V CORSO GUCH

Il paziente adulto con cardiopatia congenita

La cardiochirurgia nel Fallot adulto

La via destra

Carlo Pace Napoleone

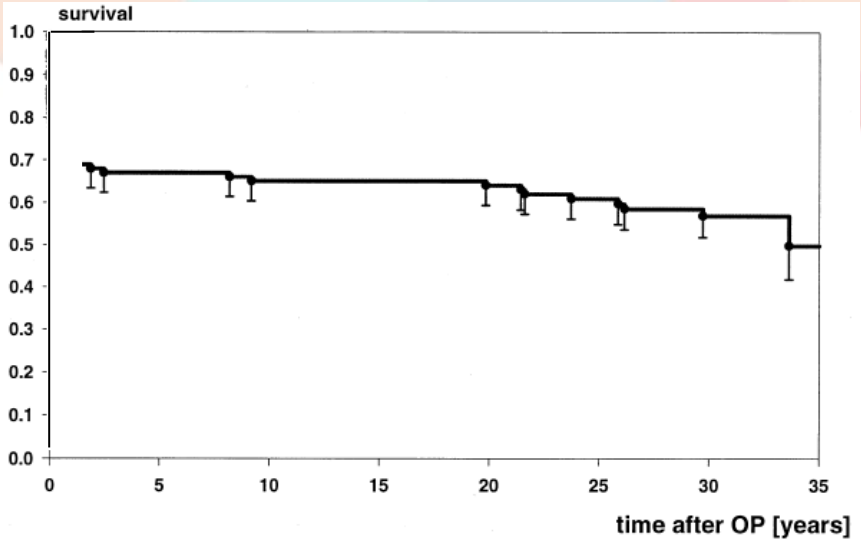
Long-Term Survival in Patients With Repair of Tetralogy of Fallot: 36-Year Follow-Up of 490 Survivors of the First Year After Surgical Repair

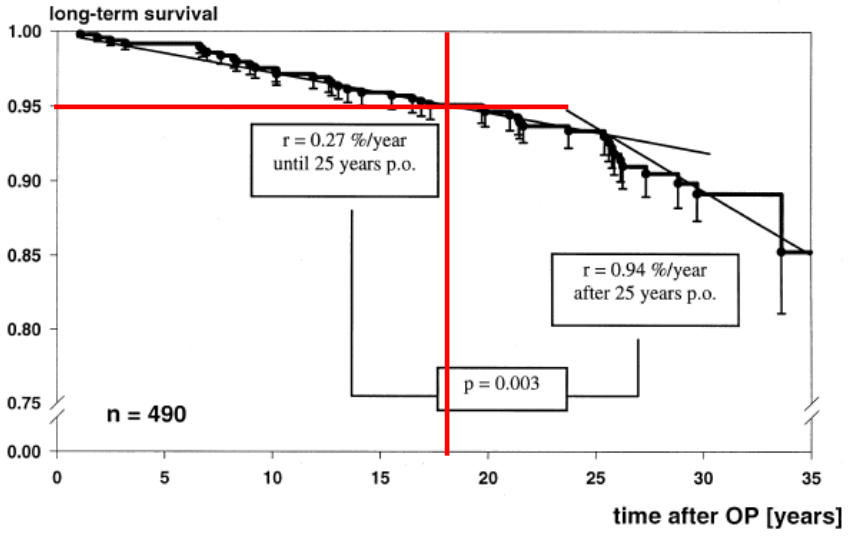
GEORG NOLLERT, MD,* TEDDY FISCHLEIN, MD, STEFAN BOUTERWEK, DMD, CHRISTINE BÖHMER, WERNER KLINNER, MD, BRUNO REICHART, MD

Munich, Germany

(J Am Coll Cardiol 1997;30:1374–83)

This study reviews our historic (1958 to 1977) long-term experience in the correction of TOF and pulmonary stenosis in 739 patients. Correction was performed predominantly in children 10 years old, and previous palliation was common. The large patient group is homogeneous because mainly all patients were operated on by one surgeon (W.K.) using the same technique, without changes in operative management.





Aspetti anatomici

Deviazione del setto conale

- Difetto interventricolare
- Destroposizione aortica
- Stenosi della via di efflusso destra
- Ipertrofia ventricolare destra

Intervento chirurgico correttivo

Chiusura del difetto interventricolare

- Per via trans-ventricolare
- Per via trans-atriale

Allargamento via di efflusso ventricolare destra

- Resezione ipertrofia ventricolare destra
- Ricostruzione della via di efflusso destra con patch eventualmente salvando la valvola polmonare se di calibro e morfologia adeguata

Chiusura difetto interventricolare

Per via trans-ventricolare infundibolare

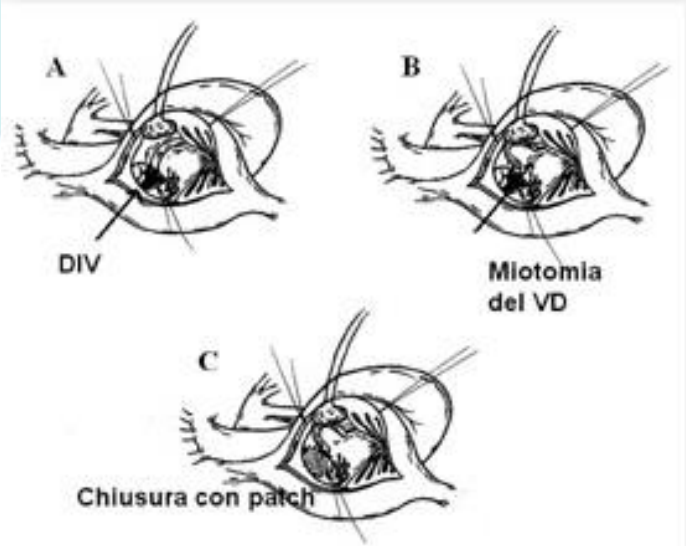
- Migliore visualizzazione del DIV
- Necessita di una ventricolotomia più ampia

Per via trans-atriale

- DIV più indaginoso da visualizzare
- Eventuale ventricolotomia limitata alla necessità di eseguire la resezione e l'allargamento infundibolare
- Maggior rispetto per l'infundibolo

Senza ventricolotomia

- Ipertrofia muscolare importante
- Ipoplasia minima dell'infundibolo
- Anulus polmonare di buon calibro
- Lembi valvolari normali



- Resezione muscolare per via trans-tricuspidalica

Con patch infundibolare

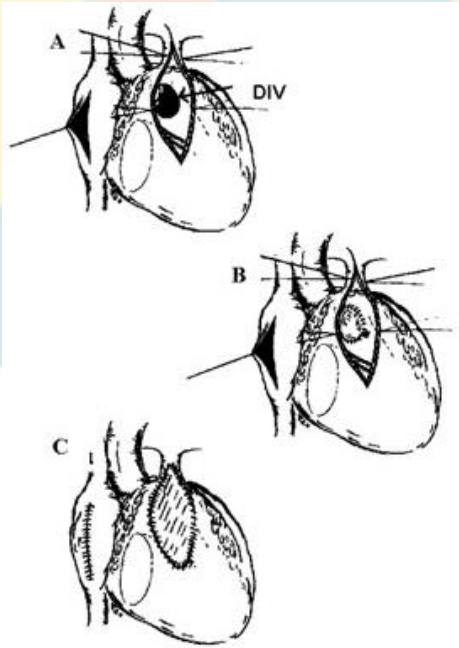
- Ipertrofia muscolare importante
- Ipoplasia severa dell'infundibolo
- Anulus polmonare di buon calibro
- Valvola polmonare normale

Patch infundibolare

- Incisione infundibolare, resezione muscolare per via trans-ventricolare e patch di allargamento infundibolare

Con patch transanulare

- Ipertrofia muscolare importante
- Ipoplasia severa dell'infundibolo
- Anulus polmonare di calibro ridotto
- Valvola polmonare displasica



- Incisione infundibolare estesa al tronco polmonare
- asportazione della valvola polmonare
- resezione muscolare per via trans-ventricolare
- patch di allargamento transanulare

Reinterventi sulla via destra

Ostruzione via di efflusso destra

Insufficienza polmonare severa

The role of primary surgical repair technique on late outcomes of Tetralogy of Fallot: a multicentre study

Massimo A. Padalino [a,*], Nicola Pradegan[a], Danila Azzolina[b], Lorenzo Galletti [c], Carlo Pace Napoleone [d], Salvatore Agati[e], Gaetano Palma[f], Stefano Maria Marianeschi[g], Francesco Seddio[c], Maria Teresa Cascarano[d], Cristina Carro[g], Dario Gregori [b], Vladimiro Lorenzo Vida[a] and Giovanni Stellin[a]

European Journal of Cardio-Thoracic Surgery 57 (2020) 565–573

720 patients undergoing primary ToF repair between 6/1990 and 12/2015 in 6 Italian Centers

	TV (n = 287)	TA (n = 433)	P-value
FU data			
FU duration (years), median (range)	4 (1–9)	4 (2–9)	0.20
Interventional procedure, n (%)	29 (12)	43 (13)	0.66
PDA/PAs stenting	17 (59)	37 (86)	
VSD closure	0	1 (2)	
PV implantation	0	0	
Surgical procedure, n (%)	19 (8)	20 (6)	0.43
PV replacement	3 (17)	2 (11)	
Residual RVOT stenosis resection	5 (28)	11 (58)	
PAs reconstruction	3 (17)	4 (21)	
Late death, n (%)	7 (3)	3 (1)	0.07

