Techniques of annulus sparing repair in TOF

Dr. Thomas Mathew
Star Hospital, Hyderabad

IACTSCON 2024, Bhubaneshwar

Definitions & confusions

- Valve sparing repair - used synonymously with annulus sparing
    - *A misnomer, nobody “spares” the valve (unless thin and trileaflet)*

- Infundibulum sparing repair - used synonymously Trans RA PA repair
    - *Again a misnomer, nobody “spares” the infundibulum*

- Limited Transannular patch—???

- **ToF repair - No ventriculotomy**
- **ToF repair - Ventriculotomy, Annulus intact**
- **ToF repair - Transannular patch**

Topics

- Patient selection
- Anatomy
- Techniques
- Evidence for annulus sparing ToF Repair and current trends

Patient selection

Pre-operative

Review

Validity of Pulmonary Valve Z-Scores in Predicting Valve-Sparing Tetralogy Repairs—Systematic Review †

Children **2019**, *6*, 67; doi:10.3390/children6050067

- Echo : Z scores of the annulus
    - Just a guide- more than -2, - 2 to -4, less than -4

Like traffic lights , these are only suggestions !!!

- Age <3 months be cautious

- Noonan's syndrome— Inspite of good annulus leaflet thickening is the limiting factor

Intra-operative

One can see only with(in) the heart!!

- DON’T
  - For the obviously small annulus
  - Coronary crossing at the annulus (not at the infundibulum)
  - Most Doubly committed VSD
  - Dysplastic thickened leaflets
  - A thick fibrous ring under the Pulmonary valve

Anatomy

Pediatric Cardiac Surgery Annual

The First Stella Van Praagh Memorial Lecture: The History and Anatomy of Tetralogy of Fallot

Richard Van Praagh

Stella Van Praagh, MD (1927-2006) of Children's Hospital Boston was one of the greatest pediatric cardiologists and pediatric cardiac pathologists of the 20th and early 21st centuries. Née Stella Zacharioudaki from Crete, Greece, in addition to her stellar professional attainments, she was also an outstanding cuisinière, hostess, linguist, philosopher, and philanthropist. In 1962, she married Richard Van Praagh, MD, beginning a life-long collaboration that was in every sense an *affaire de coeur*. They had three children and seven grandchildren. Dr Stella was the author of more than 110 scientific publications which helped to clarify the pathologic anatomy, the clinical and laboratory diagnosis, and often the surgical management of many different forms of congenital heart disease, including dextrocardia, single ventricle, truncus arteriosus, tetralogy of Fallot (TOF), transposition of the great arteries, double-outlet right ventricle, sinus venosus defect, anomalous pulmonary venous drainage, the heterotaxy syndromes with asplenia or polysplenia, juxtaposition of the atrial appendages, and apical muscular ventricular septal defect. In 1999, Dr Stella Van Praagh received the Distinguished Achievement Award of the Society for Cardiovascular Pathology, and in 2004, she was honored with the Paul Dudley White Award of the American Heart Association. Dr. Stella Van Praagh was that vanishingly rare combination of brilliant clinician, internationally renowned medical scientist, and deeply cultivated humanist. The anomaly now known as the TOF was first described by Niels Stensen in 1671, with other early reports by Edouard Sandifort (1777), William Hunter (1784), and many others. In 1888, Etienne-Louis Arthur Fallot published five serialized contributions in *Marseille Médical* concerning what he called the "blue malady," in which he described the now classical tetralogy of pulmonary outflow tract obstruction, ventricular septal defect, aortic overriding, and right ventricular hypertrophy. The other outstanding feature of Fallot's report was its emphasis on clinicopathologic correlation. In 1924, Maude Abbott coined the term "tetralogy of Fallot." In 1970, Van Praagh and colleagues presented the concept that the TOF is basically just one anomaly, a failure of normal expansile growth of the subpulmonary infundibulum and its sequelae. The anatomy of TOF is presented angiocardiographically, diagrammatically, and anatomically. A morphometric study of typical neonatal TOF is presented, based on 16 autopsied heart specimens with age-matched normal controls. The morphometric study documents that TOF is characterized by a low-volume subpulmonary infundibulum. The diagnostic and surgical significance of these findings is highlighted. Two rare and recently discovered forms of TOF are presented: tetralogy {S,D,I}, and tetralogy {I,D,S}. Because tetralogy {I,D,S} has atrioventricular discordance, in addition to a standard TOF repair, such patients also need an inverted (mirror-image) atrial switch operation (inverted Senning or inverted Mustard procedure). Because associated malformations can be very important to the surgical outcome of patients with tetralogy, the associated anomalies found in 100 randomly selected autopsied cases are presented.
Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 12:19-38 © 2009 Published by Elsevier Inc.

KEYWORDS Stella Van Praagh, Tetralogy of Fallot, history, anatomy, morphometry, associated malformations, TOF {S,D,I}, TOF {I,D,S}

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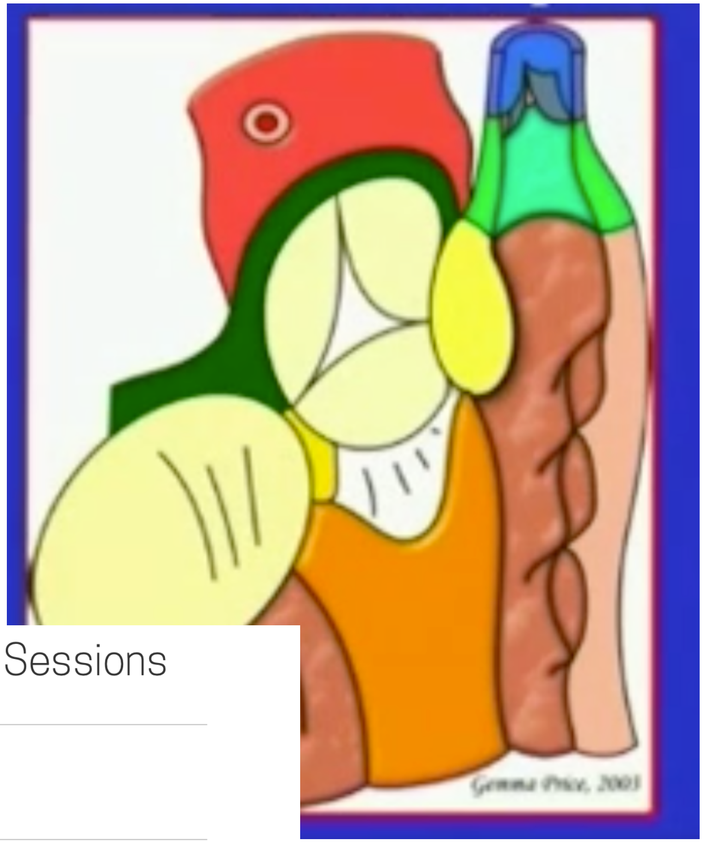
1092-9126/09/$-see front matter © 2009 Published by Elsevier Inc.
doi:10.1053/j.pcsu.2009.01.004



