

Replacement of the Ascending Aorta in Early Childhood: Surgical Strategies and Long-Term Outcome

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European **Reference** Network

> for rare or low prevalence complex diseases

Network Respiratory Diseases (ERN-LUNG)

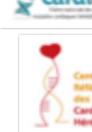


European Reference Network

for rare or low prevalence complex diseases

Network Heart Diseases (ERN GUARD-HEART)







Association pour la Recherche en Cardiologie du Fœtus à l'Adulte

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I have no conflict of interest to declare





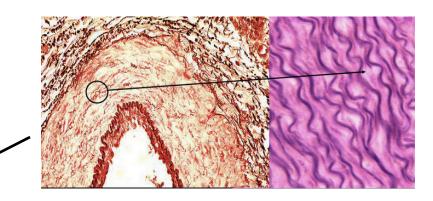




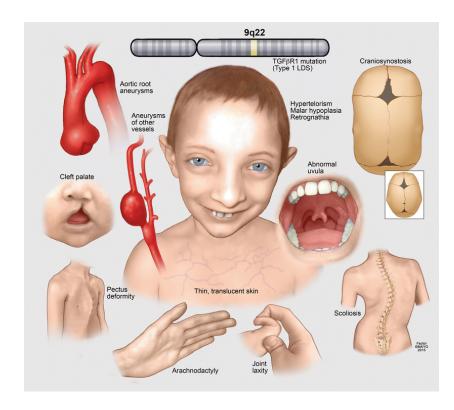
Aortic root dilatation: rare in young children



Connective Tissue Diseases







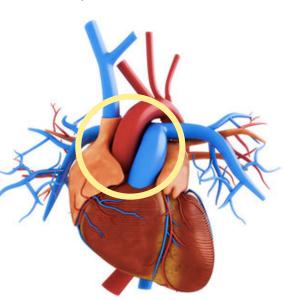
Background

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Isolated Congenital Heart Diseases





