

# Morphology – Imaging – Surgical Correlates: Ebstein's Anomaly

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Hospital for Sick Children

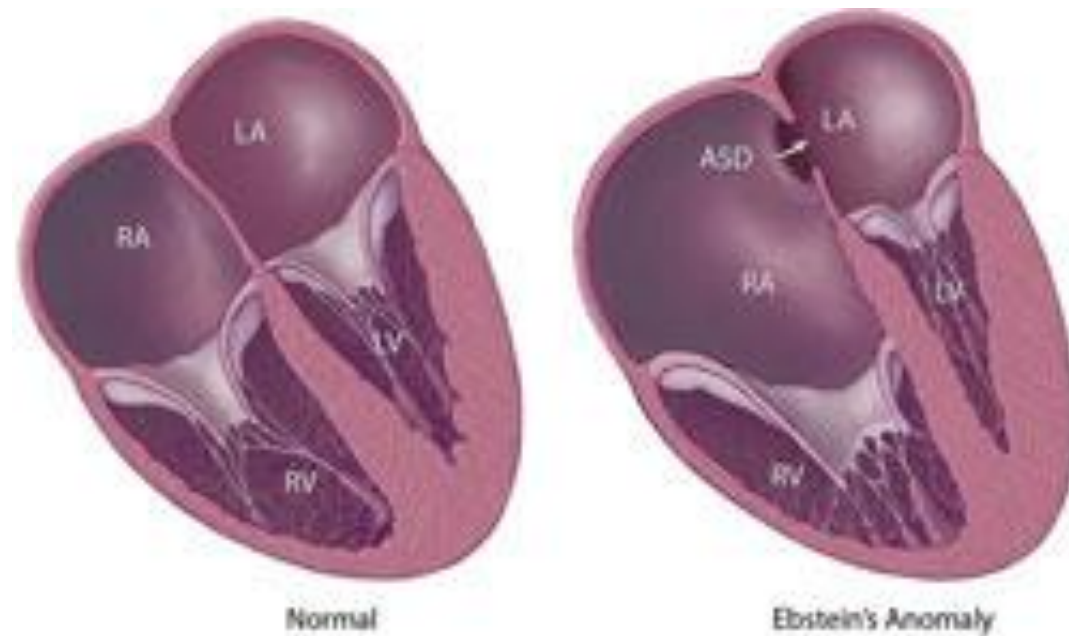
The 2<sup>nd</sup> Contemporary Morphology Course with Specimens and 3D Print Models

