VI CORSO GUCH

NON SOLO LA VIA DESTRA

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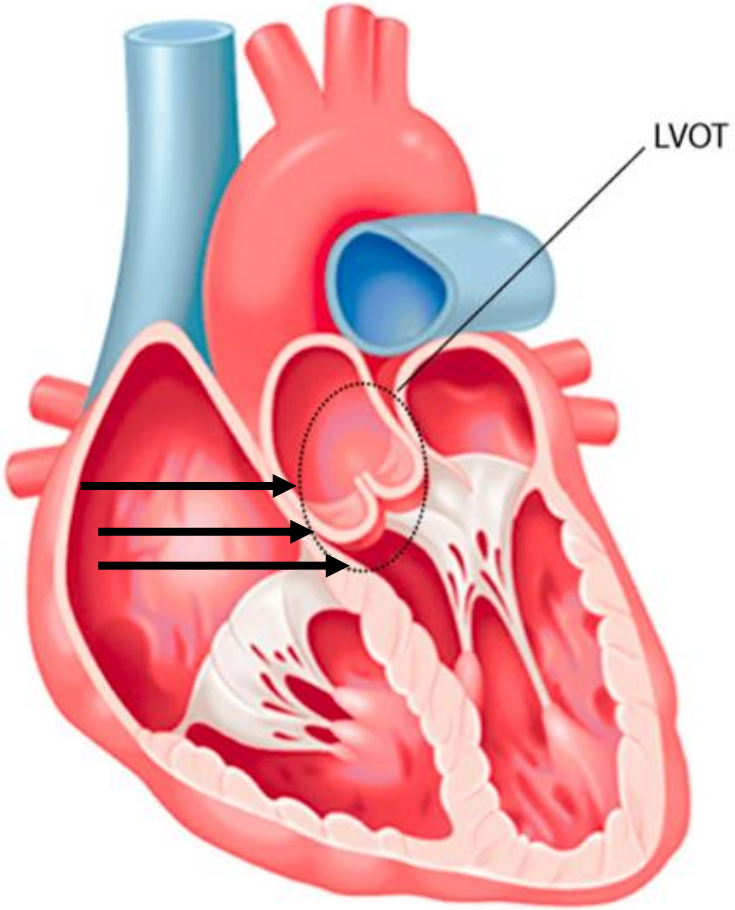
Left Ventricular Outflow Tract (LVOT) in Tetralogy of Fallot



Stenosis

- Subvalvular (subaortic membrane 1%)
- Valvular (bicuspid aortic valve 0.2%)
- Supravalvular (coarctation 0.04%)

Aortic Regurgitation (12.5%) due to dilatation of the aortic root and/or ascending aorta (28-69%)

Changela, V., John, C. & Maheshwari, S. Unusual Cardiac Associations with Tetralogy of Fallot—A Descriptive Study. *Pediatr Cardiol* **31**, 785–791 (2010). https://doi.org/10.1007/s00246-010-9701-1

Subaortic membrane

Only 1% of TOF present also a subaortic membrane.

It can be :

- primitive
- secondary to coexisting lesions or after surgical correction

The obstruction may become hemodynamically relevant and unmasked only after closure of the VSD

SELECTED REPORTS

Tetralogy of Fallot with Discrete Fibrous Subaortic Stenosis*

*John H. Sanders, Jr., M.D.; Richard Van Praagh, M.D.; and Robert M. Sade, M.D.***

This is the first reported case of discrete fibrous subaortic stenosis in tetralogy of Fallot. Transaortic excision of the subaortic membrane was readily accomplished. To detect this and other forms of left ventricular outflow-tract obstruction in tetralogy of Fallot before surgery, routine selective left ventriculography is advocated.

CHEST, 69: 4, APRIL, 1976

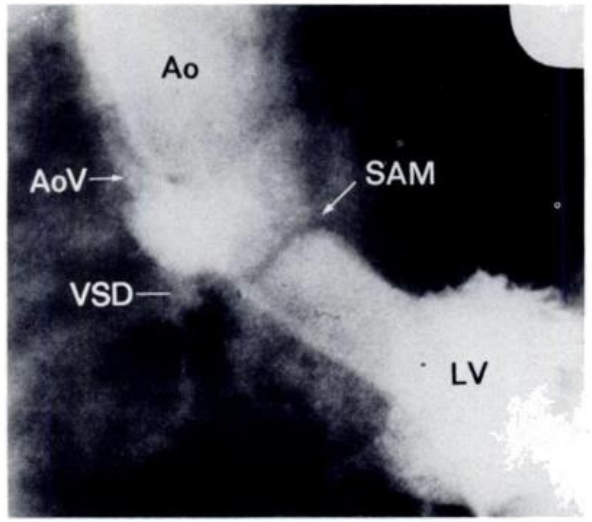


FIGURE 1. Selective left ventriculogram showing subaortic linear lucency typical of discrete fibrous subaortic stenosis. Note moderate thickening and doming of aortic leaflets. Ao, Aorta; AoV, aortic valve; LV, left ventricle; SAM, subaortic membrane; VSD, ventricular septal defect.

John H. Sanders, Richard Van Praagh, Robert M. Sade,Tetralogy of Fallot with Discrete Fibrous Subaortic Stenosis,Chest,Volume 69, Issue 4,1976,Pages 543-544,ISSN 0012-3692,

"Acquired" Subvalvular Aortic Stenosis after Repair of a Ventricular Septal Defect*

Maria Paola Cicini, M.D.; Salvatore Giannico, M.D.; Bruno Marino, M.D.; Fiore S. Iorio, M.D.; Antonio Corno, M.D.; and Carlo Marcelletti, M.D.

Postsurgical SAS appeared to be an uncommon complication in patients with VSD and tetralogy of Fallot, with frequency of 3.2% and 2.1% respectively.

Causes SAS may be:

- Excessive extension of the VSD patch into the LVOT
- Extreme calcification of the VSD patch with formation of a septal ridge protruding into the LVOT

Table 1—*Occurrence of Postsurgical SAS*

Congenital Condition*	No. of Patients Who Underwent Repair	No. of Patients with Post-surgical SAS (%)	Morphologic Features
TF	186	4 (2.1)	Fibromuscular ridge and/or accessory mitral tissue
VSD	153	5 (3.2)	Fibromuscular ridge and/or accessory mitral tissue
DORV	14	3 (21.4)	Left ventricular infundibular fold apposed to fibrous tissue around the patch