Congenital Morphology Video Sessions

Sunday, March 17, 2013
By **Robert Anderson**

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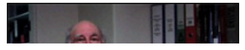
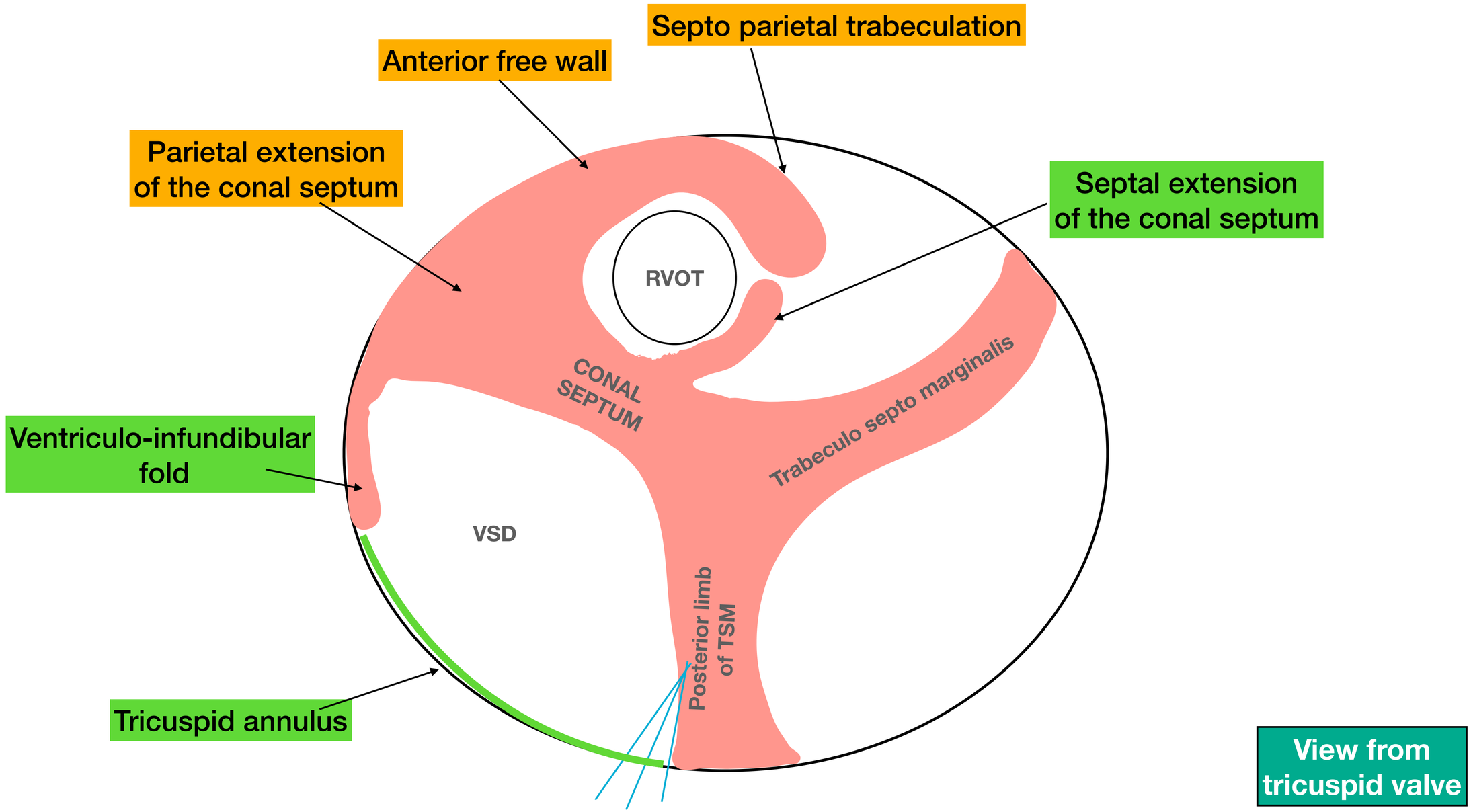
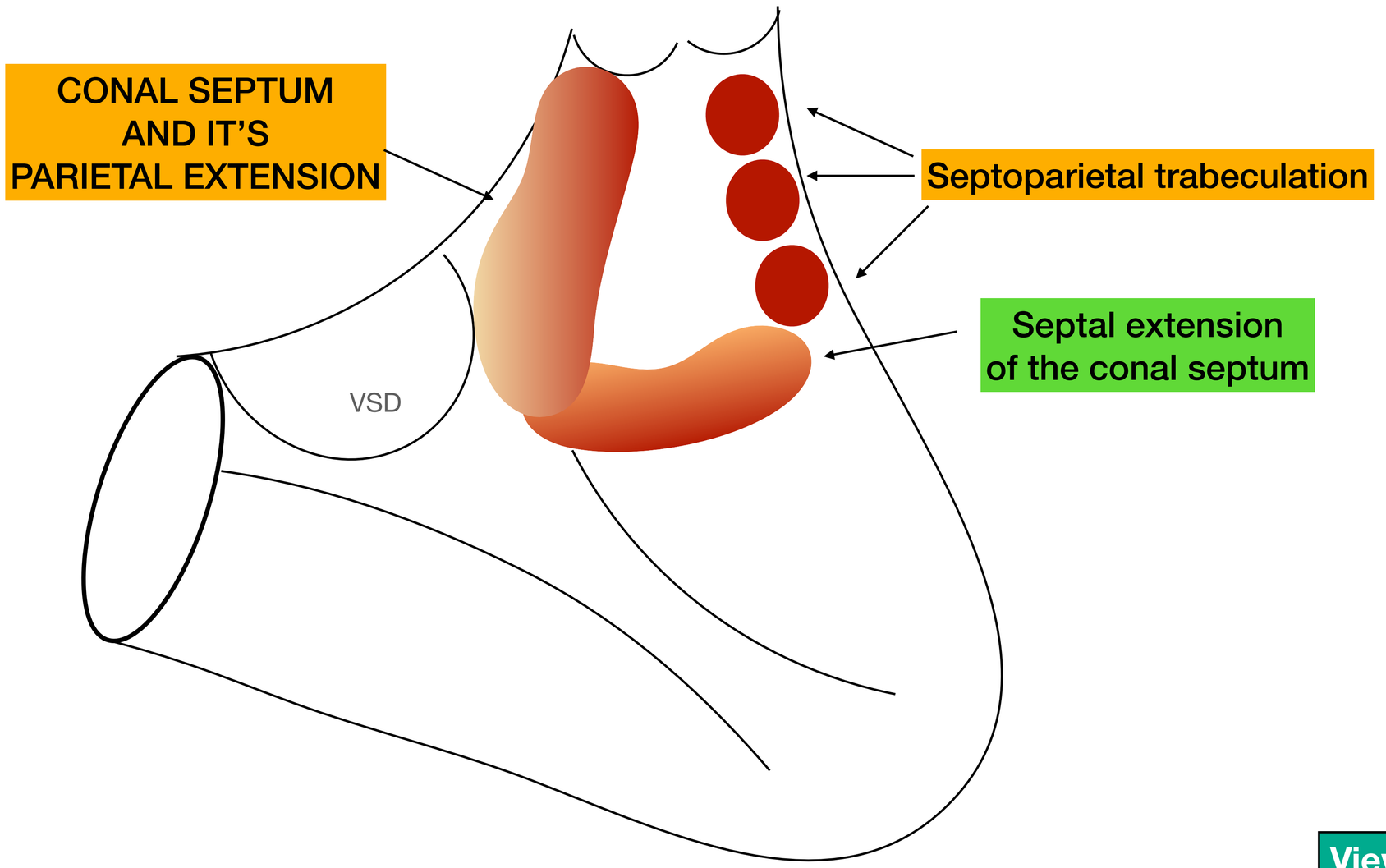
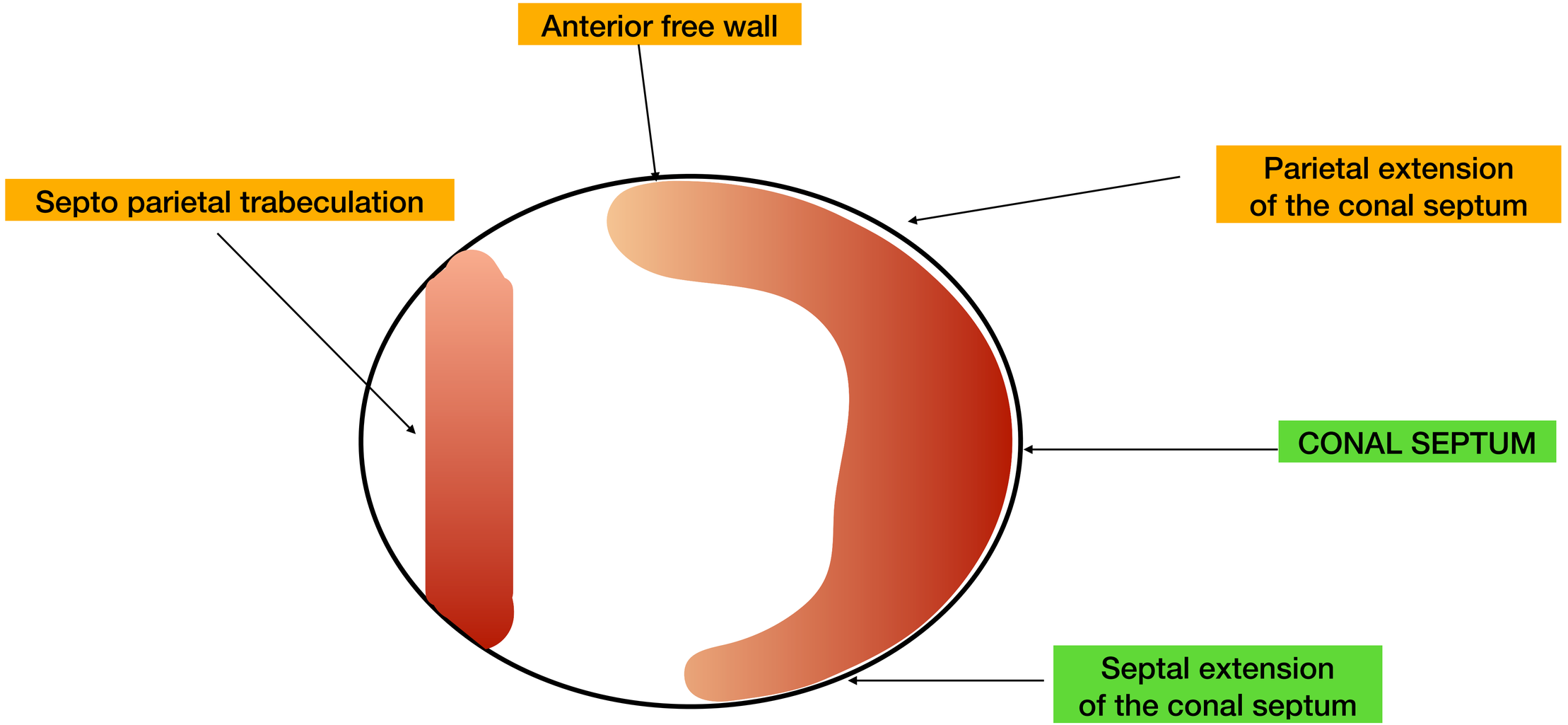
Introductory Remarks