	No TAP (n = 249)	TAP (n = 471)	P-value
FU data, n (%)			
Interventional procedure	17 (8)	55 (15)	0.013
PDA/PAs stenting	10 (59)	44 (80)	
Residual VSD closure	0	1 (2)	
Percutaneous PV implantation	0	0	
Surgical procedure	8 (4)	31 (8)	0.029
PV replacement		5 (17)	
Residual RVOT stenosis resection	5 (62)	11 (38)	
PAs reconstruction	3 (38)	4 (14)	
Pulmonary regurgitation >2+ at FU	50 (30) (n = 161)	172 (60) (n = 283)	<0.001
Death at FU	3 (1)	7 (2)	0.65

The role of primary surgical repair technique on late outcomes of Tetralogy of Fallot: a multicentre study

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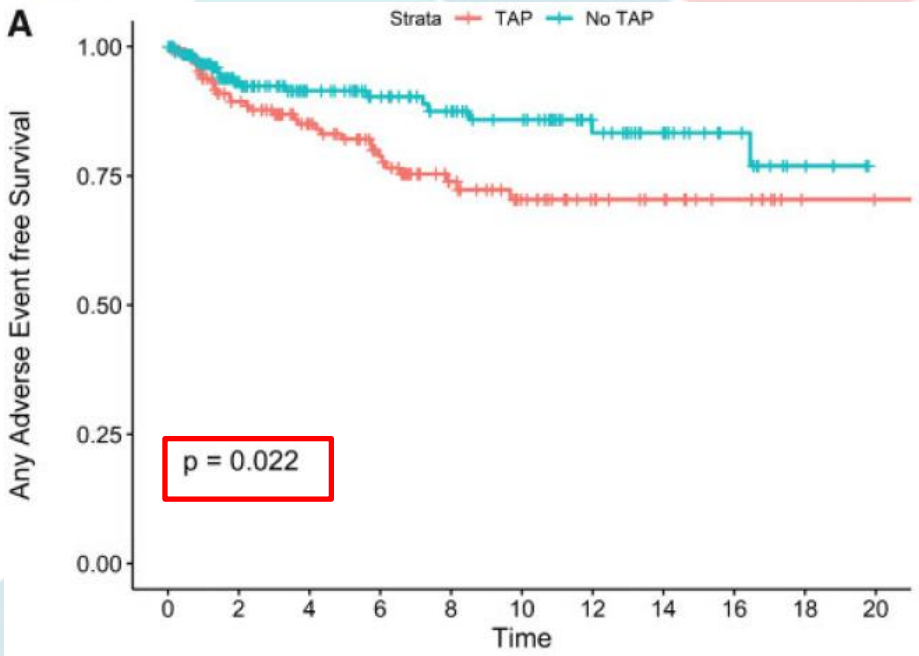
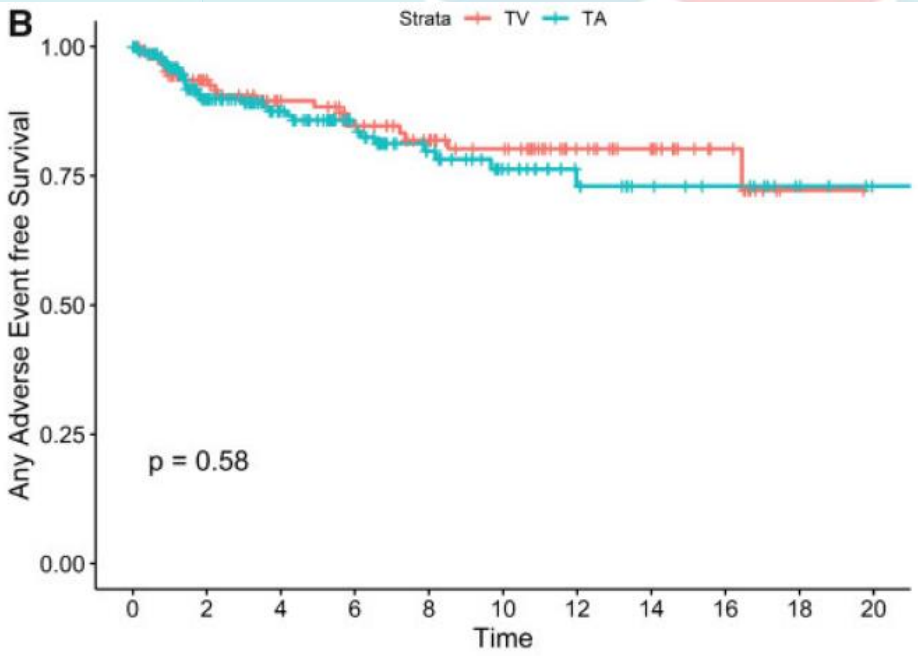
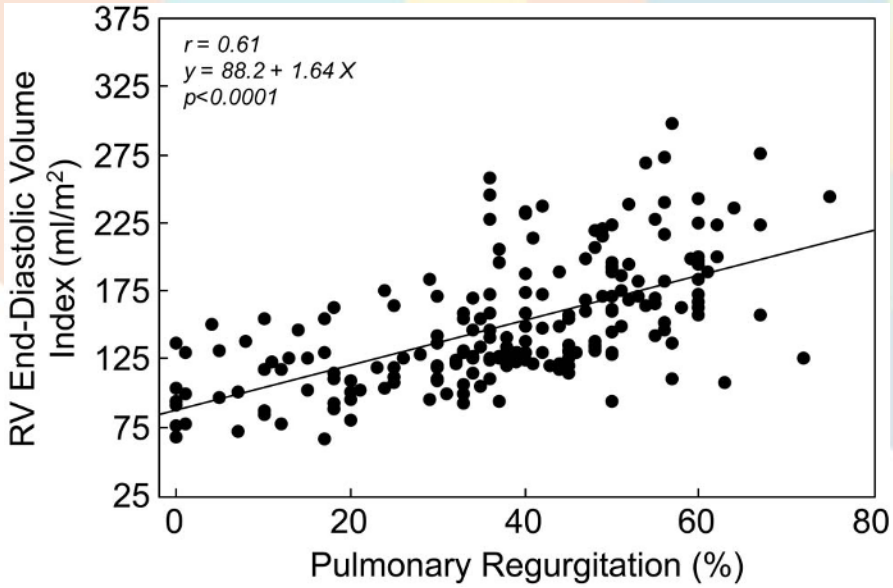
European Journal of Cardio-Thoracic Surgery 57 (2020) 565–573

