Surgical Approach in DORV with Subpulmonary Conus

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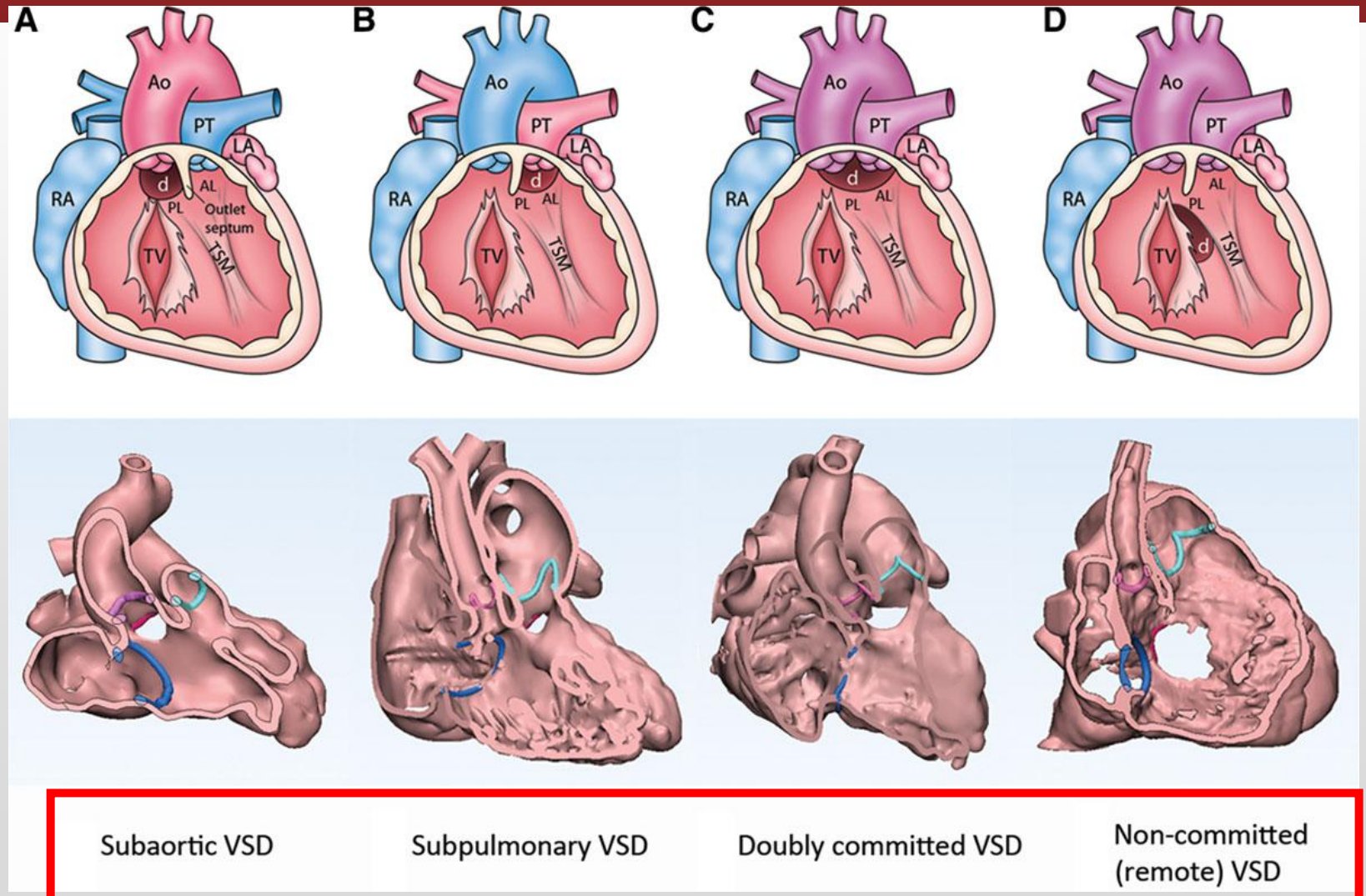
Surgical Intervention for DORV



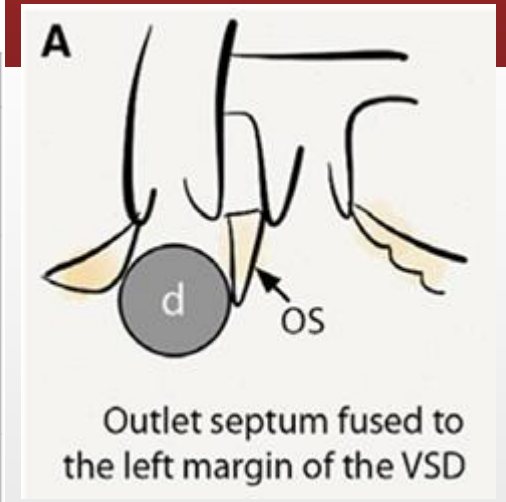
Different Types of DORV dictate different surgical approaches

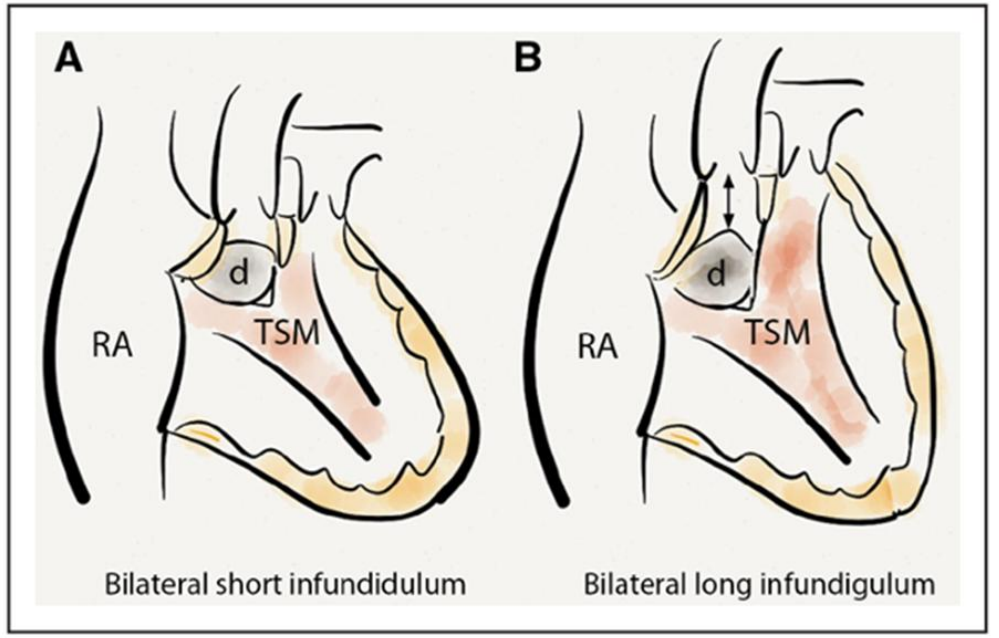
Circ Cardiovasc Imaging 2018 11

Its not as simple as 4 types based on the VSD

Table. List of Essential Modifiers of Surgical Anatomy of Double Outlet Right Ventricle

Features	Primary	Secondary
Relationship of the atrioventricular conduction axis to the VSD margin	Perimembranous VSD Nonperimembranous VSD Atrioventricular septal defect	
Location of the VSD seen from the right ventricle	Predominantly outlet Predominantly inlet Confluent inlet and outlet Predominantly apical trabecular Confluent involving all 3 parts	Relationship of the VSD to the tricuspid valve annulus: Along <upper 1/3 Along upper 1/3 to 2/3 Along >upper 2/3
Size and multiplicity of the VSD	Unrestrictive Restrictive No identifiable VSD	Single Multiple
Orientation of the outlet septum relative to the VSD margin	To the left margin of the VSD To the right margin of the VSD Parallel with the plane of the VSD Not related to the VSD margin Deficient or vestigial	
Muscular infundibulum	Subaortic Subpulmonary Bilateral Bilaterally deficient	Ex
Great arterial relationship	Normally related Mirror-image of normal Dextro-malposed Levo-malposed Side-by-side with aorta on the right Side-by-side with aorta on the left	





Its not as simple as 4 types based on the VSD

Outflow tract stenosis	Subaortic stenosis Aortic valvar stenosis Subpulmonary stenosis Pulmonary valvar stenosis Pulmonary valvar atresia	Aortic arch Unobstructed Tubular hypoplasia Coarctation Interruption
Type of DORV	VSD location per Lev et al's[9] classification: Subaortic Subpulmonary Doubly committed Noncommitted or remote Aligned with the subaortic outflow Aligned with the subpulmonary outflow Aligned with neither outflow	STS-EACTS-AEPC class: VSD type Tetralogy type TGA type Noncommitted VSD type AVSD
Atriovenricular valve abnormalities	Stenosis of the tricuspid or mitral valve Straddling or over-riding of the tricuspid or mitral valve Insertion of the atrioventricular valve tension apparatus to the margin of the VSD or outlet septum	
Ventricular volumes	Right ventricular volume Enough space for intraventricular baffling Too little space for intraventricular baffling	Left ventricular volume Normal Borderline hypoplasia Too small
Other findings and associated abnormalities	Anomalous systemic venous connection Anomalous pulmonary venous connection Juxtaposition of the atrial appendages Coronary arterial origins and distribution	

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How common is DORV with Subaortic VSD and what associated issues are there?

CONGENITAL: DOUBLE OUTLET RIGHT VENTRICLE

Repair of double outlet right ventricle: Midterm outcomes

Olubunmi Oladunjoye, MBBS, MPH,[a,b] Breanna Piekarski, BSN, RN,[a] Christopher Baird, MD,[a] Puja Banka, MD,[c] Gerald Marx, MD,[c] Pedro J. del Nido, MD,[a] and Sitaram M. Emani, MD[a]

JTCVS

Characteristic	N = 238	Primary BiV repair (n = 158)	Staged BiV repair (n = 80)
Anatomy type			
Doubly committed VSD	18 (7.6)	12 (7.7)	6 (7.6)
Noncommitted VSD	80 (33.6)	30 (19.2)	50 (63.3)
Subaortic VSD	78 (32.8)	62 (39.7)	16 (20.3)
Subpulmonary VSD	59 (24.8)	52 (33.3)	7 (8.9)

Approx 1/3 of patients have DORV with subaortic VSD
21% had an intervention before their definitive repair

JTCVS 2020 159:254-264

How common is DORV with Subaortic VSD and what associated issues are there?

Characteristic	Subaortic
Heterotaxy	2 (2.6)
Dextrocardia	5 (6.4)
Age at surgery (mo)	3.7 (1.5-11.3)
VSD enlargement	12 (15.4)
Atrial switch	3 (3.9)
CPB time (min)	140 (108-174)
Crossclamp time (min)	88 (66-119)
Ventilation time (d)	2 (1-5)
ICU length of stay (d)	5 (3-9)
Hospital length of stay (d)	9 (6-14)

Outcomes not reported for each sub-type of DORV repair

How common is DORV with Subaortic VSD and what associated issues are there?

Surgical Results in Patients With Double Outlet Right Ventricle: A 20-Year Experience

John W. Brown, MD, Mark Ruzmetov, MD, Yuji Okada, MD, Palaniswamy Vijay, PhD, MPH, and Mark W. Turrentine, MD

Section of Cardiothoracic Surgery, James W. Riley Hospital for Children, and Indiana University School of Medicine, Indianapolis, Indiana

ANNALS OF THORACIC SURGERY

124 patients over 20 yrs 1980-2000

Of the 57 patients with a subaortic VSD, 28 patients had tetralogy of Fallot anatomy.

Pulmonary outflow tract obstruction including pulmonary atresia was present in 65 patients and was most prevalent in the group with subaortic VSDs.

Ann Thorac Surg 2001;72:1630 –5

How common is DORV with Subaortic VSD and what associated issues are there?

Surgical outcomes of 380 patients with double outlet right ventricle who underwent biventricular repair

Shoujun Li, MD, Kai Ma, MD, PhD, Shengshou Hu, MD, Zhongdong Hua, MD, Keming Yang, MD, Jun Yan, MD, and Qiuming Chen, MD, PhD

380 patients with DORV repair over 8 years 2005-2012

58% had DORV with subaortic type VSD

Variables	Subaortic VSD (n = 219)
Patient characteristics	
Mean age at BVR (y)	1.2 ± 1.1
Mean weight at BVR (kg)	9.6 ± 5.0
Great arteries	
Normal relation	130 (59.3%)
Side by side	12 (5.5%)
Anterior aorta	77 (35.2%)
Subaortic conus	117 (53.4%)
Pulmonary stenosis	96 (43.9%)
Coronary anomalies	25 (11.5%)
Arch obstruction	3 (1.4%)
Pulmonary arterial hypertension	43 (19.6%)

J Thorac Cardiovasc Surg 2014;148:817-24

Characteristics of patients with postdischarge left ventricular outflow tract obstruction

	Age at repair (mo)	Systolic pressure gradient (mm Hg)	Duration from biventricular to LVOTO (mo)	VSD position
Early LVOTO				
1	20	43	Immediately after BVR	Noncommitted
2	6	66	Immediately after BVR	Noncommitted
3	9	50	Immediately after BVR	Noncommitted
4	10	75	Immediately after BVR	Subaortic
5	12	55	Immediately after BVR	Subaortic
6	5	30	Immediately after BVR	Noncommitted
Late-onset LVOTO				
7	11	106	44	Doubly committed
8	11	98	35	Noncommitted
9	7	39	48	Noncommitted
10	7	33	35	Subaortic
11	5	43	50	Noncommitted
12	6	113	69	Noncommitted
13	12	35	87	Noncommitted
14	24	136	97	Subaortic
15	4	67	36	Noncommitted

How common is DORV with Subaortic VSD and what associated issues are there?

Current outcomes of live-born children with double outlet right ventricle in Norway

Mads Holten-Andersen [a,b,*], Matthias Lippert[b,c], Henrik Holmstrøm[b,d], Henrik Brun [c,d] and Gaute Døhlen[d]

87 children had surgery over 14 yrs from 2003 – 2017

49% had DORV with subaortic type VSD

DORV type	VSD type	Fallot type
Total, *n* (%)	18 (21)	24 (28)
Sex		
Females	4 (4.6)	10 (11)
Males	14 (16)	14 (16)
Birth weight (kg)[a]	3.0 (2.8–3.7)	2.9 (2.3–3.2)
Syndrome/malformation	7 (8.1)	8 (9.2)
AVSD	0 (0)	6 (6.9)
Doubly committed VSD	0 (0)	1 (1.2)
Age at 1st surgery (months)	2.7 (1–5.4)	17 (1.8–33)
Age at repair (months)	2.7 (0.9–6.3)	28 (13–46)
Patients surgical route		
Primary BiV repair	17 (20)	13 (15)
Staged BiV repair	0 (0)	8 (9.2)
UniV repair	1 (1.2)	3 (3.4)

European Journal of Cardio-Thoracic Surgery 2022, 63(1)

How common is DORV with Subaortic VSD and what associated issues are there?

DORV type	VSD type	Fallot type
Cross-clamp time (min)	60 (36–115)	59 (44–80)
Ventilation time (days)	1.5 (1–5)	1.5 (1–4)
ICU stay (days)	5.5 (2–13)	7 (4–9)
Hospital stay (days)	18 (7–26)	20 (11–26)
Deaths, *n* (%)	3 (3.5)	2 (2.3)
Early (<30 days)	2 (2.3)	0 (0)
Late (≥30 days)	1 (1.2)	0 (0)
Very late (>1 year)	0 (0)	2 (2.3)
Catheter procedures	8 (7.9)	32 (32)
Diagnostic	7 (6.9)	15 (15)
Interventional	1 (1.0)	17 (17)

70% had a cath intervention pre surgery

How common is DORV with Subaortic VSD and what associated issues are there?