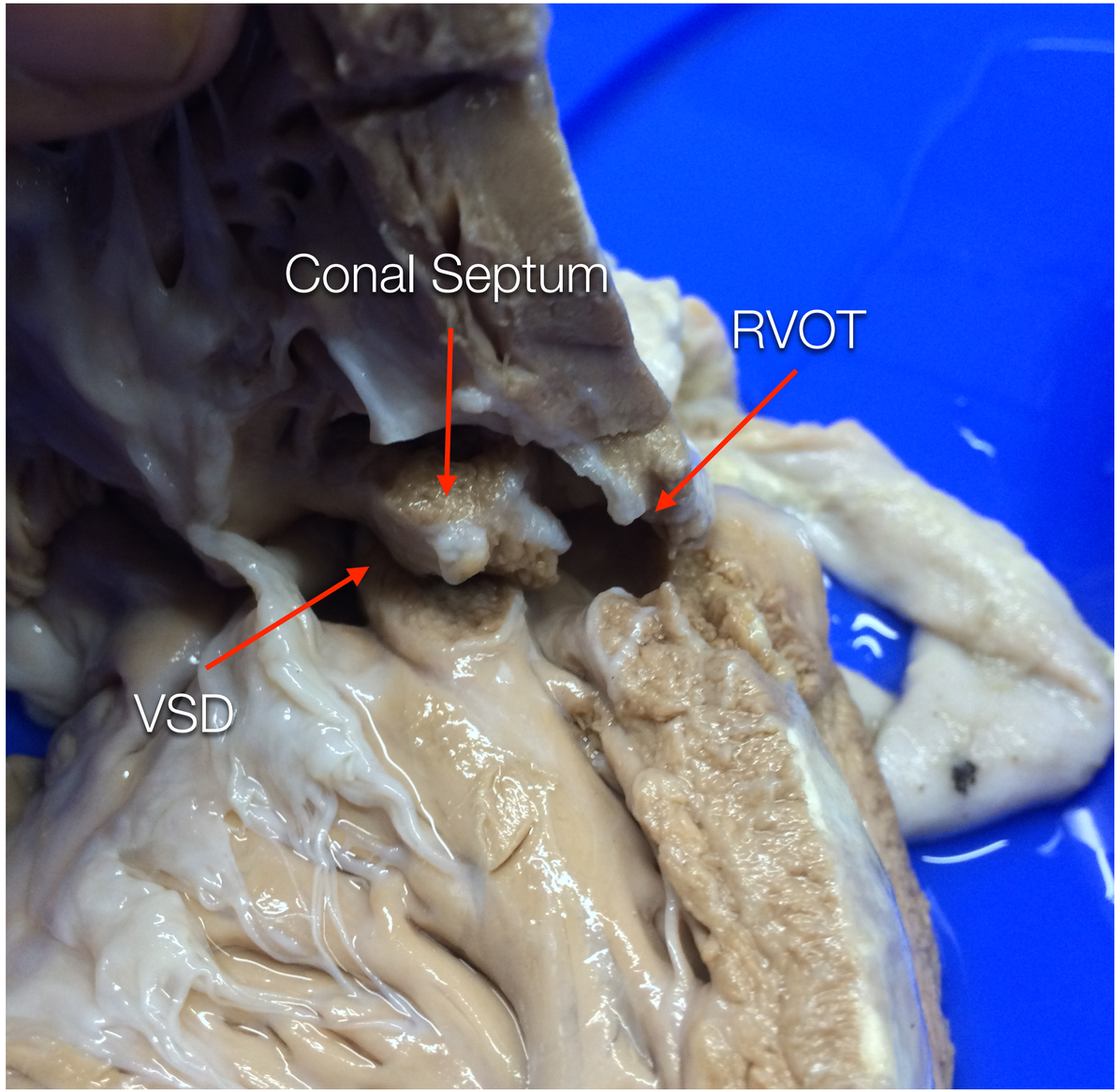


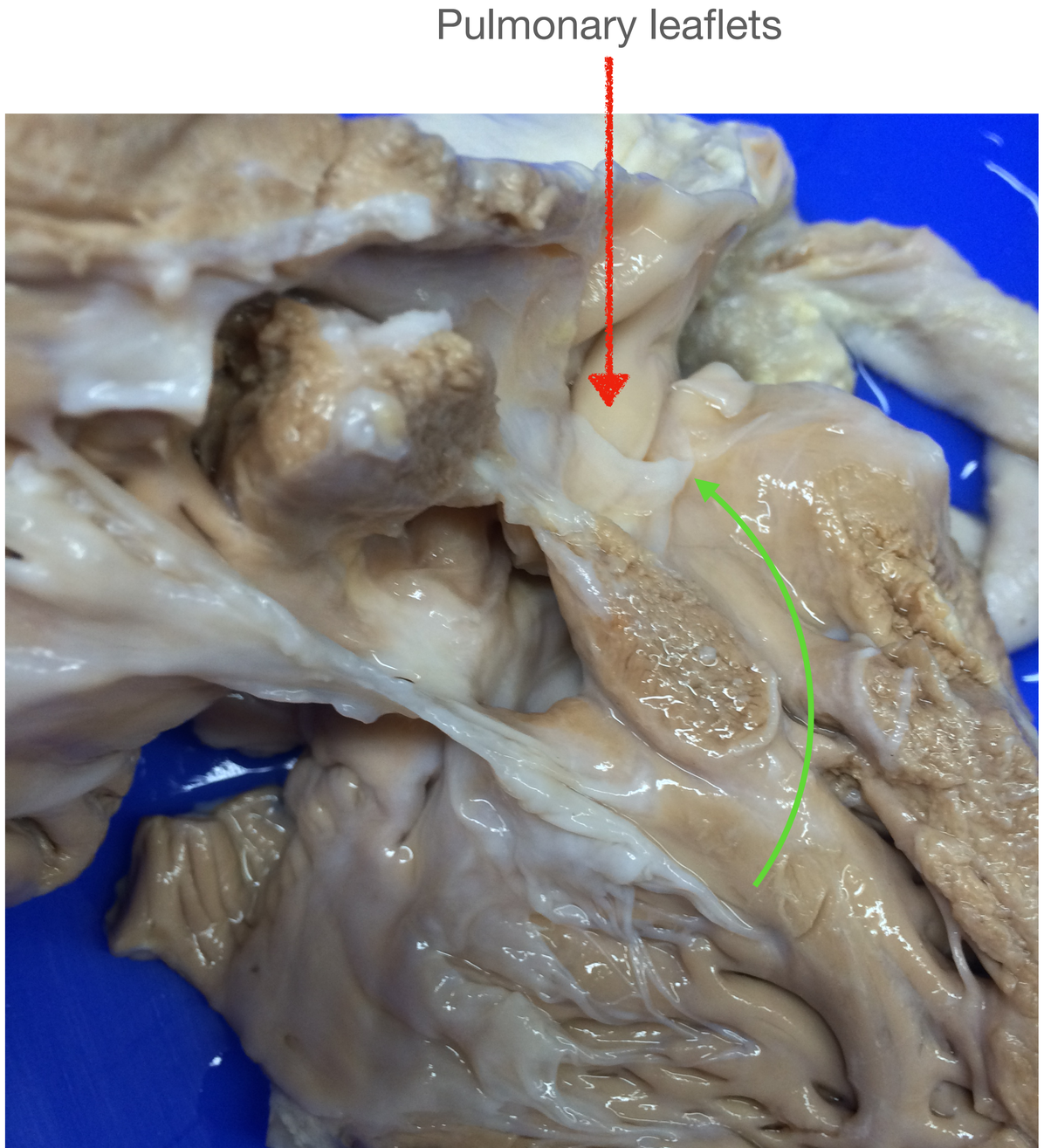
The following collection of video presentations has been generated by **Professor Robert H. Anderson**, using


Septo parietal trabeculation
Anterior free wall
Parietal extension of the conal septum
Septal extension of the conal septum
RVOT
CONAL SEPTUM
Trabeculo septo marginalis
Ventriculo-infundibular fold
VSD
Posterior limb of TSM
Tricuspid annulus
View from tricuspid valve


CONAL SEPTUM
AND IT'S
PARIETAL EXTENSION
Septoparietal trabeculation
Septal extension
of the conal septum
VSD
View from
Anterior


Anterior free wall
Parietal extension of the conal septum
Septo parietal trabeculation
CONAL SEPTUM
Septal extension of the conal septum
View from Pulmonary valve


Conal Septum
RVOT
VSD


Pulmonary leaflets

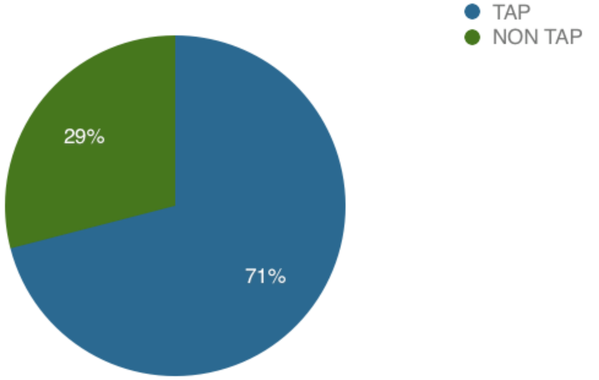
Operative Techniques

Off the table

- Mindset
  - Be prepared to revise
- Headlight
- Tenotomy scissors

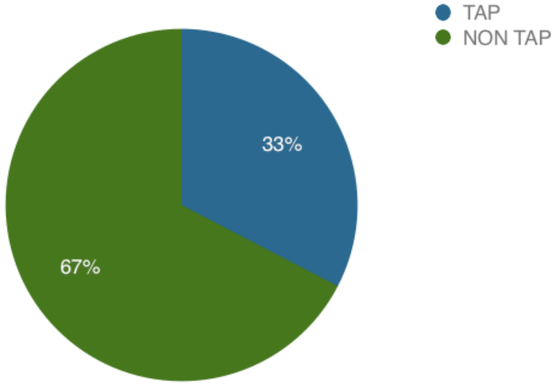
Pre-Iyer (Jul-Sept)

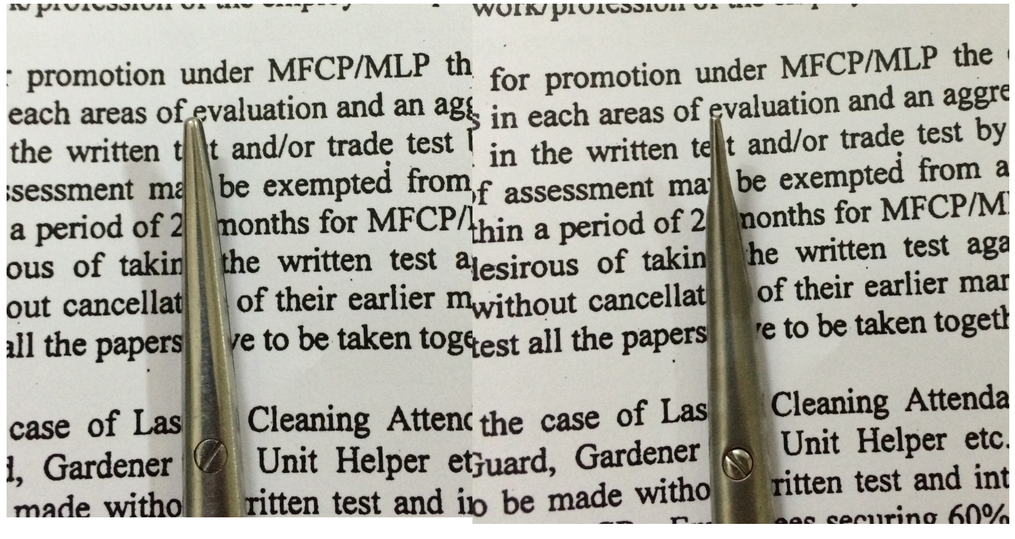
PROCEDURE	NO.
TAP	22
NON TAP	9



Post-Iyer (Oct-Dec)

PROCEDURE	NO.
TAP	15
NON TAP	31





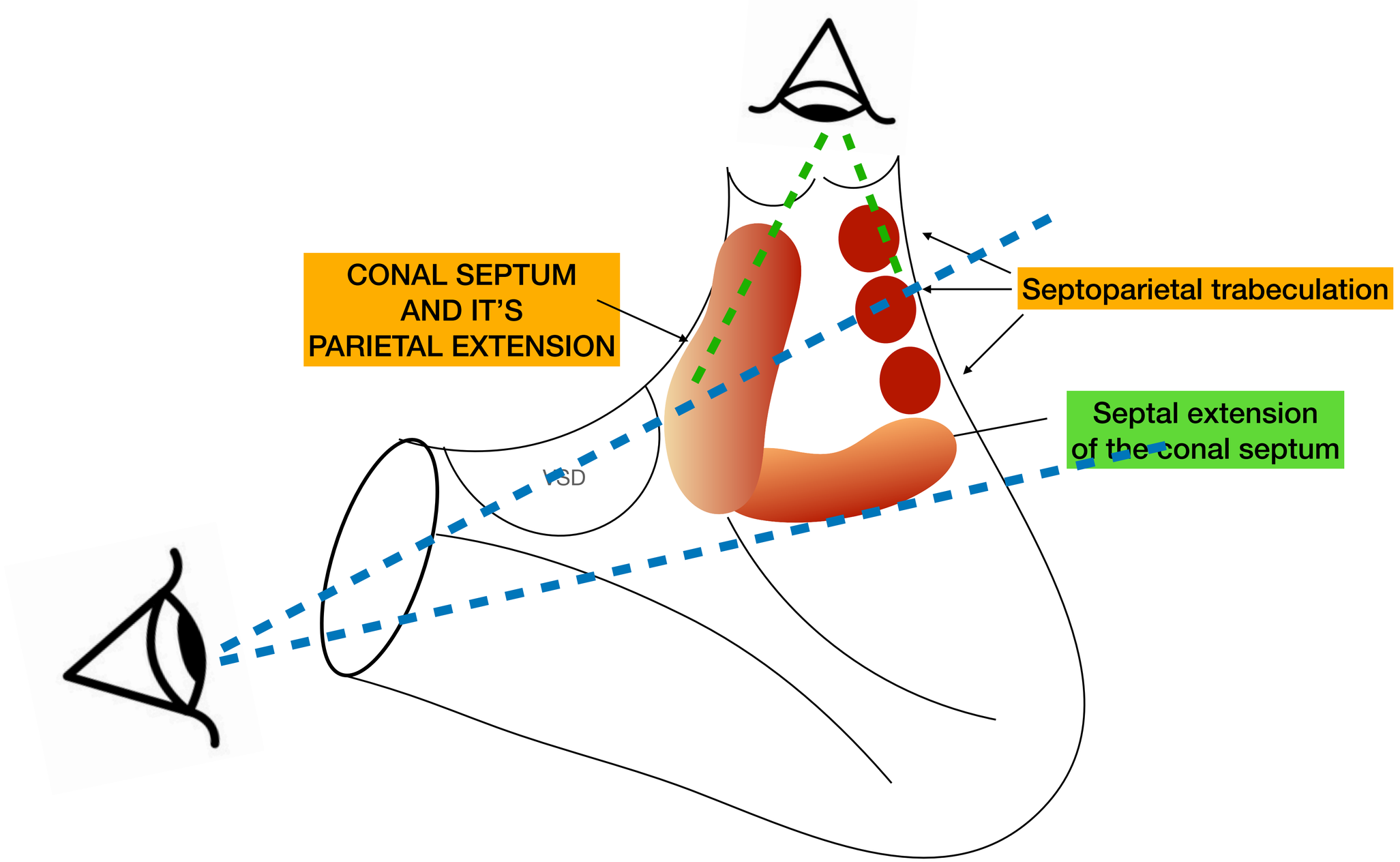
Approach to infundibular resection

Three tiered approach (RA— —PA— —RVOT)

- Trans RA VSD closure - with RV approach tendency to place a longer incision for VSD exposure

- Trans RA resection - takes off the lower bundles, parietal extension of the conal septum and lower septa-parietal bundles

