Management of Ascending Aortic Aneurysms
DECLARATION OF CONFLICT OF INTEREST

☐ None
Pathophysiology of Aortic Aneurysms

- **Mechanisms**
  - Congenital aortic fragility from genetic predisposition
  - Mechanical stress

- **Aortic media affected by damage and repair events**
  - Excessive injury: Valve dysfunction, Hypertension, Age
  - Impaired repair: Connective tissue disorders
Aetiology of Ascending Aortic Aneurysms

- Degenerative
- Syndromic
  - Marfan syndrome
  - Loeys-Dietz
  - Ehlers-Danlos
  - Turner / Noonan
- Family (non-syndromic)
- BAV
- Aortic valve disease
Management of Ascending Aortic Aneurysms

- Medical Treatment
- Follow-up by Imaging Techniques
- Predictors of Complications
- Surgical Treatment Indications
- Type of Surgery
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Medical Management

• Aggressive blood pressure control

Beta-blocker therapy preferred.

If beta blockers are contraindicated, poorly tolerated, or additional blood pressure control required:
- angiotensin-converting enzyme inhibitors
- angiotensin receptor blockage (ARBs)

• Follow-up of aortic size by close imaging technique tests

Deficiency of fibrillin-1

Excessive signaling TGF-β

Angiotensin II-receptor blocker (ARB) inhibits TGF-β signaling

Several trials are coming
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Follow-up by Imaging Techniques

Aortic root:
2D-echo annually and every 6 m when absolute size >45 cm or significant AR.

Tubular AA:
MRI and CT in advanced dilatation and to evaluate the entire aorta.
In clinical practice:

- not significant change if < 3-4 mm
- do not take surgical indications using reports

Frequent mistakes:

- Not to use the same imaging technique

- Axial measurements may exaggerate diameter > 20% in elongated AA
  
  *Use reconstruction so that it be perpendicular to the longitudinal axis of the aorta*

- Motion artifacts can adversely affect the resolution of CT images, producing changes as great as 7.5% to 27.5%.
  
  *Gated tomographic angiography*
Accurate measurement of ascending aorta diameter

Requirements:
- Same imaging technique
- Same aorta level
- Side by side measurement
- Include/exclude the aorta wall
- Multiplanar modality
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Yearly rates of rupture, dissection, or death related to thoracic aortic size
Survival with Thoracic Aneurysms

Eleftiarides et al. Radiology 1999, 211:889
• Limited value of aortic size
• < 50 mm surgery would fail to prevent 40% of AD
• Dilatation is only one manifestation of aortic wall weakness
Risk of Aortic Rupture related to Diameter and BSA

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= low risk (~4% per yr)
= moderate risk (~8% per yr)
= severe risk (~20% per yr)

Indices for normalising diameters

- Z scores > 3
- Aortic root diameter ratio: Observed/maximum predicted > 1.3
- AA area / height > 10
- AA diameter / BSA > 2.75 mm/cm²
514 patients, AA>35 mm. 70 BAV, 445 TAV
Aortic growth rate (1.9 vs 1.3 mm/y; p<0.01)

Despite faster rates of growth, BAV has a similar rate of aortic complications than TAV!
642 patients
Follow-up: 9y

Mortality 4% vs 3%
Cardiac surgery 22%
Aortic dissection 0.4%

Outcomes in Adults With Bicuspid Aortic Valves

JAMA 2008; 300 (11) : 1317

Nikolaos Tzemos, MD
Judith Therrien, MD
James Yip, MD
George Thanassoulis, MD
Sonia Tremblay, MD
Michal T. Jamorski, BSc
Gary D. Webb, MD
Samuel C. Siu, MD, SM
Aortic complications of BAV and implications on management

- Dilation of the aorta is an independent risk factor for surgery.

- Real aortic complication is aortic dissection.

- Risk of dissection with smaller diameters?
Marfan Syndrome vs BAV

- Marfan S has a high lifetime risk (40%) of aortic dissection.

- BAV disease carries a 6.1% lifetime risk of aortic dissection (9-fold higher than general population).

- BAV is 100 times more common than Marfan S
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Indications for elective surgery in Ascending Aortic Aneurysm

- Aortic diameter > 55 mm
- Evaluate comorbidities
- Risk of surgery
- Age
Indications for elective surgery in the Marfan Syndrome

Ascending aortic diameter

≥ 50mm

≥ 45 mm with risk factors:

First degree family relative with AA dissection / rupture
Concomitant indication for aortic valve surgery
Ratio of aortic diameter to BSA ≥ 2.75 cm/m² (children)
Expansion rate ≥ 2 mm/y *

≥ 40 mm if pregnancy is desired and aortic valve replacement is not required.
Indications for elective surgery in the Bicuspid Aortic Valve

Aortic diameter ≥ 55 mm

Aortic diameter ≥ 50 mm if:

- Aortic coarctation, corrected or not
- First-degree family relative with Ao dissection/rupture
- Small body size:
  - Aortic area/height > 10 cm²/m
  - Aortic diameter / BSA > 2.75 cm/m²
- Severe AS or AR without surgical criteria (?)
- Expansion rate ≥ 2 mm/y *

Aortic diameter > 45 mm with concomitant indication for elective AVR
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Dilated ascending aorta

Diameter <55 mm
(<50 mm Marfan syndrome and bicuspid valves) *

- β blocker treatment
- Control of associated risk factors
- Serial imaging follow-up

Growth rate: ≥ 2 /year

Diameter ≥55 mm

Surgical treatment

Supravalvular aortic aneurysm

Supracoronary tube graft

Aortic root aneurysm

Macroscopically normal valves

Valve sparing

Structural valve deterioration

Composite graft replacement

Diameter ≥55 mm
Results of surgery for aortic root aneurysm in patients with Marfan syndrome

Nilto Carias de Oliveira, MD
Tirone E. David, MD
Joan Ivanov, PhD
Susan Armstrong, MSc
Maria J. Eriksson, MD
Harry Rakowski, MD
Gary Webb, MD

Similar survival
Lower rates of valve related complications

J Thorac Cardiovasc Surg 2004;125,789-96
Conclusions

- Although both Marfan syndrome and BAV present wall abnormalities that cause aortic wall weakness and its progressive dilatation, the risk of dissection or aortic rupture is clearly higher in Marfan Syndrome.

- In Marfan syndrome, prophylactic surgery of the ascending aorta has changed the natural history of this disease. The indication for surgery when the diameter is 50 mm appears to be appropriate. Nevertheless, in patients with risk factors, surgery should be considered when diameter is 45 mm.

- In BAV, without severe dysfunction of the valve, timing of ascending aorta surgery (50 vs 55 mm) should be individualised considering the presence of aortic coarctation, body size, progressive dilatation, age and comorbidities. With concomitant indication of AVR, aortic surgery should be performed when aortic diameter is > 45 mm.
Conclusions

- Crucial points in the correct management of ascending aortic aneurysms are:

  - a) Adequate clinical assessment of risk factors, scheduled follow-up and medical treatment.

  - b) Excellent imaging techniques using the best methodology to improve accurate measurements of the aorta size. “Surgery should not be indicated based only on test reports”

  - c) Experts surgeons with high skill in this elective aortic surgery.