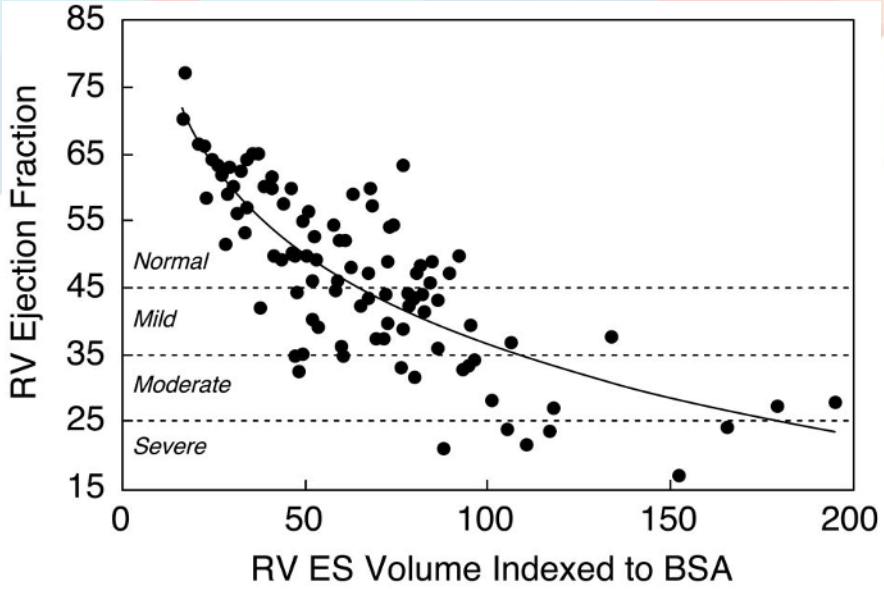
Overall adverse events at FU





Correlazione IP-dilatazione-disfunzione VDx



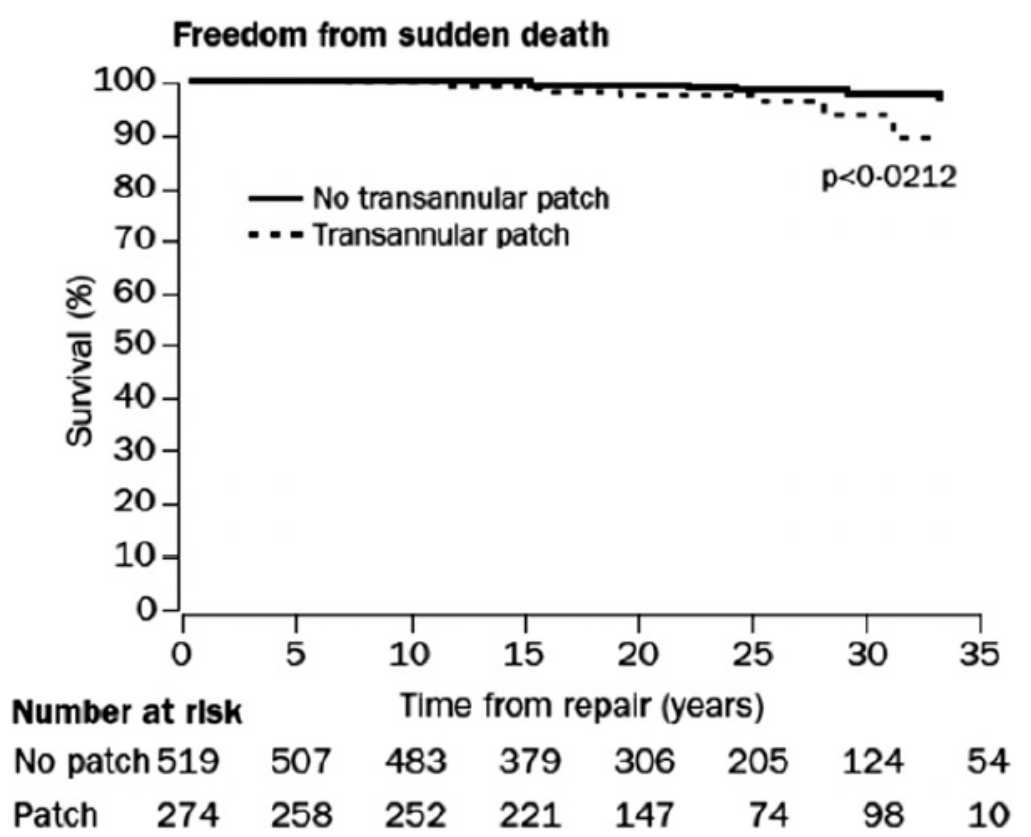


Repaired tetralogy of Fallot: the roles of cardiovascular magnetic resonance in evaluating pathophysiology and for pulmonary valve replacement decision support

Tal Geva

Journal of Cardiovascular Magnetic Resonance 2011, **13**:9

La differenza???



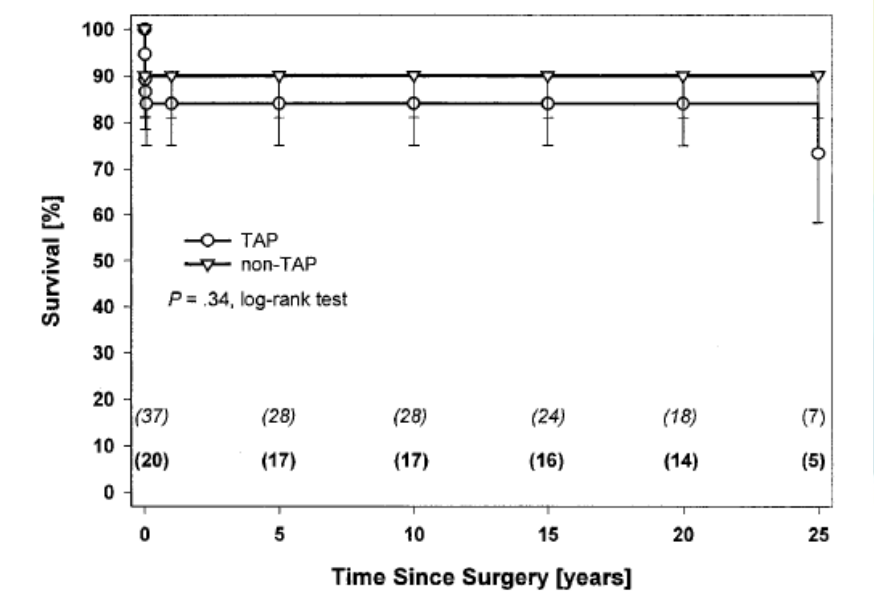
La via di efflusso destra

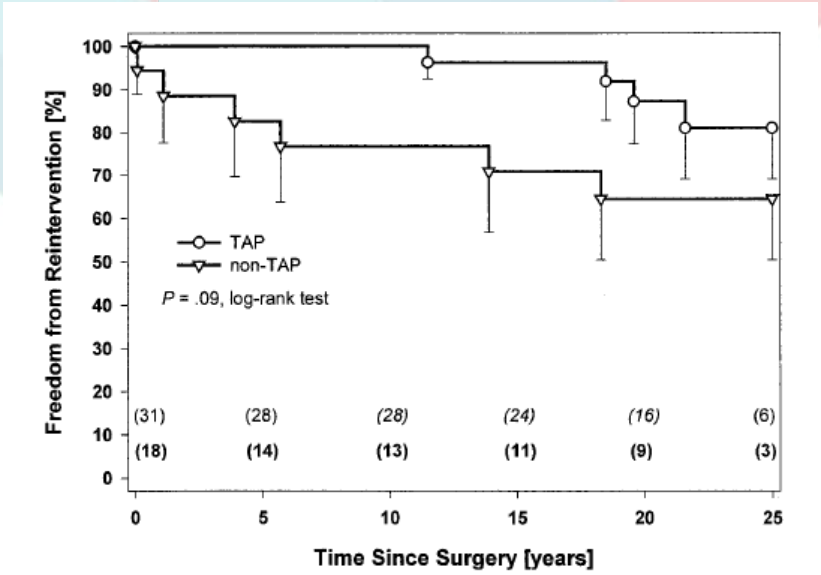
Timing and Technique of Pulmonary Valve Replacement in the Patient With Tetralogy of Fallot

James S. Tweddell,[a,b] Pippa Simpson,[c] Shun-Hwa Li,[c] Jennifer Dunham-Ingle,[b] Peter J. Bartz,[b,d] Michael G. Earing,[b,d] and Andrew N. Pelech[b,d]

Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 15:27-33 © 2012

…anche se…





Long-term results after early primary repair of tetralogy of Fallot

Emile A. Bacha, MD[a]
Albertus M. Scheule, MD[a*]
David Zurakowski, PhD[b]
Lars C. Erickson, MD[c]
Judy Hung, MD[c]
Peter Lang, MD[c]
John E. Mayer, Jr, MD[a]
Pedro J. del Nido, MD[a]
Richard A. Jonas, MD[a]

J Thorac Cardiovasc Surg 2001;122:154-61

Guidelines

Recommendations	Class[a]	Level[b]
PVRep is recommended in symptomatic patients with severe PR[c] and/or at least moderate RVOTO.[d]	I	C
In patients with no native outflow tract,[e] catheter intervention (TPVI) should be preferred if anatomically feasible.	I	C
PVRep should be considered in asymptomatic patients with severe PR and/or RVOTO when one of the following criteria is present. • Decrease in objective exercise capacity. • Progressive RV dilation to RVESVi ≥80 mL/m^2, and/or RVEDVi ≥160 mL/m^2 [f], and/or progression of TR to at least moderate. • Progressive RV systolic dysfunction. • RVOTO with RVSP >80 mmHg.	IIa	C

2020 ESC Guidelines for the management of adult congenital heart disease

The Task Force for the management of adult congenital heart disease of the European Society of Cardiology (ESC)

COR	LOE	Recommendations
Therapeutic		
I	B-NR	5. Pulmonary valve replacement (surgical or percutaneous) for relief of symptoms is recommended for patients with repaired TOF and moderate or greater PR with cardiovascular symptoms not otherwise explained.[S4.3.5-9–S4.3.5-11]
IIa	B-NR	6. Pulmonary valve replacement (surgical or percutaneous) is reasonable for preservation of ventricular size and function in asymptomatic patients with repaired TOF and ventricular enlargement or dysfunction and moderate or greater PR.[S4.3.5-1,S4.3.5-9,S4.3.5-12–S4.3.5-14]
IIa	B-NR	7. Primary prevention ICD therapy is reasonable in adults with TOF and multiple risk factors for SCD.[S4.3.5-15–S4.3.5-17]
IIb	C-EO	8. Surgical pulmonary valve replacement may be reasonable for adults with repaired TOF and moderate or greater PR with other lesions requiring surgical interventions.
IIb	C-EO	9. Pulmonary valve replacement, in addition to arrhythmia management, may be considered for adults with repaired TOF and moderate or greater PR and ventricular tachyarrhythmia.