Table 2—*Course of Patients with Postsurgical SAS

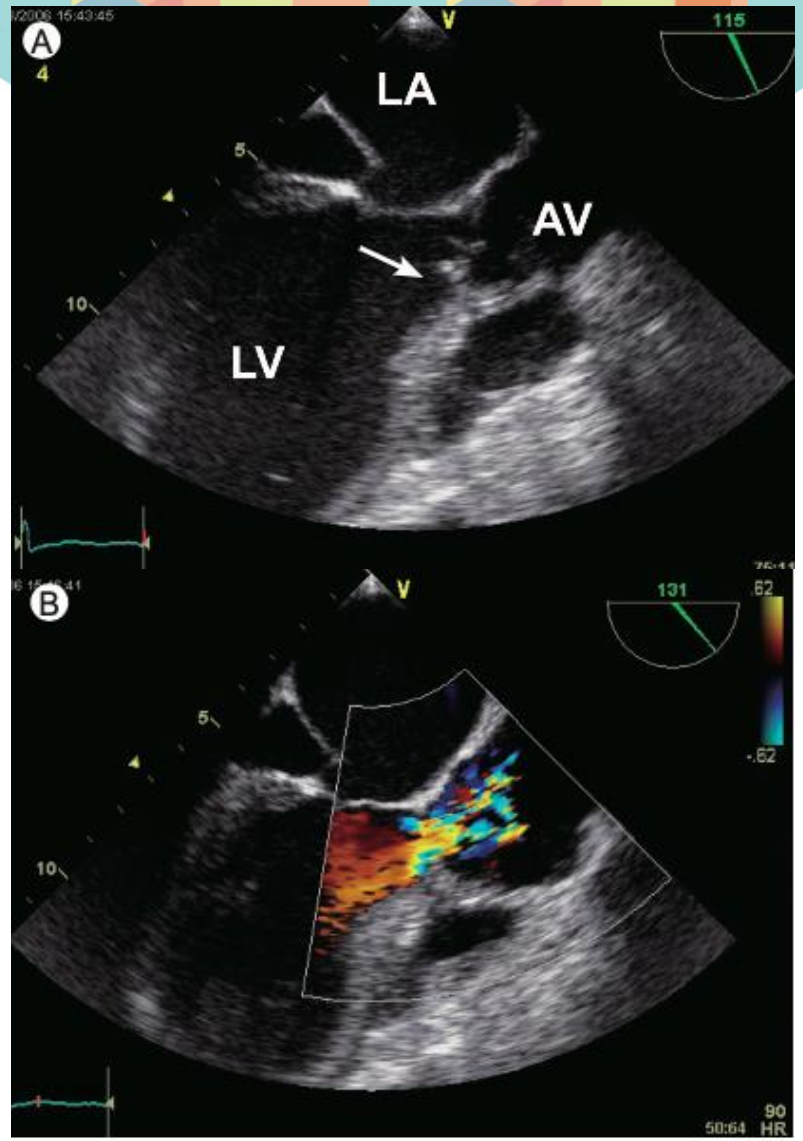
Patient	Date of Birth	Diagnosis	Date of Repair	SAS Diagnosis			SAS Treatment	
				Date	Modality	LV-Ao, mm Hg	Date	Modality
1	01/81	VSD	05/81	03/87	E2D	45	05/87	Resection
2	12/83	VSD	01/85	12/86	Cath	100	01/87	Resection
3	05/81	VSD	11/82	09/86	E2D	50	04/88	Resection
4	03/82	TF	02/84	06/86	Cath	70	09/86	Resection
5	10/89	VSD	02/87	10/89	E2D	30		
6	03/81	DORV	09/81	07/86	E2D	15		
7	11/84	TF	04/86	03/90	E2D	30		
8	02/87	TF	06/89	10/89	Cath	25		
9	08/85	VSD	09/87	03/90	E2D	25		
10	02/83	DORV	11/84	02/88	Cath	60	07/88	PBA (20)
11	10/82	DORV	11/83	10/85	Cath	40	11/85	Resection
12	10/84	TF	08/86	07/90	E2D	15		

(Chest 1992; 101:115-18)

Subaortic membrane

Discrete subaortic stenosis is linked to important complications:

- bacterial endocarditis
- significant aortic regurgitation; flow turbulence distal to the obstructing subaortic membrane plays a crucial role in progressive damage to the aortic valve with resultant thickening of its leaflets and AoR



Kim KH, Kim HK, Chang SA, Oh S, Kim KH, Sohn DW. Subaortic membrane late after surgical correction of tetralogy of Fallot. Korean J Intern Med. 2012 Dec;27(4):455-8. doi: 10.3904/kjim.2012.27.4.455. Epub 2012 Nov 27. PMID: 23269888; PMCID: PMC3529246.

Case report

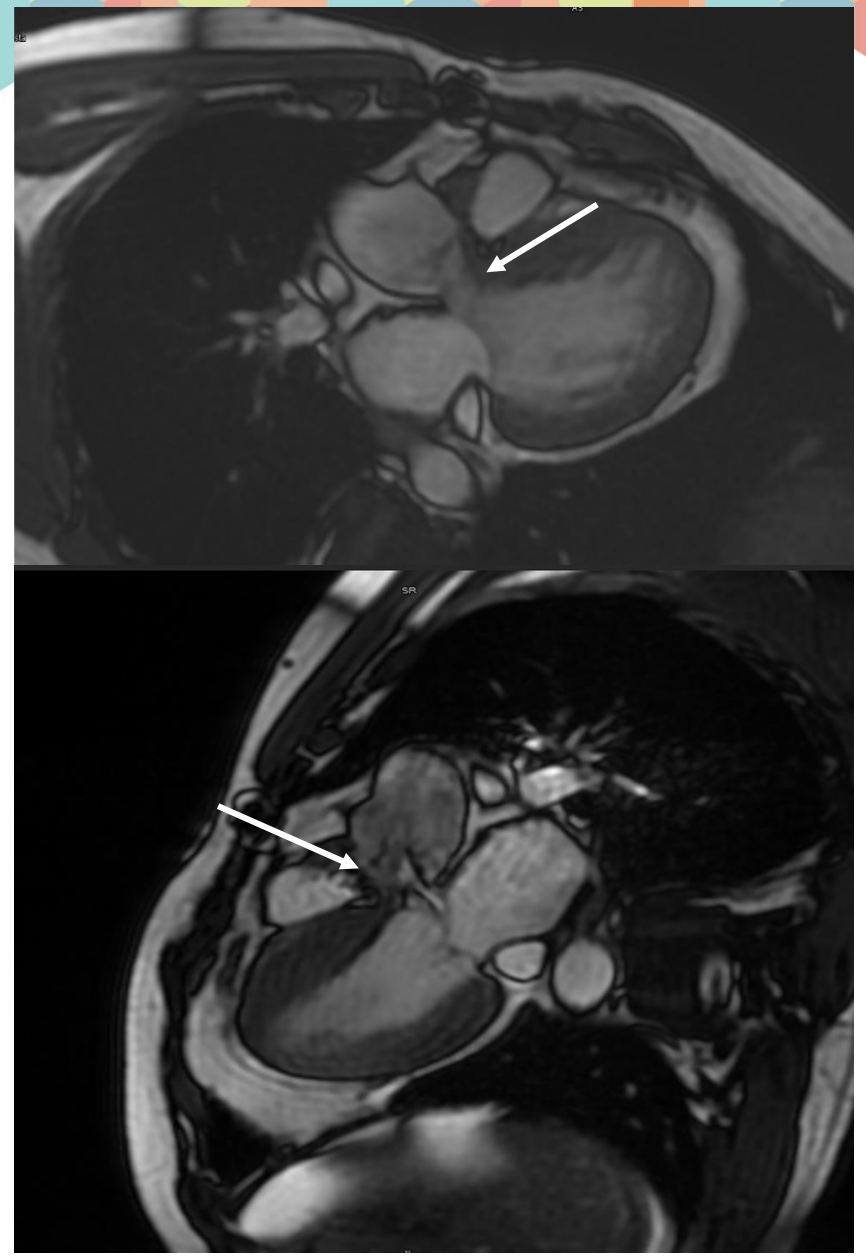
40 years old patients with impaired exercise tolerance.

Medical history:

1. 1984 (2 yo) TOF Correction (VSD closure with dacron patch, enlargement of RVOT and closure with patch of the right ventriculotomy)
2. 1986 (4 yo) pulmonary infundibulum stenosis and subaortic stenosis; correction with infundibular patch and subaortic membrane asportation

Pre-op TTE: subaortic membrane grad max/med 38/26 mmHg, v max 3 m/s, after exertion 54/38 mmHg , v max 3,8 m/s. AoR 1-2+/4+. LVEDD 51, EF 60%, SIV 12.

Pre-op cardiac MRI: subaortic stenosis with flow acceleration, 17 mm below the aortic valve



Case report

The patient underwent subaortic membrane resection and myectomy sec. Morrow.

Intraoperatively, abundant fibrous tissue was observed extending toward LVOT and especially in the interventricular patch area. After resection, the aortic valve was checked, which was continent and free.