Summary:

Approx 1/3 of DORV patients are of the subaortic type and about ½ of these will be “Tetralogy-like”

Surgery Features:

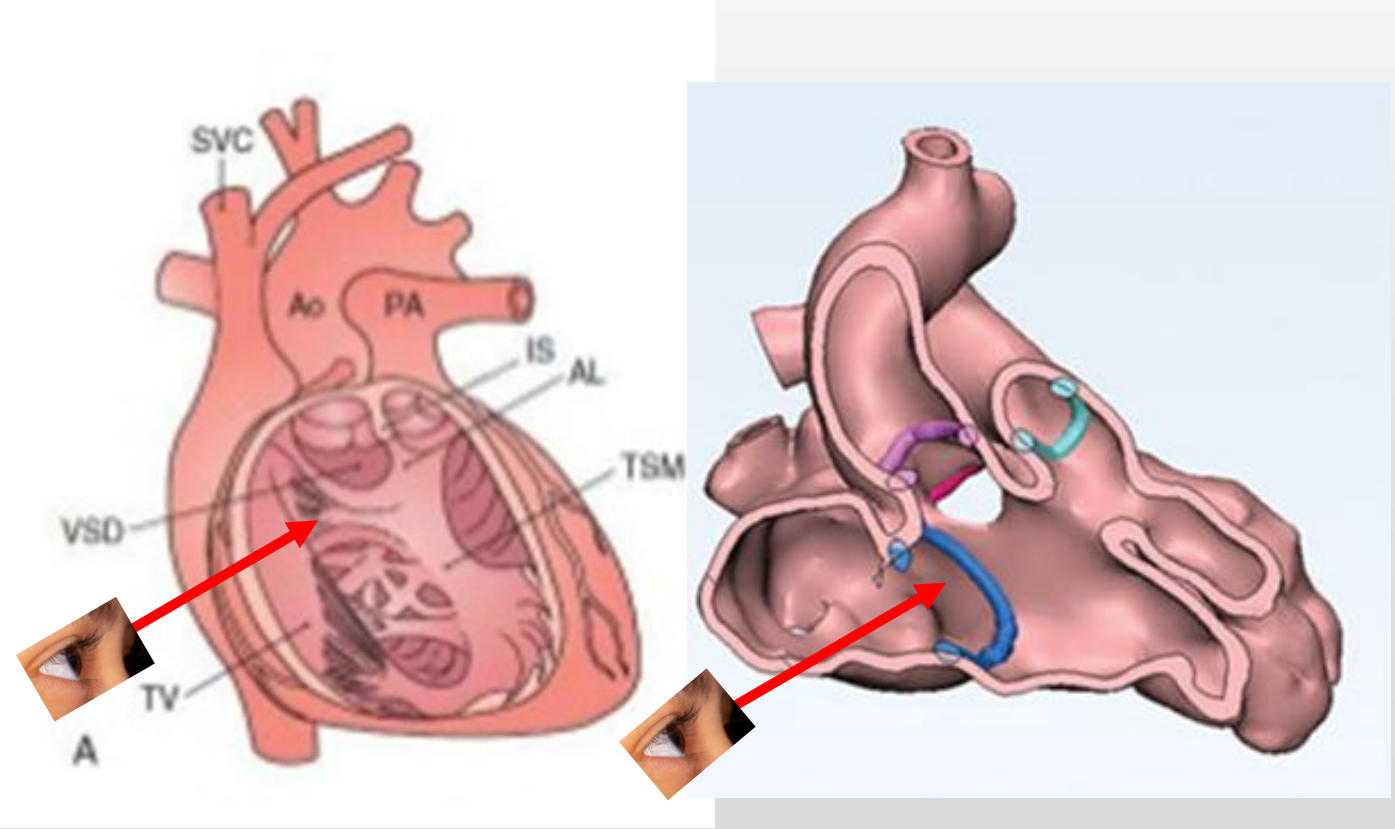
About 30 -70% of the “Tetralogy-like” DORV patients will need an intervention to augment pulmonary blood flow before their definitive surgery.

Up to 15% of DORV patients with subaortic VSD will need a VSD enlargement at repair

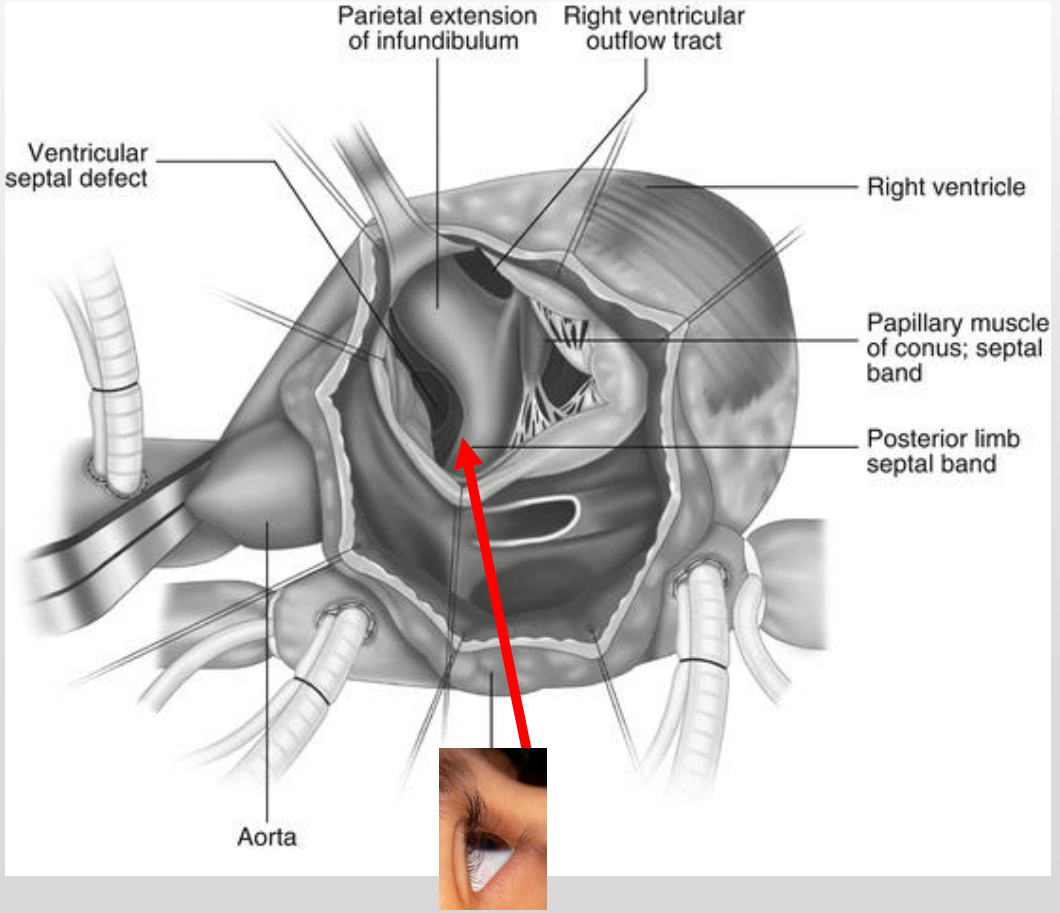
Recurrent LVOT obstruction is uncommon, but can occur post-op.

What are the issues with the VSD closure?

DORV with Subaortic VSD

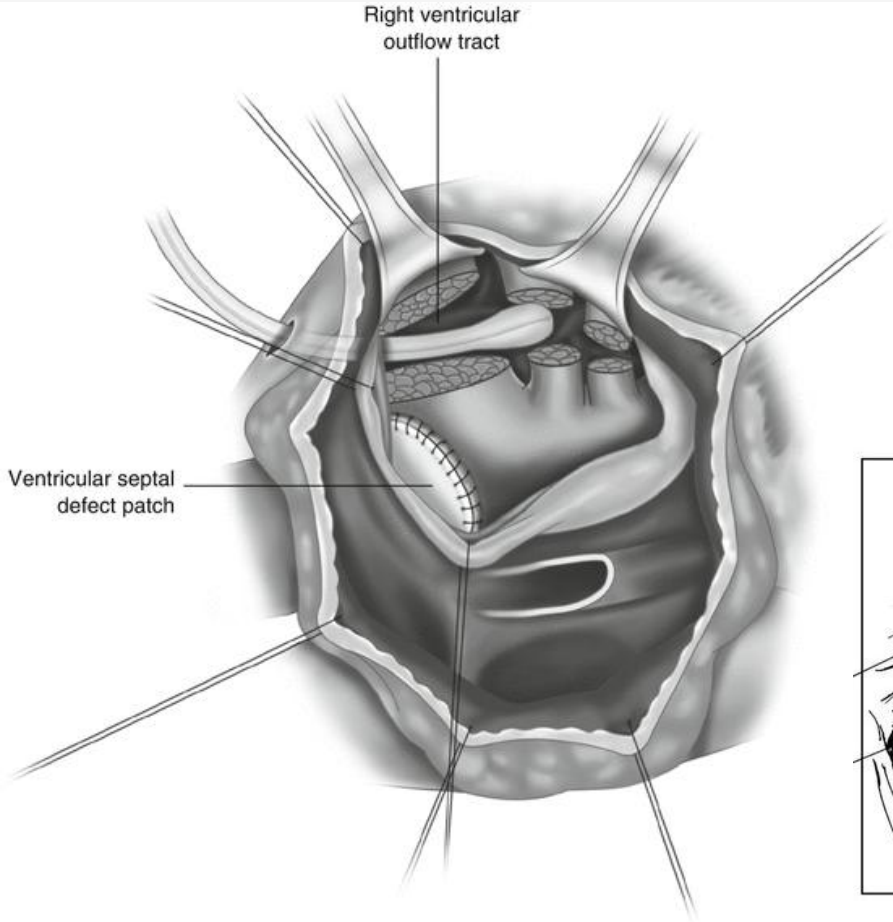


Surgical Repair:
"Tetralogy type trans-atrial repair"



What are the issues with the VSD closure?

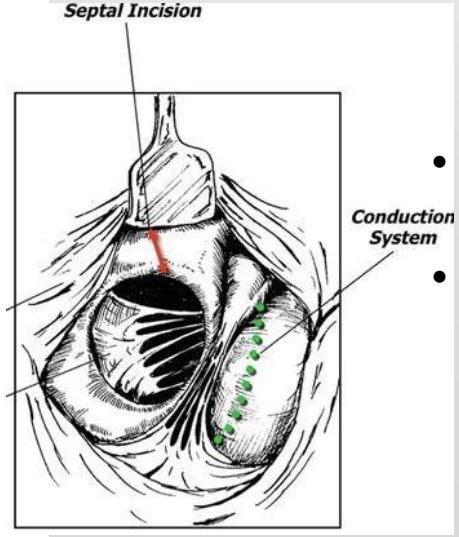
What the surgeon needs to know:



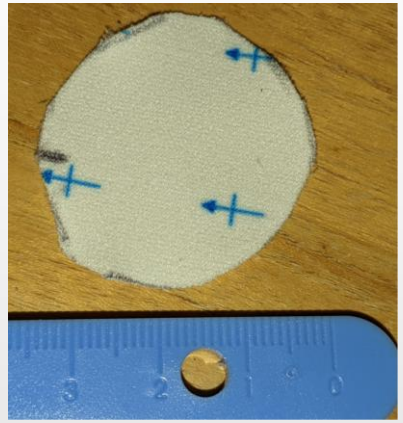
Pre-op:

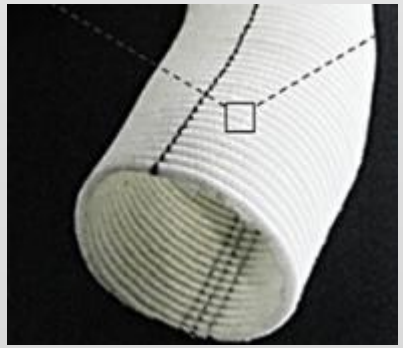
- Degree of aortic override?
- How big is the conal septum?
- How big is the VSD?
- Is there inlet extension?

Intra-op:



- How much do I enlarge the VSD?
- Do I use a patch or curved baffle?





Is it important how we approach RVOT / Pulmonary Valve?

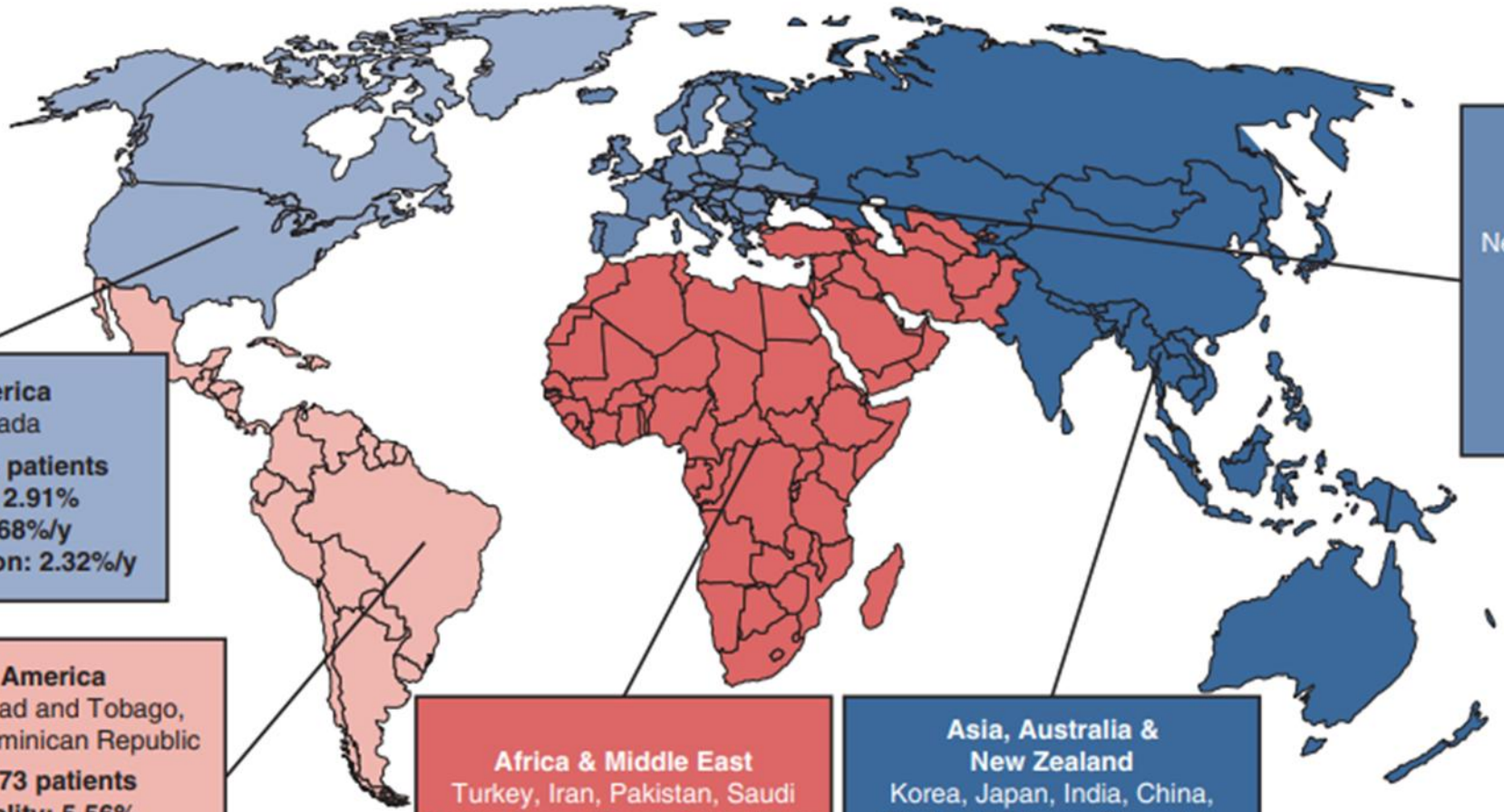
Outcome after surgical repair of tetralogy of Fallot: A systematic review and meta-analysis