AHA/ACC GUIDELINE

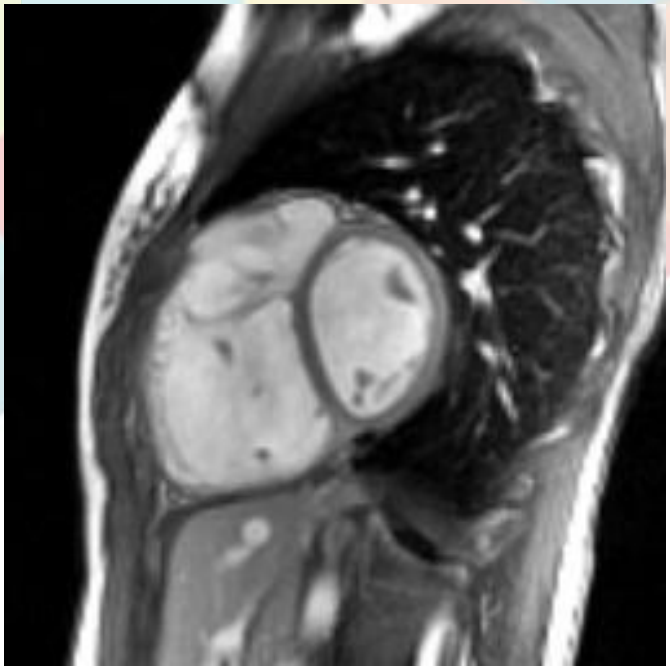
2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease

A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines

Indicazioni all'impianto di valvola polmonare

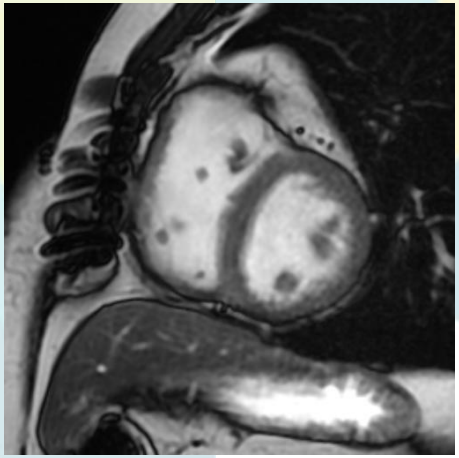
- Sintomatologia

- Risonanza magnetica
  - VTDVDx > 160 ml/m^2
  - VTSVDx > 80 ml/m^2



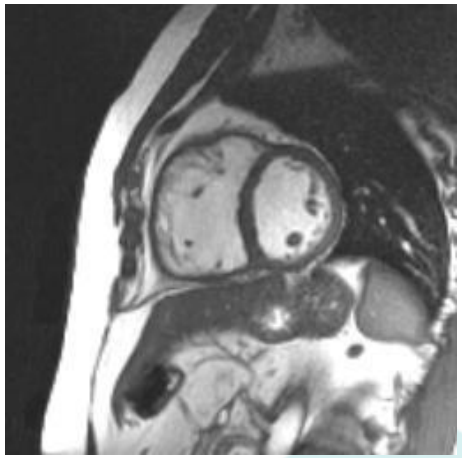
- Test cardiopolmonare
  - Riduzione della VO_2 max al di sotto del 60% del teorico

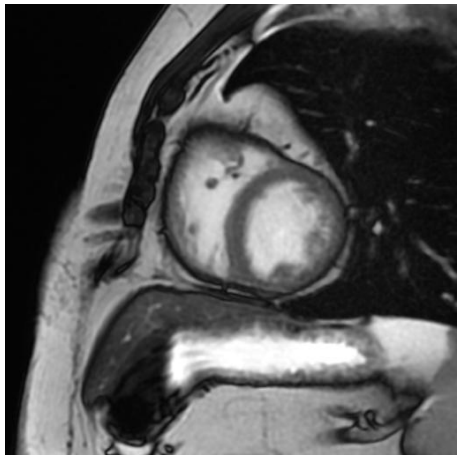
RM



Pre

Post





Valvole disponibili

…perchè abbiamo la BANCA!!!

graft?

sostituto della

…con ampia disponibilità!!!

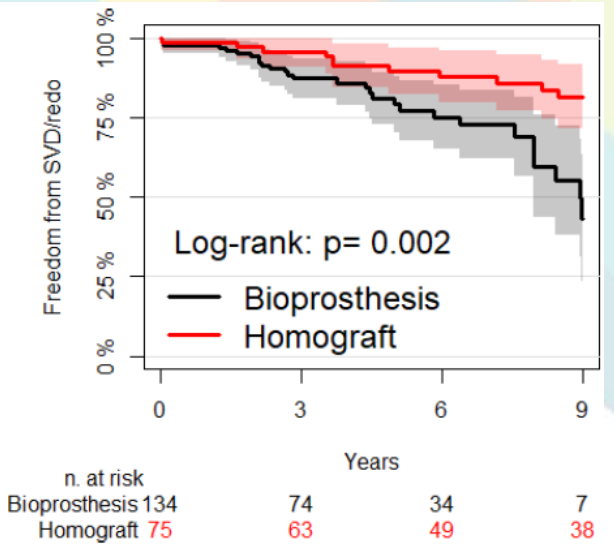
- Può essere
  valvolazion
- …e poi…

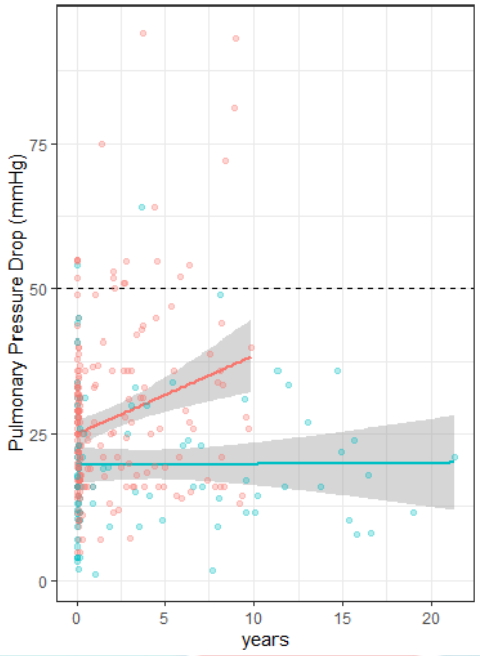
Characteristics	Homograft	Bioprosthesis	*P* Value
No.	75	134	. . .
Age, median (IQR), y	23 (12–35)	24 (18–34)	0.16
Male, n (%)	40 (53.3)	78 (58.2)	0.59
Chromosomal abnormality, n (%)	6 (8.0)	17 (12.7)	0.42
Smoking history, n (%)	14 (18.7)	26 (19.4)	1.0
LVEF <50%, n (%)	8 (10.7)	24 (17.9)	0.23
BSA, mean (SD), m^2	1.59 (0.44)	1.72 (0.30)	0.02
Concomitant TVR, n (%)	3 (4.0)	21 (15.7)	0.02
Concomitant RVOT reconstruction, n (%)	10 (13.3)	15 (11.2)	0.81
Concomitant PA plasty, n (%)	16 (21.3)	29 (21.6)	1.0
Time from repair, median (IQR), y	20 (12–27)	22 (16–31)	0.02
Long-term antiplatelet therapy, n (%)	31 (41.3)	55 (41.0)	1

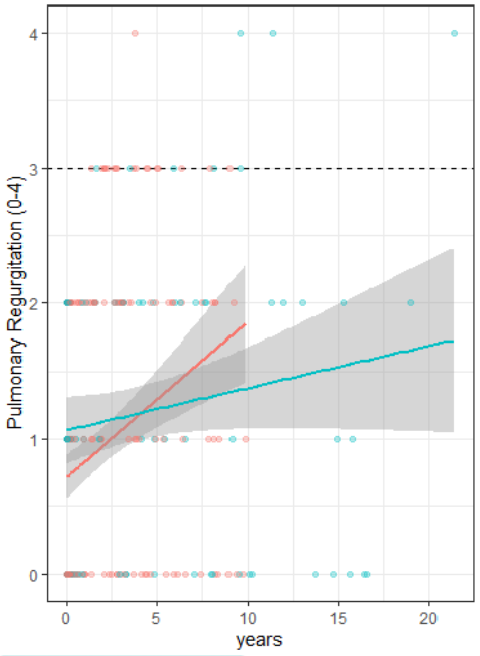
Long-Term Comparison Between Pulmonary Homograft Versus Bioprosthesis for Pulmonary Valve Replacement in Tetralogy of Fallot

Lucia Cocomello, MD; Marco Meloni, MD; Filippo Rapetto, MD; Mai Baquedano, MSc; Maria Victoria Ordoñez, MD; Giovanni Biglino, PhD; Chiara Bucciarelli-Ducci, MD, PhD; Andrew Parry, MD; Serban Stoica, MD; Massimo Caputo, MD

