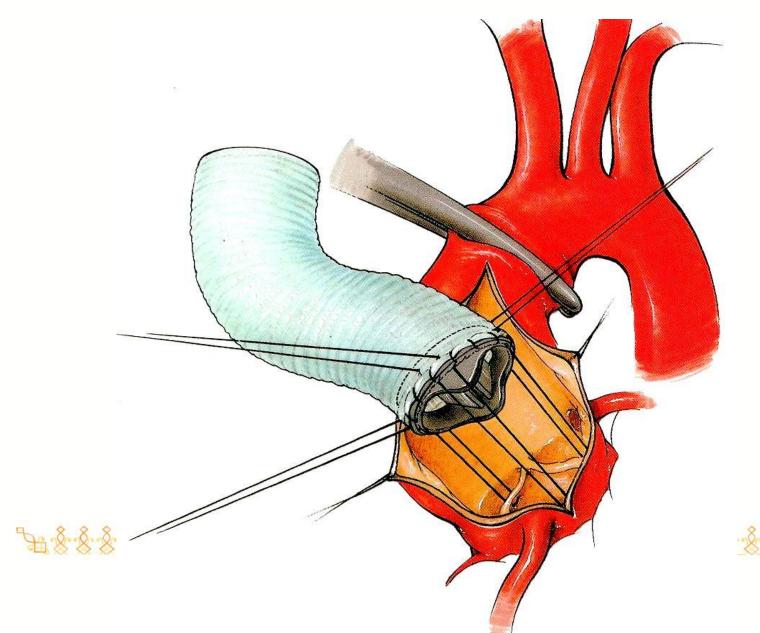


 Composite homograft aortic root replacement

• If aneurysm is long a Dacron tube can be interposed.







Composite replacement of the ascending aorta and aortic valve (Mechanical prosthesis)



ENDOVASCULAR PERCUTANEOUS THERAPY?!

- Access issue
- Elastic structure
- Landing zone
- Need for debranching



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Vascular Disease

Endovascular Repair of Ascending Aortic Dissection

A Novel Treatment Option for Patients Judged Unfit for Direct Surgical Repair

Qingsheng Lu, MD, Jiaxuan Feng, MD, Jian Zhou, MD, Zhiqing Zhao, MD, Junmin Bao, MD, Rui Feng, MD, Liangxi Yuan, MD, Xiang Feng, MD, Lefeng Qu, MD, Yifei Pei, MD, Zhijun Mei, MD, Zaiping Jing, MD, PhD

Shanghai, China

Objectives This paper sought to report the outcomes of patients who are considered unfit for urgent surgical repair of as-

cending aortic dissections (AADs) who were treated using a novel endovascular repair strategy.

Background AAD is best treated by direct surgical repair. Patients who are unable to undergo this form of treatment have poor prognoses. Previously, clinical case reports related to endovascular repair of AAD have been controversial.

Methods

Between May 2009 and January 2011, 41 consecutive patients with AAD were treated in our institution. Fifteen patients were considered poor candidates for direct surgical repair and subsequently underwent the endovascu-

lar repair.

Results The nature of the referral process to our tertiary care facility made the median time from aortic dissection onset

to treatment 25.5 days (range: 6 to 353 days). Dissections in 5 patients (33.3%) were considered acute, and those in 10 patients (66.7%) were considered chronic. The rate of successful stent-graft deployment was 100%, and there were no major morbidities or deaths in the perioperative period. Median follow-up was 26 months (range: 16 to 35 months). One new dissection occurred in the aortic arch at 3 months and was treated with a branched endograft. Significant enlargements of true lumens and decreases of false lumens and overall thoracic

aorta were noted after the procedures.