## CONGENITAL HEART DISEASES IN YOUR HANDS

♥ Abnormalities of the Atrioventricular Junction ♥



# Ebstein's Anomaly Of the Tricuspid Valve

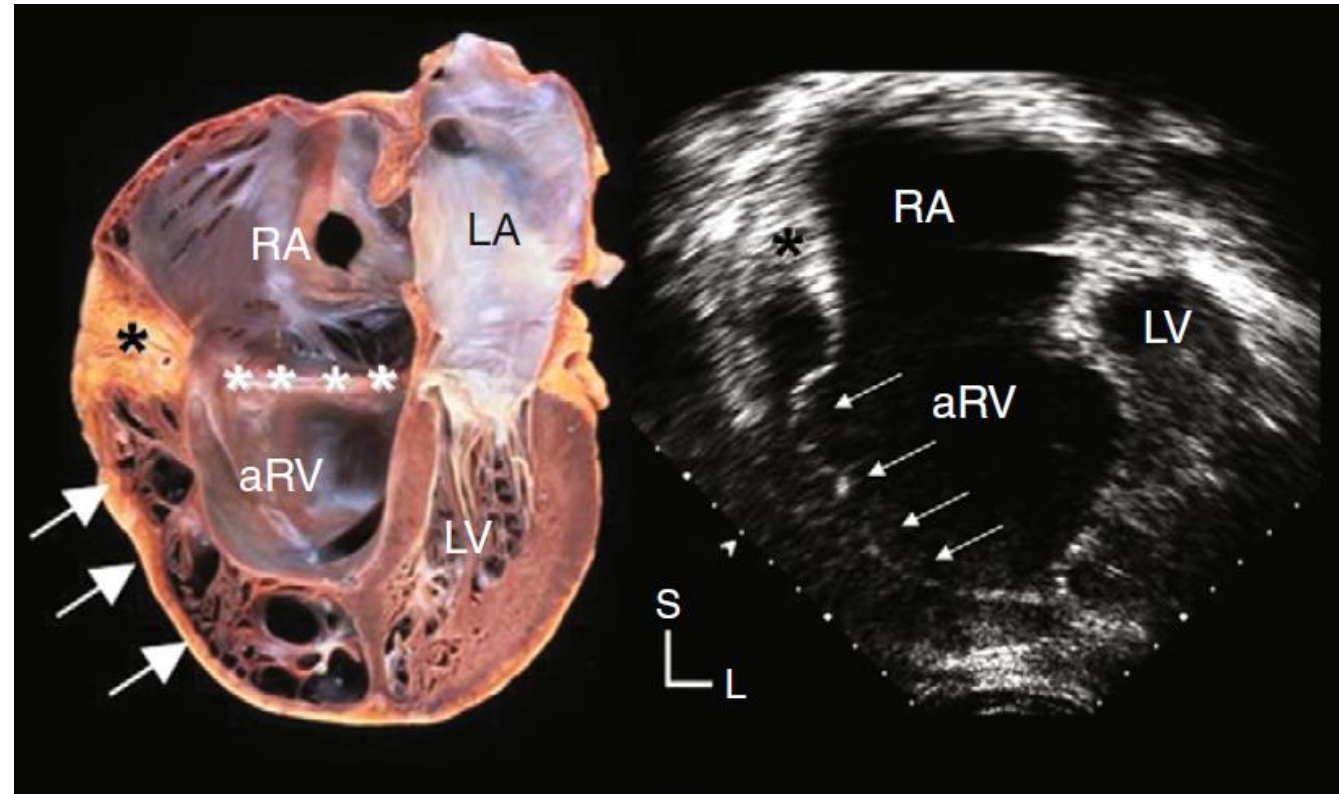


