Surgery for Aortic Root Dilatation Following Repair of Congenital Heart Disease

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Nemours

No disclosures

Root aneurysm / CHD

- Increasing recognition during follow up of patients undergoing interventions for conotruncal anomalies
- Surgical indication not well defined due to unknown natural history
  - risk of rupture or dissection
- Multiple challenges
  - Technically complex
  - Exposure
  - Multiple operations
  - Challenging physiology
  - Surgical risk?

Aortic program

Neo-Aortic Root Dilation and Valve Regurgitation Up to 21 Years After Staged Reconstruction for Hypoplastic Left Heart Syndrome

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Philadelphia, Pennsylvania

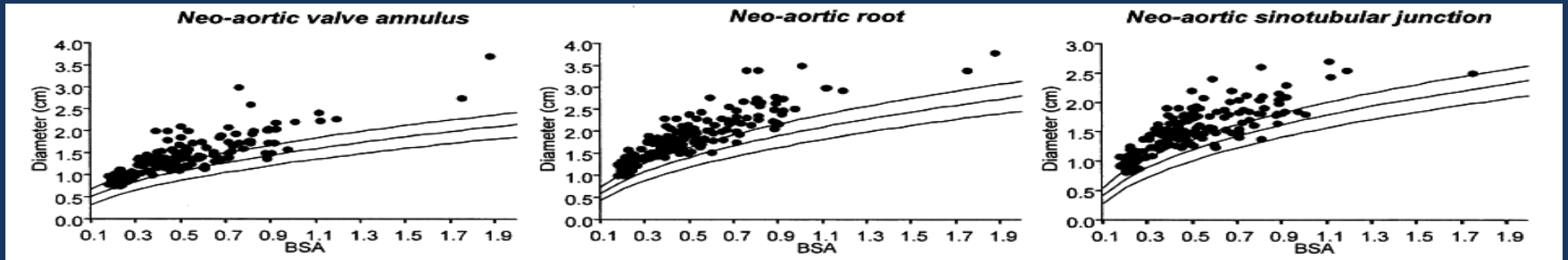
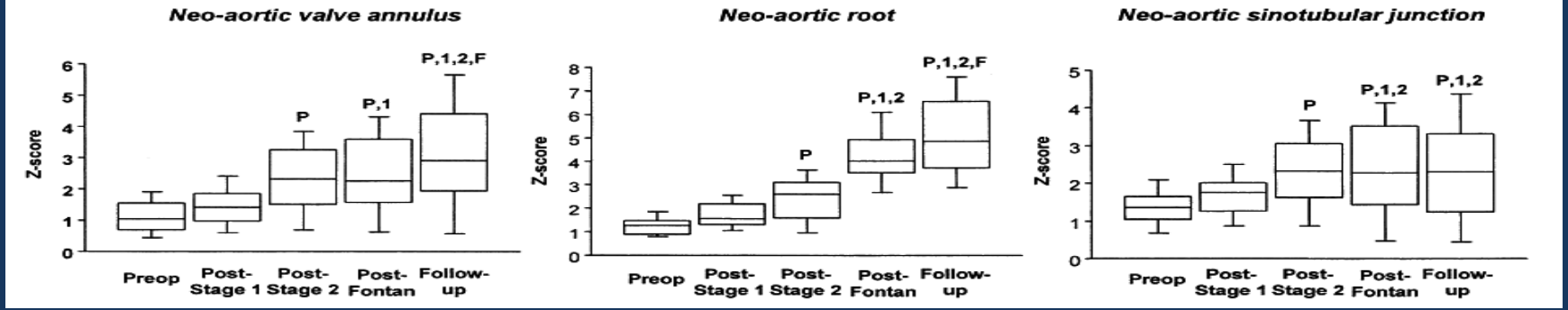