- Trans PA for - upper bundles and pulmonary valve intervention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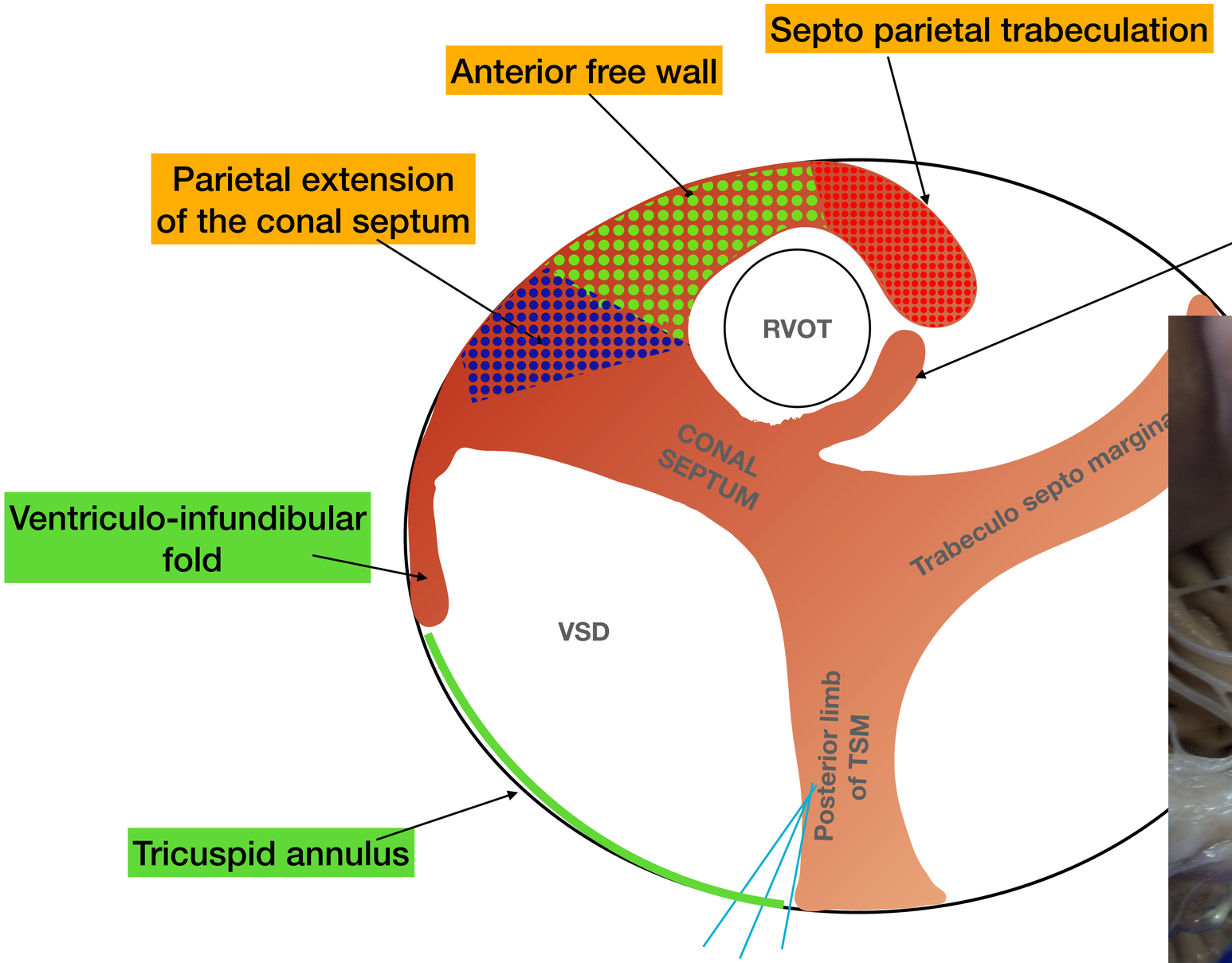
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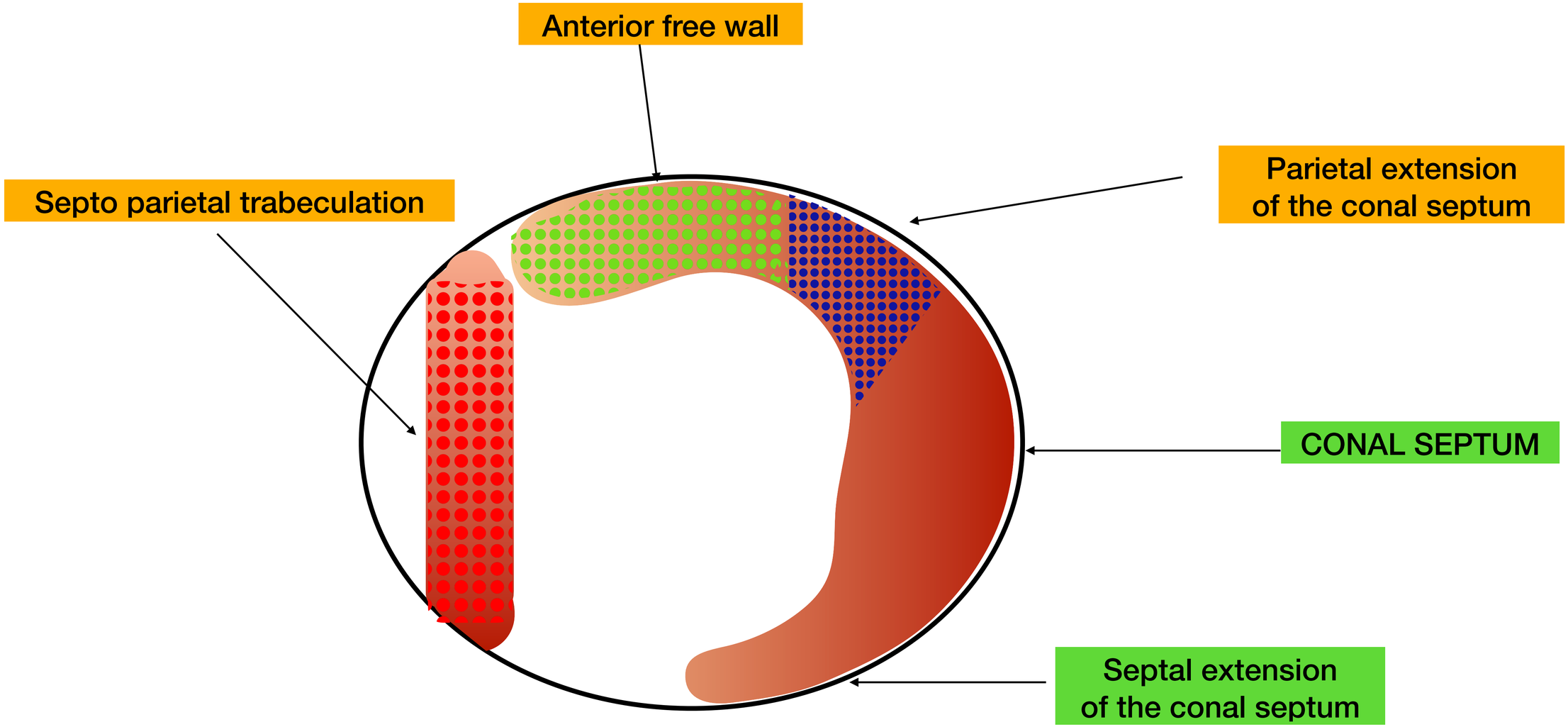
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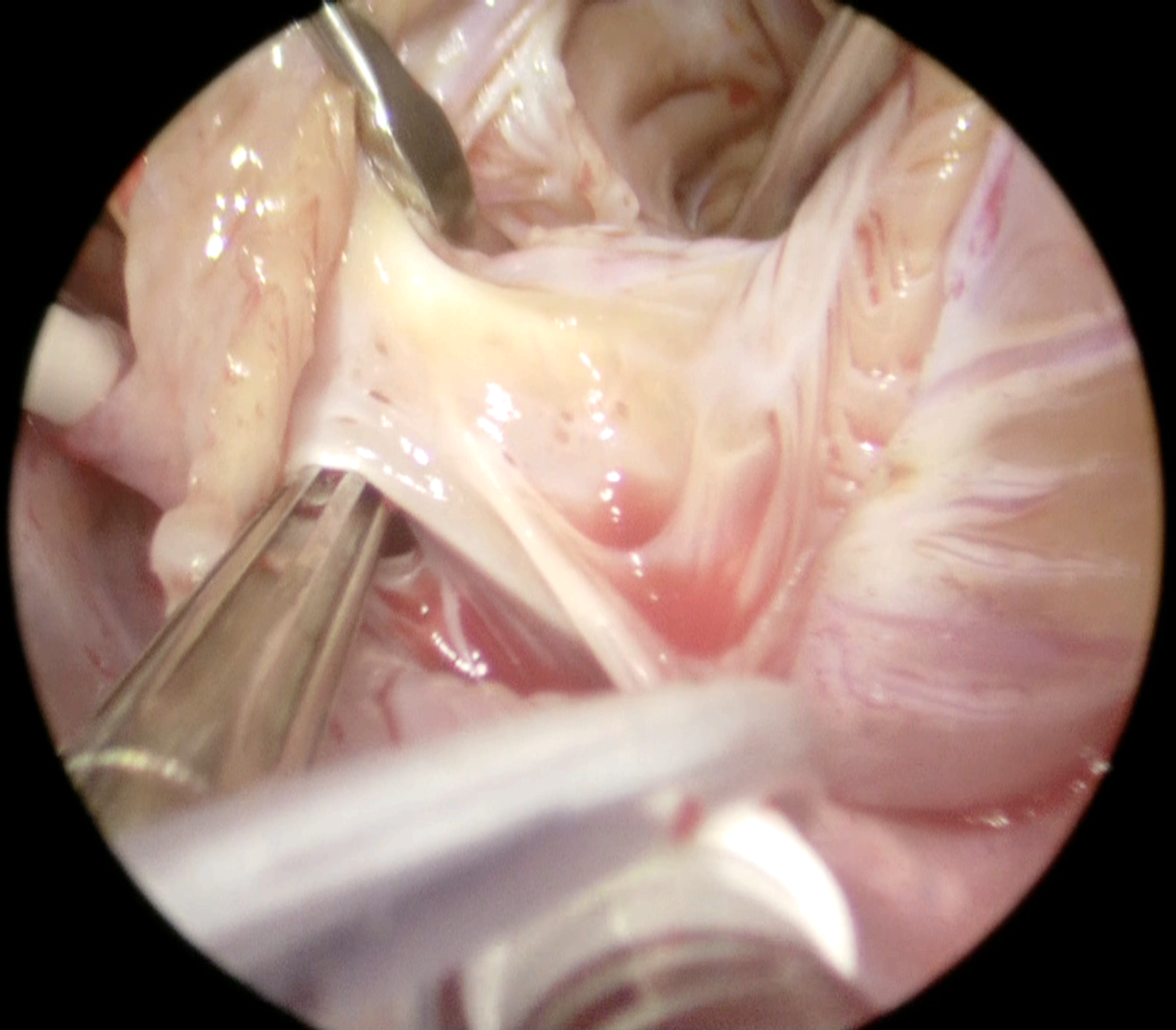
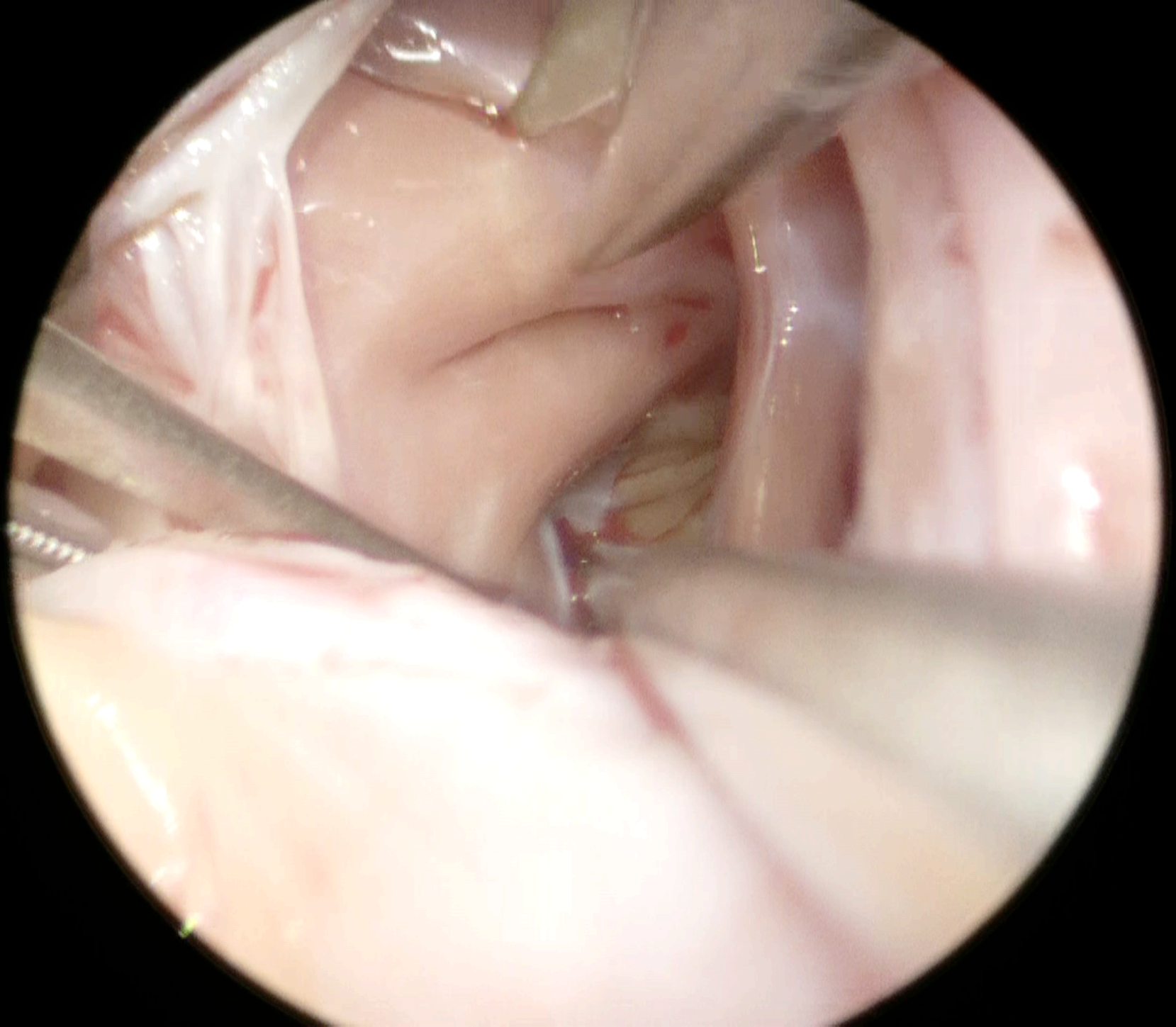
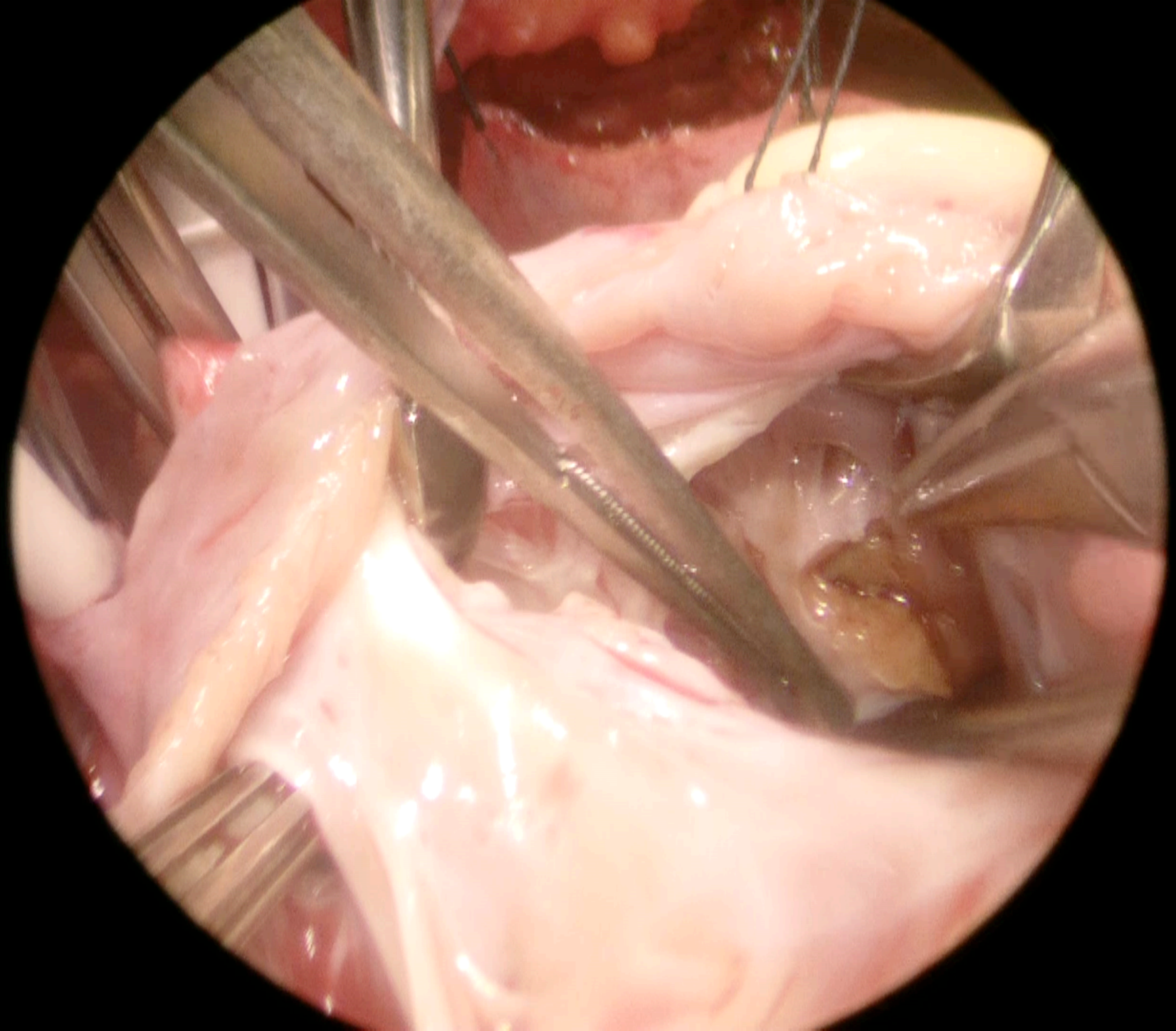
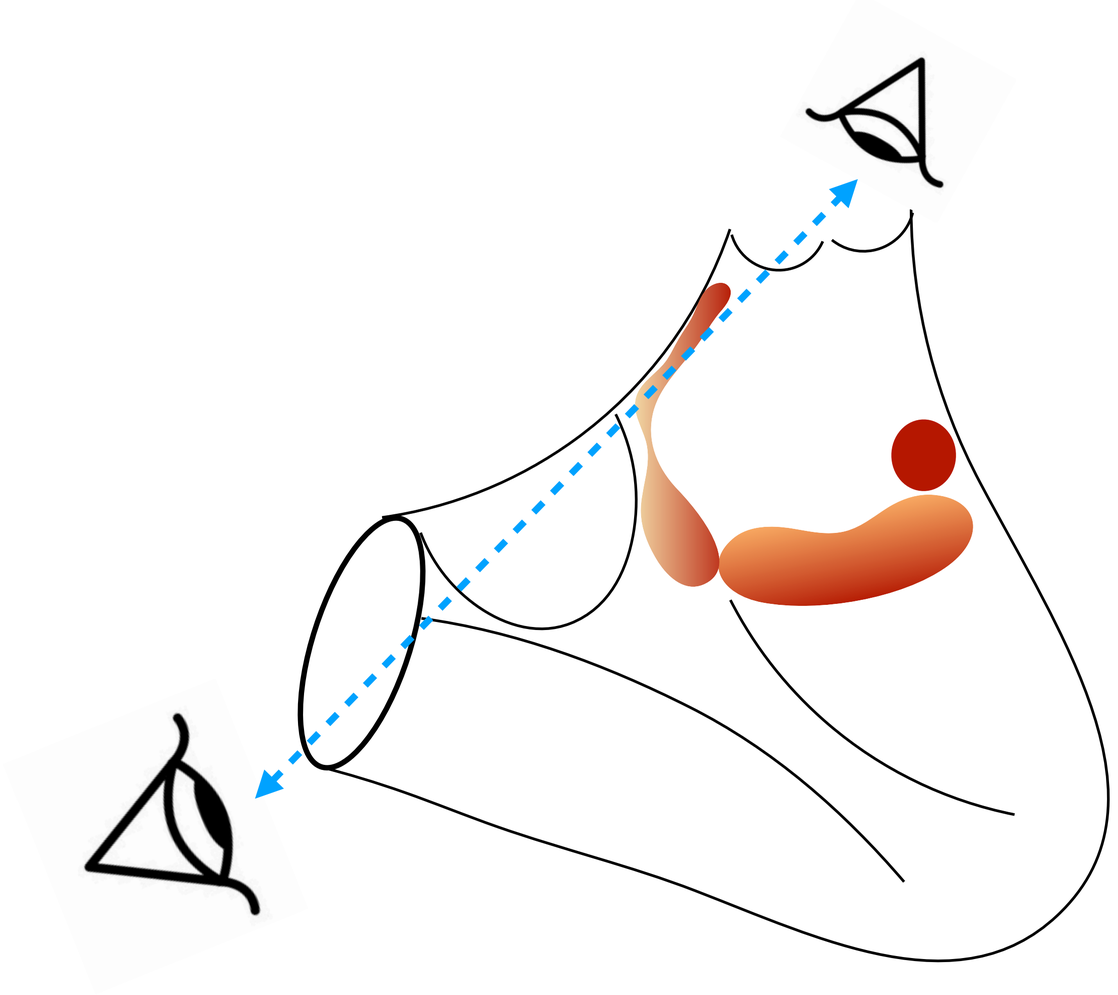