*Wilhelm Ebstein 1866*

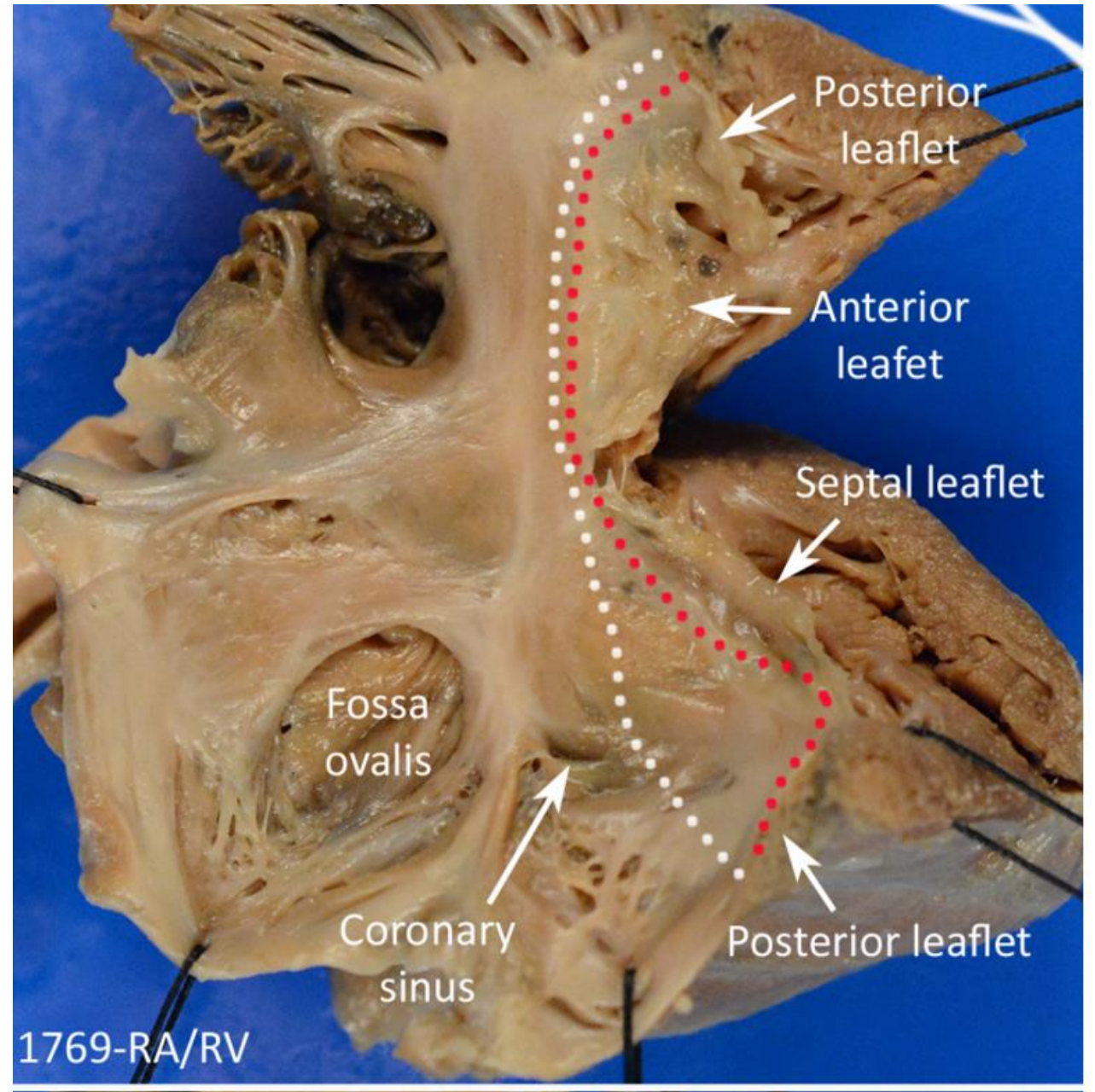
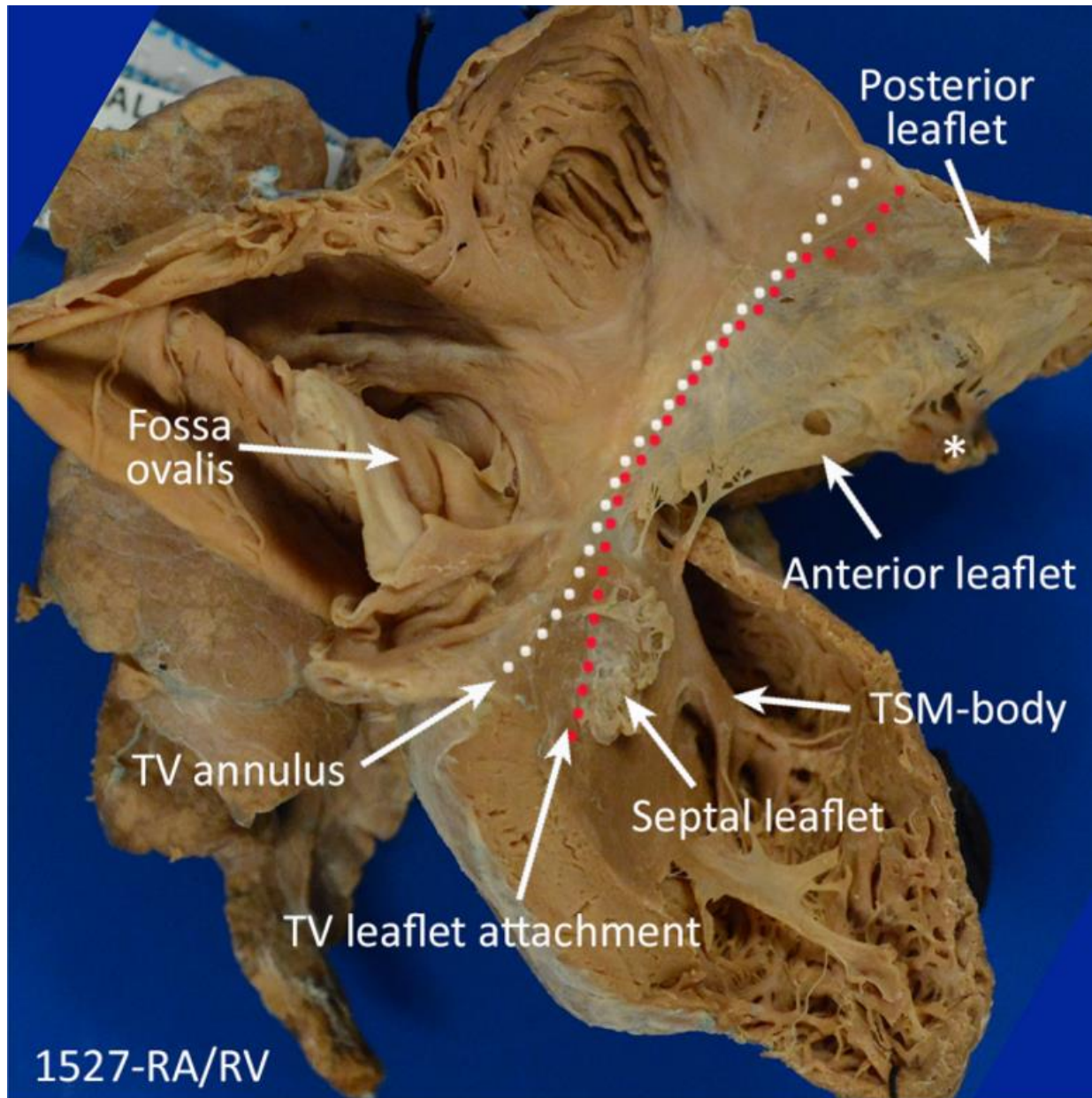
**Downward displacement of the septal leaflet of the tricuspid valve**