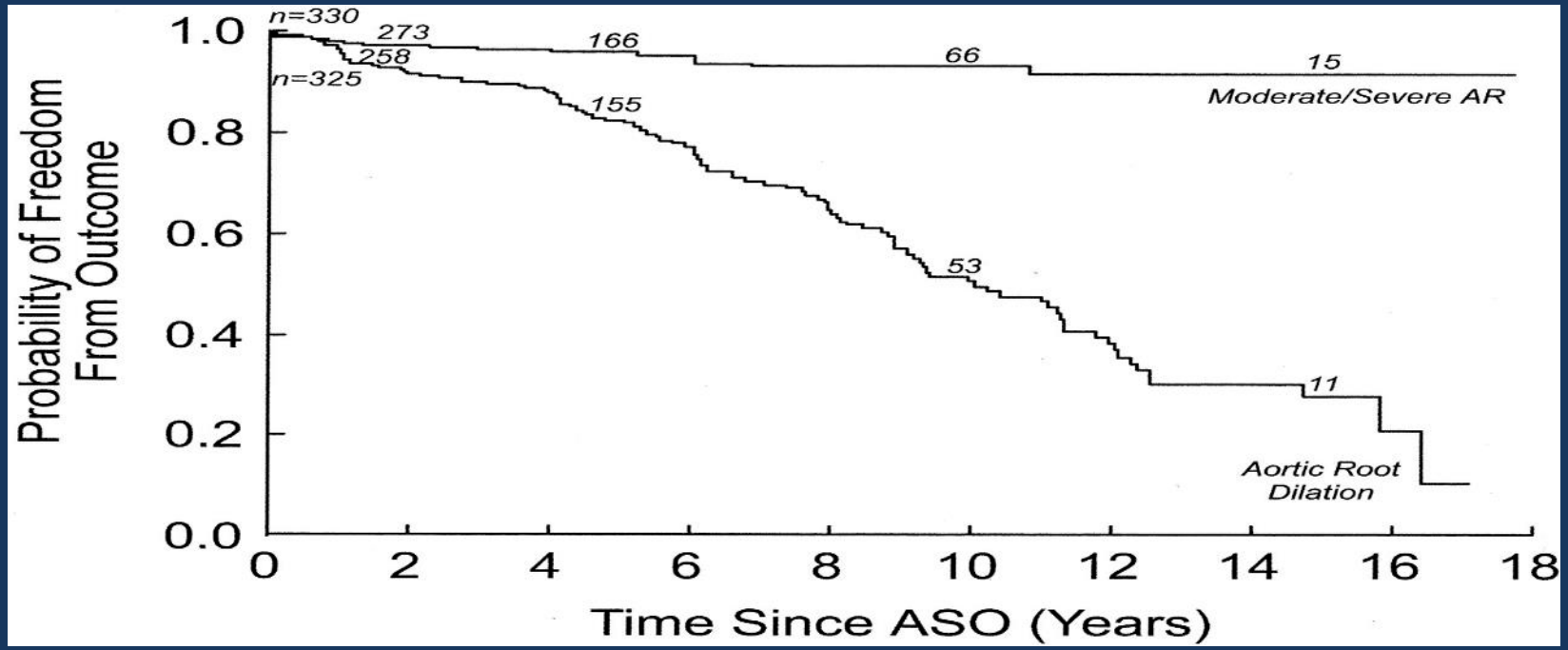
Figure 2. The diameters of the neo-aortic valve annulus **(left)**, root **(middle)**, and sinotubular junction **(right)** are plotted against body surface area (BSA) and compared with the normal distribution (mean with 95% confidence intervals).



JACC 2003;42:533-40

Long-Term Predictors of Aortic Root Dilation and Aortic Regurgitation After Arterial Switch Operation

Marcy L. Schwartz, MD; Kimberlee Gauvreau, ScD; Pedro del Nido, MD; John E. Mayer, MD; Steven D. Colan, MD



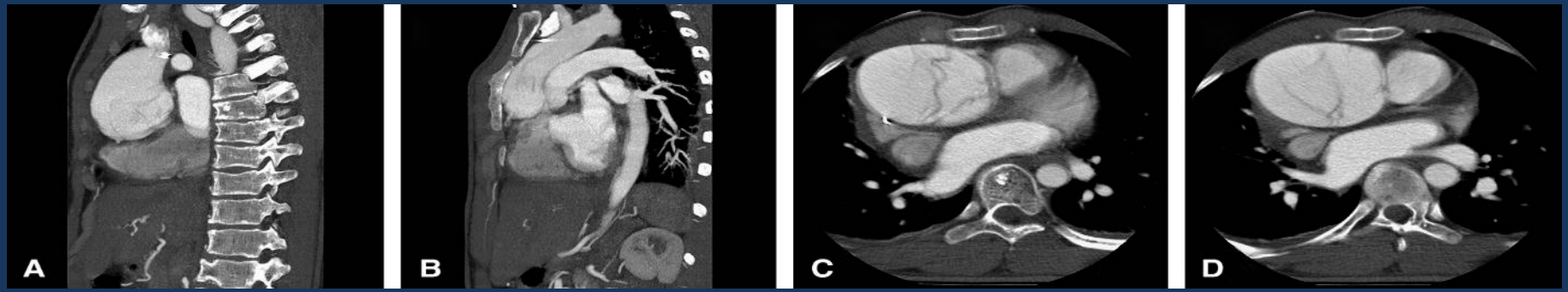
Schwartz, M. L. Circulation 2004;110:II-128-II-132

Congenital Heart Disease

Aortic Root Dilatation in Adults with Surgically Repaired Tetralogy of Fallot

A Multicenter Cross-Sectional Study

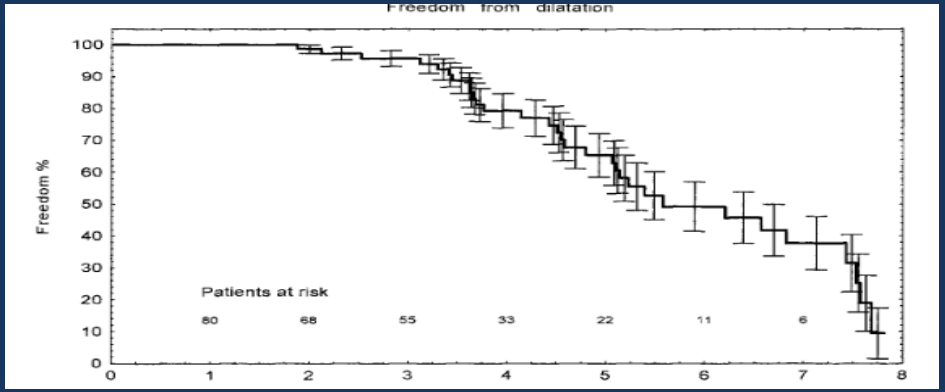
- Aortic root dilatation prevalence (30% > 4 cm; O/E >1.5 is 6.6%). Asc aorta >4 cm 19%
- Associated with older age at surgery, pulmonary atresia and mod- severe AI
- Mod-severe AI in 3.5% cases
- Histology strikingly similar to Marfan syndrome.
- Several case reports of dissection