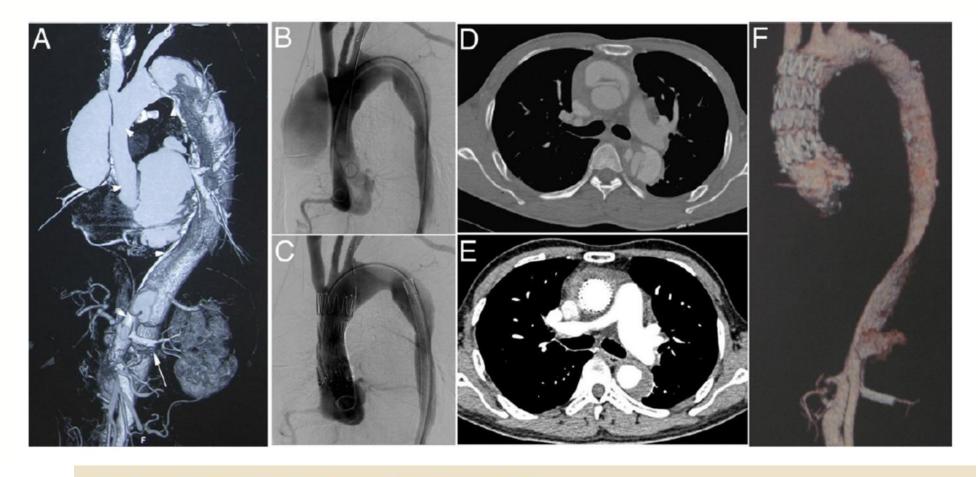
Conclusions Endovascular repair of AAD was an appropriate treatment option in patients who were considered poor candi-

dates for traditional direct surgical repair by the clinical criteria used in our institution. A larger series of cases with longer follow-up is needed to substantiate these results. (J Am Coll Cardiol 2013;61:1917-24)

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Ascending Aortic Dissection with Compromised Branch Artery





Acquired Cardiovascular Disease

Roselli et al

Endovascular stent grafting for ascending aorta repair in high-risk patients

Eric E. Roselli, MD, Jahanzaib Idrees, MD, Roy K. Greenberg, MD, Douglas R. Johnston, MD, and Bruce W. Lytle, MD

Objectives: Standard treatment of ascending aortic pathology is open repair, but some patients are too high risk. Thoracic endovascular aortic repair (TEVAR) of the ascending aorta has been used as an alternative. Our objectives were to characterize patients, describe repair methods, and assess outcomes.

Methods: From 2006 to 2014, 22 patients underwent supracoronary ascending TEVAR for acute Type A dissection (n = 9), intramural hematoma (n = 2), pseudoaneurysm (n = 9), chronic dissection (n = 2), or aorta-cardiac fistula (n = 2). Mean age was 71 ± 13 years and the maximum proximal aortic diameter was 6 ± 1 cm. Devices were delivered via a transfemoral (n = 10), transapical (n = 7), or axillary (n = 5) artery approach. The proximal landing zone was at the sinotubular junction in 14 patients, mid to distal ascending aorta in 3 patients, and surgical graft from previous ascending repair in 5 patients. More than 1 device was used in 15 patients. Imaging and engineering analysis was performed for all patients.

Results: There were 3 hospital deaths (13.6%) (tamponade in 1 patient, bleed from left atrial fistula in another patient). One patient had partial occlusion of the left coronary artery requiring open conversion and died later from multiorgan failure. One patient required early open conversion for retained delivery system. There were 3 strokes, 2 myocardial infarctions, and 2 tracheostomies, but there was no new-onset renal failure. Median follow-up was 12 months. Six patients developed type 1 endoleak: 2 were treated endovascularly, 1 with open repair, 1 resolved, 1 refused treatment, and 1 is being watched. In 2 patients, initial TEVAR was performed as a bridge for ruptured high-risk dissection and were later converted to open repair. Reoperations also included removal of stent graft due to distal migration and repair of left ventricular pseudoaneurysm. There were 3 late deaths. Actual survival at 30 days, 1 year, and 5 years was 86%, 80%, and 75%, respectively.

Conclusions: Ascending TEVAR is a feasible alternative to medical therapy for repair of acute and chronic ascending disease in high-risk patients. Development of devices dedicated to treat ascending aortic pathology is needed to improve outcomes. (J Thorac Cardiovasc Surg 2015;149:144-54)





THANK YOU!