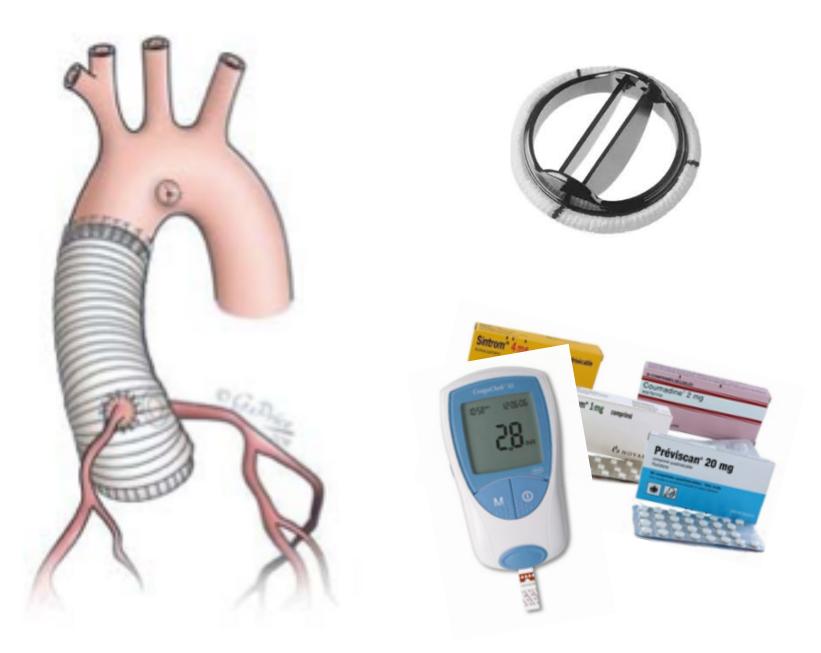




Background

Ascending Aorta Replacement: surgical techniques

Composite graft procedure



Pictures from Michael Gatzoulis

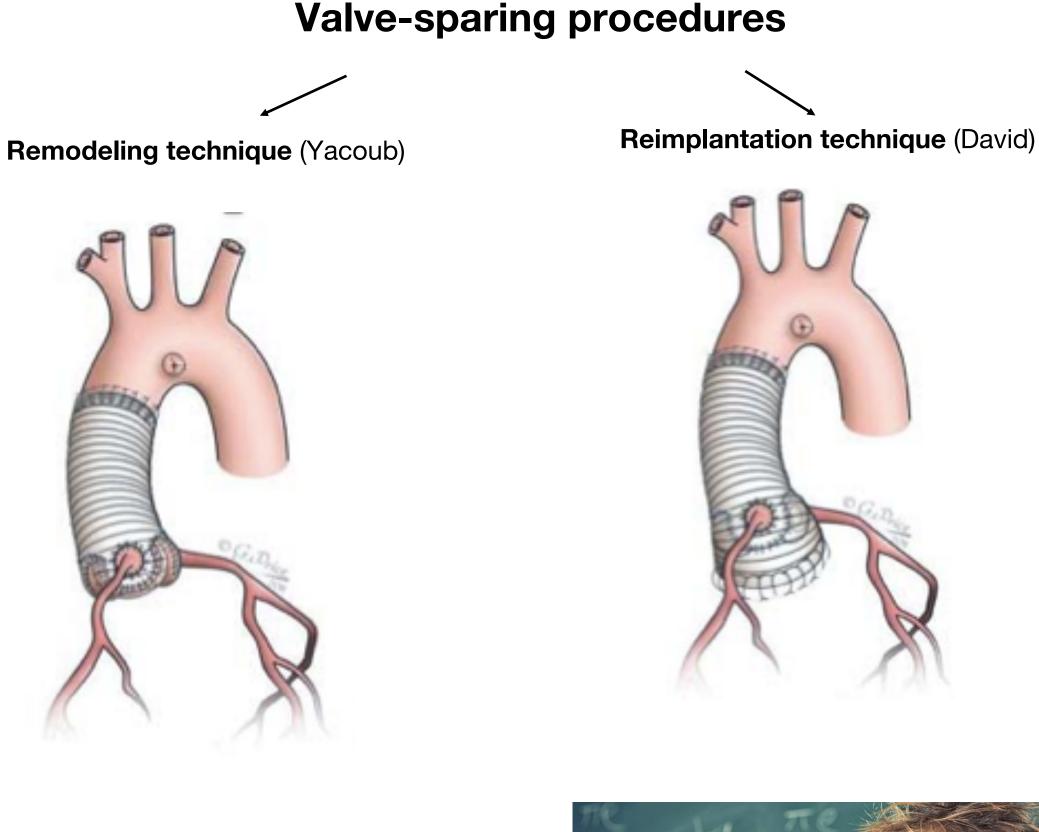
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root surgery by com Results of aortic valve-sparing operations: experience with remodeling and reimplantation Flynn CD¹, Tian DH², Wilson-Sn procedures in 65 patients.

Bethea BT¹, Fitton TP, Alejo DE, Barreiro CJ, Cattaneo SM, Dietz HC, Spevak PJ, Lima JA, Gott VL, Cameron DE.











- **Retrospective study** in Necker hospital (Paris, France) between 1995 and 2017
- Ascending aorta replacement (AAR): 32 children < 10 year-old
 - Median age : 5.3 years (range, 7 months to 9.7 years)
 - Median weight : 18 kg (range, 5.8 to 55 kg) \bullet
- Main **population features**
 - n = 22 (69%) had a connective tissue disease

 - n = 10 (31%) associated a bicuspid aortic value

Methods



n = 7 (22%) had undergone a neonatal surgery including outflow tract (Ross procedure, arterial switch operation,...)

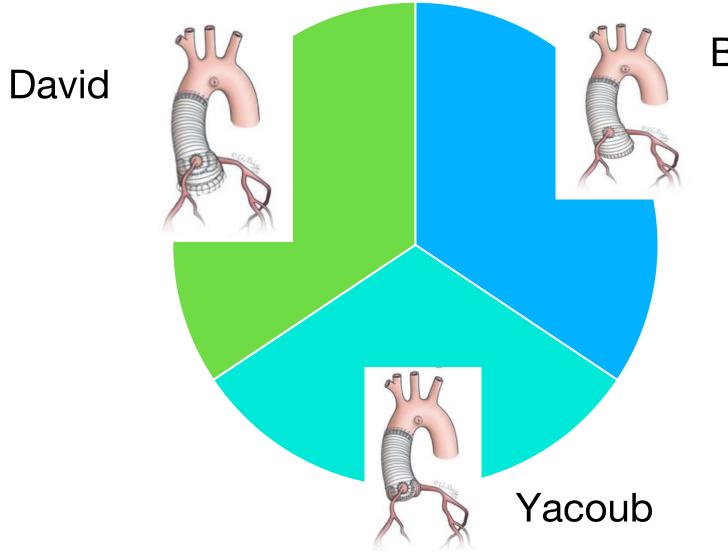




Methods

AAR procedures

- n = 11 composite mechanical valve graft procedures (Bentall operations)
- n = 21 valve-sparing procedures
 - n = 10 remodeling procedures (Yacoub operations)
 - n = 11 reimplantation procedures (David operations) •