Mongeon. Circulation. 2013;127:172-179

Fate of the Aortic Root Late After Ross Operation

Giovanni Battista Luciani, MD; Gianluca Casali, MD; Alessandro Favaro, MD;
Maria Antonia Prioli, MD; Luca Barozzi, MD; Francesco Santini, MD; Alessandro Mazzucco, MD



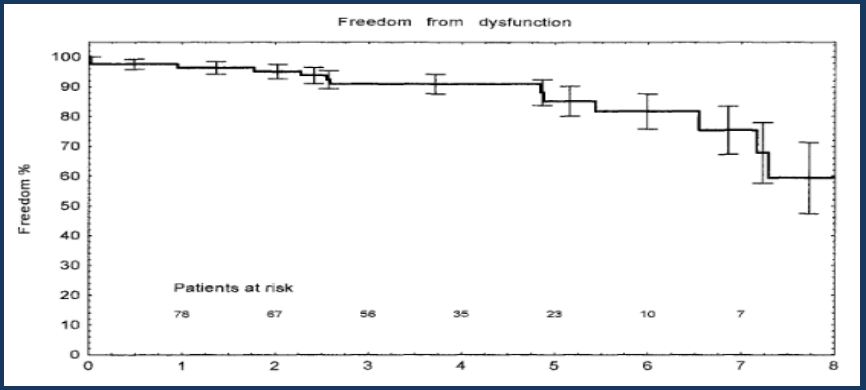


TABLE 3. Risk Factors for Autograft Dilatation

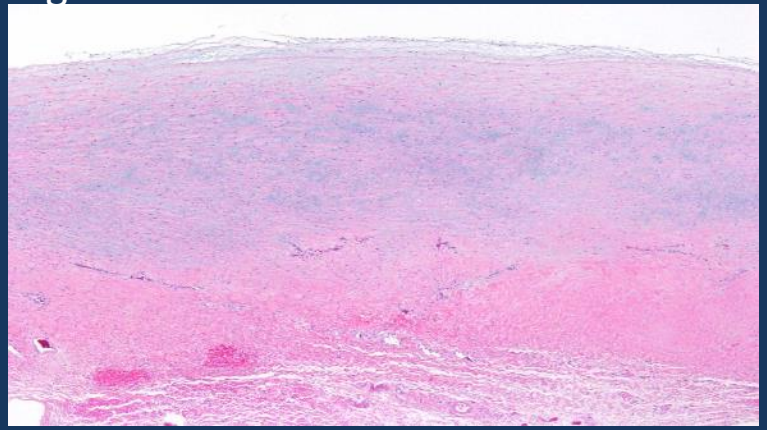
Cox proportional hazard	Beta factor	Standard Error	*P*
Age	−0.07	0.04	0.05
Preoperative sinus Valsalva diameter	0.24	0.12	0.02
Root replacement technique	2.80	1.27	0.03
Pericardial strip buttressing	−2.61	1.33	0.04

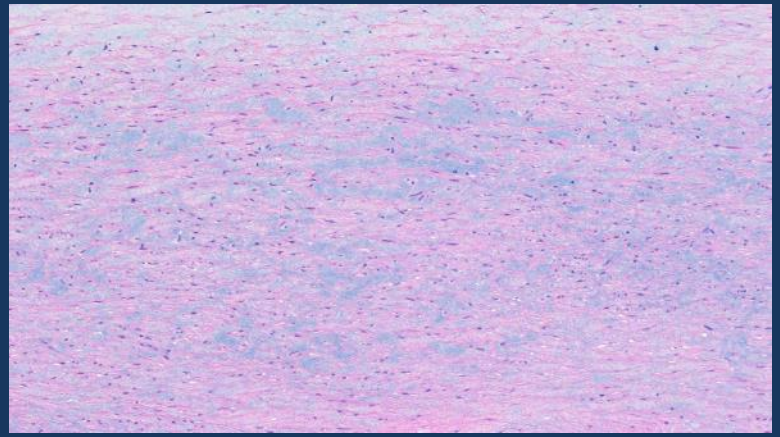
TABLE 4. Risk Factors for Autograft Dysfunction

Cox proportional hazard	Beta Factor	Standard Error	*P*
Sex (female)	3.51	1.14	0.002
Preoperative Sinus Valsalva diameter	0.34	0.16	0.04
Follow-up Sinus Valsalva diameter	0.63	0.21	0.003
Follow-up sinotubular junction diameter	0.77	0.32	0.02

Luciani Circ 2003;108[suppl II]:II-61-II-67

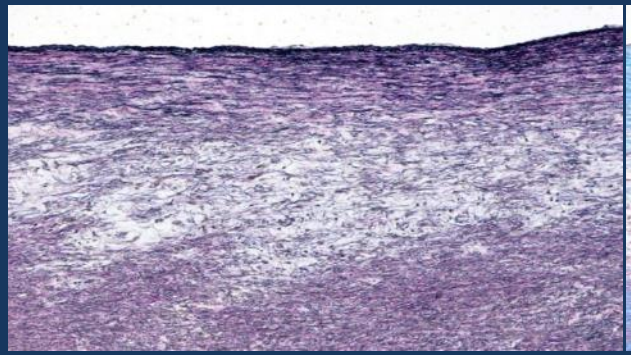
Aortic program





Sections of aorta show accumulation of myxoid material (extracellular ground substance) within the media.