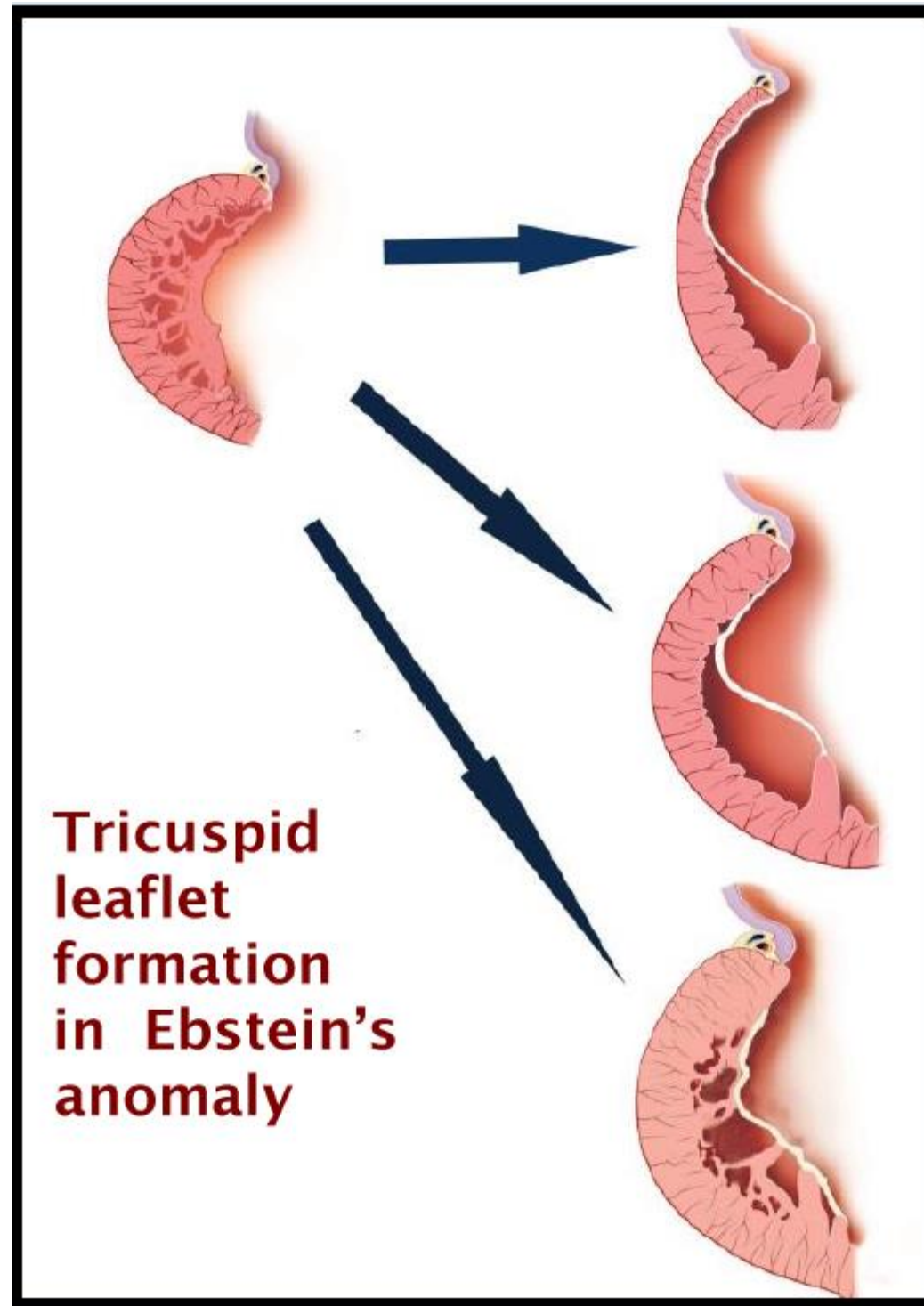
**‘Atrialisation’ of the inferior wall of the right ventricle**