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Bentall





Methods-Population

AAR procedures

Characteristics	$\begin{array}{l} \textbf{Bentall} \\ \textbf{n} = 11 \end{array}$	David n = 11	Yacoub n = 10
Mean age (years)	6.8 +/- 2.8	5.3 +/- 2.4	3.8 +/- 2.6
Mean weight (kg)	21.1 +/- 6.6	20.2 +/- 12.7	14.1 +/- 6.9
Male	9 (82%)	6 (55%)	6 (60%)
IMS	1 (9%)	5 (45%)	9 (90%)
Preoperative $AR > 1+$	10 (91%)	3 (27%)	2 (20%)
Preoperative aortic root (mm)	36.3 +/- 7.6	39.3 +/- 6.4	37.8 +/- 6.3
Preoperative aortic root Z-score	+5.9 +/- 2.1	+7 +/- 1.5	+7.6 +/- 1.7
Previous cardiac surgery	7 (64%)	0	3 (30%)
Mean cross-clamp time (min)	112 +/- 33	112 +/- 22	114 +/- 52
Associated surgery	4 (36%)	6 (55%)	3 (30%)
Mean aortic root graft (mm)	22.4 +/- 1.2	21.1 +/- 3.3	17.5 +/- 2.6

Characteristics	IMS n = 15	LDS n = 7	Others n = 10	<i>p</i> Value
Mean age (years)	4.4 +/- 2.5	4.4 +/- 2.9	7.5 +/- 2.3	0.017
Mean weight (kg)	18.6 +/- 12.4	14.7 +/- 6.6	22.3 +/- 5.7	0.05
Male	10 (67%)	2 (29%)	9 (90%)	0.035
Preoperative $AR > 1+$	3 (20%)	3 (43%)	9 (90%)	0.002
Preoperative aortic root (mm)	40.2 +/- 7.4	36.9 +/- 4.6	34.9 +/- 6.1	ns
Preoperative aortic root Z-score	+7.5 +/- 1.5	+7.4 +/- 1.6	+5.3 +/- 1.8	0.026
Previous cardiac surgery	2 (13%)	0	8 (80%)	0.003
Aortic valve sparing procedure	14 (93%)	5 (71%)	2 (20%)	< 0.001
Mean cross-clamp time (min)	121 +/- 40	96 +/- 15	115 +/- 32	ns
Associated surgery	6 (40%)	2 (29%)	5 (50%)	ns
Mean aortic root graft (mm)	19.4 +/- 3.6	20.6 +/- 3.2	22.2 +/- 1.5	ns

Table 2: Patients and operative data according to the underlying congenital disease

IMS = Infantile Marfan Syndrome, LDS = Loeys-Dietz syndrome, AR = Aortic Regurgitation, ns = no significant

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p Value 0.052 ns ns 0.001 0.002 ns ns 0.003 ns ns

0.002

Table 1: Patients and operative data
 according to the ascending aorta procedure



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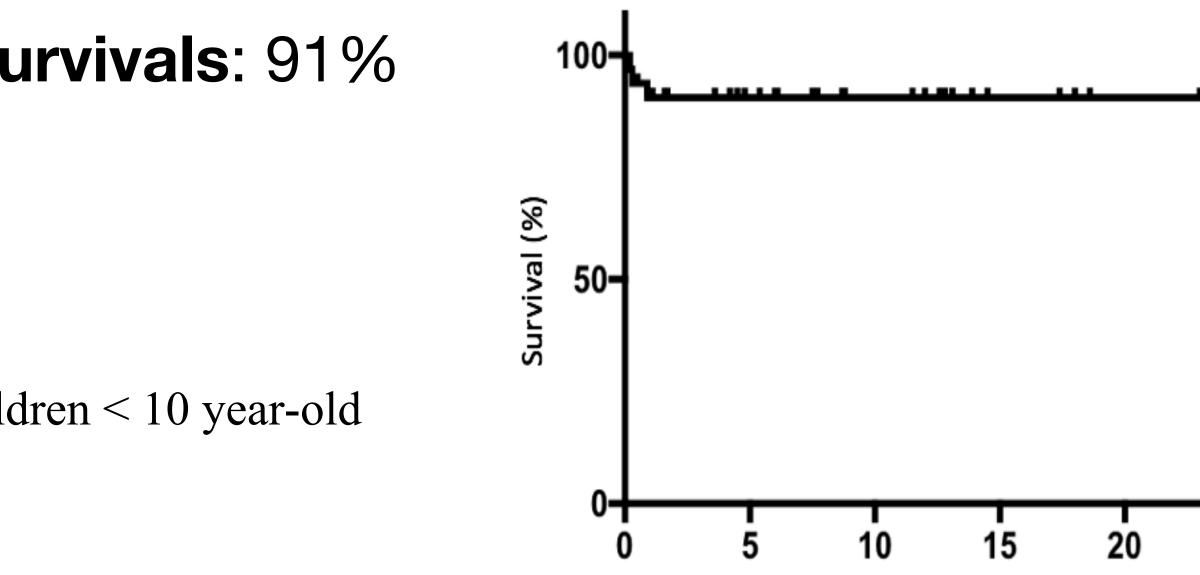