[H&E, original magnification 40X (left) and 100X (right)]



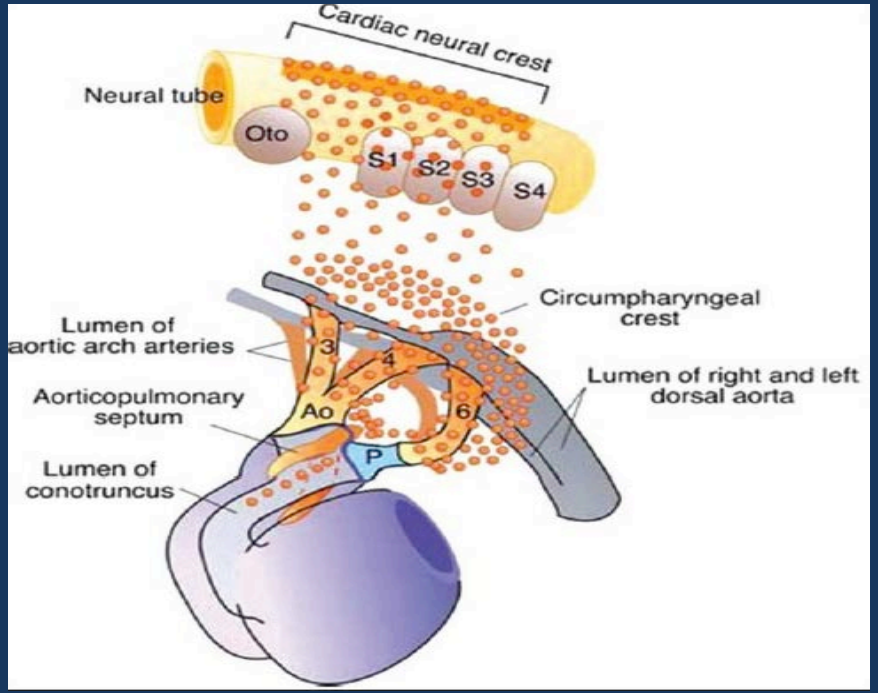
Elastin stain of aorta shows disruption and loss of elastic fibers within the media. [Elastin stain, original magnification 100X]

Aorta stained with trichrome stain (left) and smooth muscle actin immunohistochemistry (right) shows expanded zones of extracellular ground substance and disruption and loss of smooth muscle cells within the media. [Trichrome stain (left), SMA (right), original

Risk factors for Neo Aortic root dilatation

- Pulmonary artery banding
- Presence of a VSD
- Taussing-Bing anomaly
- Coronary transfer technique?
- Disruption of STJ?
- Disruption of vasa vasorum?

Etiology



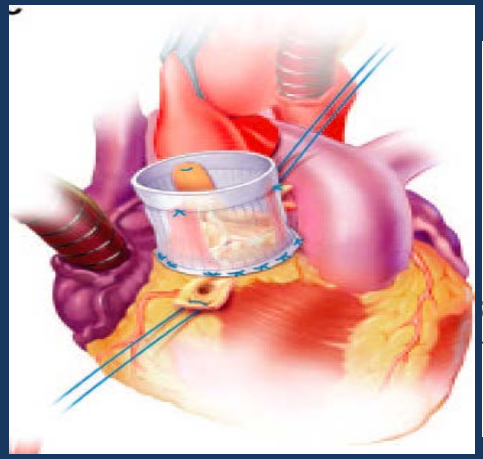
- Aneurysm panel, MYLK
- Mutations
  - TGFBR2
  - SMAD 3 mutation
  - MYH11
  - MYLK variant

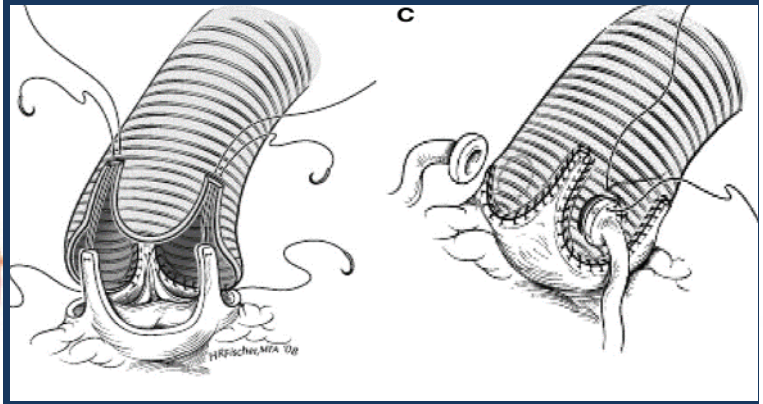
Aortic Aneurysm in CHD

- Redo sternotomy (2-4th)
- Aortic arch replacement in some cases
- Fontan physiology (preserve lung function, cannulation, blood utilization)
- Multiple systemic-pulmonary collaterals
- Increased difficulty of valve sparing procedure after Lecompte maneuver
- Consider associated conditions (decreased ventricular function)