Post-op TEE: improvement of the LVOT, grad med 15 mmHg, no AoR

Regular clinical course
1 night ICU stay

Discharged home on postoperative day 7

Subaortic membrane recurrence (8-20%)

Original Article

World Journal for Pediatric and Congenital Heart Surgery
2015, Vol. 6(3) 424-428

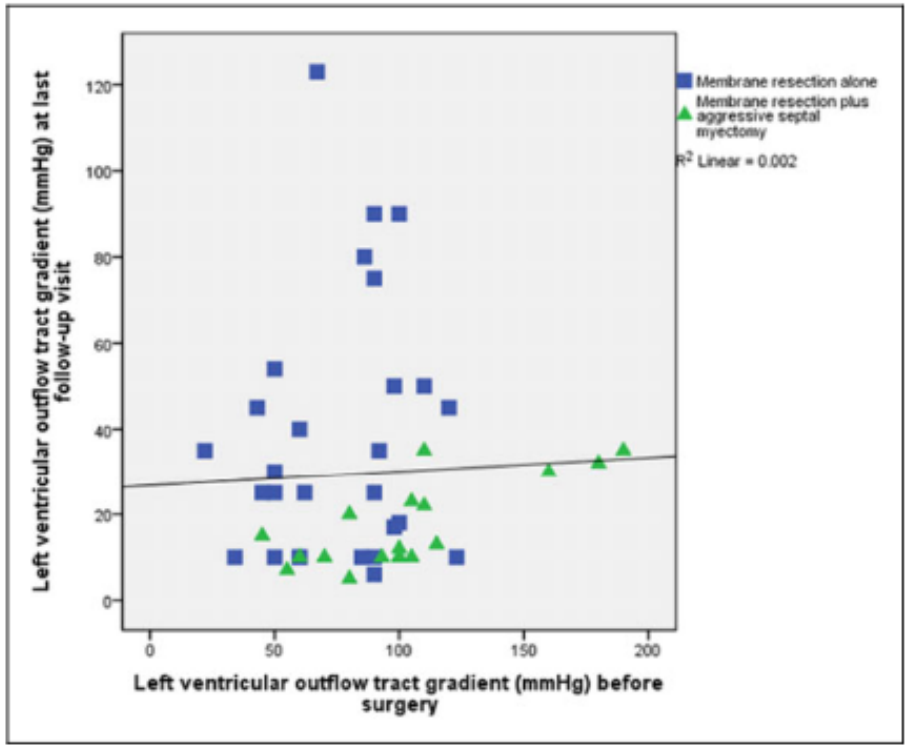
DOI: 10.1177/2150135115589789
pch.sagepub.com
SAGE

Outcome in Children Operated for Membranous Subaortic Stenosis: Membrane Resection Plus Aggressive Septal Myectomy Versus Membrane Resection Alone

Endale Tefera, MD[1], Etsegenet Gedlu, MD[1], Abebe Bezabih, MD[2], Tamirat Moges, MD[1], Tomasa Centella, MD[3], Stefano Marianeschi, MD[4], Berhanu Nega, MD[2], Carin van Doorn, MD[5], Lior Sasson, MD[6], and Michael Teodori, MD[7]

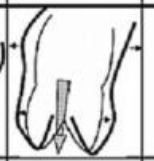
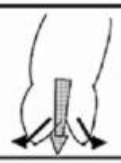
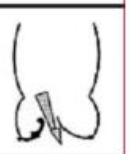
Table 2. Intraoperative and Follow-Up Data of Patients Operated for Membranous Subaortic Stenosis.

	Variables	Group I. Subaortic Membrane Resection	Group II. Subaortic Membrane Resection Plus Aggressive Septal Myectomy	*P* Value
1	Cardiopulmonary bypass time, minutes, n = 36	72.2 ± 35.7 (34-168)	85.7 ± 44.4 (46-231)	.264
2	Aortic cross-clamp time, minutes, n = 36	46.9 ± 26.4 (23-122)	60.7 ± 37.5 (27-174)	.242
3	Immediate left ventricular outflow gradient, mm Hg, n = 31	15.9 ± 15.4 (10-60)	12.7 ± 9.9 (5-40)	.476
4	ICU stay, days, n = 31	2.3 ± 1.1 (1-4)	2.8 ± 0.8 (2-5)	.136
5	Duration of follow-up, years	6.8 ± 3.2 (2-11)	3.6 ± 2.2 (1-6)	.002
6	Last follow-up left ventricular outflow gradient, mm Hg	42.3 ± 31.3 (6-123)	11.6 ± 6.3 (5-35)	<.001
7	Last follow up left ventricular ejection fraction, %, M-mode derived	69.3 ± 6.9 (57-82)	65.0 ± 3.3 (60-70)	.108



Aortic regurgitation

Dilatation of the ascending aorta or aortic root may lead to aortic regurgitation with need for cardiac surgery

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

Khoury, G El *et al*. Functional classification of aortic root/valve abnormalities and their correlation with etiologies and surgical procedures. Current Opinion in Cardiology: March 2005 - Volume 20 - Issue 2 - p 115-121

Table 2 Complex CHD associated with aortic dilatation in adults

TOF
Single ventricle with pulmonary atresia or stenosis
TAC
Transposition of the great arteries
Hypoplastic left heart syndrome
Fontan procedure

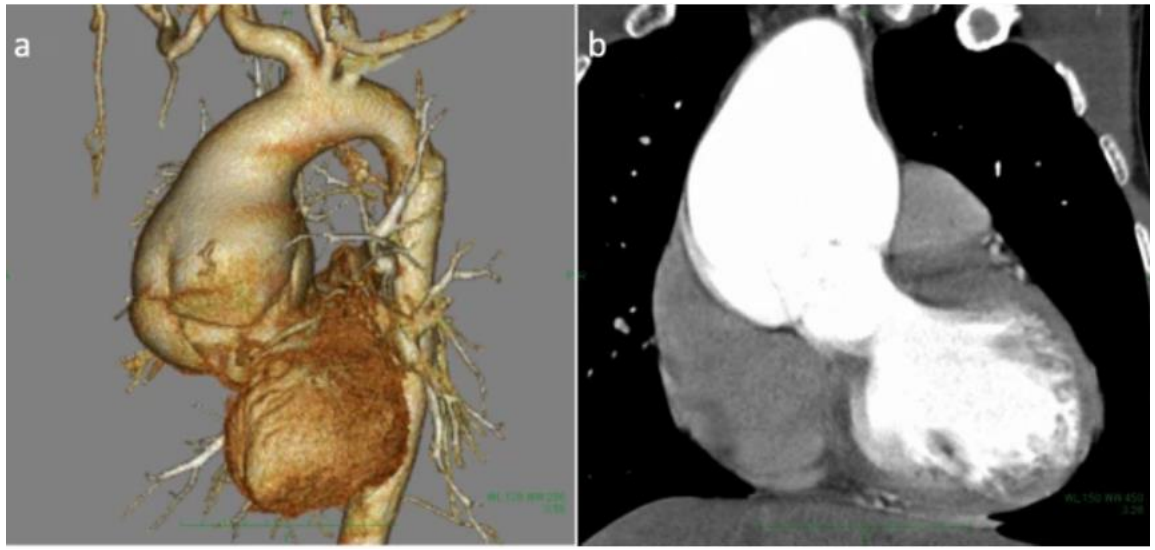
CHD, congenital heart diseases; TOF, tetralogy of Fallot; TAC, truncus arteriosus communis.

Niwa K. Aortic dilatation in complex congenital heart disease. Cardiovasc Diagn Ther. 2018 Dec;8(6):725-738. doi: 10.21037/cdt.2018.12.05. PMID: 30740320; PMCID: PMC6331370.

Aortic Root Dilatation

Aortic root dilatation is a known clinical feature of **unrepaired TOF**. These may be caused by:

- Increased and altered flow through the overriding aorta and uneven sharing of conotruncal tissue between the aorta and pulmonary artery
- Hemodynamic stress on the aortic wall before repair



Nishimura, Y *et al.* Significant delayed aortic dilatation after tetralogy of Fallot repair: a case report. *surg case rep* **6**, 173 (2020).