The Journal of Thoracic and Cardiovascular Surgery JTCVS

Litterature from 2000-2018
137 cohort studies of 21,427 patients

J Thorac Cardiovasc Surg 2020;159:220-36

Complete Correction of Tetralogy of Fallot:
a systematic review and meta-analysis of surgical outcome



North America
USA, Canada
25 cohorts / 2608 patients
30-day mortality: 2.91%
Late mortality: 0.68%/y
Late reintervention: 2.32%/y

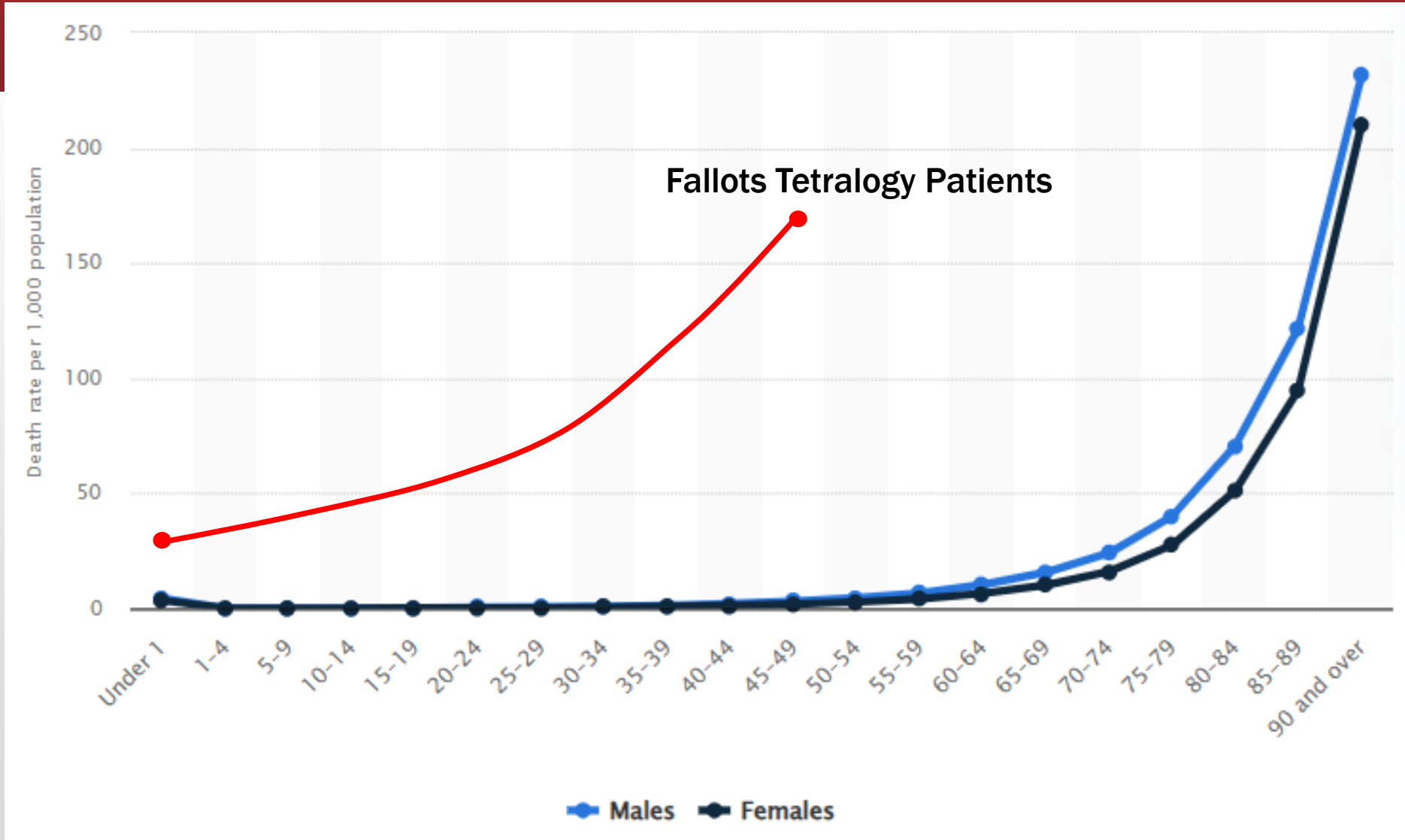
Latin America
Brazil, Trinidad and Tobago, Colombia, Dominican Republic
5 cohorts / 273 patients
30-day mortality: 5.56%
Late mortality: 2.31%/y
Late reintervention: 0.80%/y

Africa & Middle East
Turkey, Iran, Pakistan, Saudi Arabia, Israel, Egypt, Cote d'Ivoire
17 cohorts / 1746 patients
30-day mortality: 4.20%
Late mortality: 0.82%/y
Late reintervention: 3.37%/y

Asia, Australia & New Zealand
Korea, Japan, India, China, Thailand, Taiwan, Shanghai, South-Korea, Australia
43 cohorts / 7610 patients
30-day mortality: 2.36%
Late mortality: 0.32%/y
Late reintervention: 2.38%/y

Europe
Belgium, United-Kingdom, Germany, Switzerland, The Netherlands, Greece, Italy, Austria, Spain, Norway, Finland
35 cohorts / 5878 patients
30-day mortality: 3.06%
Late mortality: 0.33%/y
Late reintervention: 1.79%/y

How does our Infant Surgical Repair Technique influence life?



Death Rate per 1000 population in the UK in 2021

How does our Infant Surgical Repair Technique influence life?

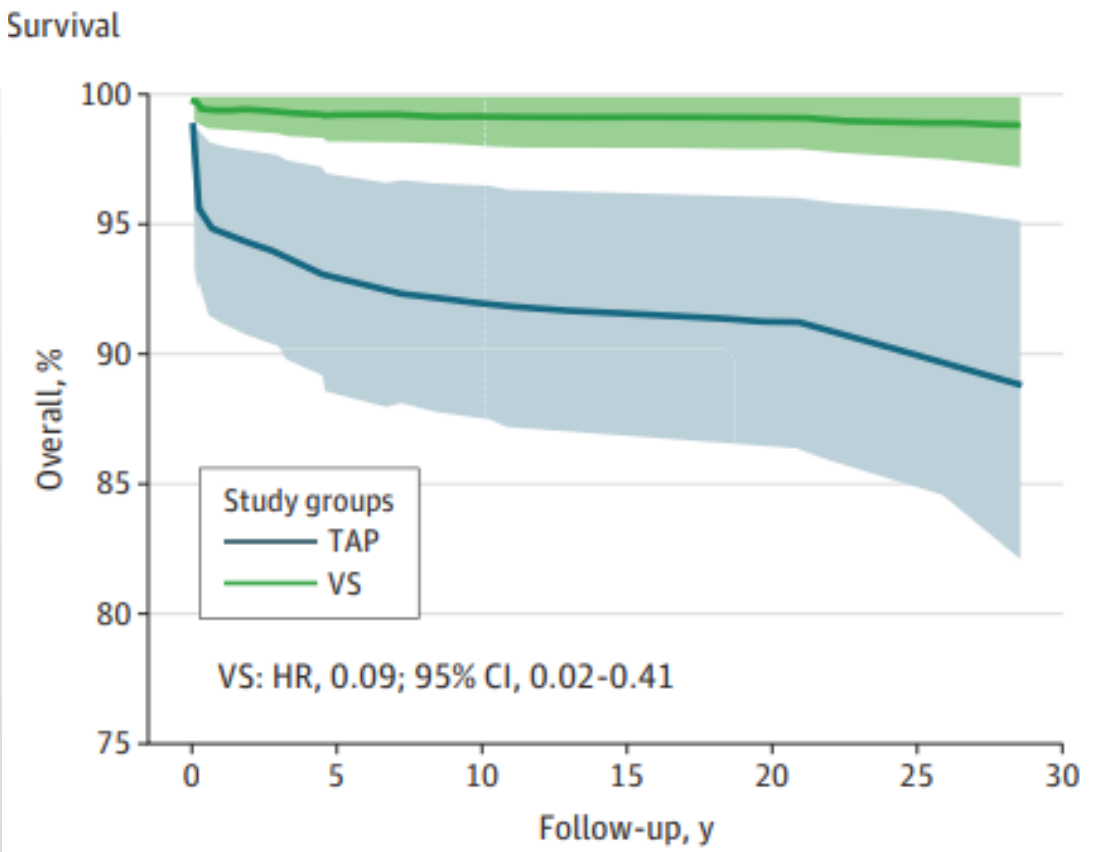
JAMA Network | Open

Original Investigation | Cardiology

Comparison of Long-term Outcomes of Valve-Sparing and Transannular Patch Procedures for Correction of Tetralogy of Fallot

- Everyone born with TOF in Quebec, Canada from 1980 to 2015 assessed in 2020 (up to 40 yrs follow-up)
- Transannular patch (TAP) vs Valve sparing (VS) procedure
- Matched based on propensity score of pre-op factors 1:1

	Transannular Patch (TAP)	Valve Sparing (VS)
30 year Survival	90.4%	99.1%
Mean number of re-interventions	2.0	0.7
Mean number of Valve replacements	1.4	0.3



JAMA 2021 4(7):1-15

Whats wrong with the transannular patch?

- <u>Free pulmonary incompetence</u>

  RV dilation and diastolic dysfunction

  RV systolic dysfunction

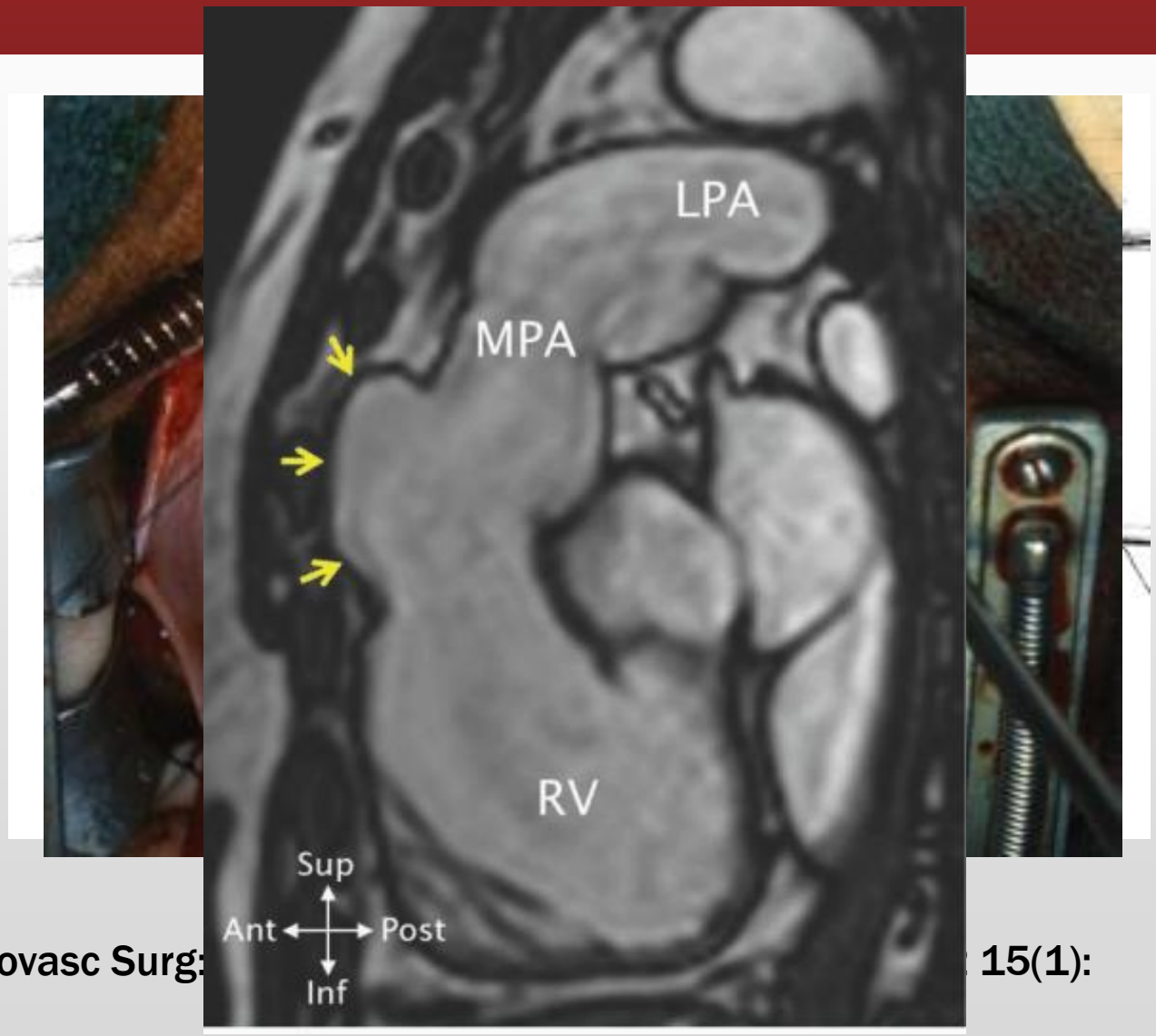
  Risk of ventricular arrhythmias

  Secondary tricuspid regurgitation

- <u>Redundant often aneurysmal RVOT</u>

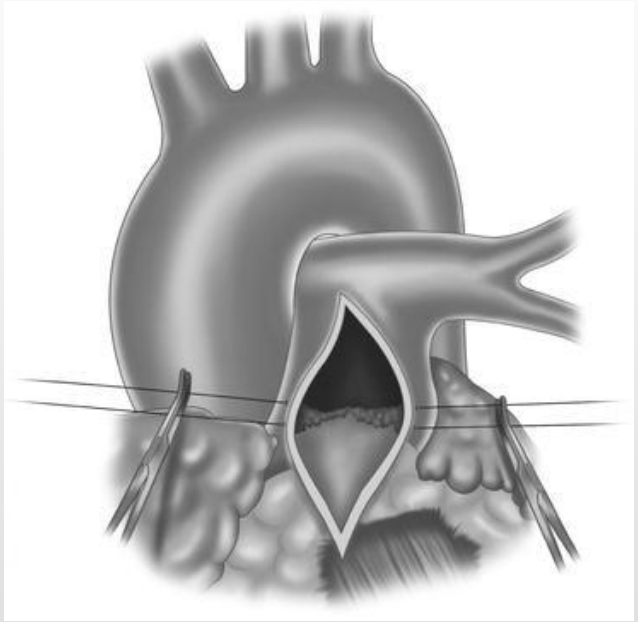
  Reduced functional ejection fraction

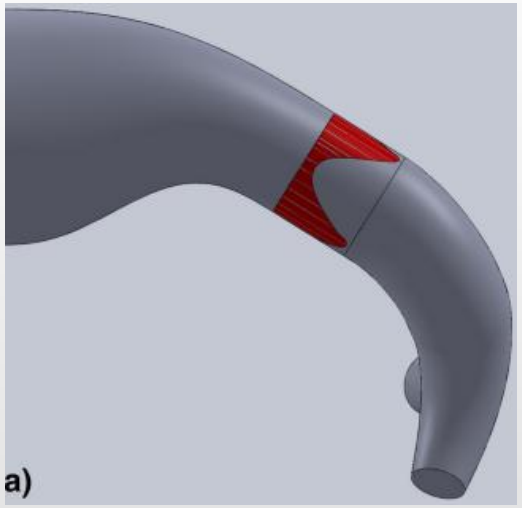
  of the RV

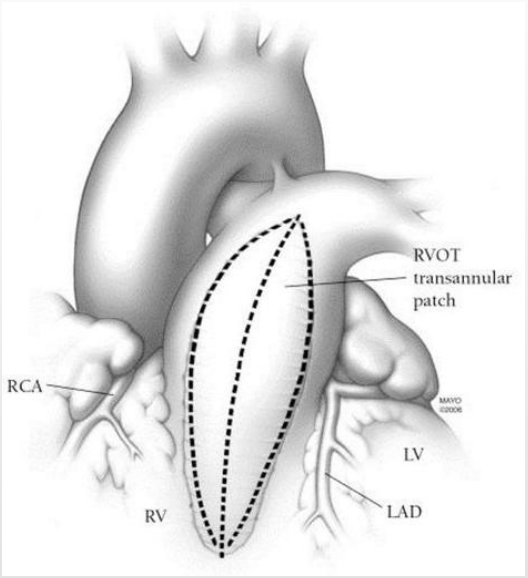


Seminars in Thorac and Cardiovasc Surg: 15(1): 24-26

Ok, but we can just put a valve in when the child is fully grown?