Intervention and Timing

- Lack of natural history data
- Do implications about aneurysm size apply?
- Lack of experience with interventions in these patient population (post arterial switch, Fontan)
- Indications are based on extrapolations from Marfan experience and/or associated pathology

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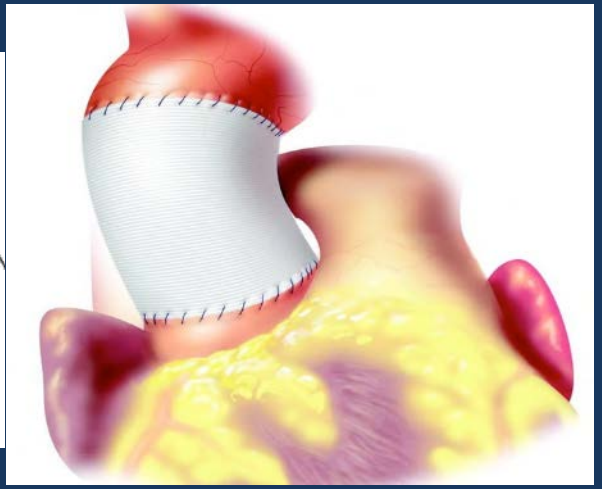
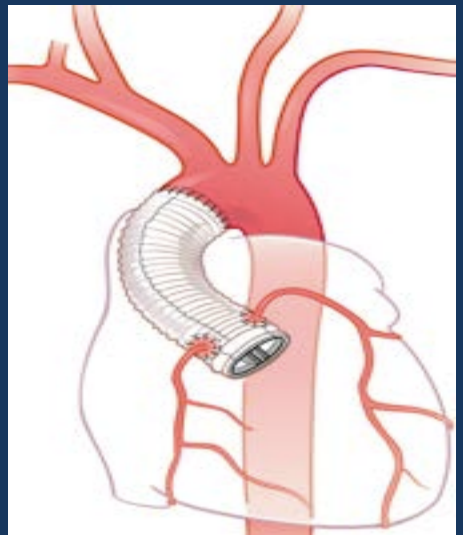
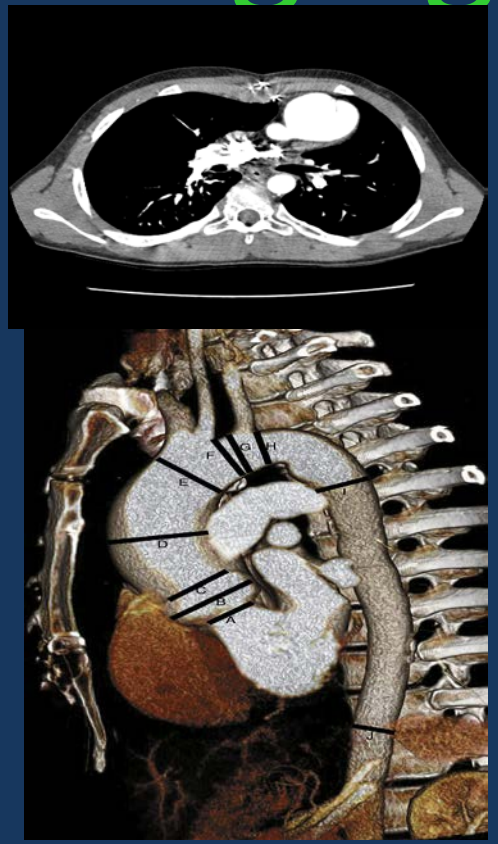




Preoperative Imaging

Echocardiogram
CT angiogram
MRI/MRA

Trend of growth
Associated valvar issues
Involvement of adjacent structures



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Technical Aspects

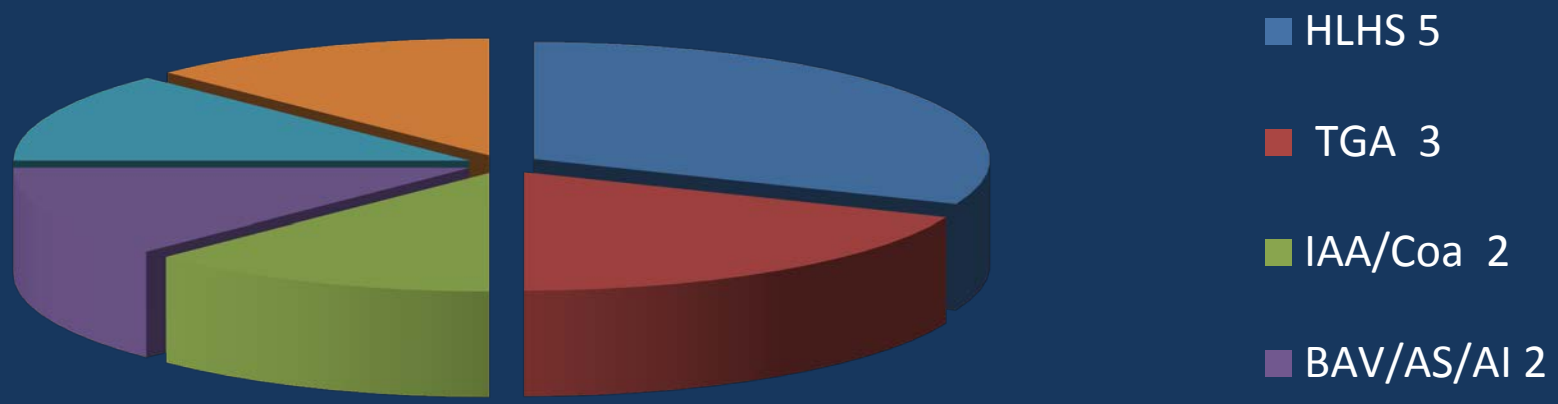
- Full root replacement
- Stabilization of the annulus (reimplantation technique/ David V)
- Arch replacement is not mandatory (Fazel et al)
- Need for routine life long surveillance (aorta at risk)