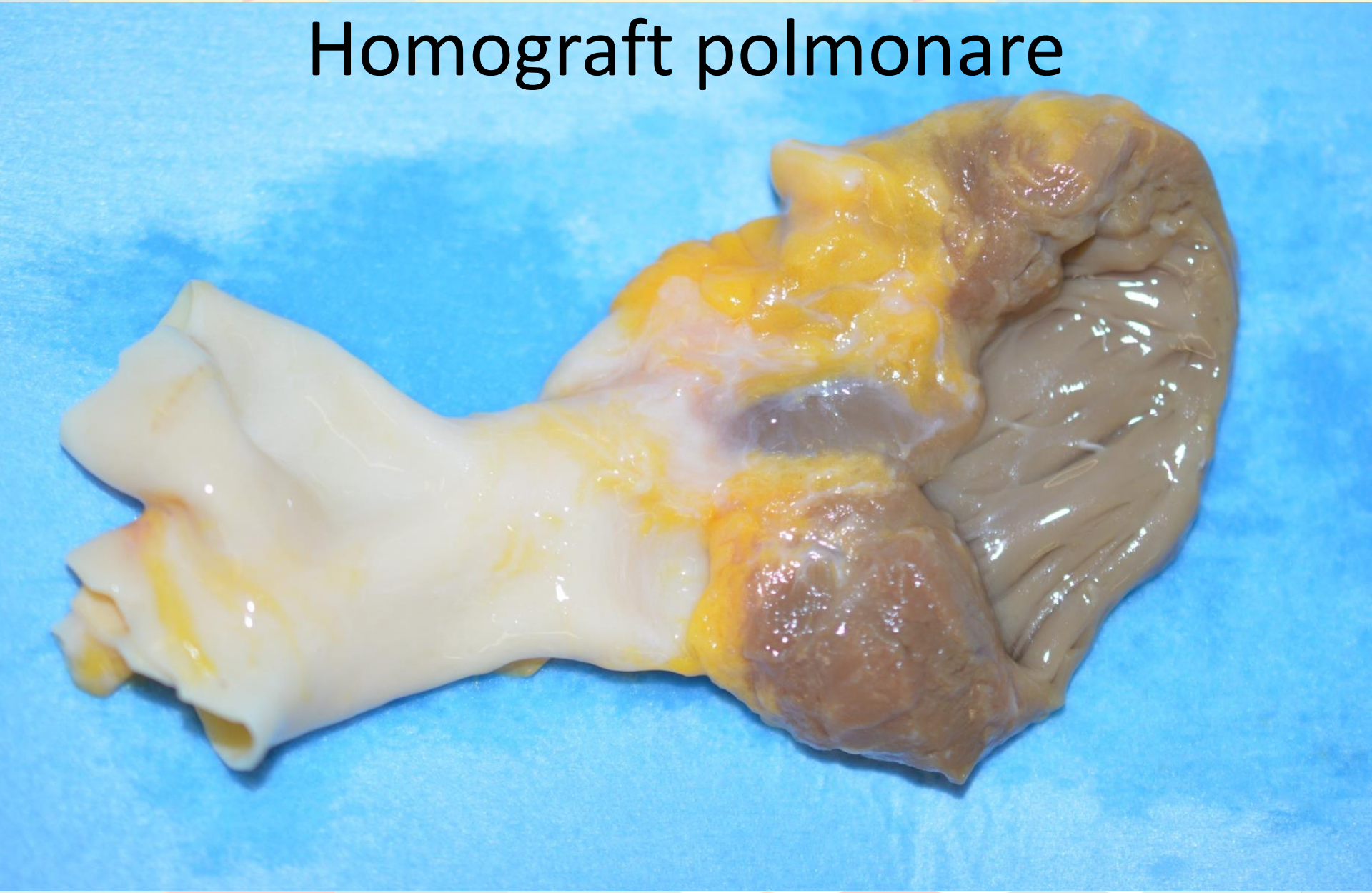
(*J Am Heart Assoc*. 2019;8:e013654.