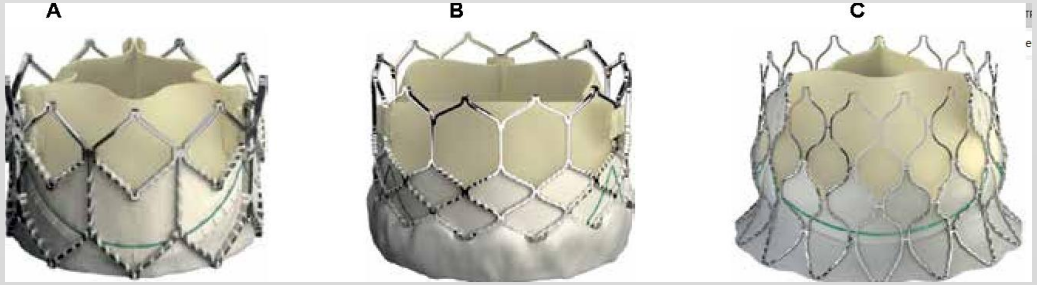


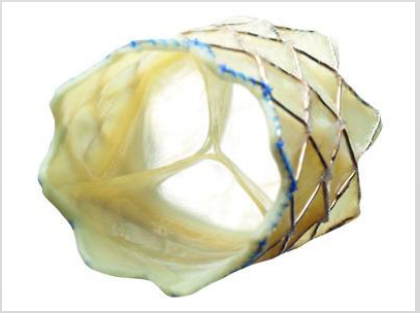




Or

Transcatheter Valve





What is the outcome of placing an adult surgical valve in a teenager?

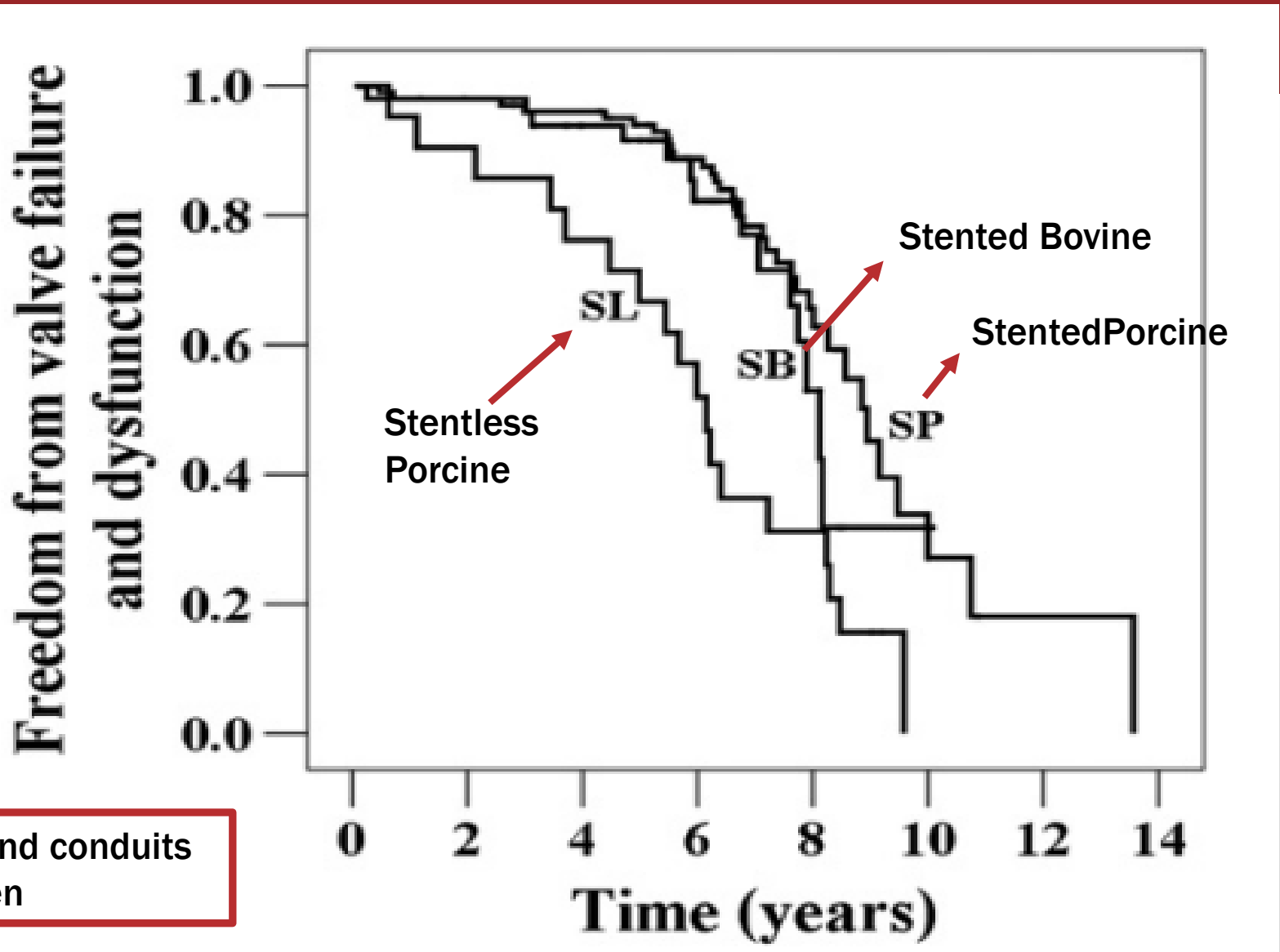
Durability of bioprosthetic valves in the pulmo
Long-term follow-up of 181 implants in patient
heart disease

The Journal of Thorac
AATS JTC

181 patients implanted
Bioprosthetic PVR
1993-2004 followed to 2011

Mean age 14 years

Median Valve size 23mm



Even surgical bioprosthetic valves and conduits are problematic in children

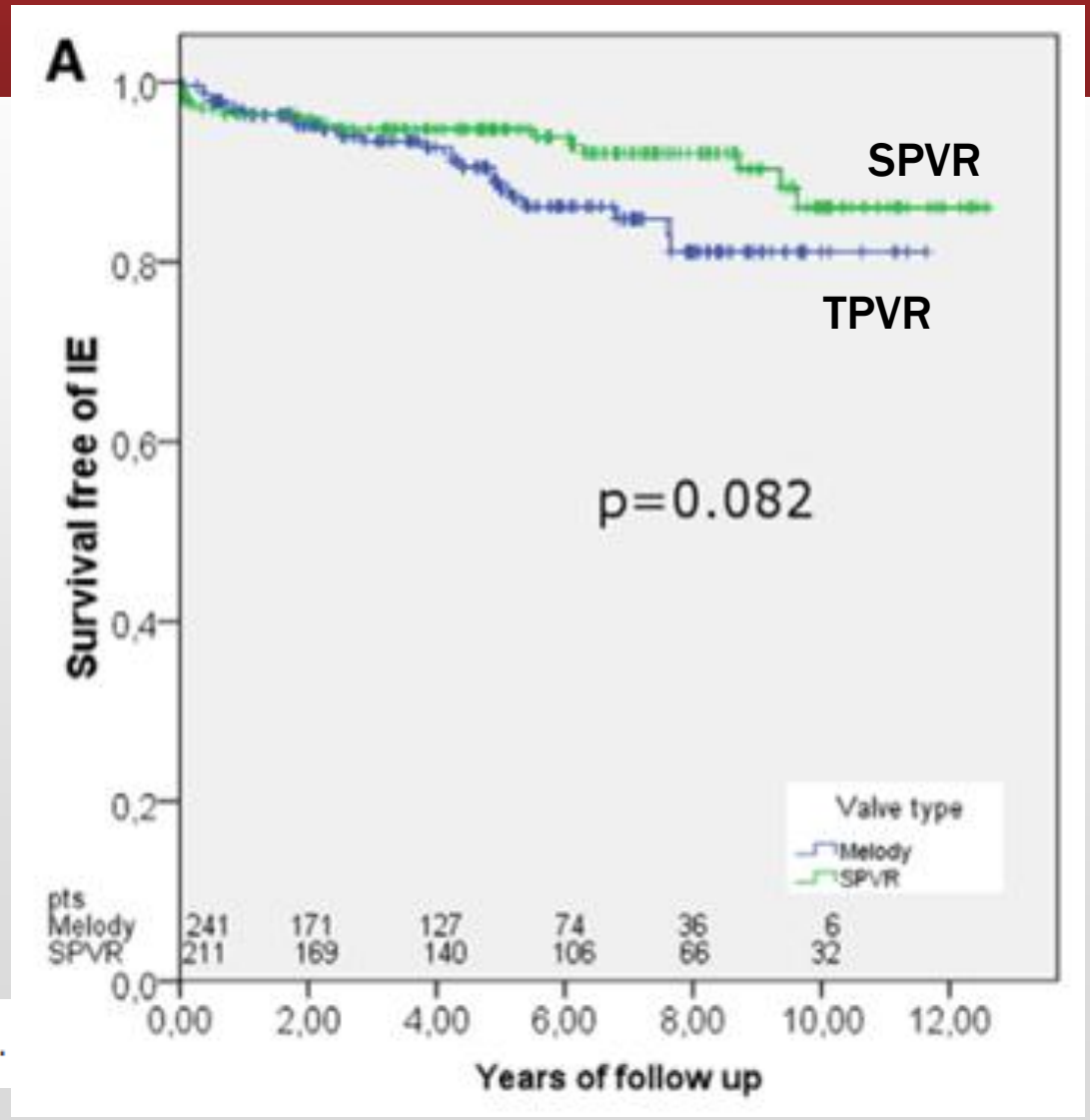
What is the outcome of placing an adult Transcatheter Valve in a teenager?

Munich Comparative Study

Prospective Long-Term Outcome of the Transcatheter Melody Valve Versus Surgical Pulmonary Bioprosthesis With Up to 12 Years of Follow-Up

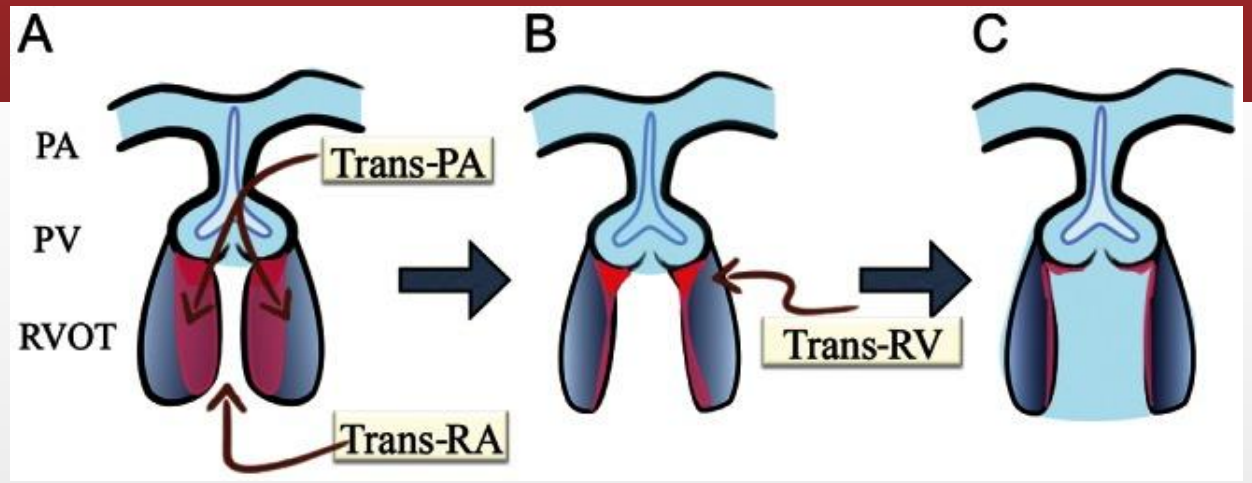
Single centre study, 2006-2018
241 transcatheter PVR patients
211 Surgical PVR patients

Survival without need for redo PVR at 10 years:
87% Transcatheter PVR
88% Surgical PVR

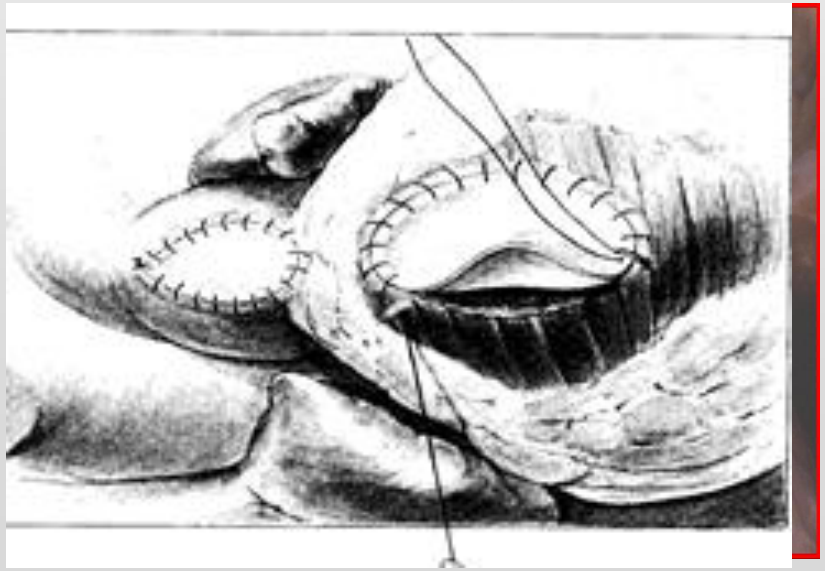


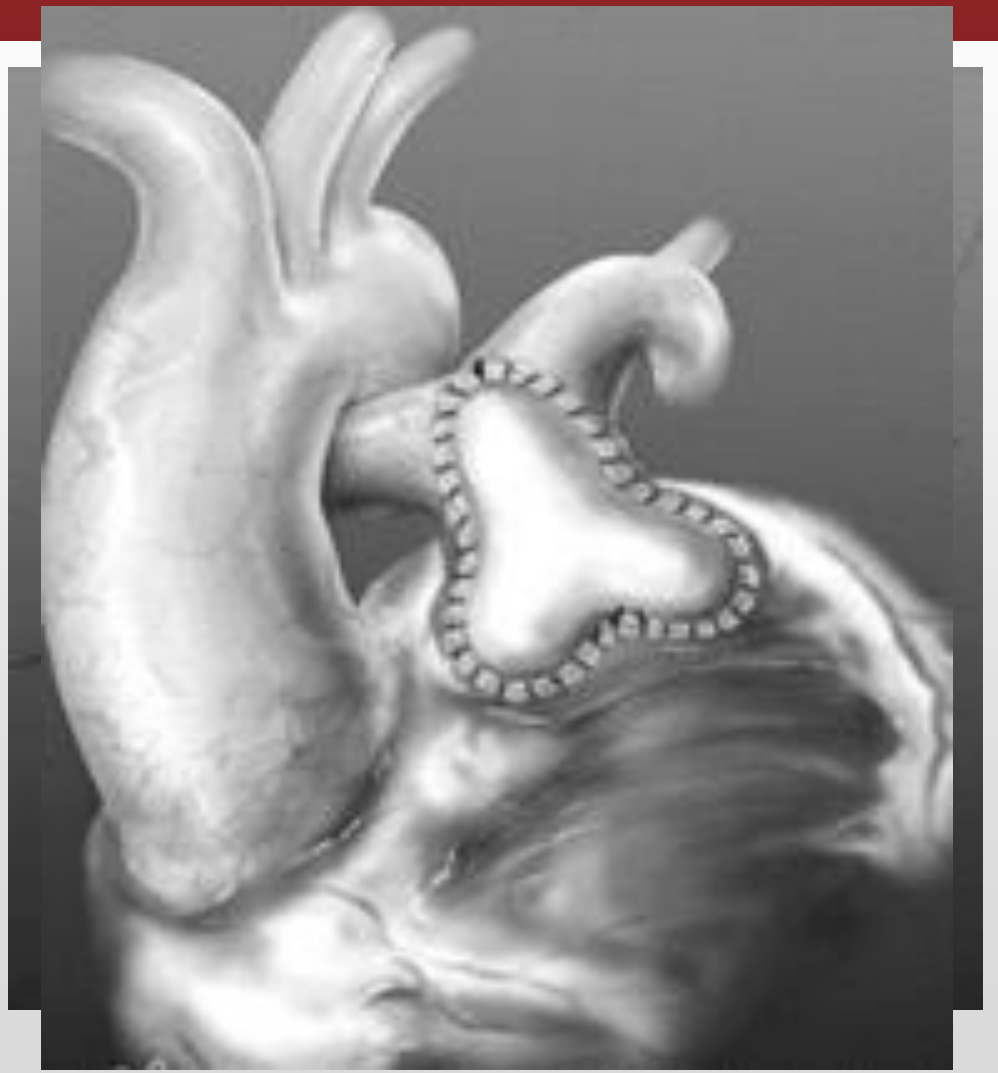
Circ Cardiovasc Interv. 2020;13:e008963.