Clinical data

Cohort included 16 patients

Age (yrs)	13.5 (5-20)
Weight (kg)	45.5 (17-103)
Height (cm)	155 (113-192)
Asc Ao (cm)	4 (2-5.1)
Asc Ao z-score	6.4 (5.4-9.5)
Ao root (cm)	3.8 (2.4-5.5)
Ao root z-score	6.2 (4.4 - 13)

Diagnosis



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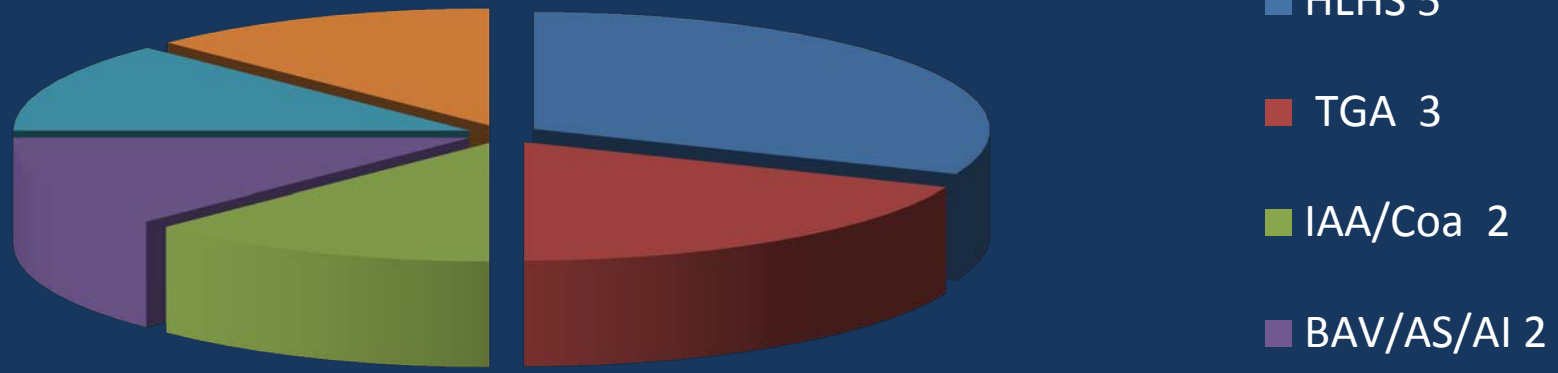
Diagnosis

Bicuspid semilunar valve in 8/16 (50%)



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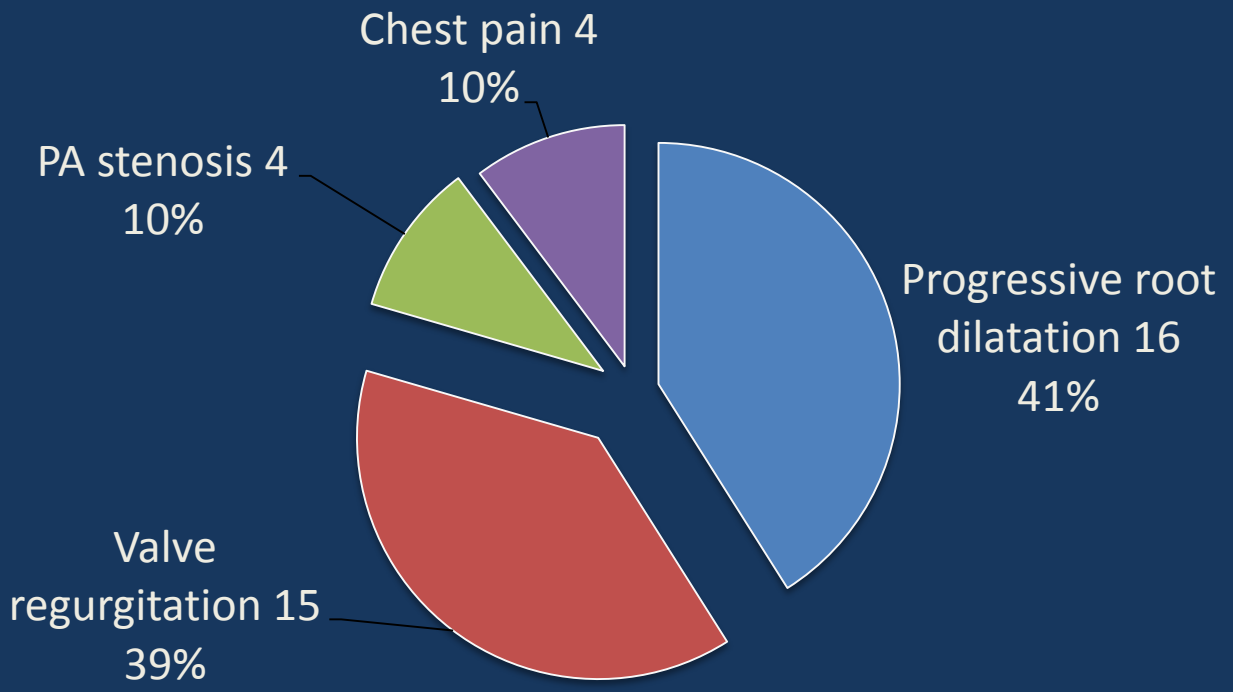
Previous interventions