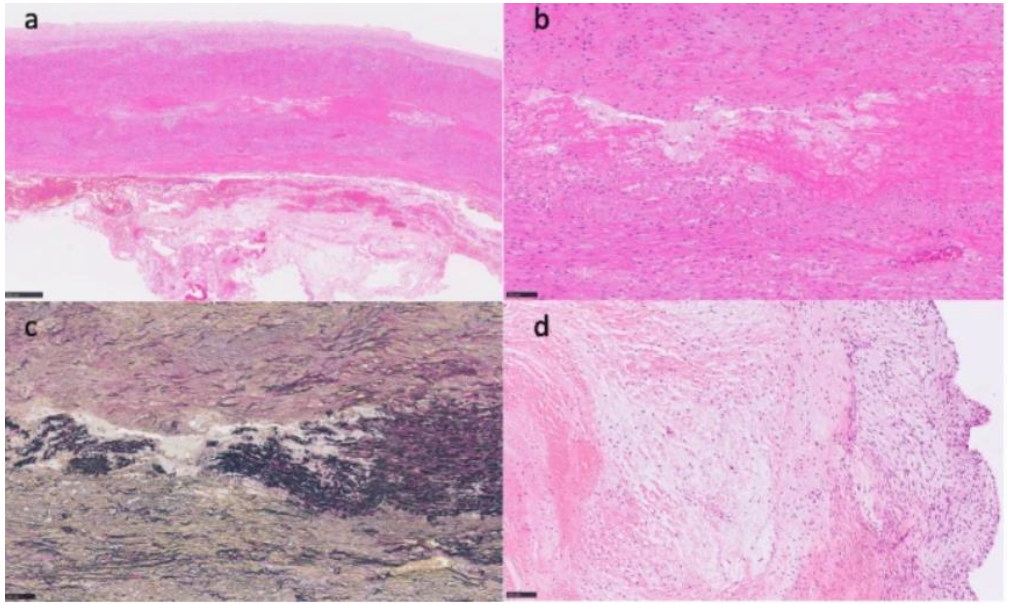
Aortic Root Dilatation

Aortic root dilatation is also observed in adult patients late **after** the repair of TOF.

The prevalence ranged from 28 to 69%.

This may result from an intrinsic aortopathy with intrinsic histologic changes such as:

- medionecrosis,
- Fibrosis
- cystic medial necrosis
- elastic fragmentation
- elastic lamella disruption



Nishimura, Y *et al.* Significant delayed aortic dilatation after tetralogy of Fallot repair: a case report. *surg case rep* **6**, 173 (2020).

ORIGINAL RESEARCH

American Heart Association.

Thoracic Aortic Dissection in Tetralogy of Fallot: A Review of the National Inpatient Sample Database

Alexander C. Egbe, MD, MPH, FACC; Juan Crestanello, MD; William R. Miranda, MD; Heidi M. Connolly, MD

The estimated TAD incidence of 6 per 10 000 (0.06%) in the current study suggests an extremely low risk of TAD in TOF patents (18353 overall patients; 11 with TOF)

TABLE 1 Literature Review of Prior Case Reports of Aortic Dissection in Tetralogy of Fallot

First Author, Year	Age, Presentation	Acute or Chronic Location and Size Histology	Treatment
Kim et al,[5] 2005	30-year-old man with chest pain	Acute Ascending aorta 7 cm Fibrous, thrombotic exudates. No cystic medial degeneration	Bentall procedure
Rathi et al,[6] 2005	36-year-old man with chest pain	Acute Ascending aorta 9.3 × 8.3 cm	Not available
Konstantinov et al,[7] 2010	18-year-old man with 22q11 deletion syndrome with chest pain	Acute Ascending aorta 6 × 7 cm	Valve sparing aortic root replacement was performed with a 30-mm Valsalva graft
Wijesekera et al,[8] 2014	60-year-old man with chest pain and dyspnea	Acute Ascending aorta 5.5 cm (4.9 cm ascending aorta 2 y earlier)	Bentall procedure
Jariwala et al,[9] 2017	30-year-old man unrepaired with dyspnea and back pain	Subacute NA	22-mm interposition graft in the ascending aorta was performed
Chow et al,[10] 2020	35-year-old man with chronic chest discomfort over past months	Acute Ascending aorta 8.8 × 7.6 cm (4.8 × 5.1 cm ascending aorta 3 y prior [stopped losartan]) Myxoid generation and focal calcification, mild cystic medial degeneration	Bentall

Egbe AC, Crestanello J, Miranda WR, Connolly HM. Thoracic Aortic Dissection in Tetralogy of Fallot: A Review of the National Inpatient Sample Database. J Am Heart Assoc. 2019 Mar 19;8(6):e011943.

Sumeet S. Vaikunth, Joshua L. Chan, Jennifer P. Woo, Michael R. Bykhovsky, George K. Lui, Michael Ma, Anitra W. Romfh, John Lamberti, Domenico Mastrodicasa, Dominik Fleischmann, Michael P. Fischbein,Tetralogy of Fallot and Aortic Dissection: Implications in Management,JACC: Case Reports,Volume 4, Issue 10,2022,Pages 581-586,ISSN 2666-0849,

Early or late repair to prevent aortic root dilatation?

Progressive Aortic Root Dilatation in Adults Late After Repair of Tetralogy of Fallot

Koichiro Niwa, Samuel C. Siu, Gary D. Webb and Michael A. Gatzoulis

Originally published 19 Aug 2002 | https://doi.org/10.1161/01.CIR.0000028462.88597.AD | Circulation. 2002;106:1374–1378

Other version(s) of this article

	Group A dilators Group A (n=32)	Group B controls Group B (n=54)	P
Sex, male:female	24:8	30:24	**0.012**
Age, y	36±8.0 (23–52)	37±9.1 (23–60)	0.49
Pulmonary atresia	6 (19)	0	**<0.0001**
Right aortic arch	16 (50)	14 (26)	**0.031**
Absent pulmonary valve	3(9)	0	**0.011**
Age at repair, y	14±8.2 (5–37)	14±9.2 (3–49)	0.61
Follow-up after repair, y	22±7.1 (4–33)	24±7.4 (4–36)	0.58
Shunt-repair interval, y	12±8.3 (3–34)	8±6.1 (2–28)	**0.048**
Aortic valve replacement and root repair	2 (6)	0	**0.023**
Aortopulmonary shunt	19	29	0.48
Blalock-Taussig shunt	16	26	
Waterston shunt	3	1	
Potts shunt	0	2	
Transannular patch	18 (56)	26 (48)	0.88
Conduit repair (for pulmonary atresia)	6 (19)	0	**<0.0001**
Residual ventricular septal defect	3 (9)	2 (4)	0.13
Blood pressure >140/90	1 (3)	0	0.12
Cardiothoracic ratio, %	58.5±5.3 (48–69)	54.6±5.9 (41–67)	**0.024**

Values are mean±SD (range) or n (%).
Significant correlations are shown in bold.

Definitive repair of TOF is being undertaken at earlier ages and repair in infancy has become the standard.

Early repair obviates the need for palliative shunts with their complications, such as pulmonary artery distortion and AoR dilatation.

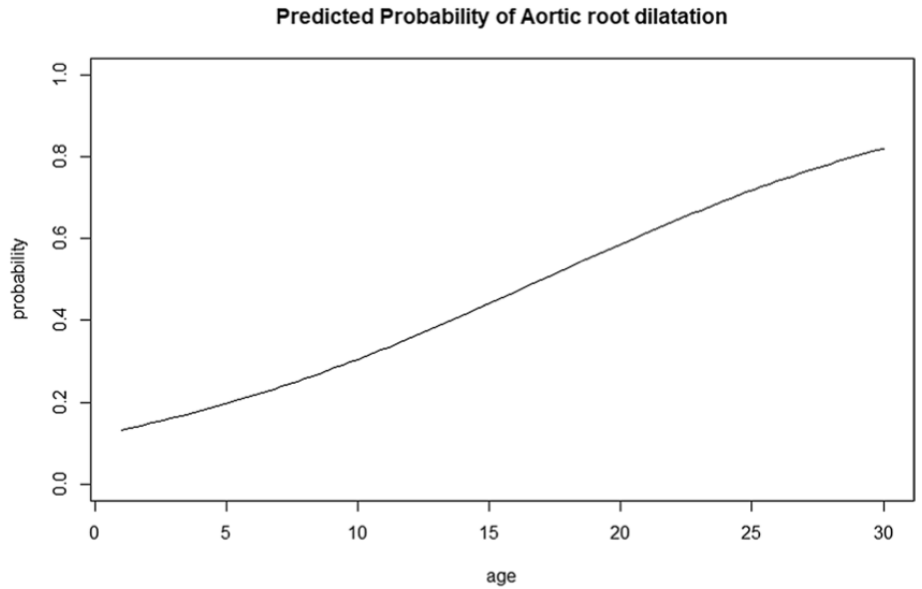
Early volume unloading of aortic outflow with definitive repair may avoid progressive AoR dilatation.