Push the limits of Valve Repair



Technical modification enabling pulmonary valve-sparing repair of a severely hypoplastic pulmonary annulus in patients with tetralogy of Fallot. Interactive CardioVascular and Thoracic Surgery 16 (2013) 802–807

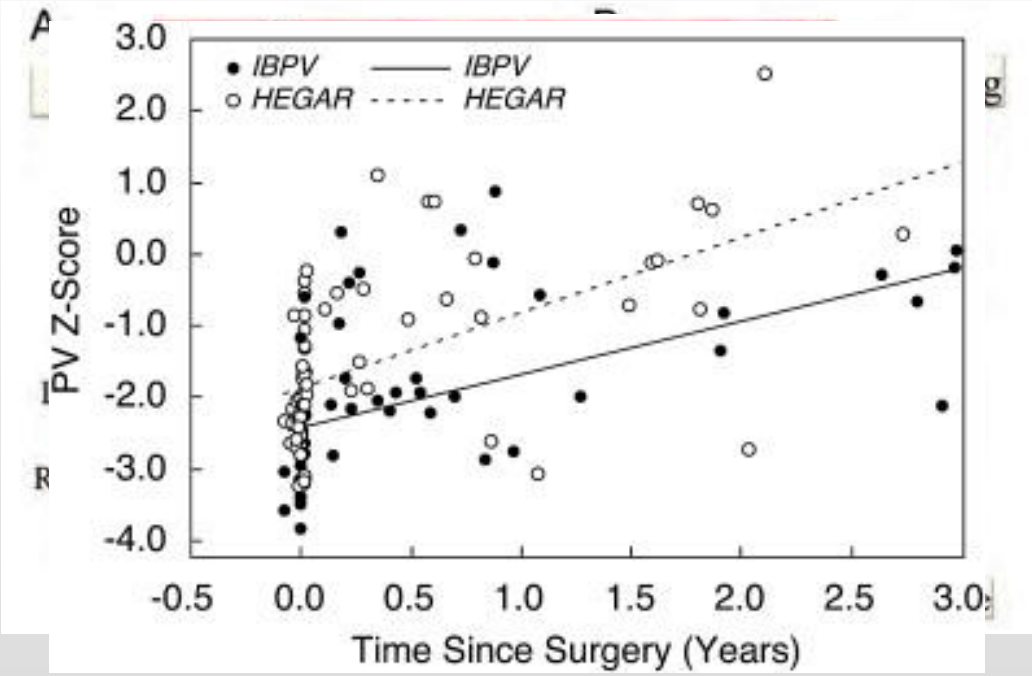




Techniques to deal with hypoplastic annulus

Intraoperative Balloon Pulmonary Valvuloplasty

- Radial annular dilation
- Commissurotomy into pulmonary artery wall allows for more effective annuloplasty
- Nominal Balloon size to annulus ratio 1.47 after dilation (8 atm)



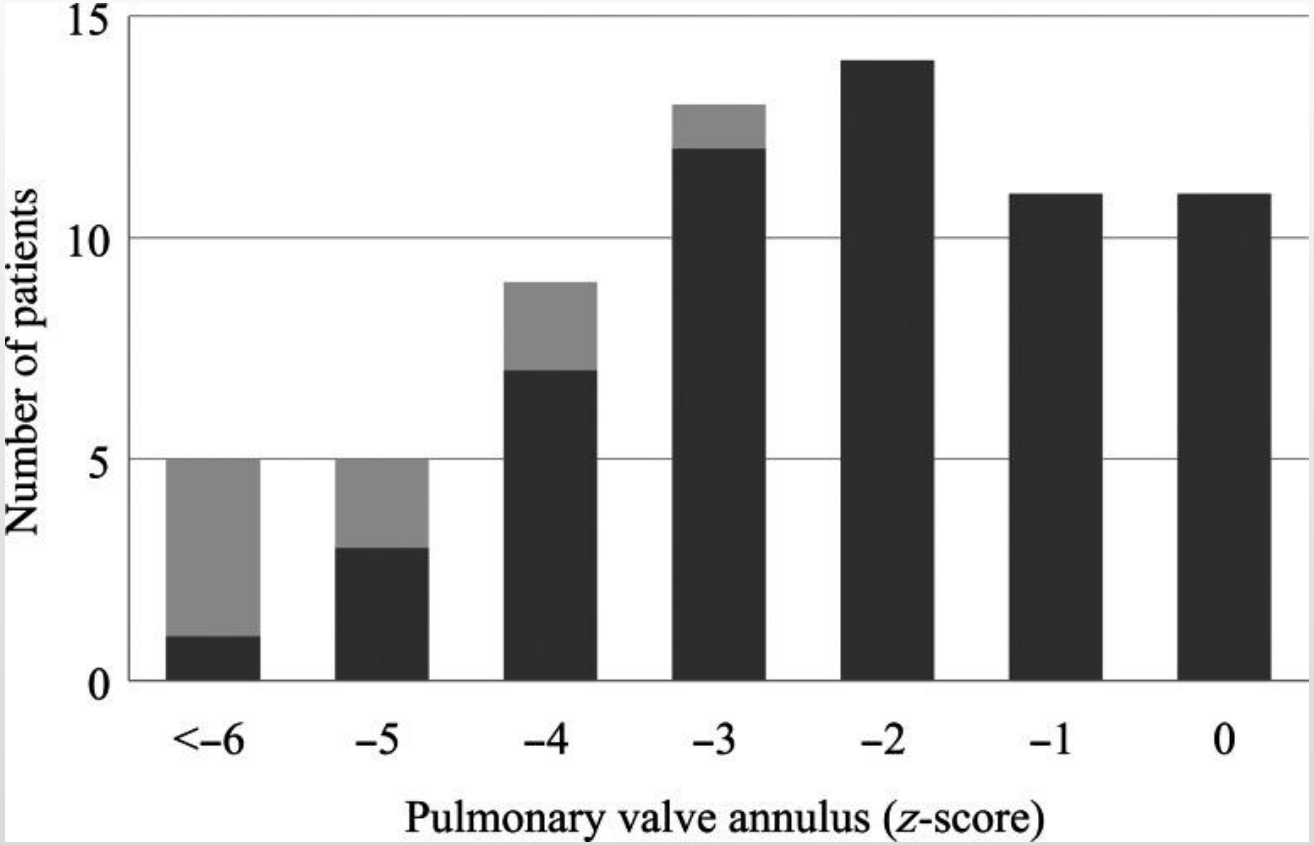
- Longitudinal growth in pulmonary annulus

The evolving role of intraoperative balloon pulmonary valvuloplasty in valve-sparing repair of tetralogy of Fallot. Journal Thorac and Cardiovasc Surg. 2011 142(6):1367-1373

Technique to deal with annular hypoplasia and leaflet restriction. Influence of pulmonary annulus z score

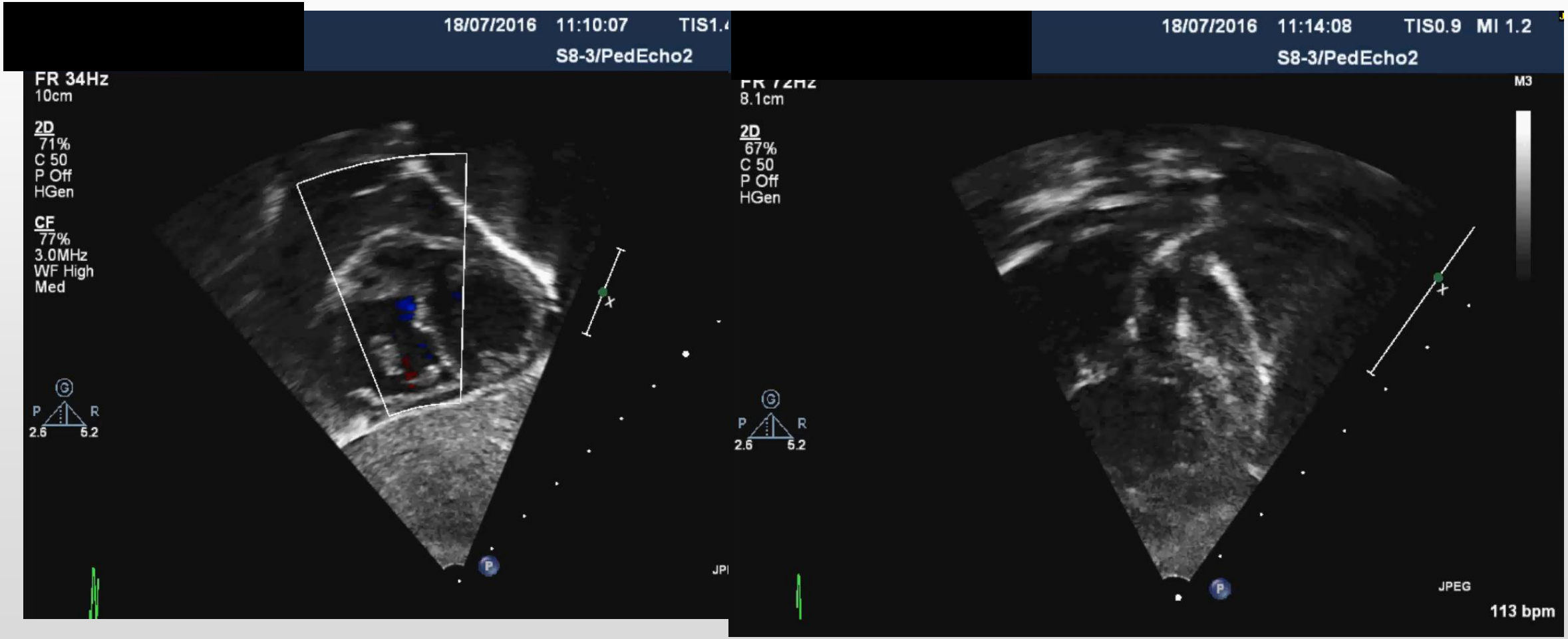
Transannular patch repair in light grey.

Pulmonary valve sparing procedure achieved in 98% with annulus Z scores >-4.

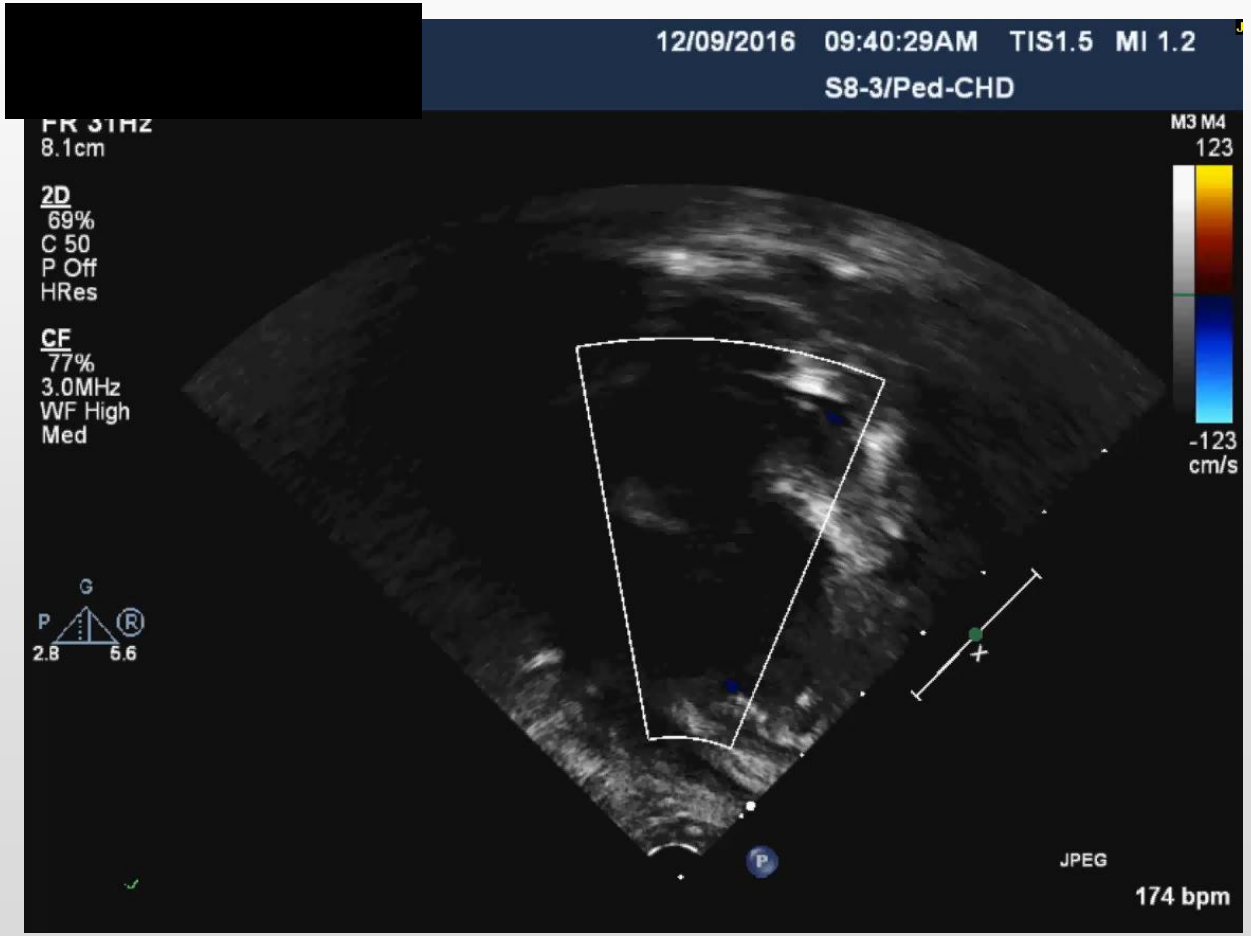


Technical modification enabling pulmonary valve-sparing repair of a severely hypoplastic pulmonary annulus in patients with tetralogy of Fallot. Interactive CardioVascular and Thoracic Surgery 16 (2013) 802–807

Pre-op Multilevel Obstruction and small Pulmonary Annulus



1. Supravalvar Y patch,
2. commissurotomy, annulus balloon dilation,
3. Subvalvar extensive muscle division and RVOT small patch