Results (1): Mortality

- No operative death
- Cardiac-related mortality: n = 2 patients (6%)
 - Related with left ventricular failure
 - 1 patient with a coronary injury during surgery
 - 1 patient with a secondary unexplained dilated cardiomyopathy
- One-year and ten-year patient survivals: 91%

Figure 1: Kaplan-Meier survival after AAR in children < 10 year-old

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Years



Results (2): Risk factors of Mortality

Risk Factors	c Factors Alive $n = 29$		p Value
Male	19 (66%)	1	ns
Mean age	5.6 +/- 2.8 y	3.2 +/- 2.5 y	ns
Mean weight	19.6 +/- 9.8 kg	12.2 +/- 4 kg	ns
Zscore	6.7 +/- 1.8	9.3 +/- 1	0.048
IMS syndrome	13 (45%)	2	ns
Connective tissue disease	19 (66%)	2	ns
Aortic regurgitation	13 (45%)	1	ns
Non-tricuspid aortic valve	12 (41%)	1	ns
Associated surgery	11 (38%)	2	ns
Previous surgery	9 (31%)	1	ns
Valve-sparing procedure	19 (66%)	2	ns
Cross-clamp time	110 +/- 26 min	169 +/- 76 min	ns

 Table 3: Cardiac-related mortality risk factors

IMS: Infantile Marfan Syndrome, ns = no significant





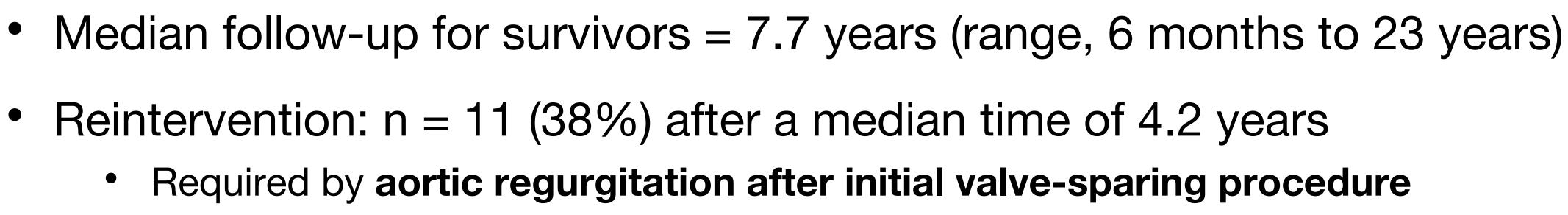


- n = 29 survivors
- Reintervention: n = 11 (38%) after a median time of 4.2 years

Patient	Connective	Age	Surgery	Aortic root	Reop. Delay	Aortic root
	Tissue Disorder	(years)		graft #1 (mm)	(years)	graft #2 (mn
1	IMS	0.8	Yacoub	-	11.1	24
2	IMS	4.8	Yacoub	20	5.0	24
3	IMS	4.6	Yacoub	14	2.0	22
4	-	9.3	Yacoub	20	0.7	22
5	IMS	2.8	Yacoub	-	4.4	28
6	IMS	6.2	Yacoub	18	4.0	24
7	IMS	3.4	Yacoub	16	5.0	24
8	IMS	5.0	David	20	4.1	22
9	IMS	4.5	David	22	1.6	24
10	IMS	4.2	David	19	8.1	28
11	IMS	9.7	David	24	2.1	28

IMS: Infantile Marfan Syndrome

Results (3): Reintervention



One-year and ten-year freedom from reintervention: 96% and 51%

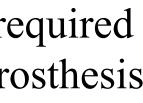
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Table 4: Surgical and demographic features of the patients who required a reintervention with a ortic valve replacement by a mechanical prosthesis