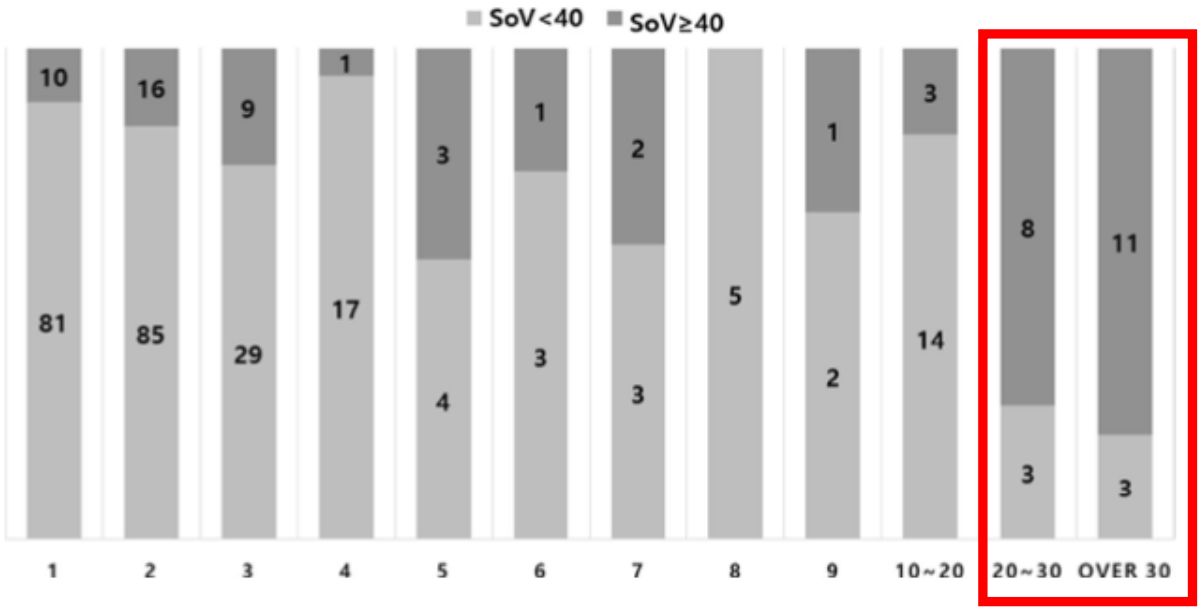
Pediatric Cardiology (2020) 41:1501–1508
https://doi.org/10.1007/s00246-020-02394-z

ORIGINAL ARTICLE

Correlation Between Total Repair Timing and Late Aortic Root Dilatation in Repaired Tetralogy of Fallot

Hyung Tae Sim[1] · Jeong-Won Kim[2] · Seong Ho Kim[3] · Su-Jin Park[3] · So-Ick Jang[3] · Chang-Ha Lee[2]





Valve-sparing aortic root replacement

European Journal of Cardio-Thoracic Surgery 50 (2016) 155–159
doi:10.1093/ejcts/ezv446 Advance Access publication 30 December 2015

ORIGINAL ARTICLE

Cite this article as: Baliulis G, Ropponen JO, Salmon TP, Kaarne MO. Valve-sparing aortic root replacement in adult patients previously operated for congenital heart defects: an initial experience. Eur J Cardiothorac Surg 2016;50:155–9.

Valve-sparing aortic root replacement in adult patients previously operated for congenital heart defects: an initial experience†

Giedrius Baliulis[a,*], Jussi O. Ropponen[b], Tony P. Salmon[a] and Markku O. Kaarne[a,b]

[1] Wessex Cardiothoracic Centre, University Hospital Southampton, Southampton, UK
[2] Department of Cardiothoracic Surgery, Helsinki University Central Hospital, Helsinki, Finland

Table 1: Patient characteristics, surgical procedures and follow-up times

Patients	Primary diagnosis	Age at VSARR (years)	Years since primary corrective surgery	Aortic root diameter (mm)	Degree of AI preoperatively	Type of VSARR procedure	Procedures for AV leaflets	Concomitant procedures	Follow-up (months)
1	TOF	36	30	52	–	David	–	PVR	78
2	TOF	41	33	52	Mild	David	Central plication (LCC, RCC)	PVR, TVA, RA Maze	74
3	TOF	50	33	55	Moderate	David	Resection of nodular fibrosis	TVA, closure of residual VSD	52
4	TOF	45	39	60	Mild	David	Central plication (LCC, NCC)	PVR, TVA, biatrial Maze	43
5	CoA, VSD, BAV	34	29	54	Moderate	David	Triangular resection (NCC)	Closure of VSD	31
6	TGA, VSD, S/P ASO	21	21	47	Moderate	David	–	Angioplasty of PA branches	32
7	TGA, VSD, CoA, S/P ASO	16	16	51	Mild	David	–	–	18
8	DILV, TGA, S/P ASO and Fontan	23	20	45	Severe	David	Central plication (all leaflets)	–	32
9	PA/IVS, S/P Fontan	19	17	68	Mild	David	Central plication (RCC)	–	28
10	Truncus arteriosus (quadricuspid valve)	20	19	46	Moderate	David	Leaflet excision	Replacement of RV-PA conduit + VSD patch, angioplasty of PA branches	30
11	VSD, AI, S/P AV repair	30	27	53	Moderate	David	Central plication (LCC, NCC)	–	12

AI: aortic insufficiency; AV: aortic valve; ASO: arterial switch operation; BAV: bicuspid aortic valve; CoA: coarctation of the aorta; DILV: double inlet left ventricle; LCC: left coronary cusp; NCC: non-coronary cusp; PA: pulmonary artery; PA/IVS: pulmonary atresia with intact ventricular septum; PVR: pulmonary valve replacement; RA: right atrium; RCC: right coronary cusp; RV: right ventricle; S/P: status post; TGA: transposition of great arteries; TOF: tetralogy of Fallot; TVA: tricuspid valve annuloplasty; VSARR: valve-sparing aortic root replacement; VSD: ventricular septal defect.

RESULTS

No mortality occurred in-hospital or during the follow-up period. During the immediate postoperative period, 2 patients underwent drainage of the pericardial collection and 1 patient required renal replacement therapy with eventual full recovery of kidney function. No other reoperations or reinterventions were needed during the follow-up period. Ten patients remain in NYHA functional class I and 1 patient with single-ventricle circulation is in NYHA class II. The most recent echocardiographic studies in 10 patients have demonstrated no or trivial AoV insufficiency. One patient, however, with severe AI preoperatively, developed mild AoV leak shortly after the operation, which has progressed to a moderate grade 1 year following the operation. The degree of AI remains moderate in this patient after 32 months of follow-up.

Baliulis G *et al.* Valve-sparing aortic root replacement in adult patients previously operated for congenital heart defects: an initial experience. Eur J Cardiothorac Surg. 2016 Jul;50(1):155-9.