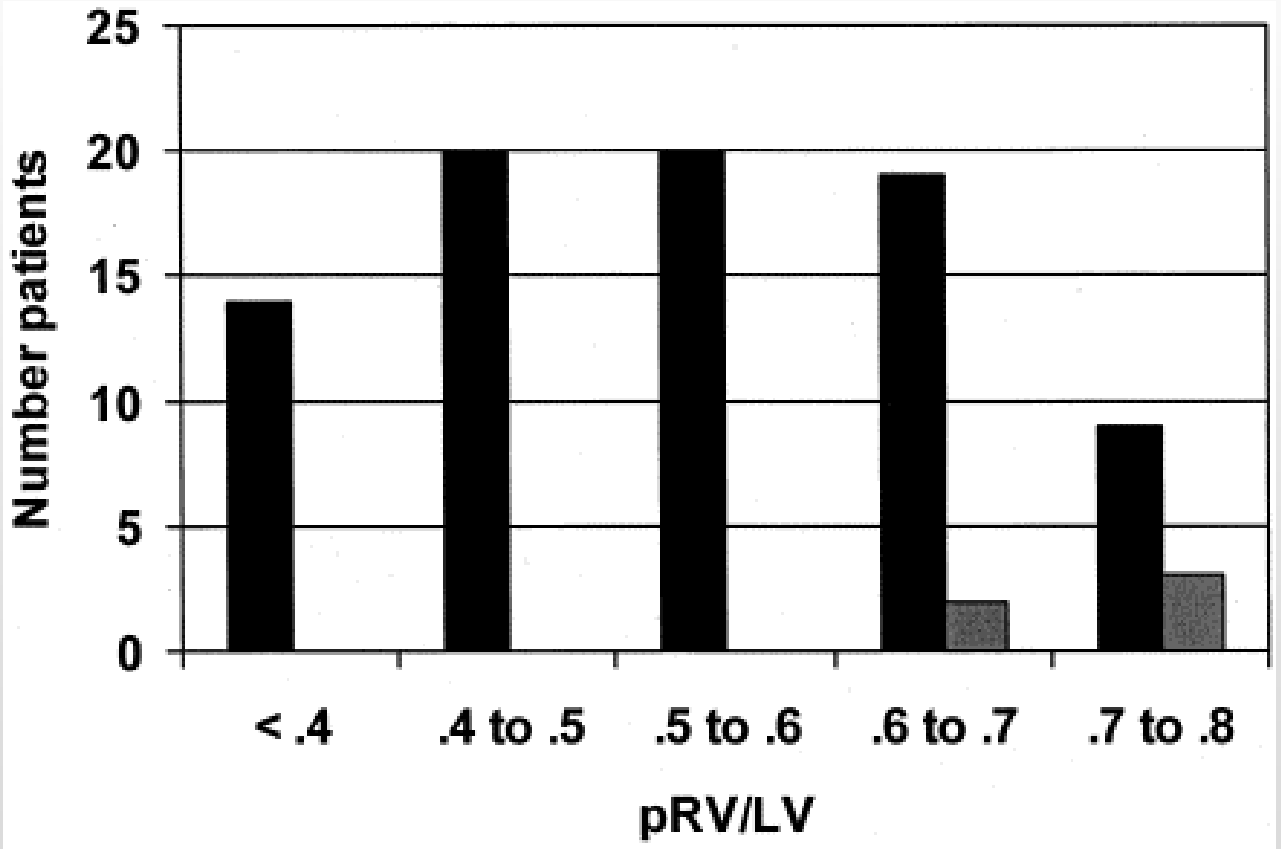
So What Factors can the Surgeon use Intra-op to decide to continue with valve sparing repair?

Factors Predicting Successful Pulmonary Valve Preservation
RV to LV pressure ratio after bypass

82 patients with pulmonary valve sparing procedure.

RV to LV pressure ratio off bypass.

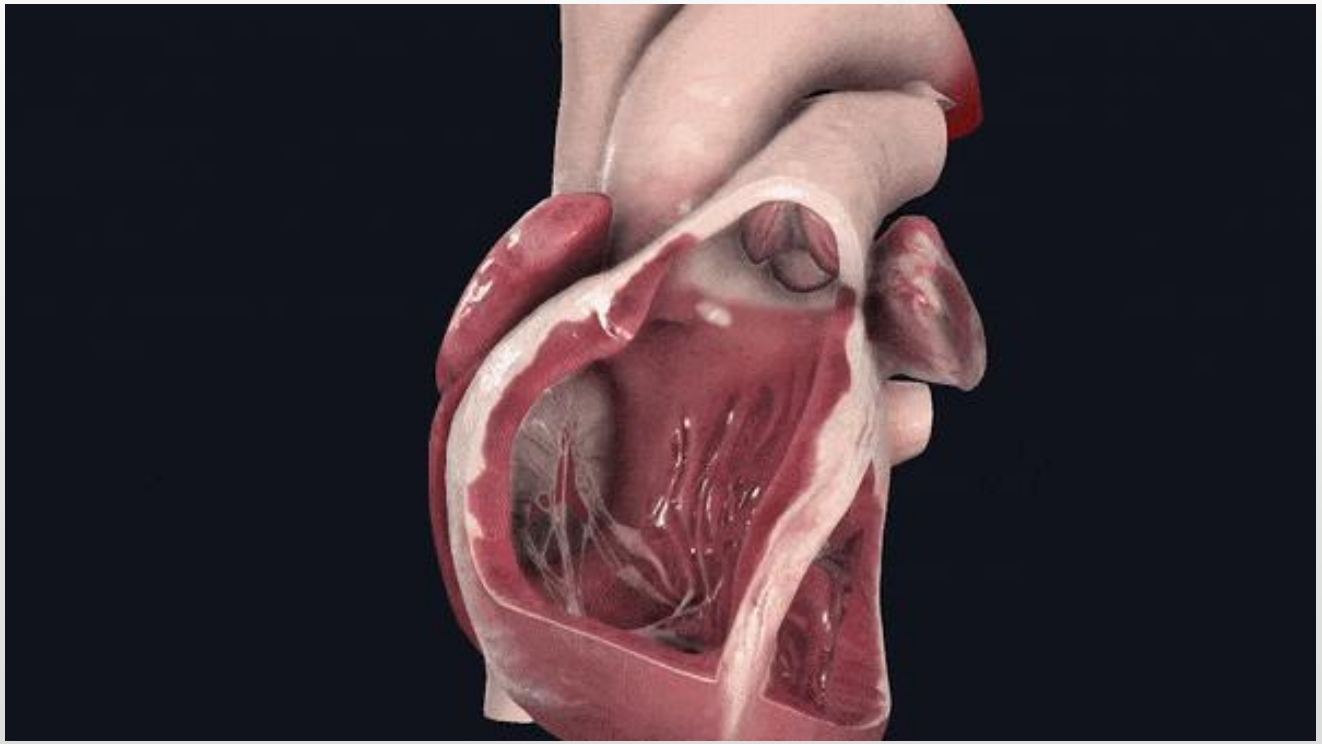
Need for redo procedure later shown in gray bars.



Tetralogy of Fallot: Results of a Pulmonary Valve-Sparing Strategy, Ann Thorac Surg 2005 80(4):1431-9

If you have to do a Transannular patch, what should be your approach?

Simple Transannular Patch



Valve Replacement With "Growing Valve"

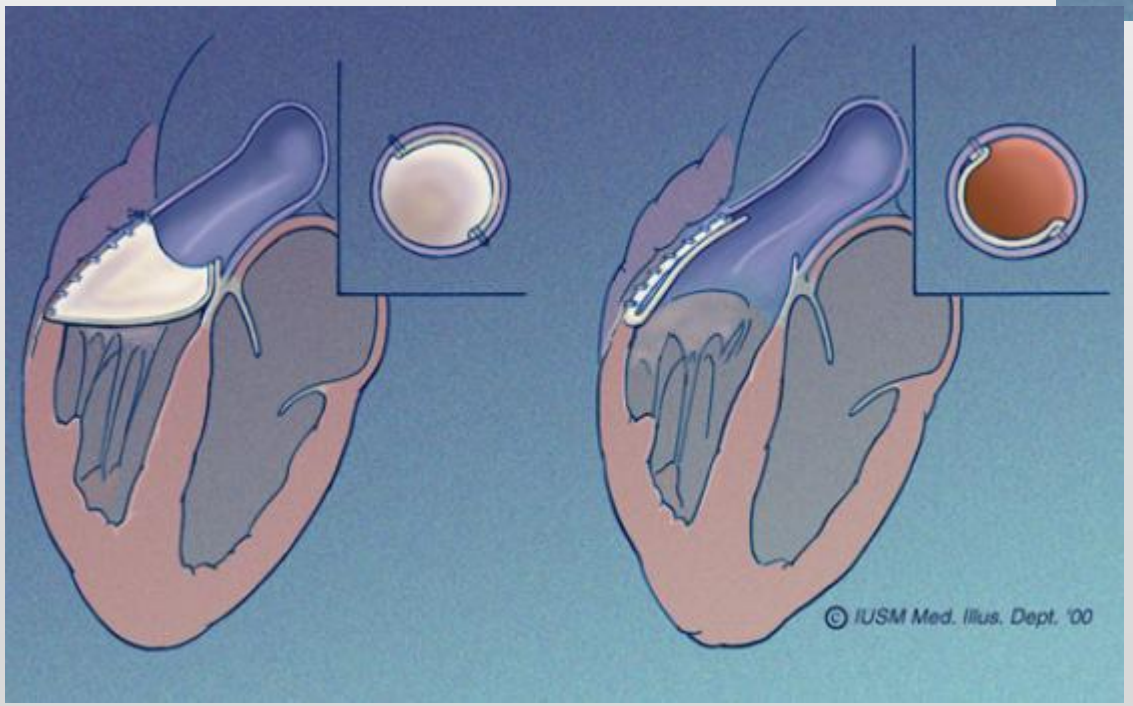
Monocusp Valve

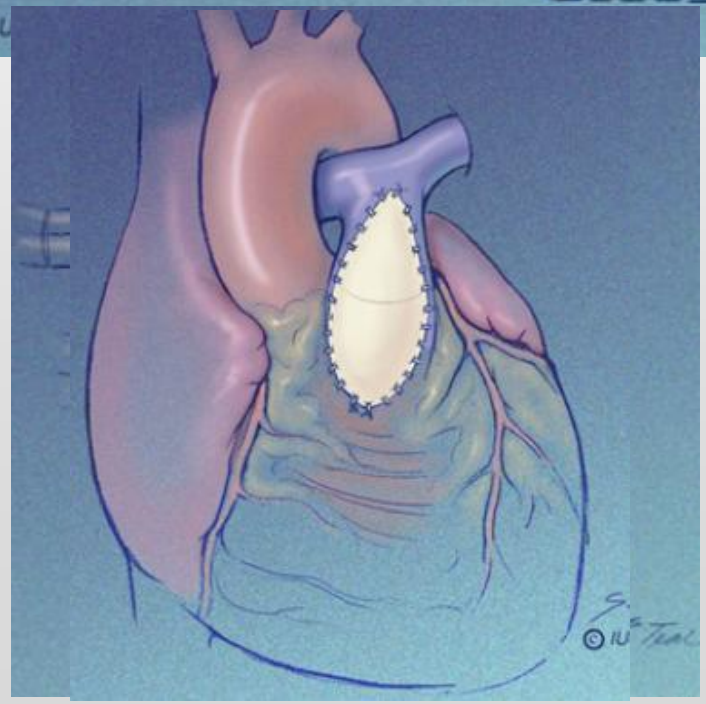
Monocusp Valves

Construction with:

- Autologous pericardium
- Bovine pericardium
- Pulmonary homograft
- PTFE(Goretex)

Ann Thorac Surg 2002 74:2202-5





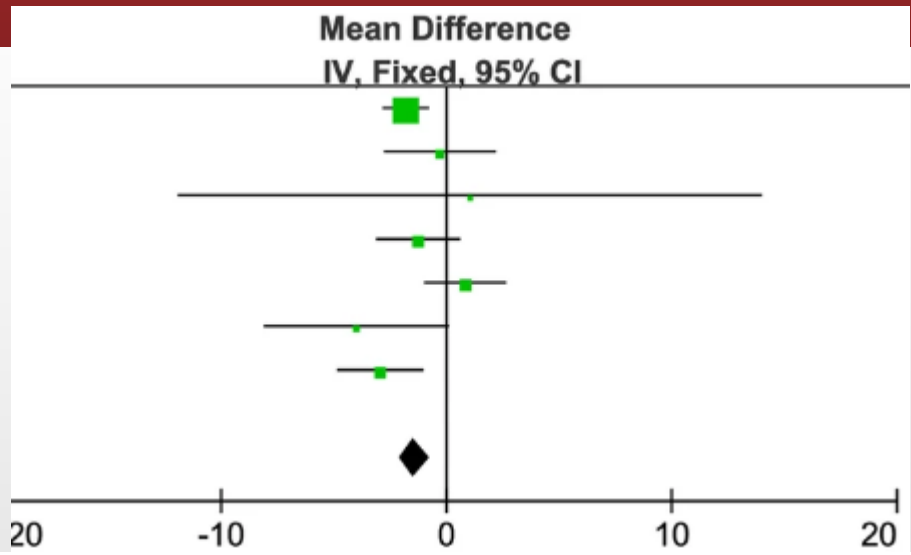
What are the Perioperative outcomes with a Monocusp Valve?

- Meta-analysis of all studies on Monocusp valves published 2000-2022
- 10 studies included
- 349 TAP with monocusp

  vs

  312 TAP without monocusp
- No difference in Perioperative Mortality
- Mean crossclamp time 14 mins longer
- Mean ICU length of stay 1.4 days shorter in Monocusp group

BMC Surgery 2022 22:18



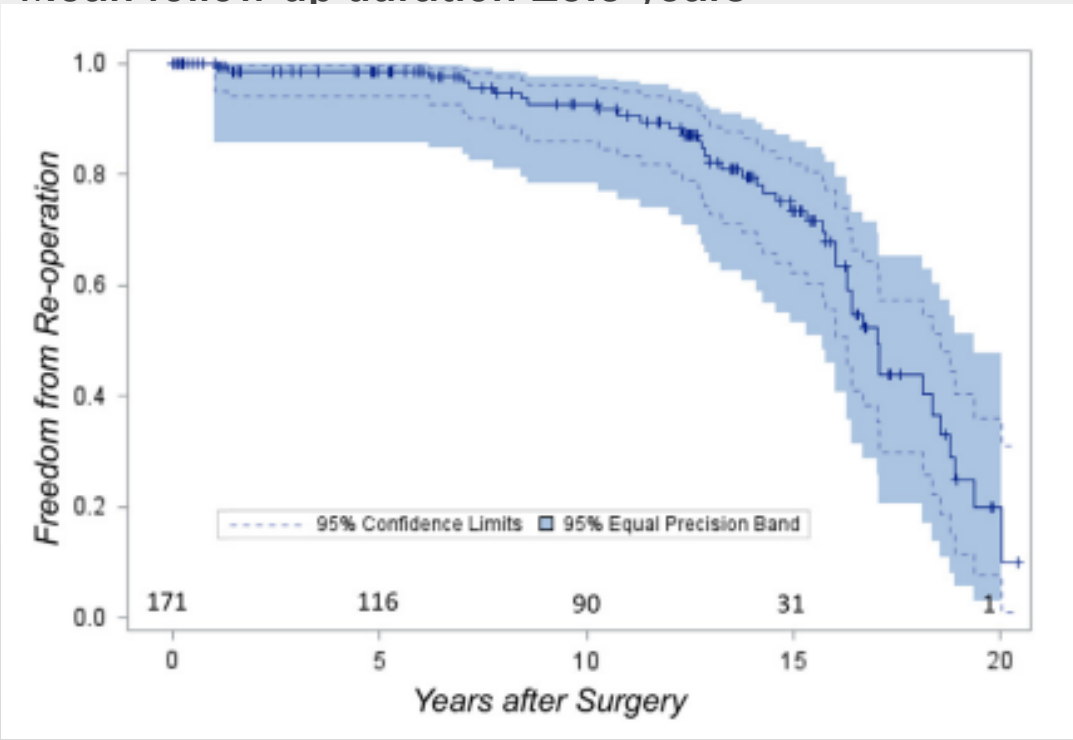
	TAP with monocusp			TAP without monocusp				Mean Difference
Study or Subgroup	Mean	SD	Total	Mean	SD	Total	Weight	IV, Fixed, 95% CI
Attanawanich, S. 2013	5.22	1.65	55	7.03	2.78	38	47.3%	-1.81 [-2.80, -0.82]
Ismail, S. R. 2010	4.5	4.5	16	4.8	3.6	48	7.8%	-0.30 [-2.73, 2.13]
Pande, S. 2010	4.04	25.62	16	3	7.5	24	0.3%	1.04 [-11.87, 13.95]
Rawat, S. 2021	4.67	1.95	15	5.93	3.01	15	14.0%	-1.26 [-3.07, 0.55]
Samadi, M. 2020	5.8	4.326	30	4.97	2.428	30	14.6%	0.83 [-0.95, 2.61]
Sasson, L. 2013	4	7.17	74	8	8.5	20	2.8%	-4.00 [-8.07, 0.07]
Sayyed, E.H.N. 2016	6.85	3.61	60	9.8	4.54	30	13.2%	-2.95 [-4.81, -1.09]
Total (95% CI)			**266**			**205**	**100.0%**	**-1.43 [-2.11, -0.76]**

Mean ICU Length of Stay

What are the Longterm outcomes with a monocusp valve?