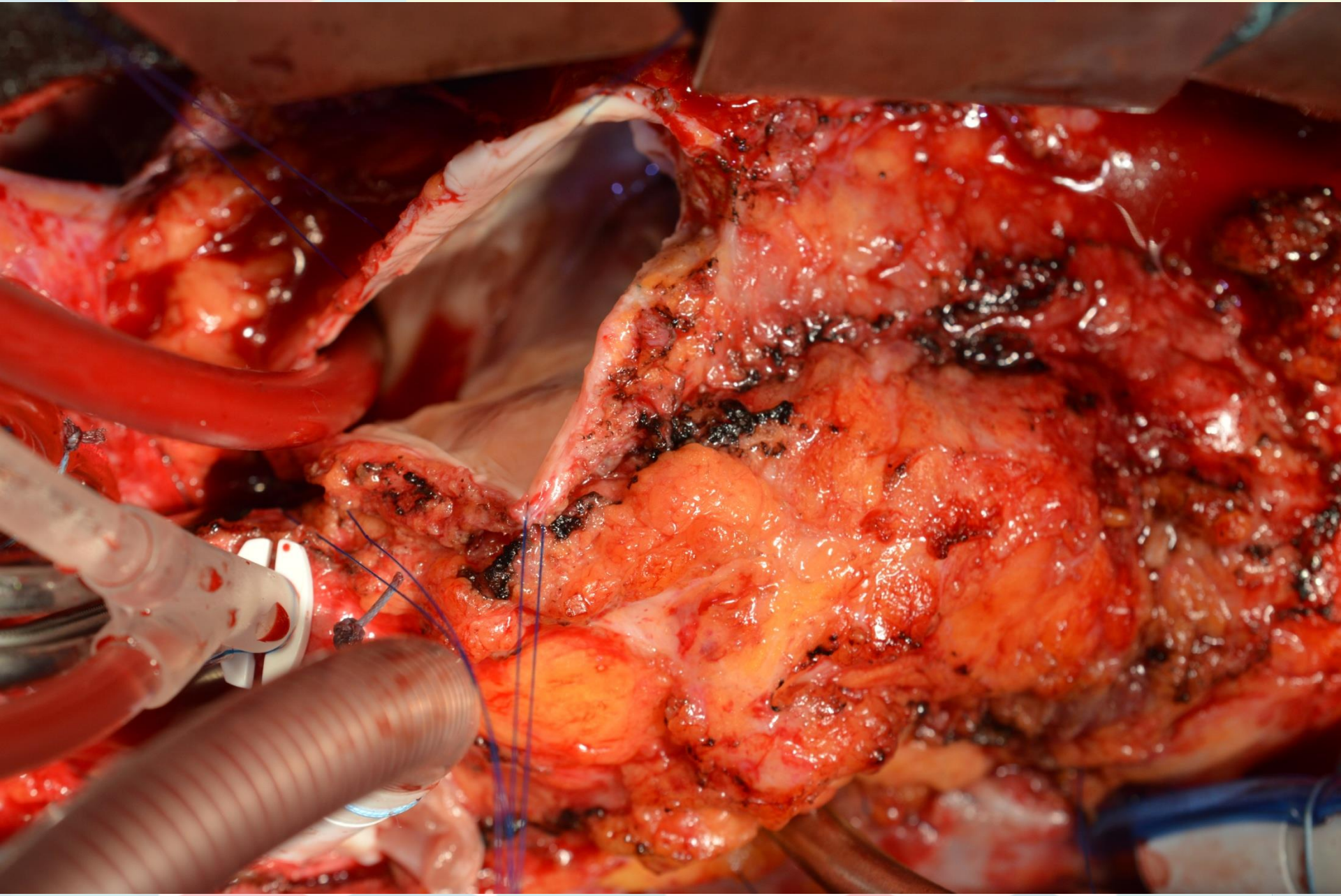


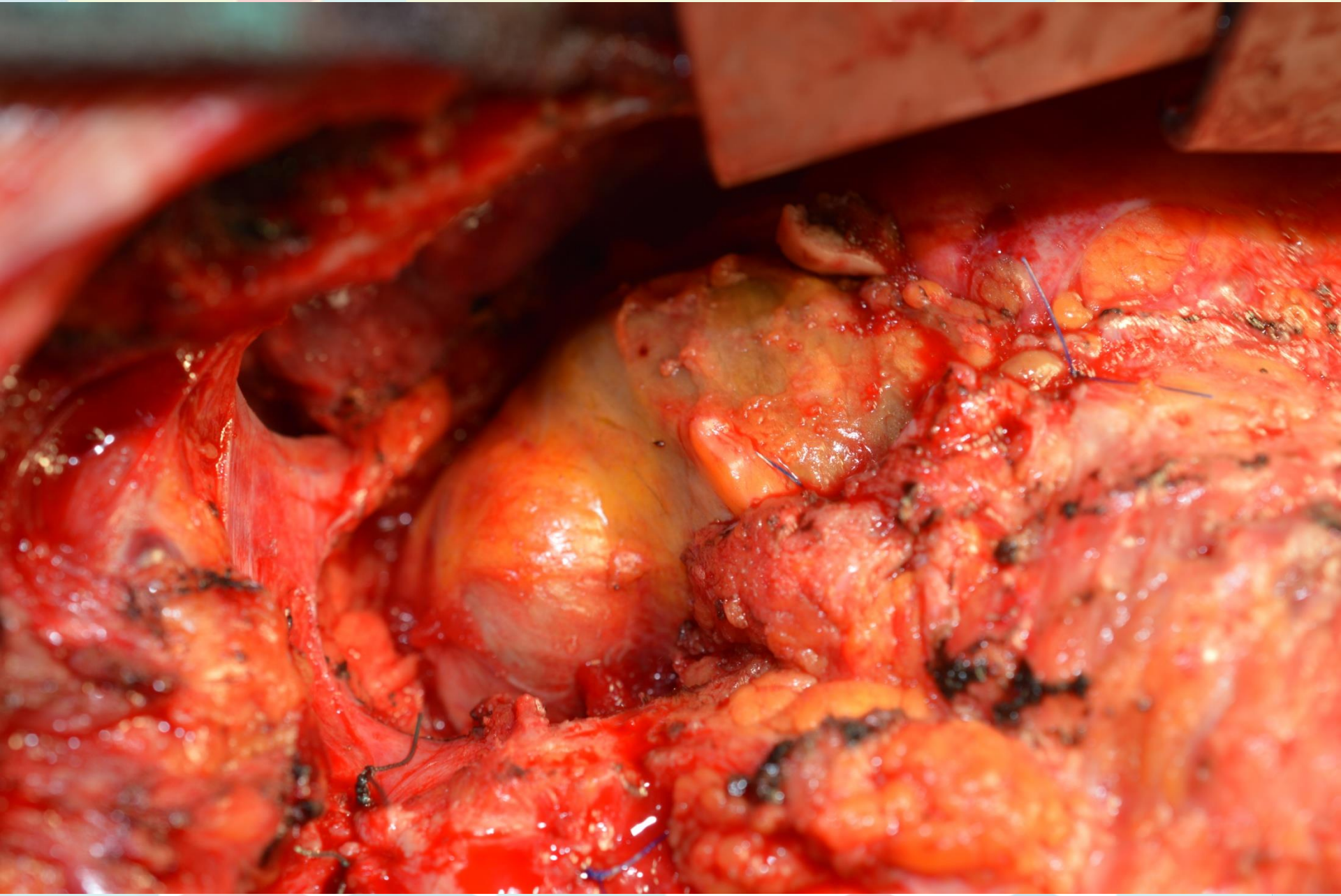




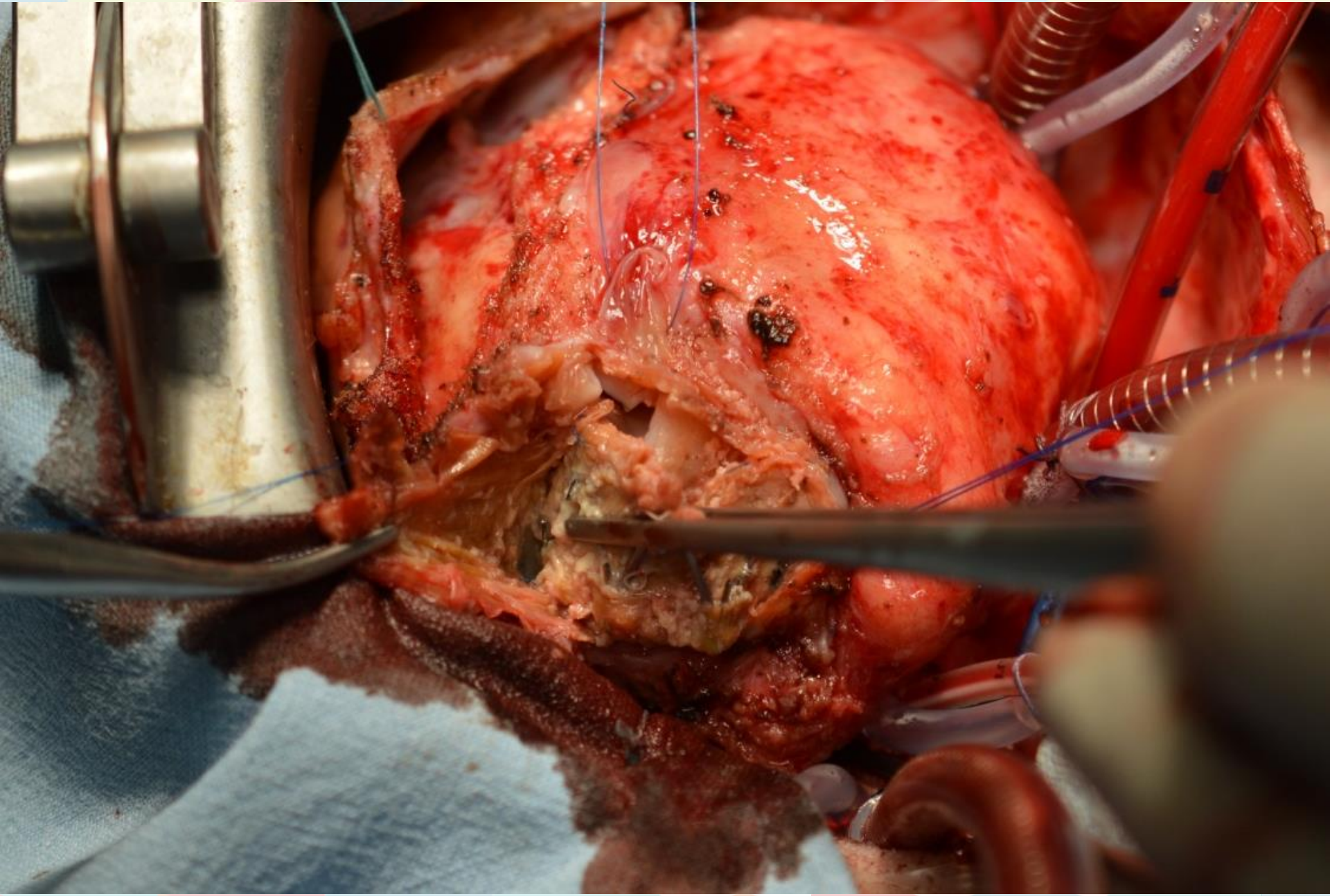
Homograft polmonare

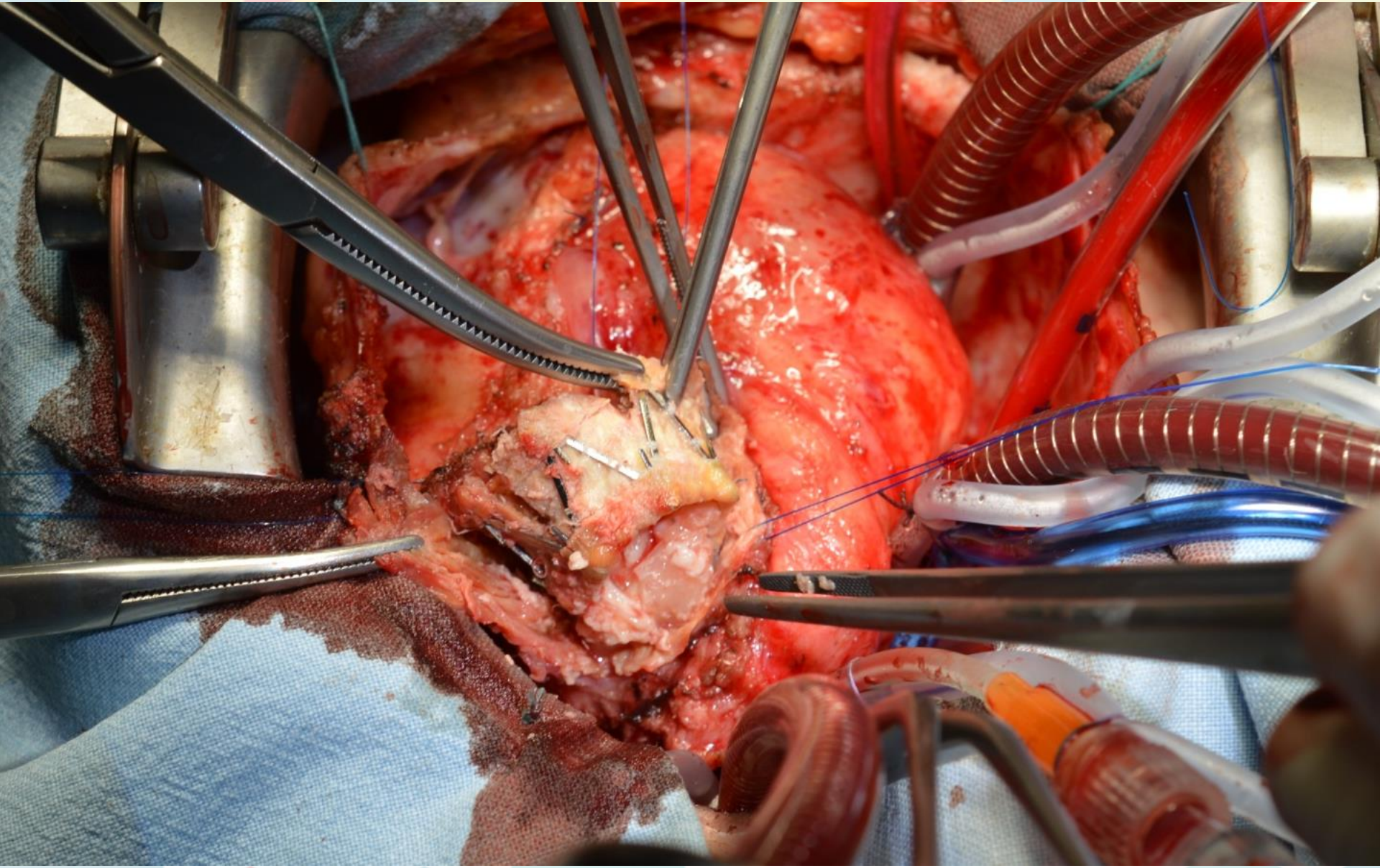


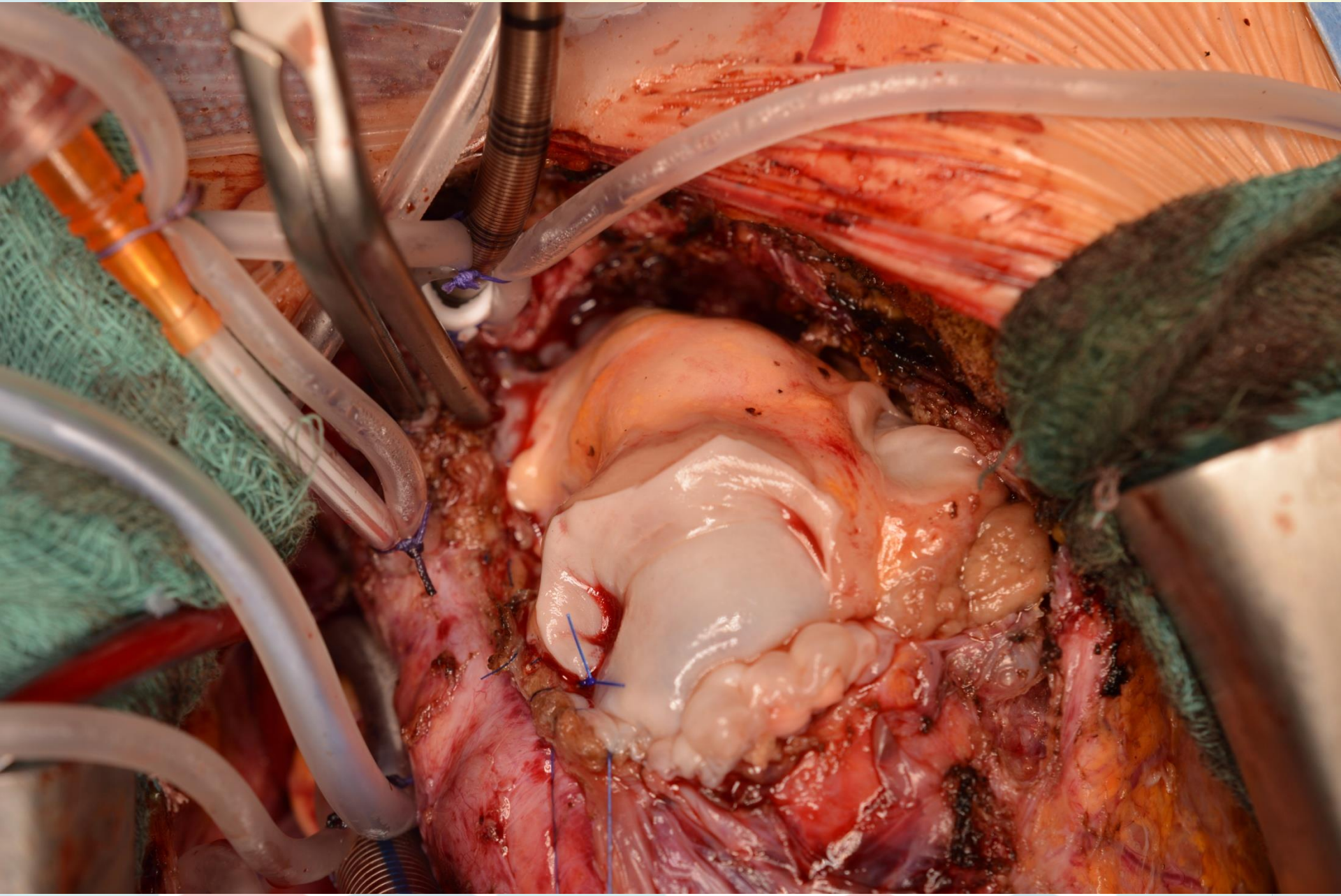




Homograft aortico

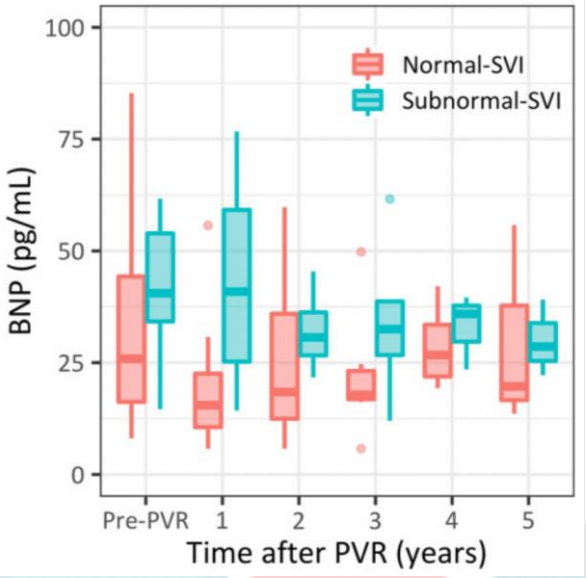






Timing

	Before PVR	After PVR	*P*-value
LVEDVI (ml/m^2)	80 [72, 89]	80 [77, 85]	0.96
LVESVI (ml/m^2)	37 [33, 47]	38 [33, 43]	0.16
LVEF (%)	51 [47, 55]	53 [50, 58]	0.052
LVSVI (ml/beat/m^2)	40 [35, 46]	43 [40, 47]	0.22
RVEDVI (ml/m^2)	203 [187, 239]	110 [97, 125]	<0.001
RVEF (%)	43 [38, 48]	38 [34, 45]	0.045



	Normal-SVI (*n* = 20)	Subnormal-SVI (*n* = 10)	*P*-value
Cardiac magnetic resonance			
LVEDVI (ml/m^2)	81 [79, 86]	77 [72, 81]	0.12
LVEF (%)	54 [52, 58]	51 [47, 57]	0.29
LVSVI (ml/beat/m^2)	44 [42, 47]	40 [34, 42]	0.038
RVEDVI (ml/m^2)	110 [103, 127]	109 [94, 117]	0.46
RVEF (%)	40 [34, 46]	38 [37, 41]	0.84
Other variables			
LVEDP (mmHg)	10 [5, 12]	7 [6, 11]	0.78
QRS duration (ms)	164 [156, 169]	145 [116, 165]	0.19
BNP (pg/ml)	16 [12, 23]	41 [25, 59]	0.002