David procedure: valve-sparing aortic root replacement

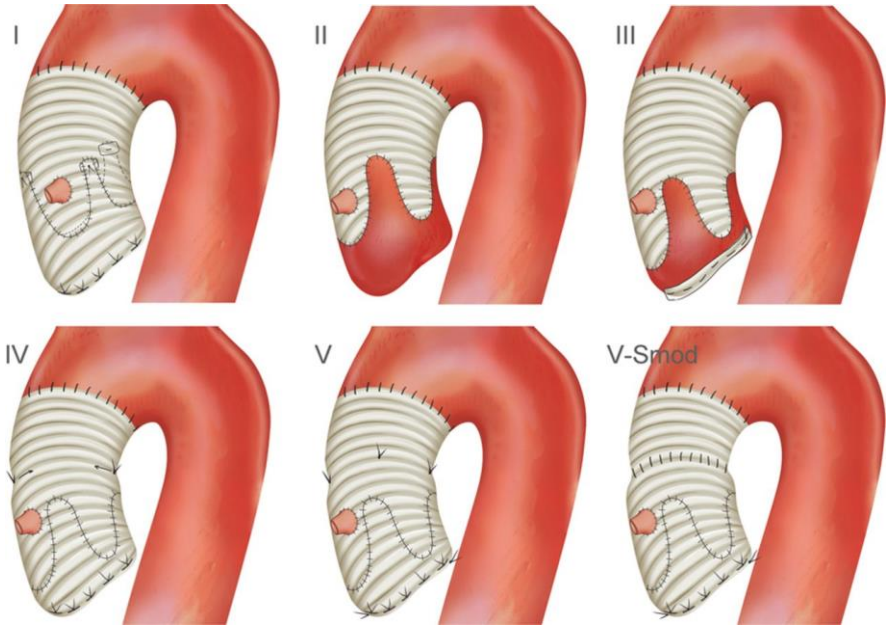
Volume 103 Number 4 April 1992

The Journal of THORACIC AND CARDIOVASCULAR SURGERY

Original Communications

An aortic valve–sparing operation for patients with aortic incompetence and aneurysm of the ascending aorta

Tirone E. David, MD, and Christopher M. Feindel, MD (by invitation),
Toronto, Ontario, Canada



David TE, Feindel CM. An aortic valve-sparing operation for patients with aortic incompetence and aneurysm of the ascending aorta. J Thorac Cardiovasc Surg. 1992 Apr;103(4):617-21; discussion 622. PMID: 1532219.

Kari FA, Siepe M, Sievers HH, Beyersdorf F. Repair of the regurgitant bicuspid or tricuspid aortic valve: background, principles, and outcomes. Circulation. 2013 Aug 20;128(8):854-63. doi: 10.1161/CIRCULATIONAHA.113.002475. PMID: 23960256.

David procedure: valve-sparing aortic root replacement

Aortic valve sparing operations: outcomes at 20 years

Tirone E. David

The Division of Cardiovascular Surgery of Peter Munk Cardiac Centre at Toronto General Hospital, University Health Network, and University of Toronto, Toronto, Canada

374 patients from 1988- 2010

Patients' survival

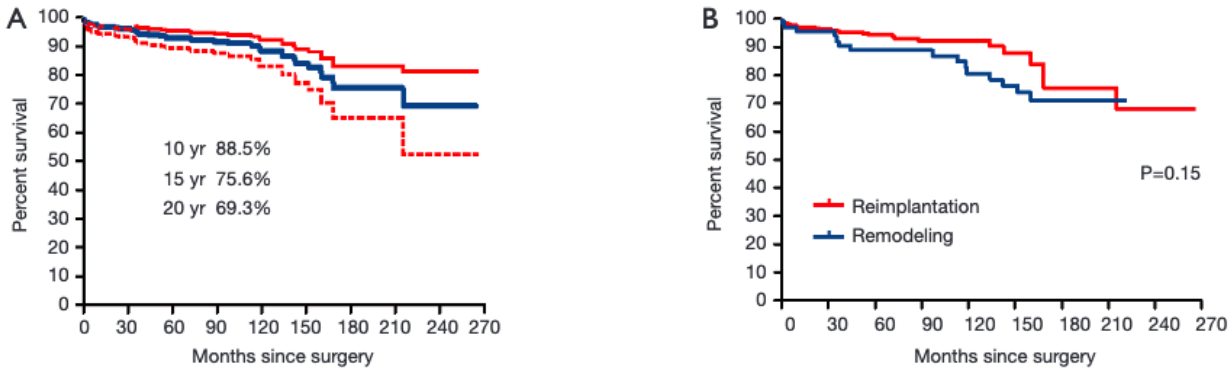


Figure 1 A. Patients' survival after aortic valve sparing operations; B. Survival after reimplantation and remodeling procedures

Freedom from reoperation

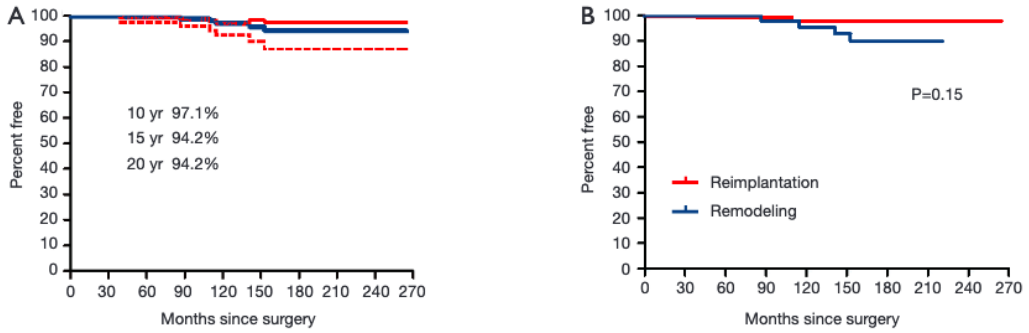


Figure 2 A. Freedom from reoperation on the aortic valve after aortic valve sparing operations; B. Freedom from reoperation after reimplantation and remodeling procedures

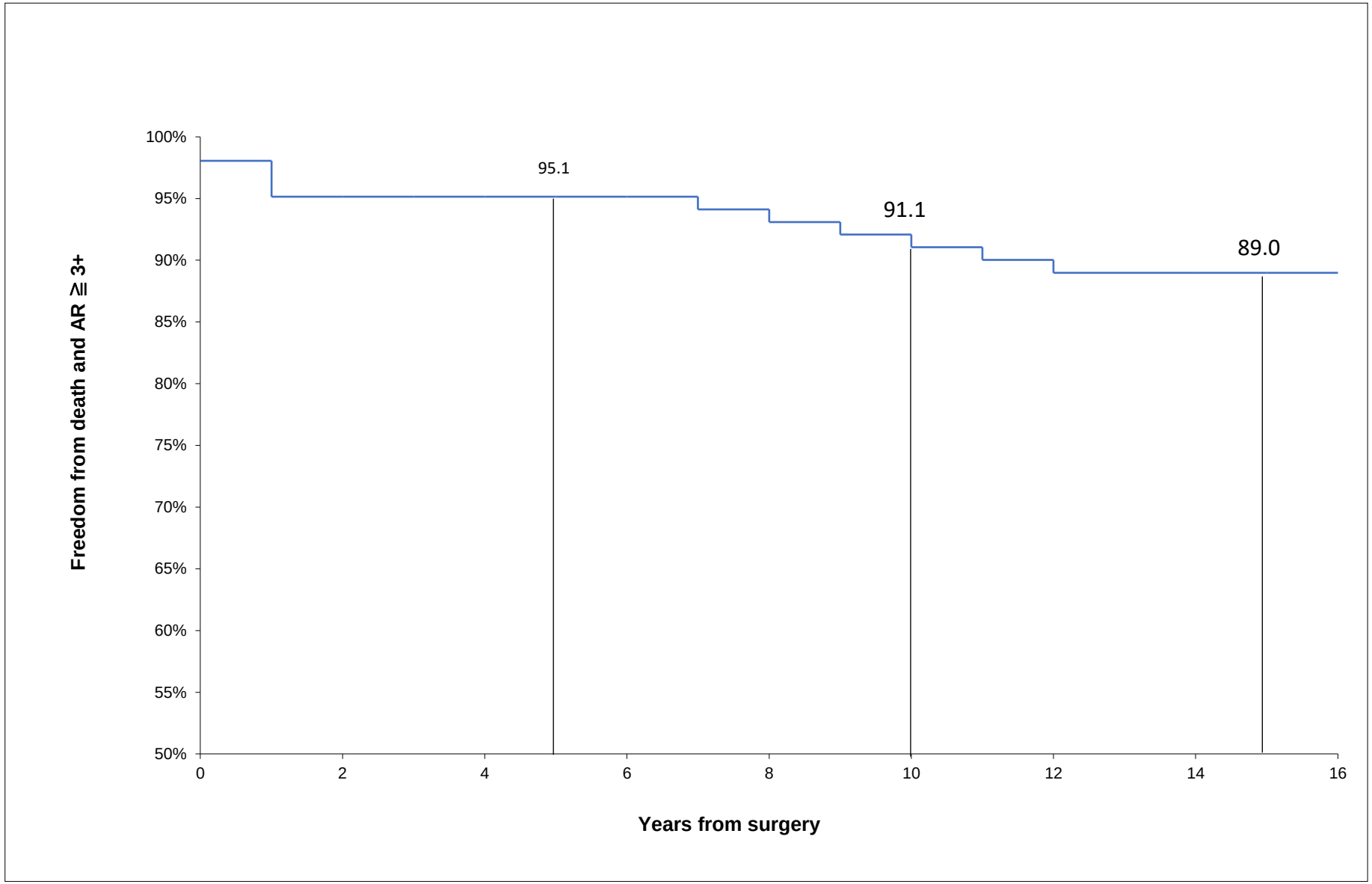
David procedures – University of Turin experience 2015-2022