Goretex monocusp – Indiana, USA

- 171 patients over 20 years RVOT reconstruction with a monocusp valve.
- Mean follow-up duration 10.9 years



171 patients

6 Deaths

25 no follow-up

98 still with monocusp

42(30%) have had a PVR mean 10 years after monocusp.

Echo follow-up, mean 9.5 years

Only 19% freedom from composite endpoint of reoperation or mod-severe PI at 10 years

Semin Thorac Surg 2016 28:463-470

Valve replacement with a “Growing Valve”

Eur J Cardiothorac Surg. 2021 Apr 13;59(3):697-704. doi: 10.1093/ejcts/ezaa374.

Surgical repair of tetralogy of Fallot using autologous right atrial appendages: short- to mid-term results

Ahmadali Amirghofran [1], Fatemeh Edraki [2], Mohammadreza Edraki [3], Gholamhossein Ajami [3],

Prof Amirghofran, Shiraz, Iran - Technique published first in 2019

2020 – 21 patients with Fallots Tetralogy, mean age 13 months

10 months follow-up

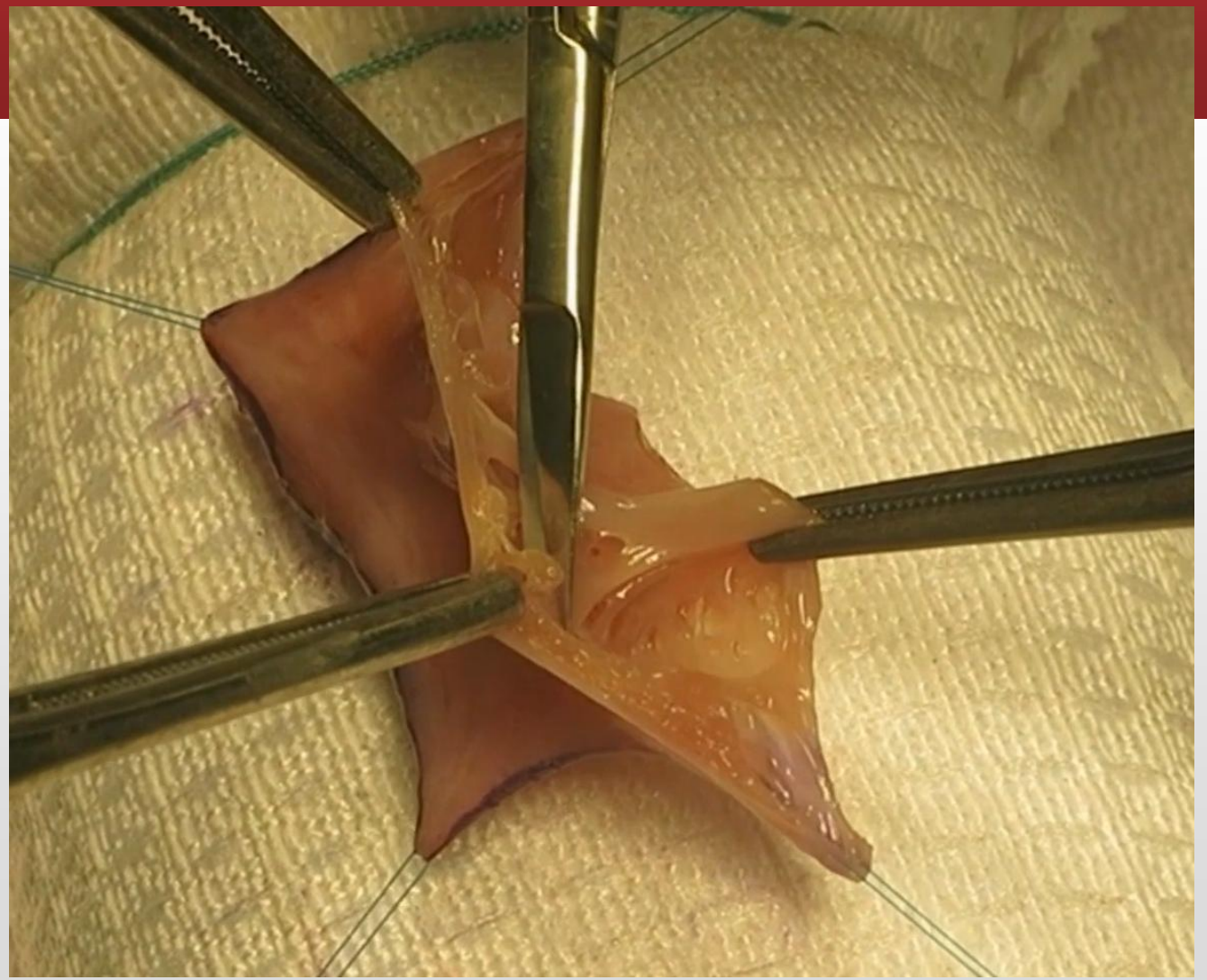
2022 – 121 patients, age range 15 days to 57 yrs

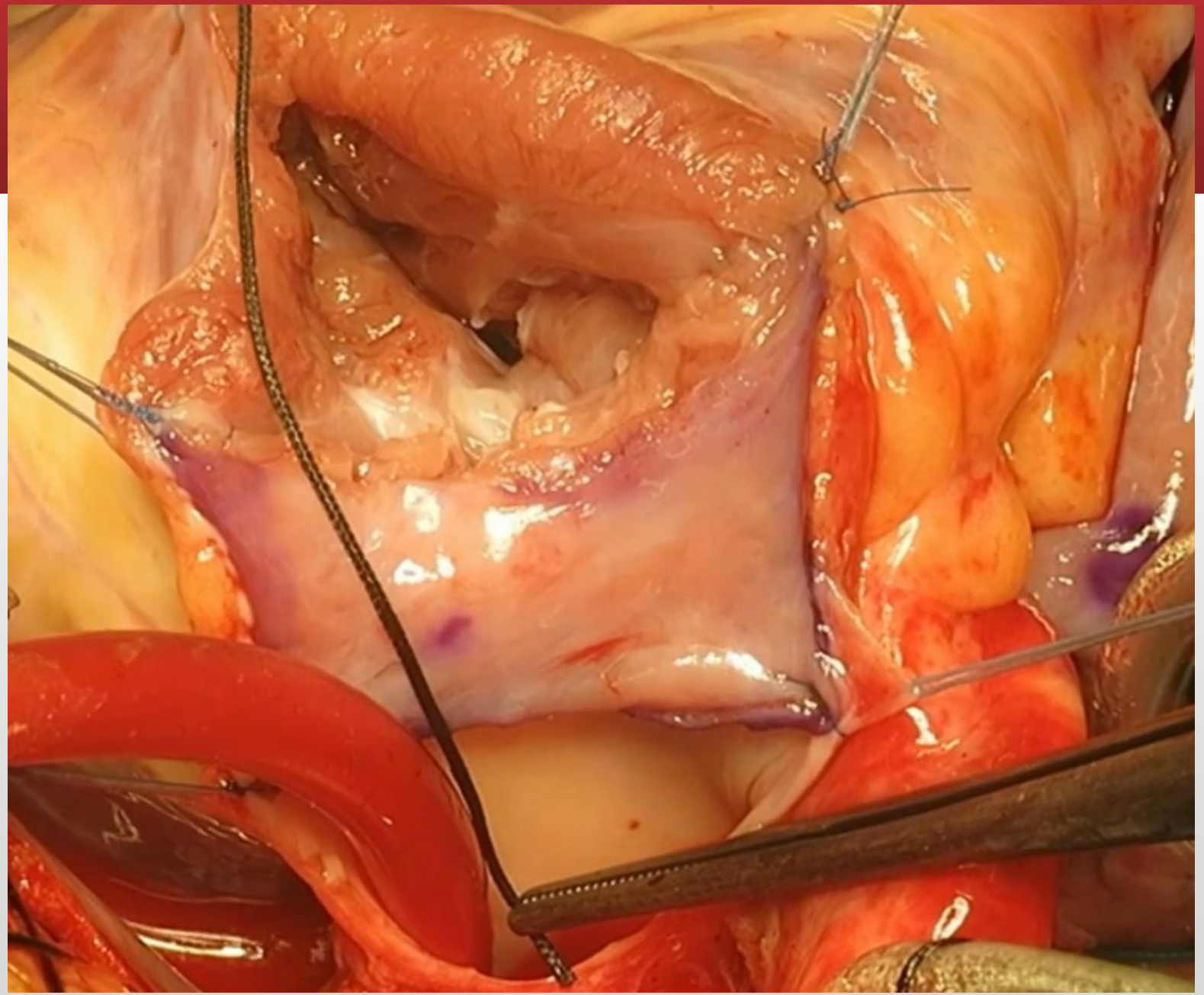
98/121 – Fallots tetralogy

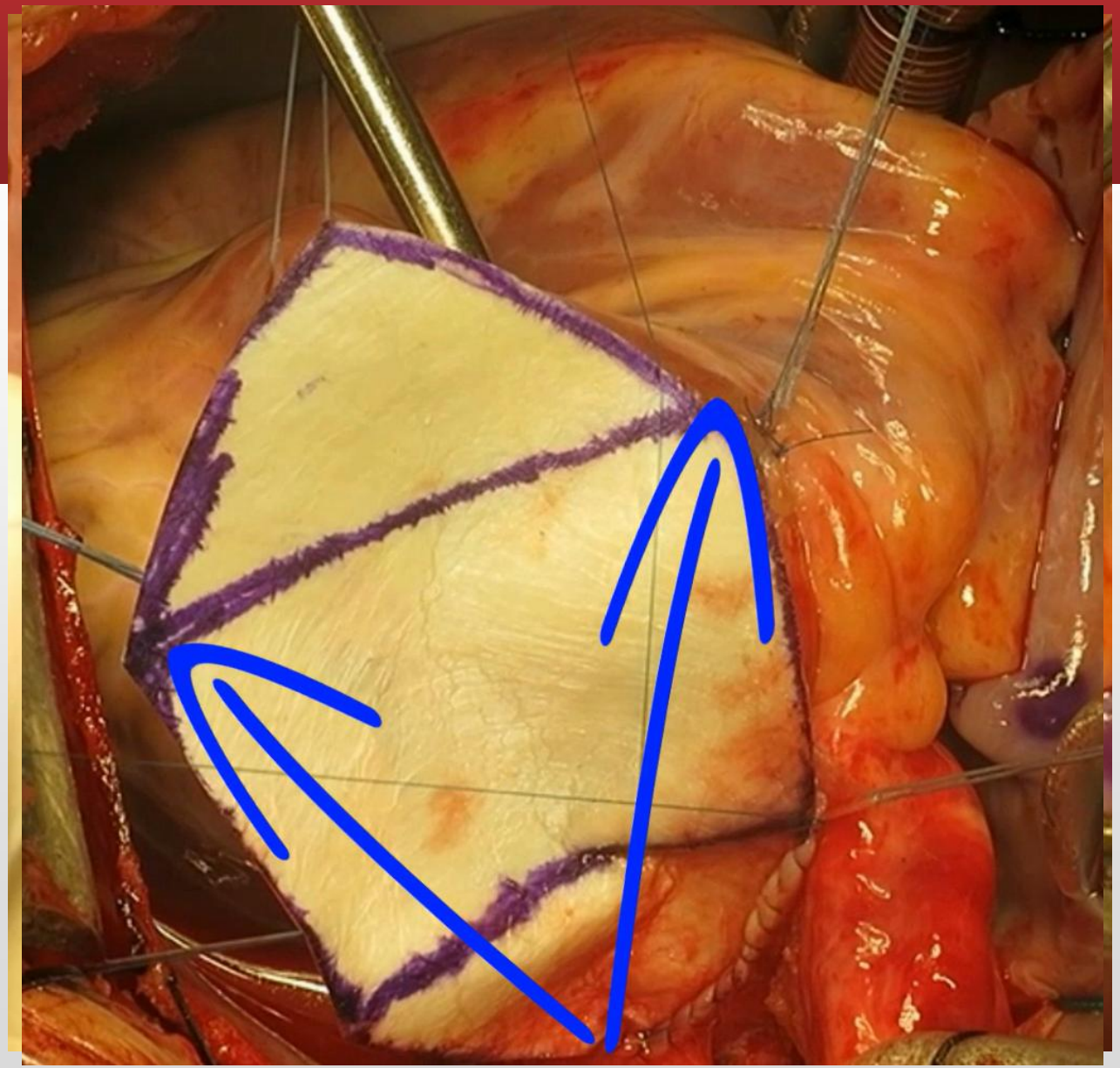
23/121 – Absent pulmonary valve, truncus, Nikaidoh

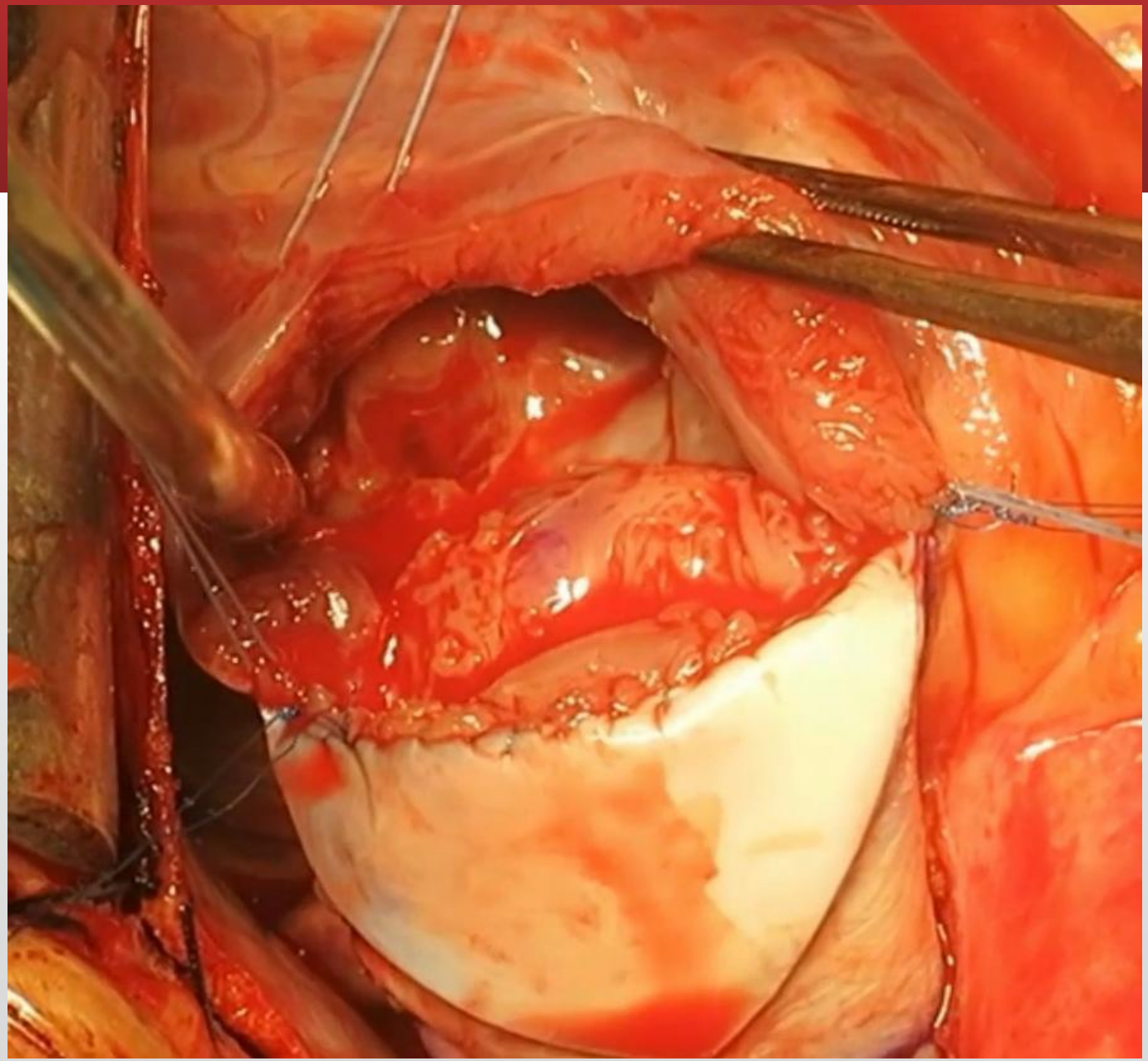
www.ctsnet.org/article/how-make-valve-rvot-right-atrial-appendage