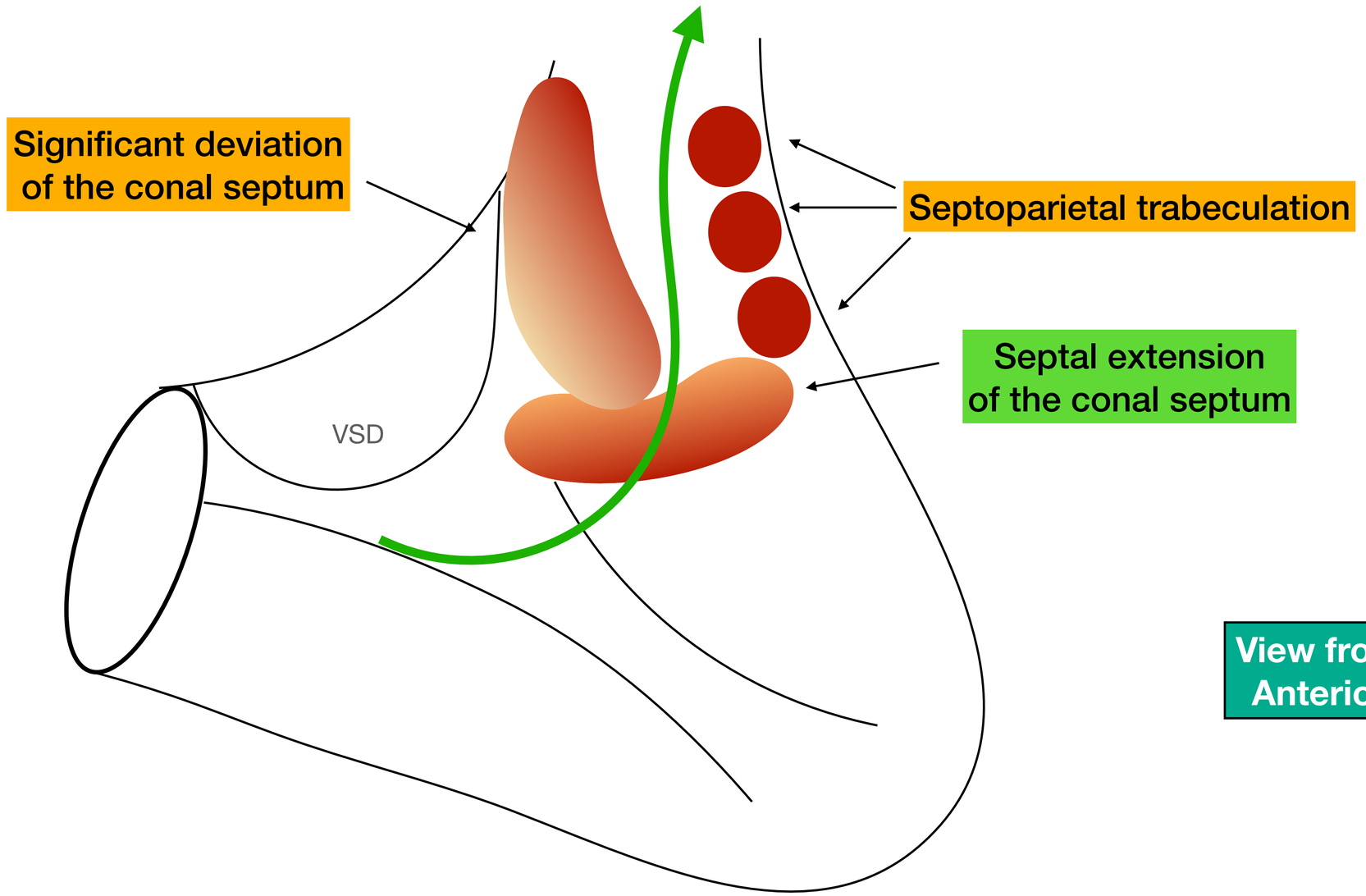
How to assess adequate resection?