Preoperative data	**104 Patients**
M (n, %)	88(84.6)
Euroscore- Log (m, SD)	1.5(1.2)
BMI (m, SD)	25.9(3.6)
Creatinine mg/dl (m, SD)	0,9(0.2)
Marfan sd (n,%)	12(11.5)
IRC (n, %)	1(0,9)
Diabetes (n, %)	5(4.8)
Hypertension (n, %)	50(48.1)
Smoke (n, %)	24(23.1)
Dyslipidemia (n, %)	21(20.1)
NYHA class 3-4 (n, %)	13(12.5)
AF (n, %)	9(8.6)
EF % (m, SD)	60.5(7.2)
DTD mm (m, SD)	54.8(6.9)
VTD mm (m, SD)	139(40.4)
Aortic annulus mm (m, SD)	26(2.6)
Aortic root mm (m, SD)	47.8(7.5)
GST mm (m,SD)	43.8(5.8)
Ascending aorta mm (m, SD)	48.9(10)
Aortic insufficiency 3-4+ (n, %)	64(61.5)
Bicuspid aortic valve (n, %)	9(8.6)
PAPs mmHg (m, SD)	27.8(6.2)

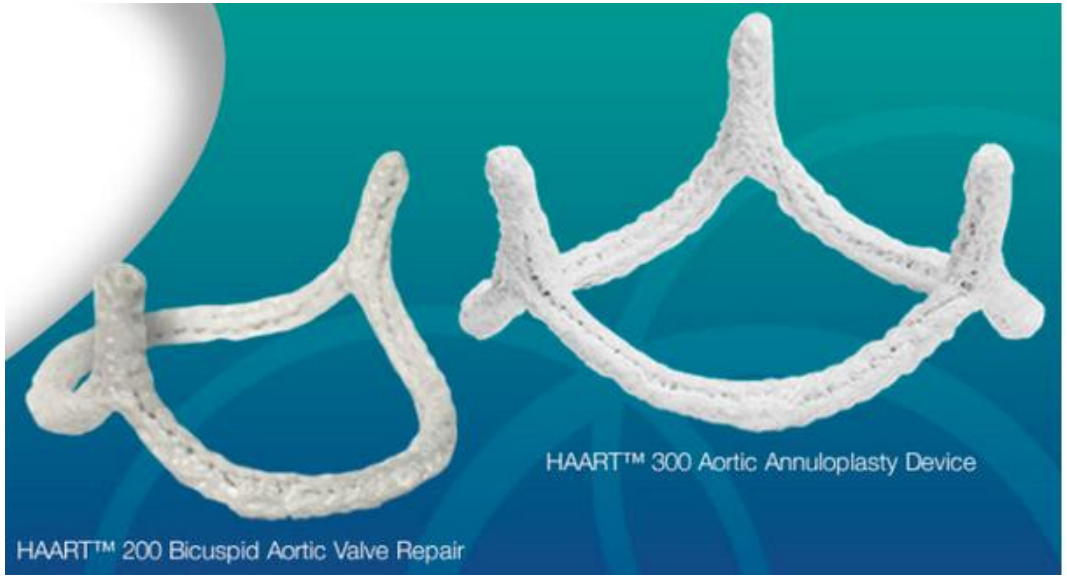
Intraoperative data	
Age at surgery Y (m, SD)	59(13.3)
CBP min (m, SD)	189.9(29.7)
Cross-clamp min (m, SD)	160.2(22)
Cusps plication (n, %)	21(20.2)
Free-edge sututure (n, %)	12 (11.5)
Associated interventions (n, %)	16(15.4)
Intraoperative conversion to Bentall (n, %)	2(1.9)

Post-operative data	**102 Patients**
ICU stay D (median, SD)	1(4.5)
VAM >72h (n, %)	2(1.9)
Respiratory insufficiency (n, %)	4(3.9)
AKI (n, %)	3(2.9)
Stroke (n, %)	0
Reoperation for bleeding (n, %)	1(0.9)
Blood transfusions (n, %)	18(17.6)
IMA (n, %)	1(0.9)
PM Implantation (n, %)	2(1.9)
Hospital stay D (m, SD)	7(5.2)
30-day mortality (n, %)	1(0.9)

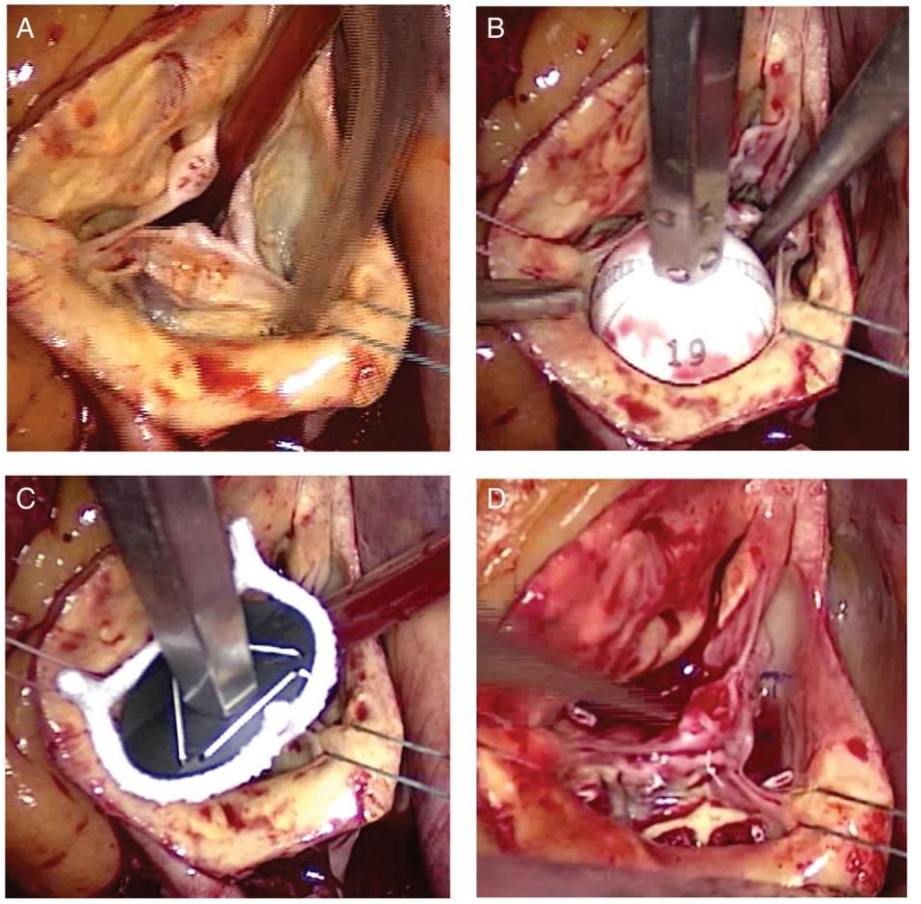
David procedures – University of Turin



…coming soon?