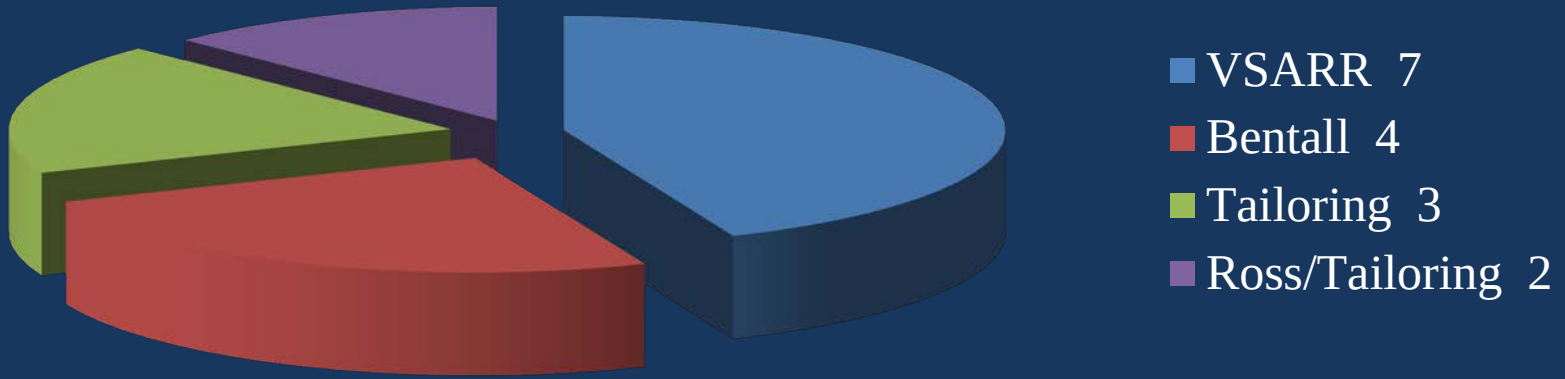
Intervention	
Fontan	5
Arterial switch	3
IAA / VSD	2
Coarctation	2
Aortic valvuloplasty	2
DORV, TOF, AVR	1

Indications



Nemours

Procedures



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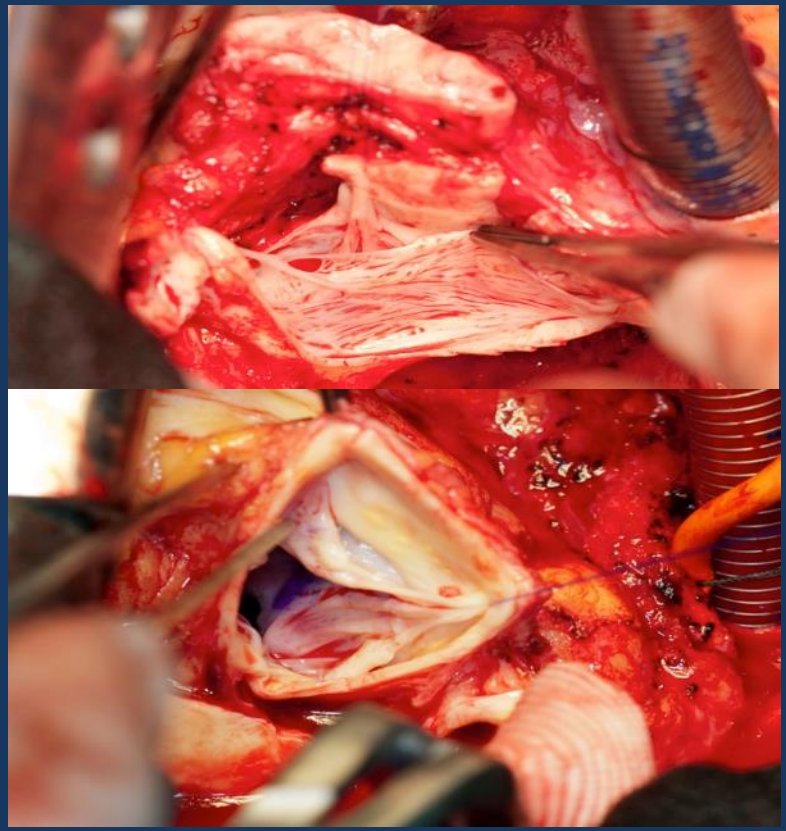
Valve preservation

- Competency
- Mechanism of regurgitation
- Anatomy / Integrity
- Annular dilatation
- Associated lesions
- Ventricular function