NEVER use Hegars to assess adequacy of resection



When to open the RVOT

Three tiered approach (RA— —PA— —RVOT)



Pulmonary valve interventions

Be aggressive, bordering destructive!!!

- A complete valvotomy and sometimes excision of one leaflet

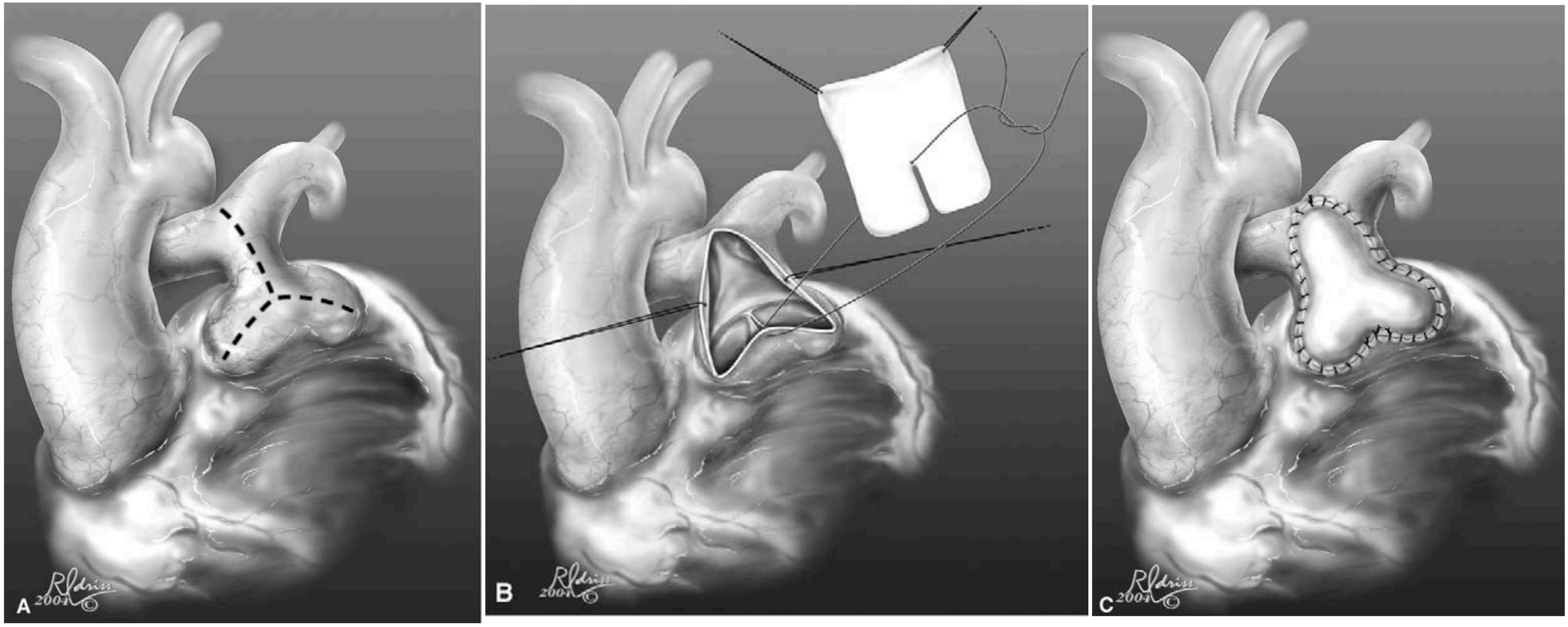
- Other techniques like leaflet thinning

MPA patching

Tetralogy of Fallot: Results of a Pulmonary Valve-Sparing Strategy

Robert D. Stewart, MD, MPH, Carl L. Backer, MD, Luciana Young, MD, and Constantine Mavroudis, MD

(Ann Thorac Surg 2005;80:1431–9)



Adequacy of the annulus

Hegar now.

- Upto one size below the required is safe

- If you have a good resection and thin leaflets, may be even 2 sizes below

When to convert to TAP?

Redo vs Regurgitation

- With hyper contractile heart, even gradient of 45-50mmHg
    - IV esmolol in some cases

- With normal hemodynamics 30-35mmHg

- If no ventriculotomy was done, the RVOT is patched.

- (I prefer to keep my redos for RVOTO to a minimum)

Does staging improve annulus sparing rates?

Unlikely

- Multiple studies for and against

- Annulus preservation rates ranged from 0% earlier studies to now 40 % in the more recent ones

Contemporary Patterns of Care in Tetralogy of Fallot: Analysis of The Society of Thoracic Surgeons Data

Nicholas S. Clarke, MD, MS,[1] Dylan Thibault, MS,[2] Diane Alejo, BA,[3] Karen Chiswell, PhD,[2] Kevin D. Hill, MS, MD,[2] Jeffrey P. Jacobs, MD,[4] Marshall L. Jacobs, MD,[3] Bret A. Mettler, MD,[3] and Danielle Gottlieb Sen, MD, MPH[3]

- Annulus preservation in this subset trends the overall annulus preservation rates

- Don’t forget the interstage mortality

Does Balloon valvotomy increase annulus sparing rates?

NO

ORIGINAL ARTICLE

Balloon pulmonary valvotomy as interim palliation for symptomatic young infants with tetralogy of Fallot

K.S. Remadevi, Balu Vaidyanathan, Edwin Francis, B.R.J. Kannan, Raman Krishna Kumar
Division of Pediatric Cardiology, Amrita Institute of Medical Sciences and Research Center, Kochi, India

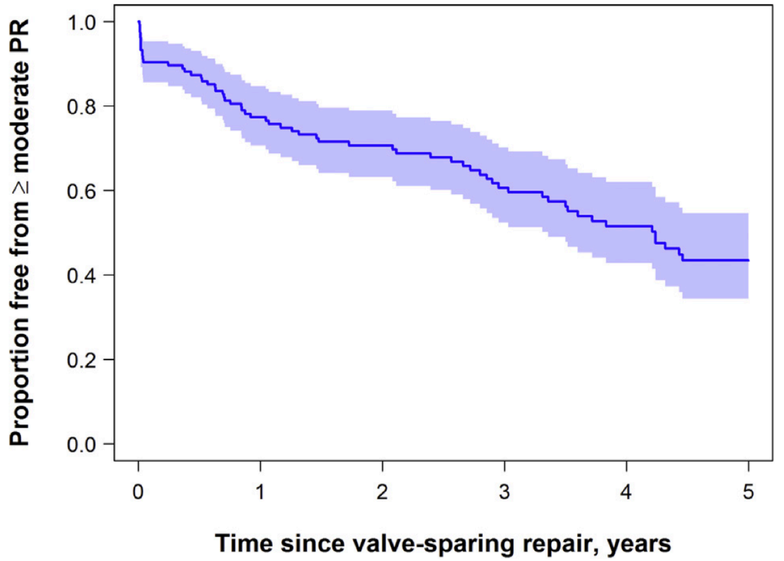
Ann Pediatr Card 2008 Vol 1 Issue 1

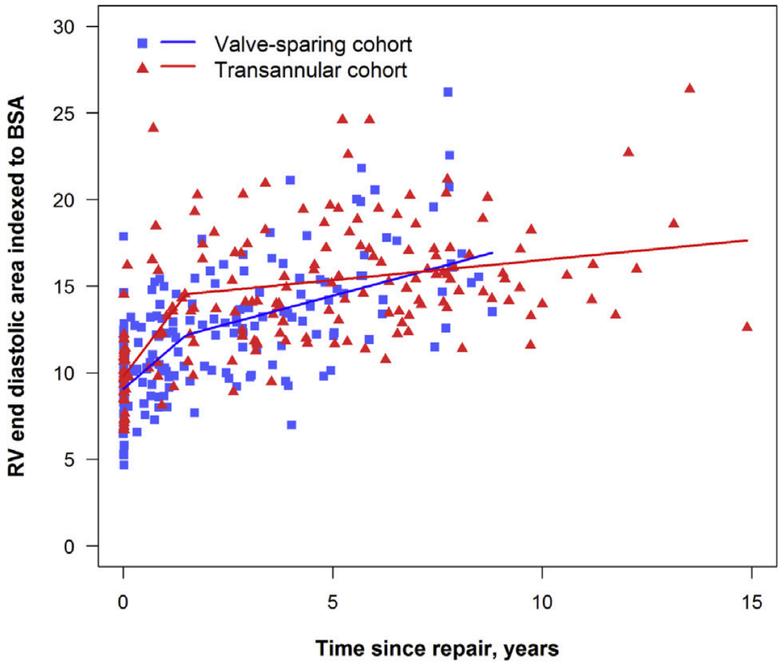
Why balloons are cosmetic!