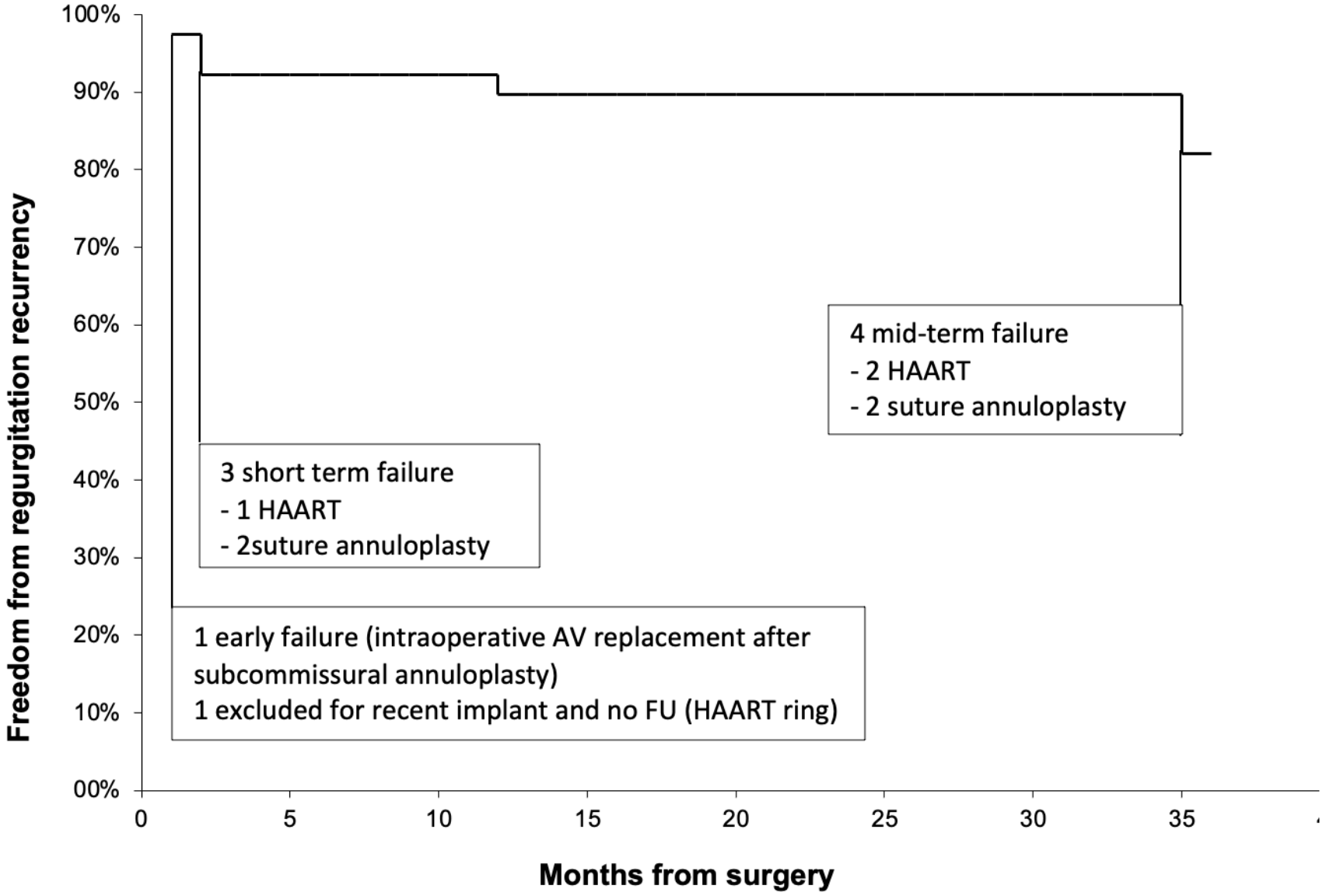
Annuloplasty Ring Placement



Eur J Cardiothorac Surg, Volume 49, Issue 3, March 2016, Pages 987–993,


75 YO F WITH 5.5 CM ASCENDING ANEURYSM AND 3+ AI
REPAIR WITH HAART RING

AV repair – University of Turin 2016-2022

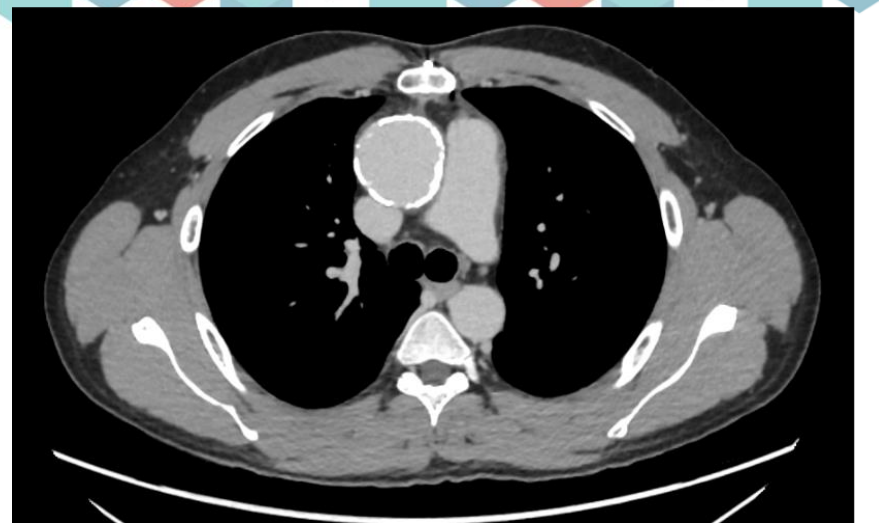


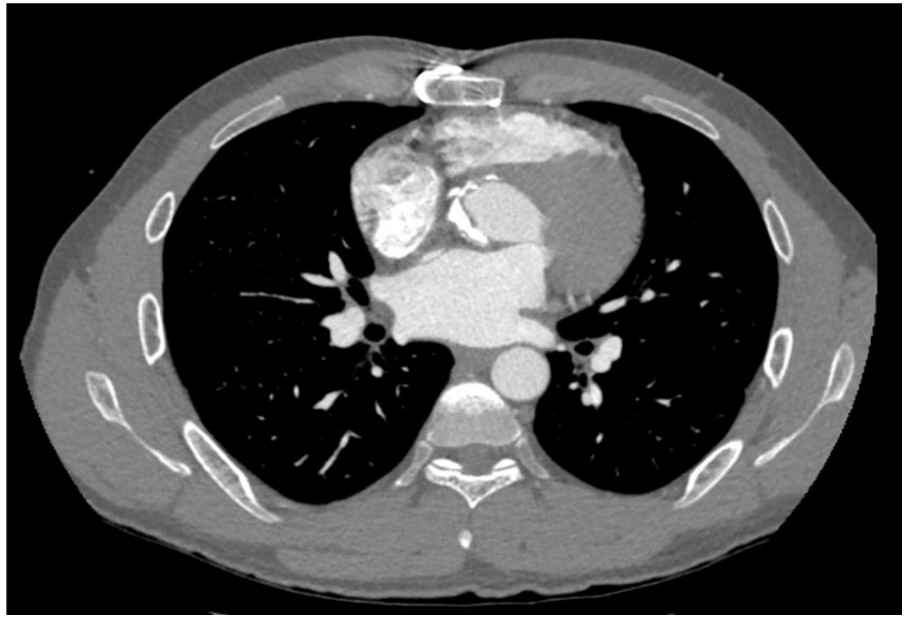
The importance of surgery planning

In GUCH patients, the majority of interventions are **re-interventions**.

Careful planning in Heart Team is mandatory to ensure a good result. Some expedients used in clinical practice are:

- careful study of medical history and previous surgeries
- CECT/Coronary CECT or cardiac MRI
- coronarography





VI CORSO GUCH

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Città della Salute e della Scienza di Torino

Thank you!