Peak VO_2 (% of predicted normal)	74 [67, 77]	64 [59, 78]	0.41

The reduced left ventricular stroke volume does not fully recover after pulmonary valve replacement in patients with repaired tetralogy of Fallot

Takashi Yasukawa [a], Takaya Hoashi [a], Kenta Imai[a], Naoki Okuda[a], Tetsuya Fukuda[b], Hideo Ohuchi[c], Kenichi Kurosaki[c] and Hajime Ichikawa[a,*]

European Journal of Cardio-Thoracic Surgery 60 (2021) 526–533

E la tricuspide?

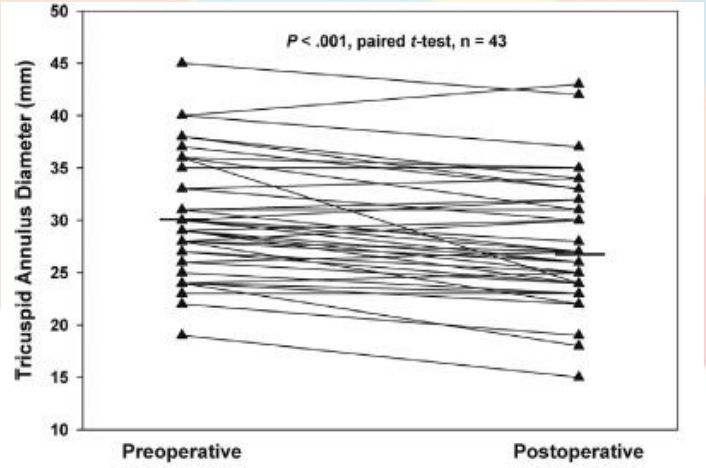
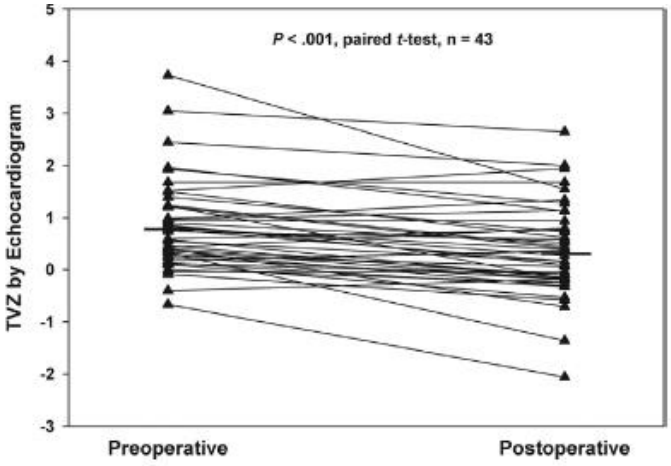
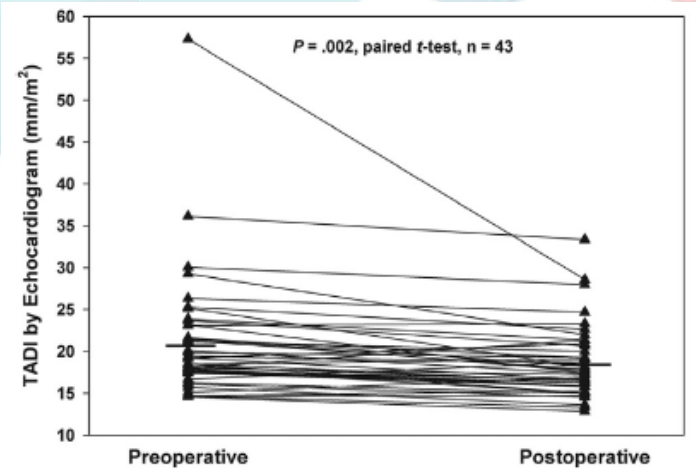


Table 2. Preoperative and Postoperative Echocardiographic Data

Parameter ($N = 43$)	Preoperative	Postoperative	P Value
TAD in mm, mean ± SD	30 ± 5	28 ± 6	<0.001
TADI in mm/m^2, mean ± SD	20.95 ± 7.24	19.39 ± 5.37	0.001
TVZ, median (IQR)	0.82 (0.26-1.22)	0.3 (−0.18 to −0.79)	<0.001
TR Grade, mean ± SD	2.1 ± 0.3	2.0 ± 0.3	0.08





Should Tricuspid Annuloplasty be Performed With Pulmonary Valve Replacement for Pulmonary Regurgitation in Repaired Tetralogy of Fallot?