Valve-sparing repair with intraoperative balloon dilation in tetralogy of Fallot: Midterm results and therapeutic implications

Sophie C. Hofferberth, MBBS,[a] Meena Nathan, MD,[a] Gerald R. Marx, MD,[a] Minmin Lu, MS,[b] Lynn A. Sleeper, ScD,[b,c] Audrey C. Marshall, MD,[b] Christopher W. Baird, MD,[a] John E. Mayer, MD,[a] Pedro J. del Nido, MD,[a] and Sitaram M. Emani, MD[a]

Patients with ToF-PS who undergo valve-sparing repair with IBD develop progressive PR. Compared with traditional TAP repair, the timing and extent of RV dilation appears similar for patients who have undergone valve-sparing repair with IBD. In patients with significant annular hypoplasia, and those younger than 3 months of age at repair, alternative surgical approaches should be explored to minimize the risk of early valve reintervention for residual PS. Although ToF patients who undergo VS-IBD repair exhibit significant longitudinal PV annular growth, the observed lack of valve leaflet remodeling warrants further investigation. The results of this study suggest that valve-sparing repair with IBD is not a suitable long-term solution to preserve PV function in patients with ToF.





Evidence and trends

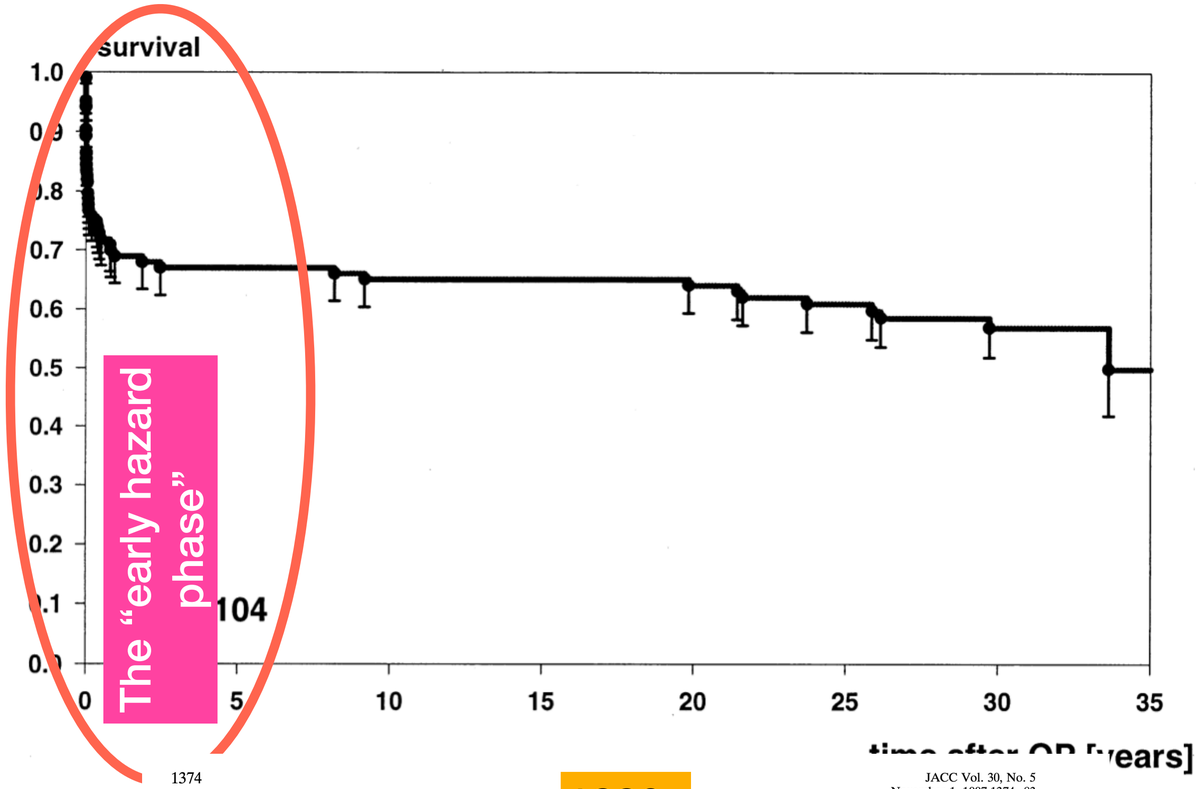
What’s the need of annulus preservation?

Questions

- Does annulus preservation reduce **Mortality** after TOF correction??

- Does annulus preservation reduce incidence of **Re-interventions** after TOF repair ??

Lower early mortality over the years.



1374

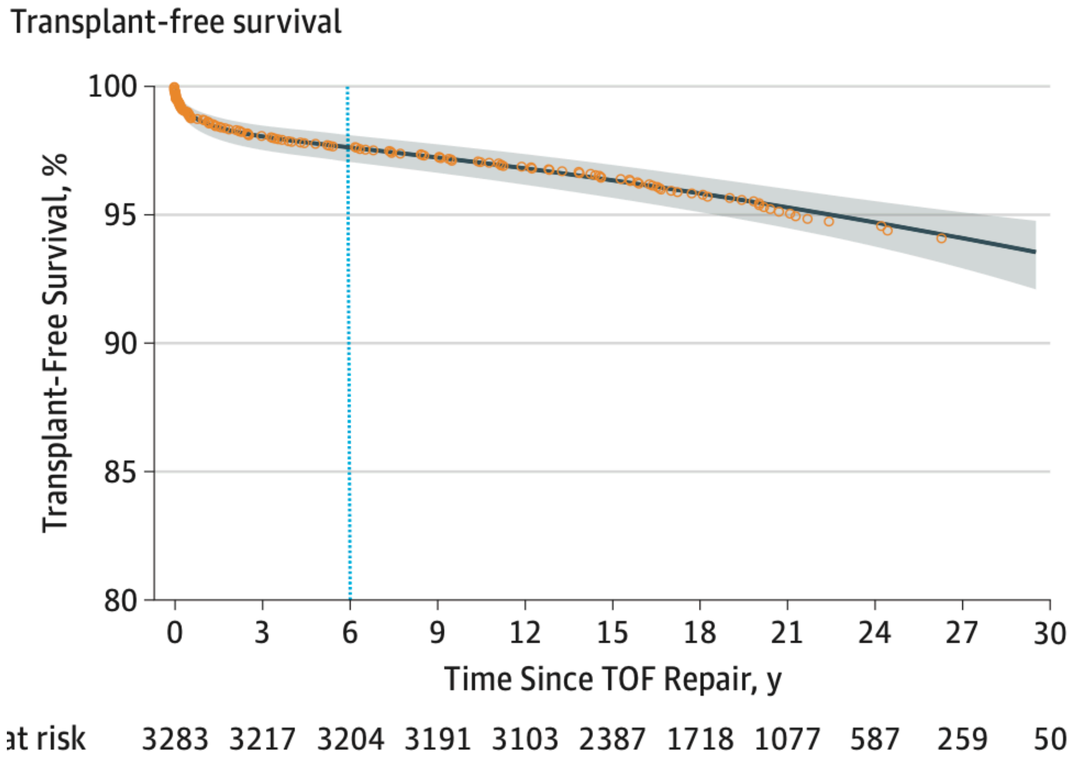
JACC Vol. 30, No. 5
November 1, 1997:1374–83

1990s

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

89% alive at 30 years



2000s

JAMA Cardiology | Original Investigation

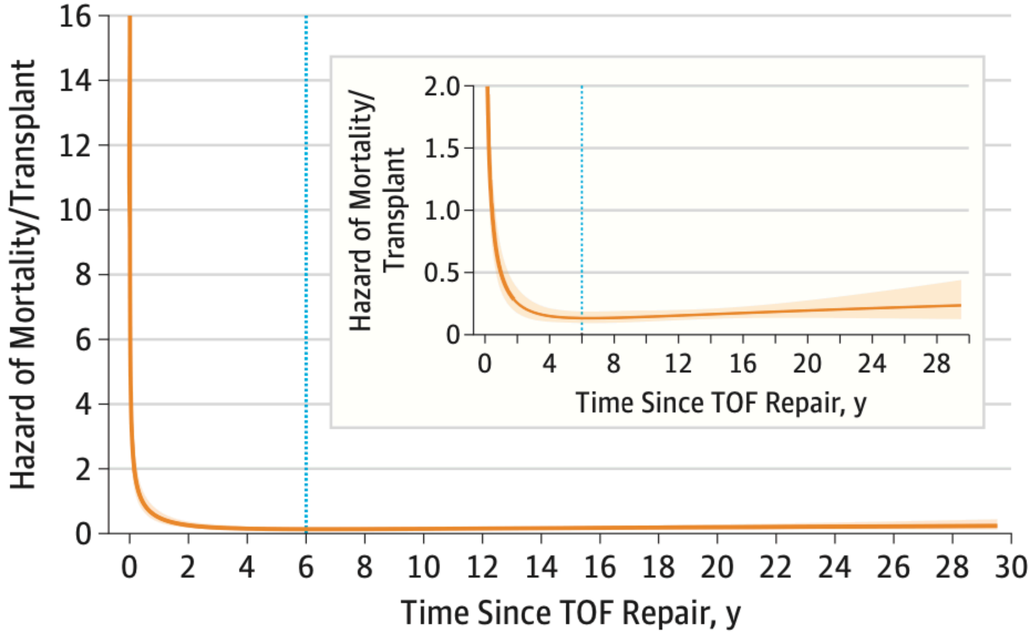
Long-term Outcomes of Tetralogy of Fallot
A Study From the Pediatric Cardiac Care Consortium

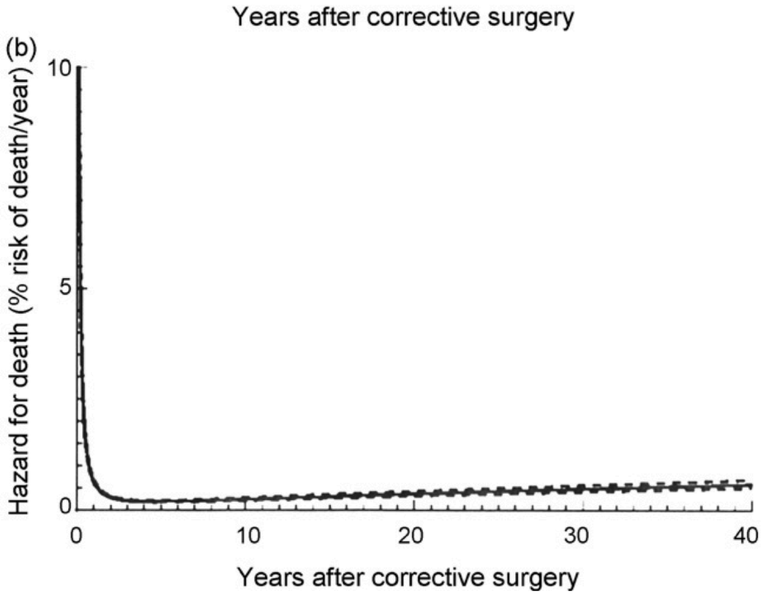
Clayton A. Smith, MD; Courtney McCracken, PhD; Amanda S. Thomas, MSPH; Logan G. Spector, PhD; James D. St Louis, MD; Matthew E. Oster, MD, MPH; James H. Moller, MD; Lazaros Kochilas, MD, MSCR

94.5% alive & transplant free at 25 years

Late mortality has not changed much.