Nemours

Associated procedures

• Arch repair	4
• PA plasty	3
• Konno	2
• MPA replacement	2
• PV replacement	1
• Ventricular aneurysm repair	1
• Cryo Maze/ pacemaker	1
• Tricuspid valvuloplasty	1

Nemours

Clinical data

AXC (min)	109 (57-183)
CPB (min)	175 (103-264)
DHCA use (%)	6/16 (38%)
DHCA (min)	29.5 (14-45)
Mech Ventilation (days)	1 (1-11)
ICU stay (days)	4 (2-15)
Hospital stay (days)	11 (6-56)

Clinical data

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Mech Ventilation	1 (1-11)
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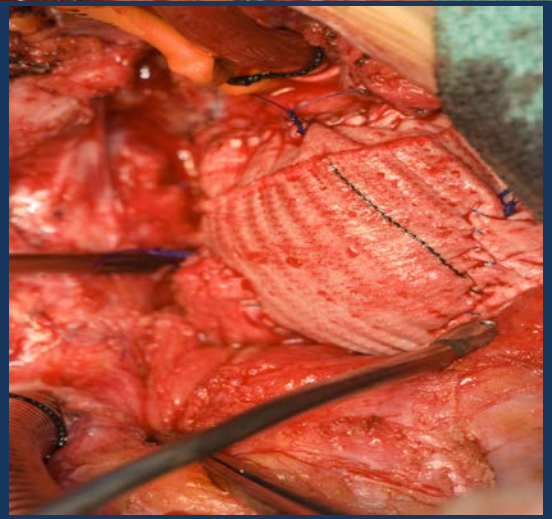
Hospital stay (days)	11 (6-56)

No operative mortality

Nemours

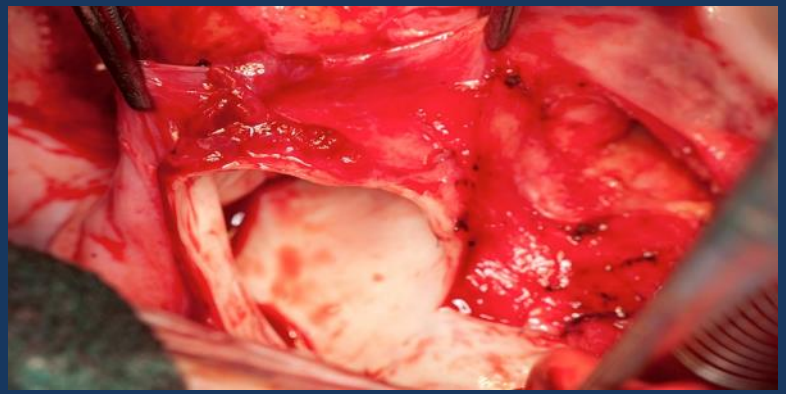
HLHS

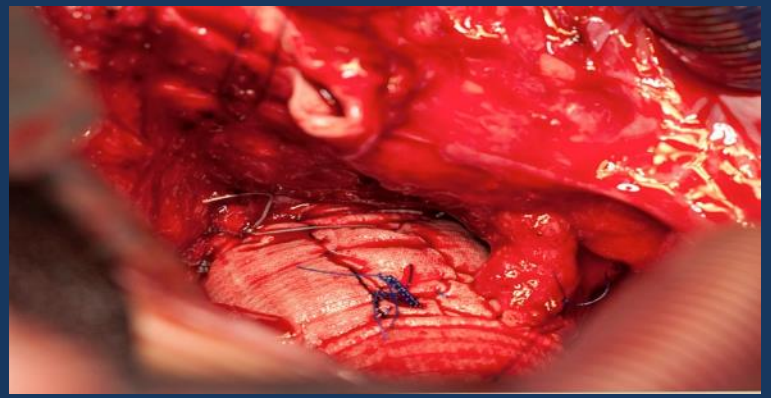
- Single ventricle physiology
- Multiple reoperations
- Myocardial preservation
- Younger patients
- Compression of the Fontan circuit
- Simpler coronary reimplantation
- Sub pulmonary conus
- Preservation of low PVR
  - CPB time
  - Blood products



Transposition of the great arteries

- Posterior location of the aorta
  - Dissection
  - Hemostasis
- Difficult assessment of the valve sparing procedure
- Coronary anatomy (imaging)
- Pulmonary artery reconstruction
- Older patients





Morbidity

Complication		
Pleural/peric effusions		2
Post op bleeding		2 (no reop)
Arrhythmia		3
Neurologic		1 (periph nerve)
Pneumonia		1
Pancreatitis		2
Sternal wound infection		1 (MSSA)
GI bleed		1

Follow up

	VSRR 7	Bentall 4	Tailoring/Ross 5
AI > mod	0	0	0
Reoperation	3 (OHT, AVR, pseudoan)	2 (AVR,OHT)	0
Late Death	2 (OHT, pseudoan)	1 (OHT)	0

Median follow up of 51.6 months (30.6-78.2)

Summary

- Despite the high complexity, surgical management of aortic dilatation in patients with CHD can achieve excellent functional outcomes.
- Indications for surgery are usually influenced by coexisting issues (valve incompetence, coarctation, pulmonary artery obstruction).
- Use of valve sparing techniques can effectively restore the aortic or neo-aortic valve competency.
- Durability of these interventions should be monitored over time
- Genetic information may inform the management of this entity

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