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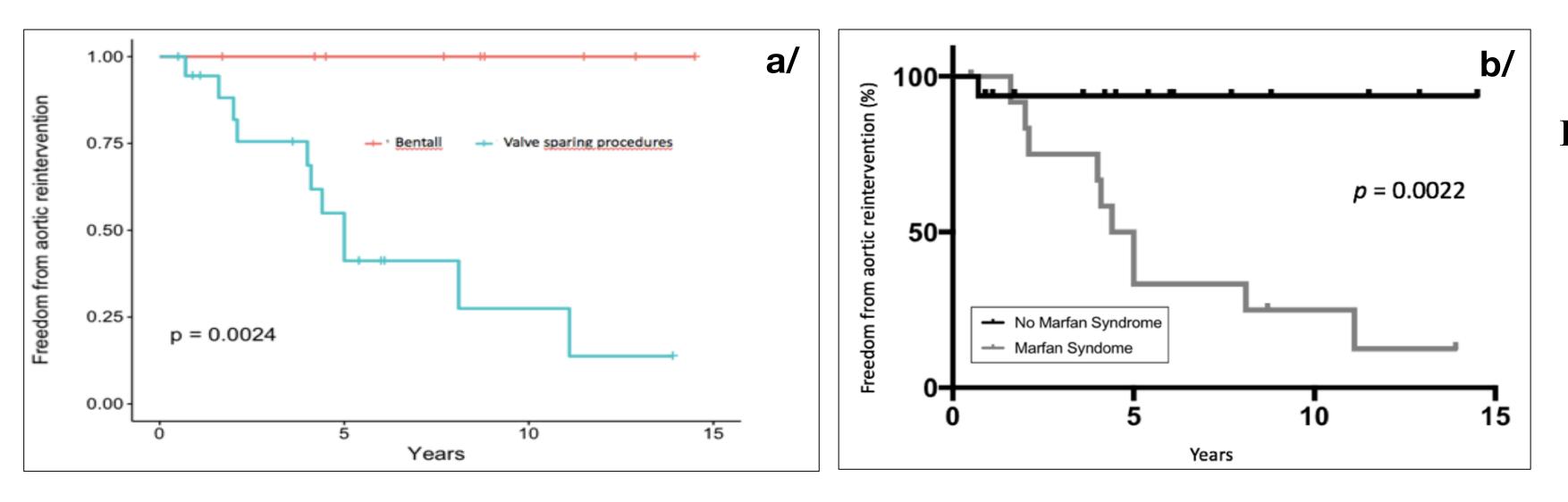




Results (4): Risk factors of Reintervention

Die	Ir Factors	Reoperation	No-reoper
KIS	k Factors	n = 11	$n = 18^{-1}$
Ma	le	8 (73%)	11 (61%)
Me	an age	5 +/- 2.6 y	5.9 +/- 3 y
Me	an weight	21.6 +/- 14 kg	18.6 +/- 7
Zsc	ore	6.9 +/- 1.7	6.6 +/- 1.9
IM	S	10 (91%)	3 (17%)
Ao	rtic regurgitation	2 (18%)	11 (61%)
	n-tricuspid aortic valve	4 (36%)	8 (44%)
Val	ve-sparing procedure	11 (100%)	8 (44%)
	Yacoub procedure vs David procedure	7 (64%)	
	IMS	10 (91%)	

IMS: Infantile Marfan Syndrome, ns = no significant



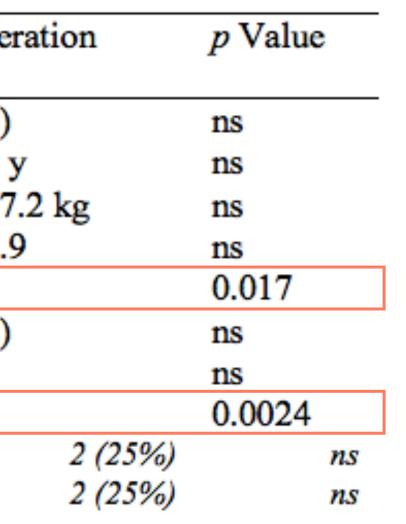


Table 5: Reoperation risk factors

Figure 1: Kaplan Meier curves according to a/ surgical procedure b/ underlying IMS



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- Acceptable long-term outcome, low late mortality
- **Bentall procedure** is safe and durable
- Data would suggest death is related with greater aortic root Zscore
- Special surgical strategy for IMS with subsequent unavoidable aortic regurgitation



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