Approximately
75% of deaths in 3 years
Touches baseline for 15 -20 years
Starts climbing at 20-25 years





Drivers for each phase of mortality

Early phase & operative

- Severity of the disease
    - TAP
- Palliation with shunt
- Residual lesions
- Extracardiac anomalies (Syndromes)

Presence of TAP- ?? indirect risk factor for mortality

Late phase

- Cardiac failure
- Sudden cardiac death
    - VT/ Heart block
- Syndromes

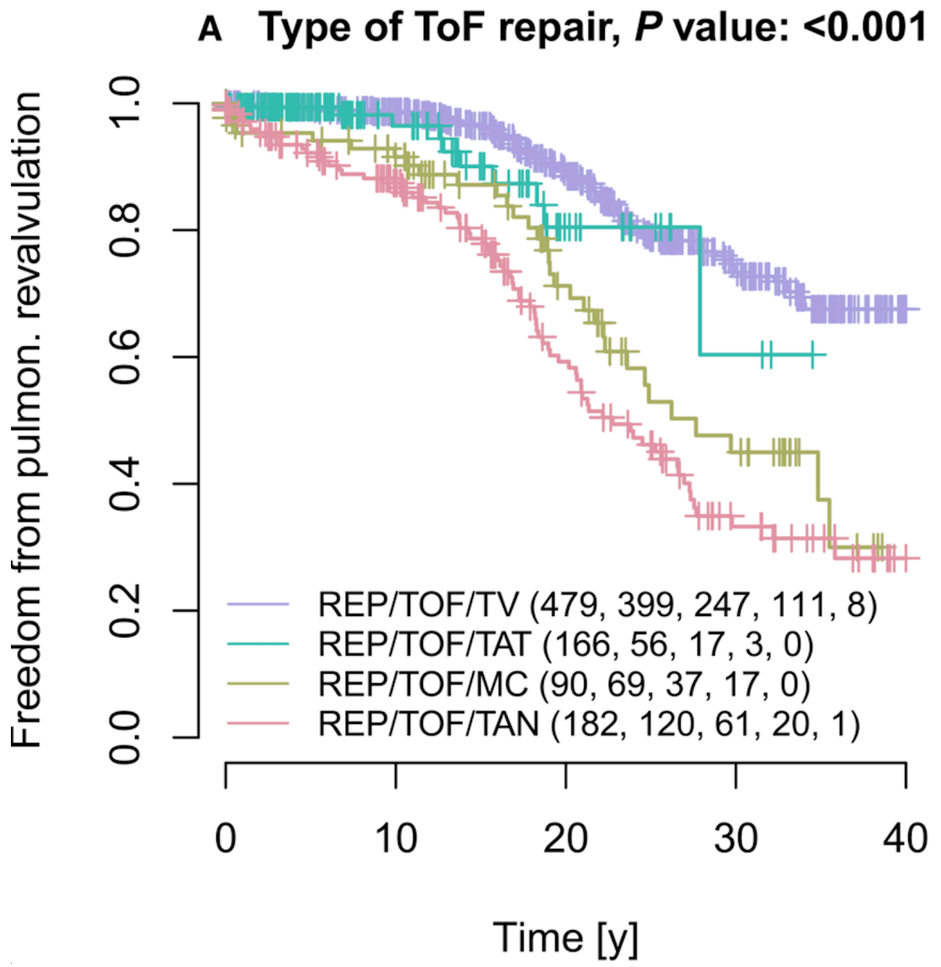
Presence of TAP- NOT a risk factor for late mortality

TAP is a risk factor for late re-intervention (PVR)

In most studies !!

- ~ 40% in 30 years

- PVR rate depends on the criteria followed

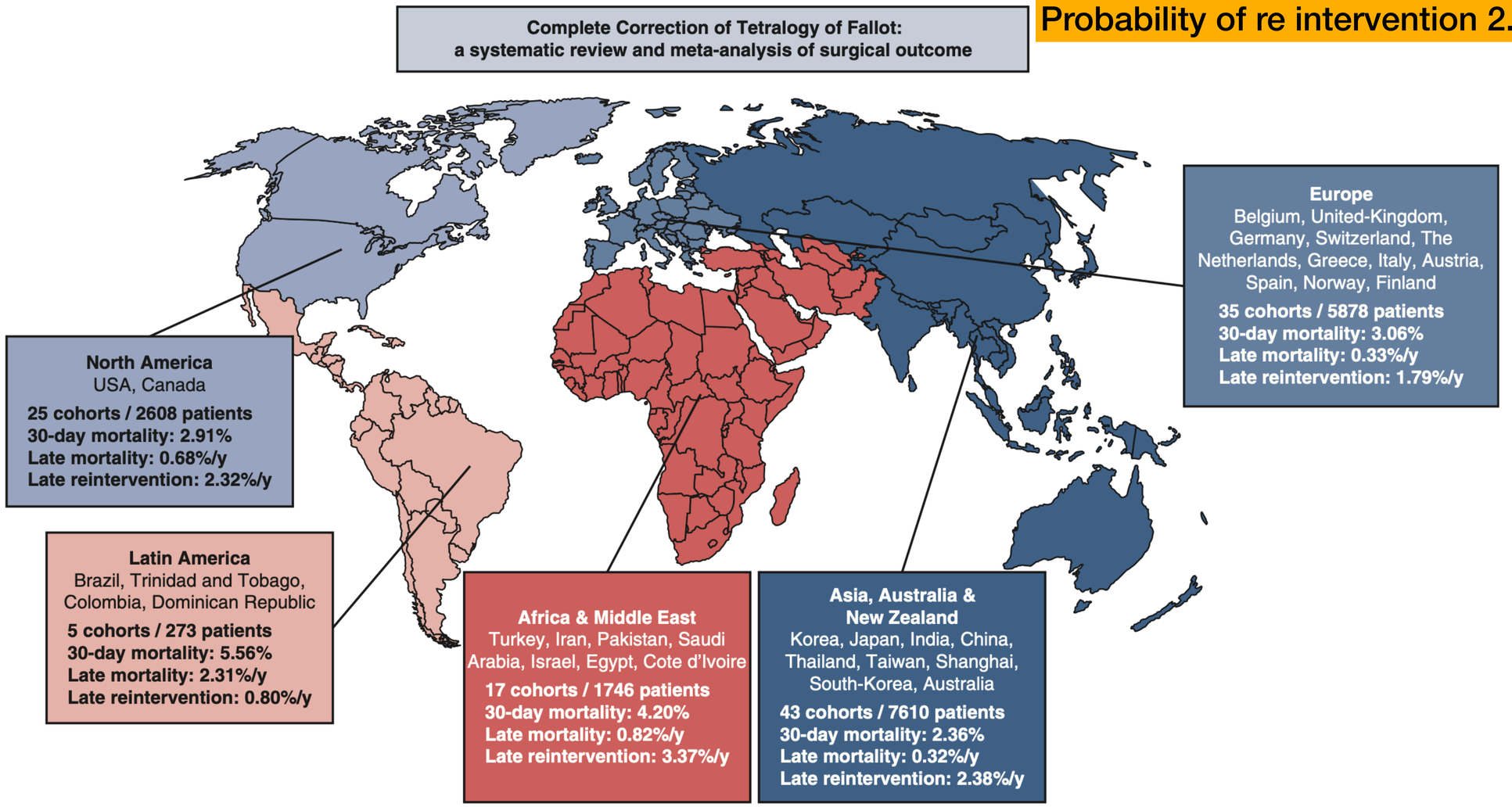


Outcome after surgical repair of tetralogy of Fallot: A systematic review and meta-analysis

J Thorac Cardiovasc Surg 2020;159:220-36)

Jamie L. R. Romeo, MD, MSc,[a] Jonathan R. G. Etnel, MD,[a] Johanna J. M. Takkenberg, MD, PhD,[a] Jolien W. Roos-Hesselink, MD, PhD,[b] Wim A. Helbing, MD, PhD,[c,d] Pieter van de Woestijne, MD,[a] Ad J. J. C. Bogers, MD, PhD,[a] and M. Mostafa Mokhles, MD, PhD[a]

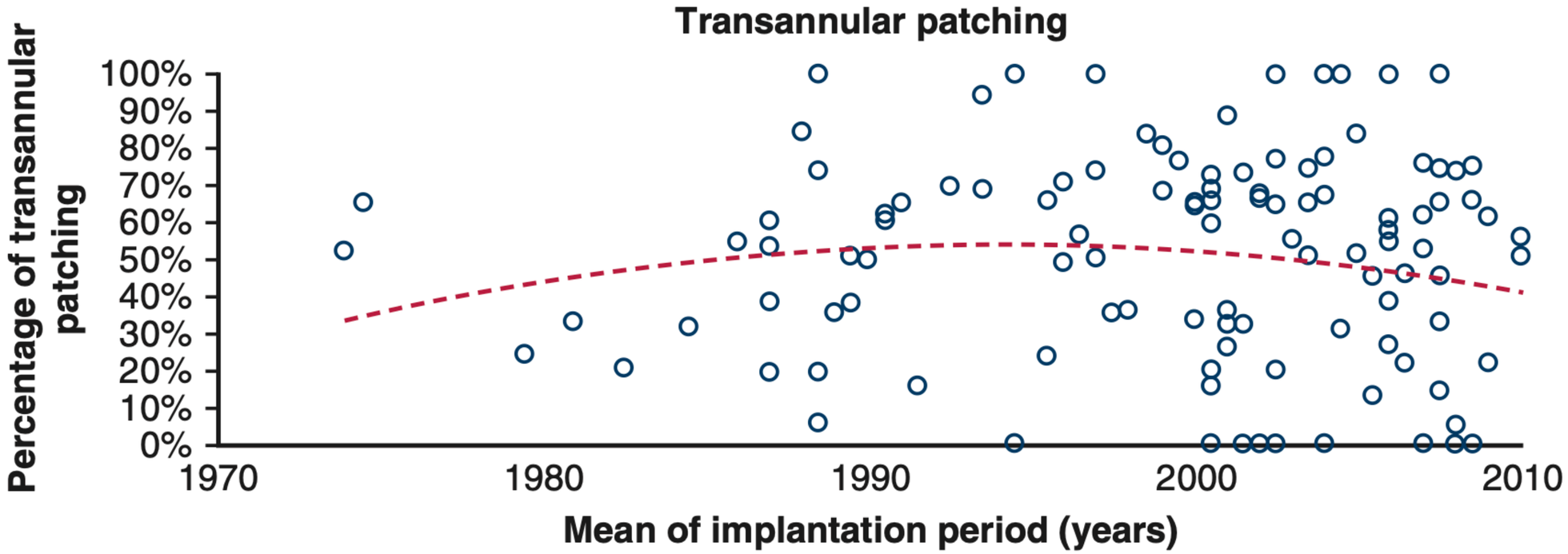
Probability of death 0.42% per year
Probability of re intervention 2.26% per year



Trends

J Thorac Cardiovasc Surg 2020;159:220-36)

TAP almost 50%, decreasing after 2000



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(Ann Thorac Surg 2023;116:768-77)

Trends

STS 2020 harvest—10,727 patients, 2010 to 2020

Contemporary Patterns of Care in Tetralogy of Fallot: Analysis of The Society of Thoracic Surgeons Data

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• ToF repair - No ventriculotomy	24.8%
• ToF repair - Ventriculotomy, Annulus intact	29.1%
• ToF repair - Transannular patch	44.6%

Truth table - how exactly will annulus sparing help?

TOF long term	***Mortalty***	***Reintervention (RVOT)***
Early (first few years)	**TAP > Non TAP (?Poor Subset)**	**Non TAP > TAP (RVOTO)**
Late (>15 years)	**TAP = Non TAP**	**TAP > Non TAP (PVR)**
SUDDEN CARDIAC DEATH unrelated to RVOT procedure ?? RV myocardial damage—RV Dysfn, LV Dysfn		

Counter-point:
Close surveillance
And timely PVR
-Low risk

Take home message

There is limit to annulus preservation, approx 60% can be preserved

- Be prepared for Redo RVOTO if you are going to be aggressive with annulus preservation

- With proper techniques annulus preservation rates can be increased
    - A complete and aggressive resection will achieve a spacious RVOT but has to balanced with the myocardial damage
    - Agressive pulmonary valvotomy will lead to higher annulus preservation but will also lead to progressive PR
- The modern annulus preservation cohorts are in the “casks” now, long term results awaited