CTSNet

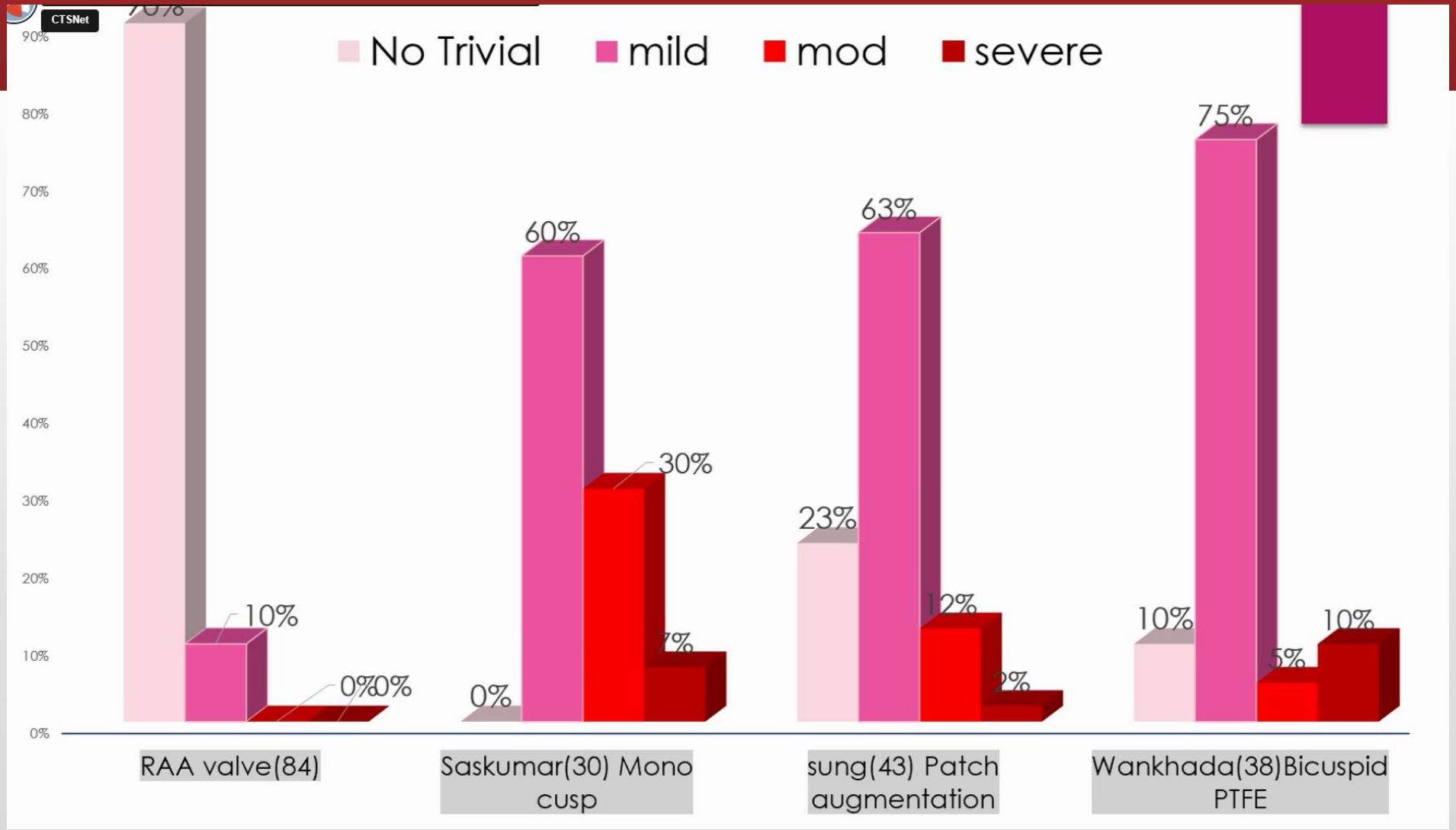






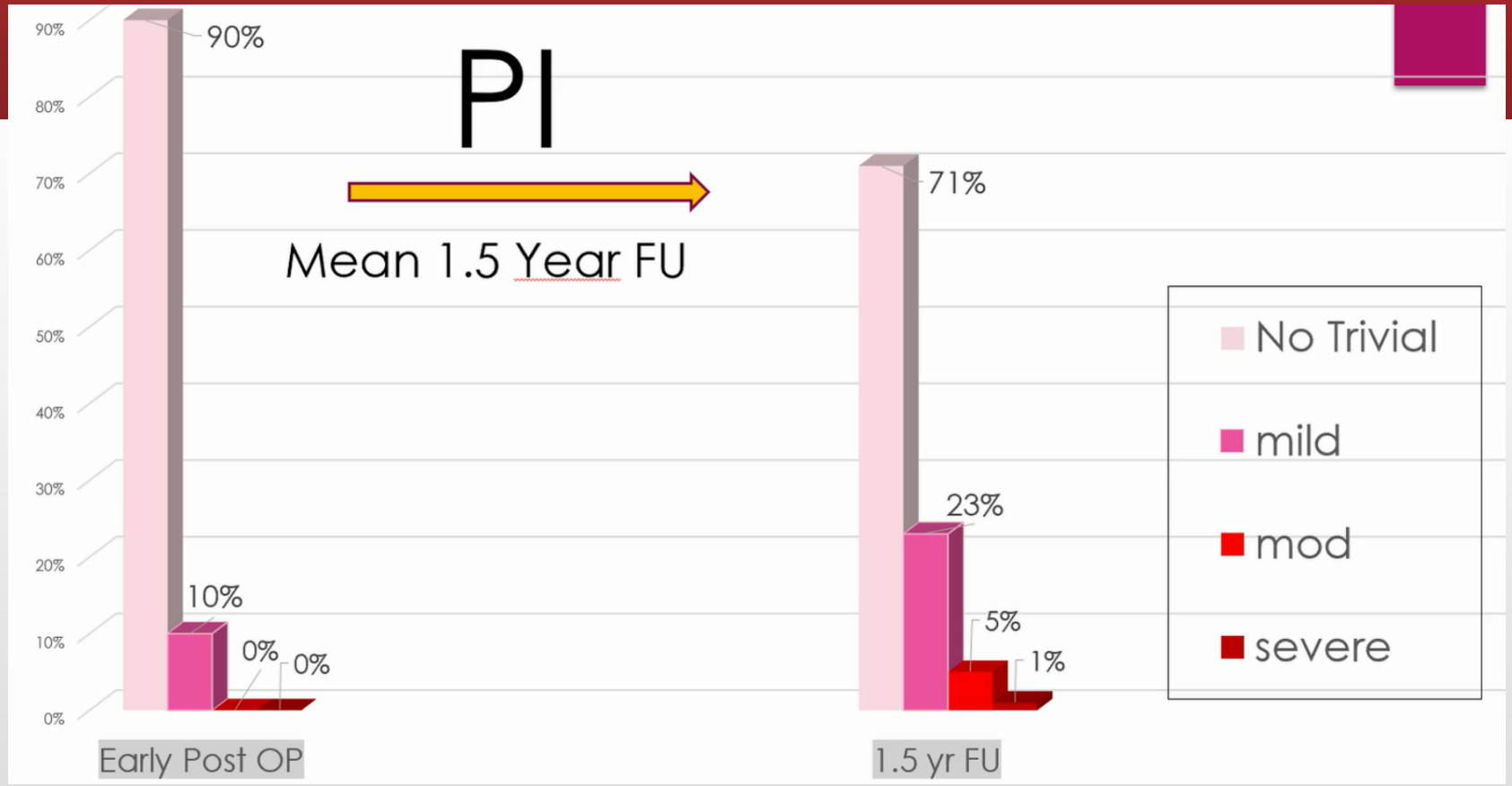


Discharge ECHO Outcomes compared to monocusps



ICU stay (days), mean ± SD	**TAP**	5.22 ± 1.98	**RAA valve**	4.22 ± 1.09

Outcomes at mean 1.5 years follow-up



Characteristics	TAP	RAA valve
Pulmonary valve pressure gradient (mmHg), mean ± SD		
Mild	60% (23.57 ± 7.11)	72% (18 ± 6.84)
Moderate	40% (43 ± 2.64)	28% (41 ± 3.45)
Severe	0%	0%

My experience – 20 patients over 2.5 years

Procedure:	Number of patients:
Fallots Tetralogy repair	8
DORV with subaortic VSD	5
DORV with AVSD	2
Severe pulmonary stenosis / Pulmonary atresia	4
Unifocalisation, VSD closure	1

Initial Intervention:	Number of Patients
RVOT stent	7
Balloon Pulmonary Valvuloplasty	4
PDA stent	3

My experience - 20 patients over 2.5 years

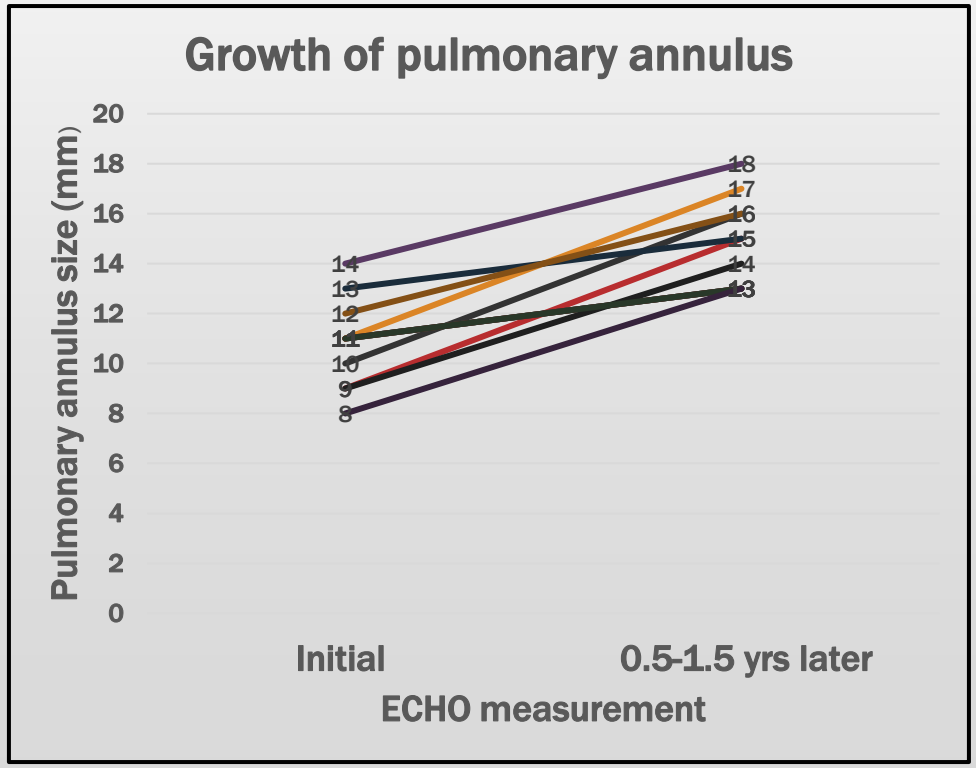
Discharge ECHO:

Mean peak gradient 18mmHg (Range 3 - 39mmHg)

ECHO 6 months – 1.5 years post-op:

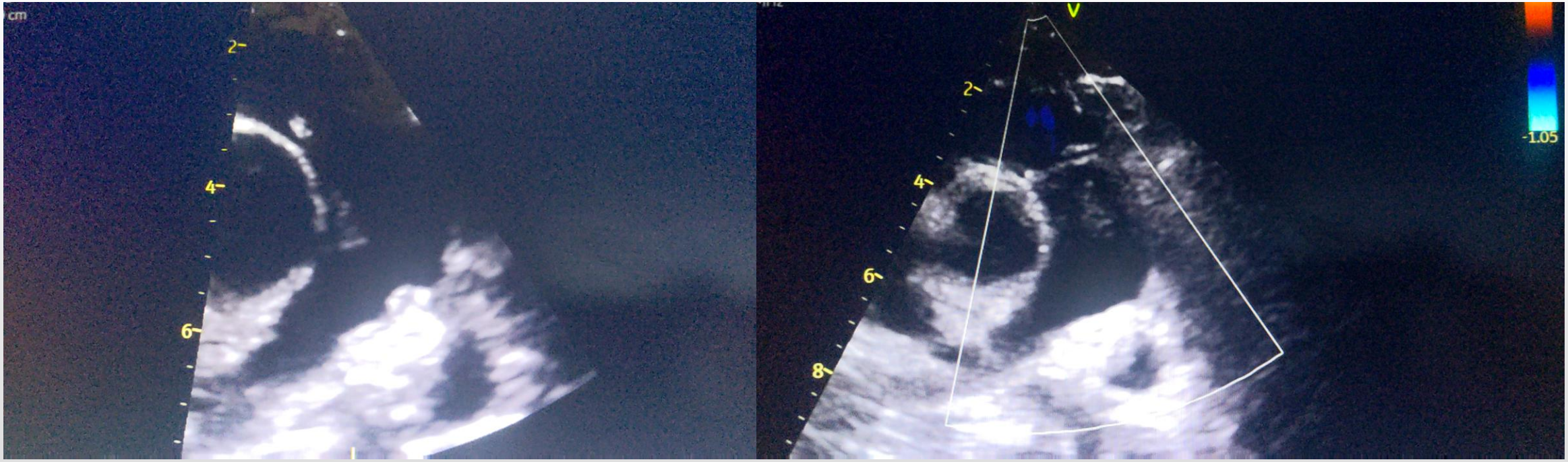
Mean Peak gradient 14mmHg (Range 6 – 22mmHg)

Initial Pulmonary Valve Leak:
None/Mild = 17/20 (85%)
Moderate/severe = 3/20 (15%)



Follow-up valve leak:
None/Mild = 12/16 (75%)
Moderate/severe = 4/16 (25%)

Post-op ECHO



Summary

Approx ½ of DORV patients with subaortic VSD will be of the Tetralogy-type

30% will need an intervention before definitive repair to augment pulmonary blood flow

15% will need a VSD enlargement

Strong consideration should be given to preserving the pulmonary valve

Monocusp and right atrial appendage valves shorten the post-op ICU length of stay by 1 day on average vs a transannular patch.

Right atrial appendage valves may have less longterm stenosis and regurgitation than a monocusp valve.