Mustafa Kurkluoglu, MD,[*] Anitha S. John, MD, PhD,[†] Russell Cross, MD,[†] David Chung, MBBS,[*] Can Yerebakan, MD,[*] David Zurakowski, PhD,[‡,§] Richard A. Jonas, MD,[*] and Pranava Sinha, MD[*]

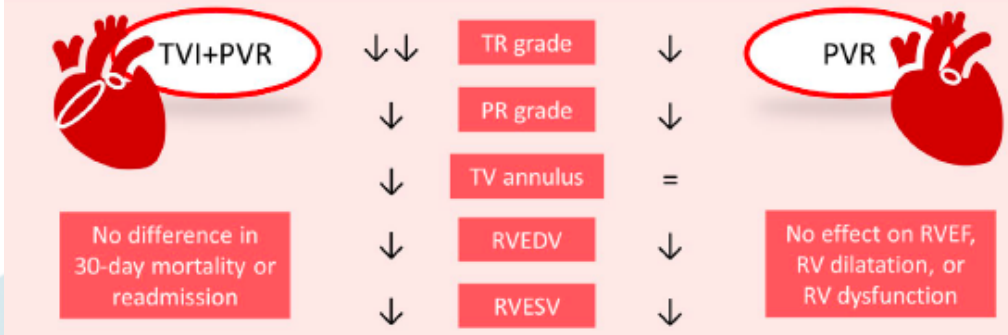
Semin Thoracic Surg 27:159–165 © 2015

Meglio ripararla?

First author	Year	Study period	Country	Design	Patient no.	FU time	TR grade
Deshaies[16]	2020	2000–2016	Canada	NP, NR, NM	542 (180 TVI+PVR, 362 PVR)	3 mo	Mild TR in 254 (19 TVI+PVR, 235 PVR), moderate TR in 192 (90 TVI+PVR, 102 PVR), severe TR in 72 (68 TVI+PVR, 4 PVR)
Taejung Kim[17]	2019	2000–2016	South Korea	NP, NR, NM	67 (38 TVI+PVR, 29 PVR)	5.5±2.7 mo	TR grade: 2.79±0.95 in TVI+PVR, 1.45±0.56 in PVR
Lueck[18]	2018	2009–2017	Germany	NP, NR, NM	28 (10 TVI+PVR, 18 PVR)	ND	TR grade: 2.0±0.77 in TVI+PVR, 1.94±0.62 in PVR
Roubertie[19]	2017	2002–2014	France	NP, NR, NM	41 (16 TVI+PVR, 25 PVR)	54.6±36.6 mo	Moderate TR in 24 (8 TVI+PVR, 16 PVR), severe TR in 17 (8 TVI+PVR, 9 PVR)
Cramer[20]	2015	1999–2012	USA	NP, NR, NM	36 (18 TVI+PVR, 18 PVR)	6 mo	TR grade: 2.7±0.5 in TVI+PVR, 2.2±0.4 in PVR
Kogon[21]	2015	2002–2008	USA	NP, NR, NM	35 (16 TVI+PVR, 19 PVR)	7.0±2.8 y	TR grade: 2.63±0.43 in TVI+PVR, 2.08±0.26 in PVR

Tricuspid Valve Intervention at Pulmonary Valve Replacement: Systematic Review and Meta-analysis

- Meta-analysis of 749 adults with congenital heart disease from 6 studies
- Comparing concomitant tricuspid valve intervention and pulmonary valve replacement (TVI+PVR) vs isolated PVR



Despite the lack of clear support of routine TVI at the time of PVR, this strategy seems to reduce TR grade to a larger extent than PVR alone

Tricuspid Valve Intervention at the Time of Pulmonary Valve Replacement in Adults With Congenital Heart Disease: A Systematic Review and Meta-Analysis

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Pulmonary Insufficiency Leads to Right Ventricular Enlargement, which Leads to Tricuspid Regurgitation, which Leads to more Right Ventricular Enlargement, which Leads to more Tricuspid Regurgitation: a Vicious Cycle?

Charles B. Huddleston, MD **Seminars in Thoracic and Cardiovascular Surgery • Volume 27, Number 2**

- L'insufficienza tricuspidalica non deriva necessariamente da una dilatazione del VDx
  - Anulus tricuspidalico distorto dal patch interventricolare
  - Le corde del lembo settale possono essere state danneggiate o distorte dalla cicatrice del patch

- Il miglioramento della tricuspide è legato alla riduzione del volume del ventricolo destro che, a sua volta, dipende dalla funzionalità della valvola polmonare…che potrebbe non essere stabile nel tempo…

Nostra esperienza

1998-2022	45 pz
Palliazione	26 pz

- 20 Shunt Ao-Po
- 6 Valvulotomie polmonari

Correzione Fallot	9 ± 12 aa (1m-36aa)

Nostra esperienza

Aritmie Pre intervento	15 pz

- 2 PM
- 13 FA-Flutter

Età reintervento	34 ± 16 aa (18 - 71)
Intervallo correzione	27 ± 13 aa (2 - 55)
Altre procedure	20 pz

- 12 VPL Tricuspide
- 6 Plastica rami polmonari
- 1 SVAo
- 1 VPL Mitrale

Nostra esperienza

Mortalità ospedaliera 3 pz (6.7%)

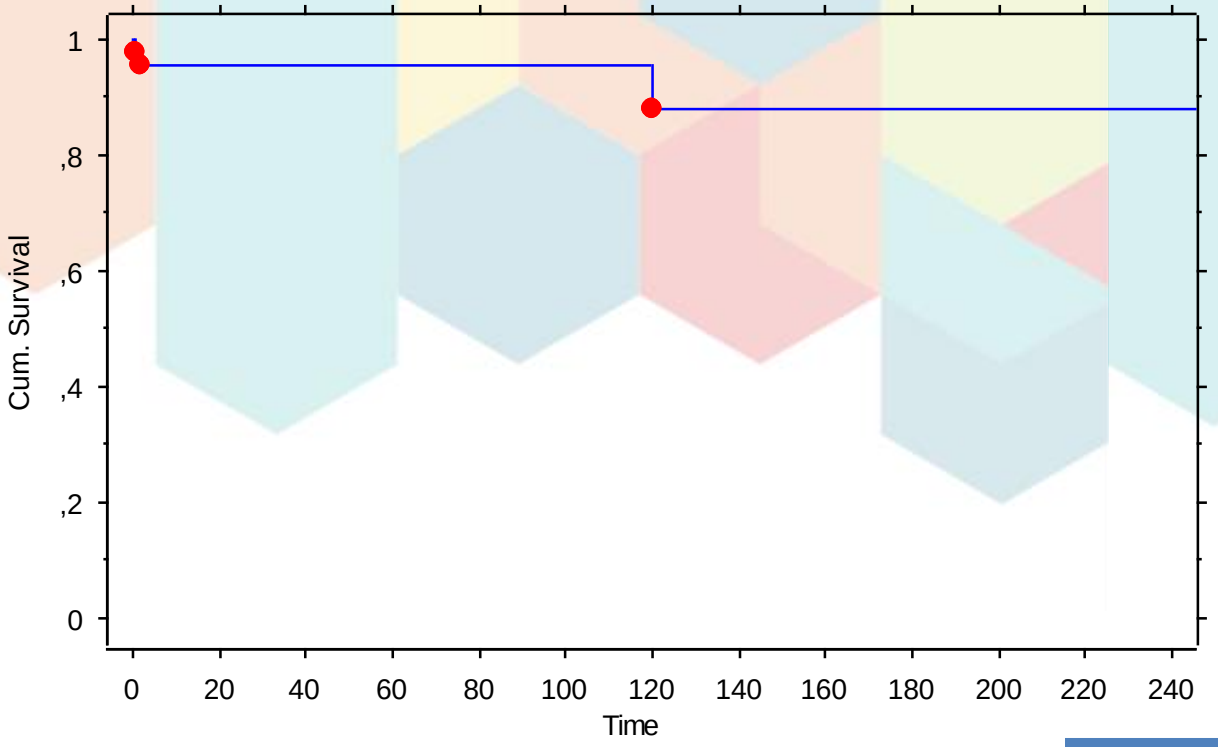
FU Medio 7 ± 8 aa (1m – 24 aa)

Mortalità al FU 1 pz (2.2%)

Interventi FU 8 pz

- 3 PM
- 2 Sostituzioni homograft
- 1 VPL Polmonare
- 2 Impianti valvola polmonare percutanea

Curva di sopravvivenza



		Pts at risk
5aa	95.5%	19
10aa	88.2%	13
20aa	88.2%	6

Quindi…

I pazienti nati con tetralogia di Fallot raggiungono l'età adulta in più del 90% dei casi

Hanno bisogno di uno stretto follow-up che intercetti in tempo le loro esigenze assistenziali

La gestione chirurgica degli esiti della correzione è complessa anche se può essere ottenuta con ottimi risultati

Questi sono strettamente correlati alla capacità di interazione tra gli specialisti dell'adulto e pediatrici

Grazie per l'attenzione

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