# Ebstein's Anomaly Of the Right Ventricle

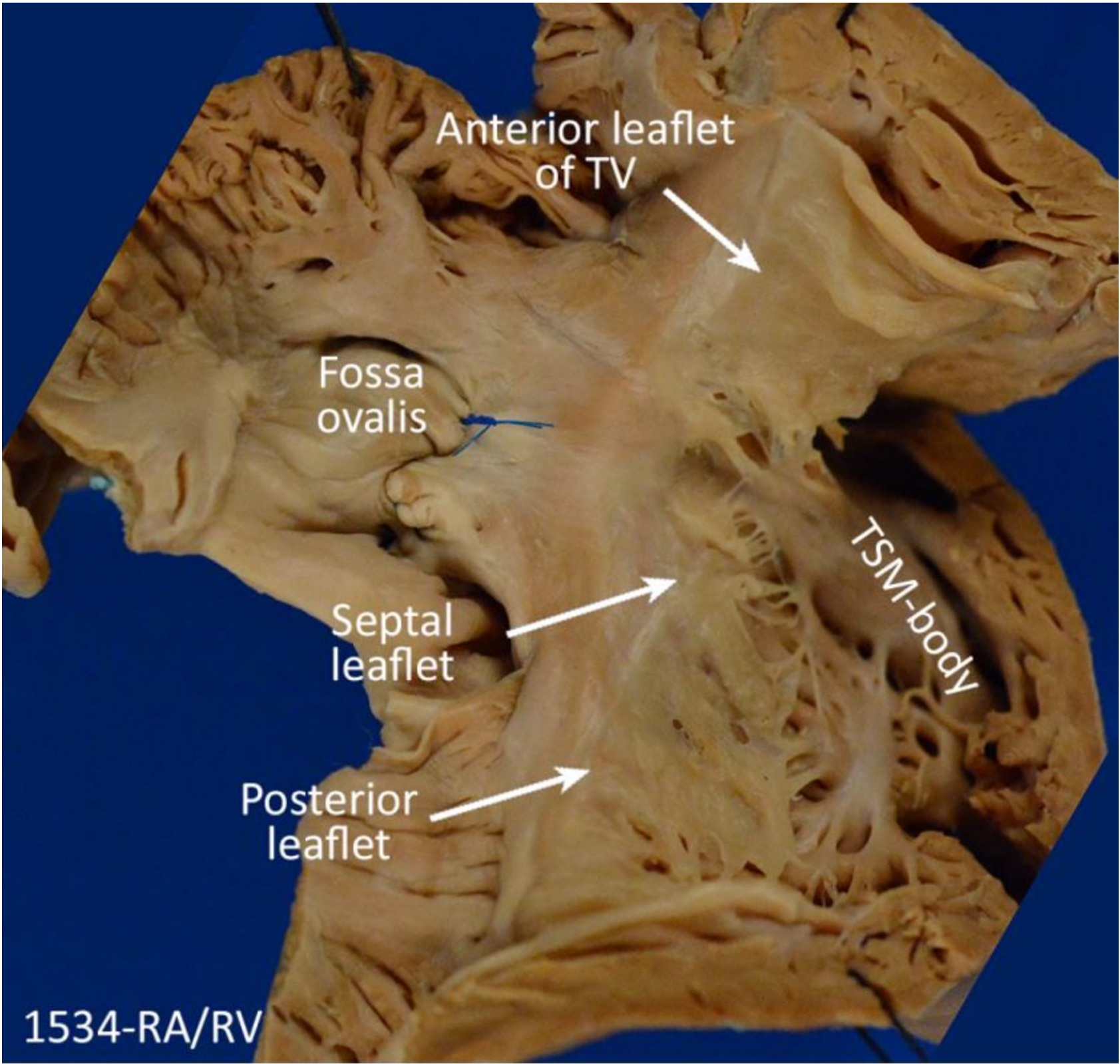


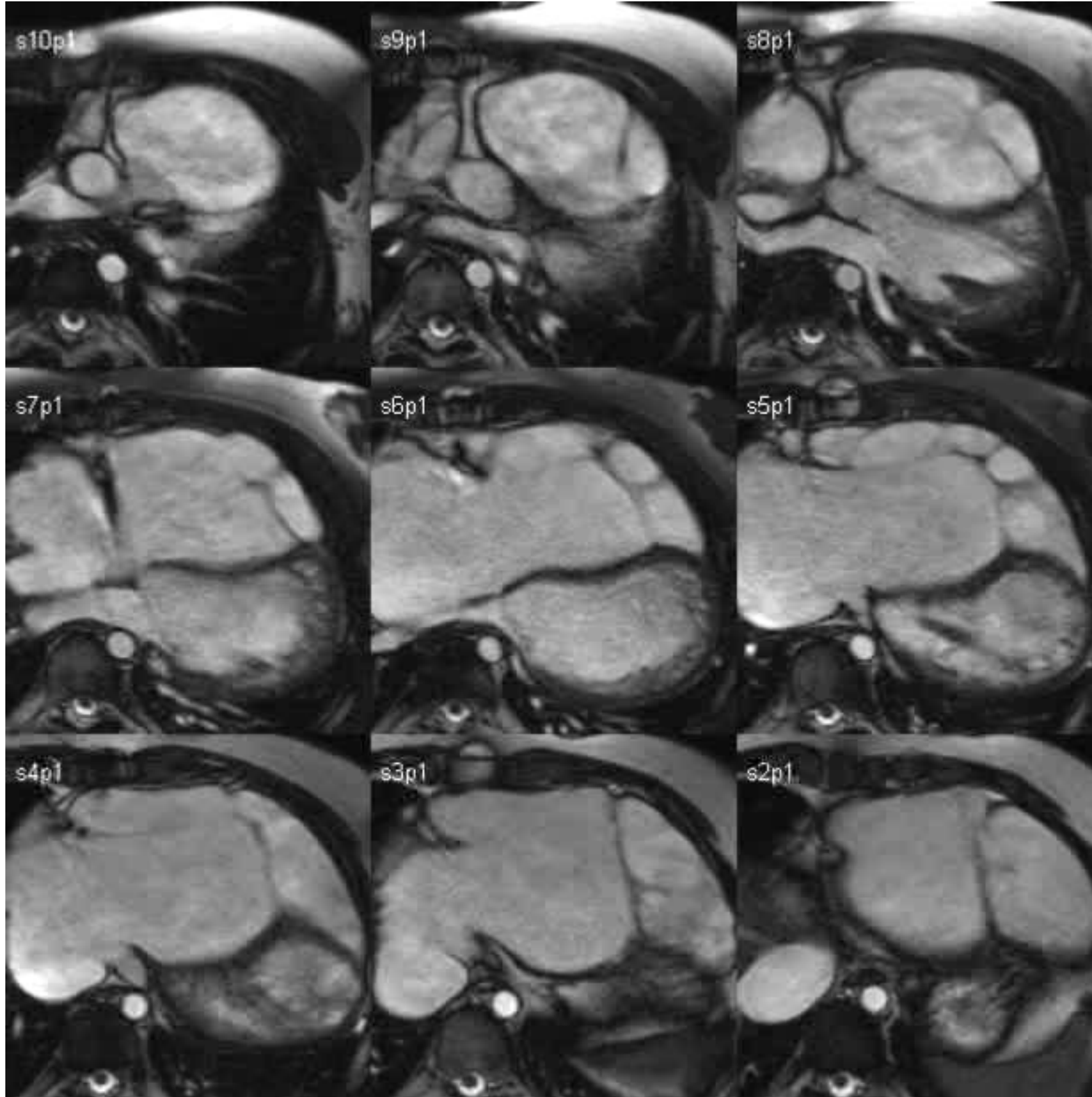
- . An abnormality that affects the whole right ventricle and its myocardium
- . Characterised by 'failed delamination' of the septal and posterior leaflets
- . A 'spiral' defect of the tricuspid apparatus
- . Anterior leaflet: variable in its degree of dysplasia, hinge point remains at the AV-junction. Usually retains many normal characteristics.



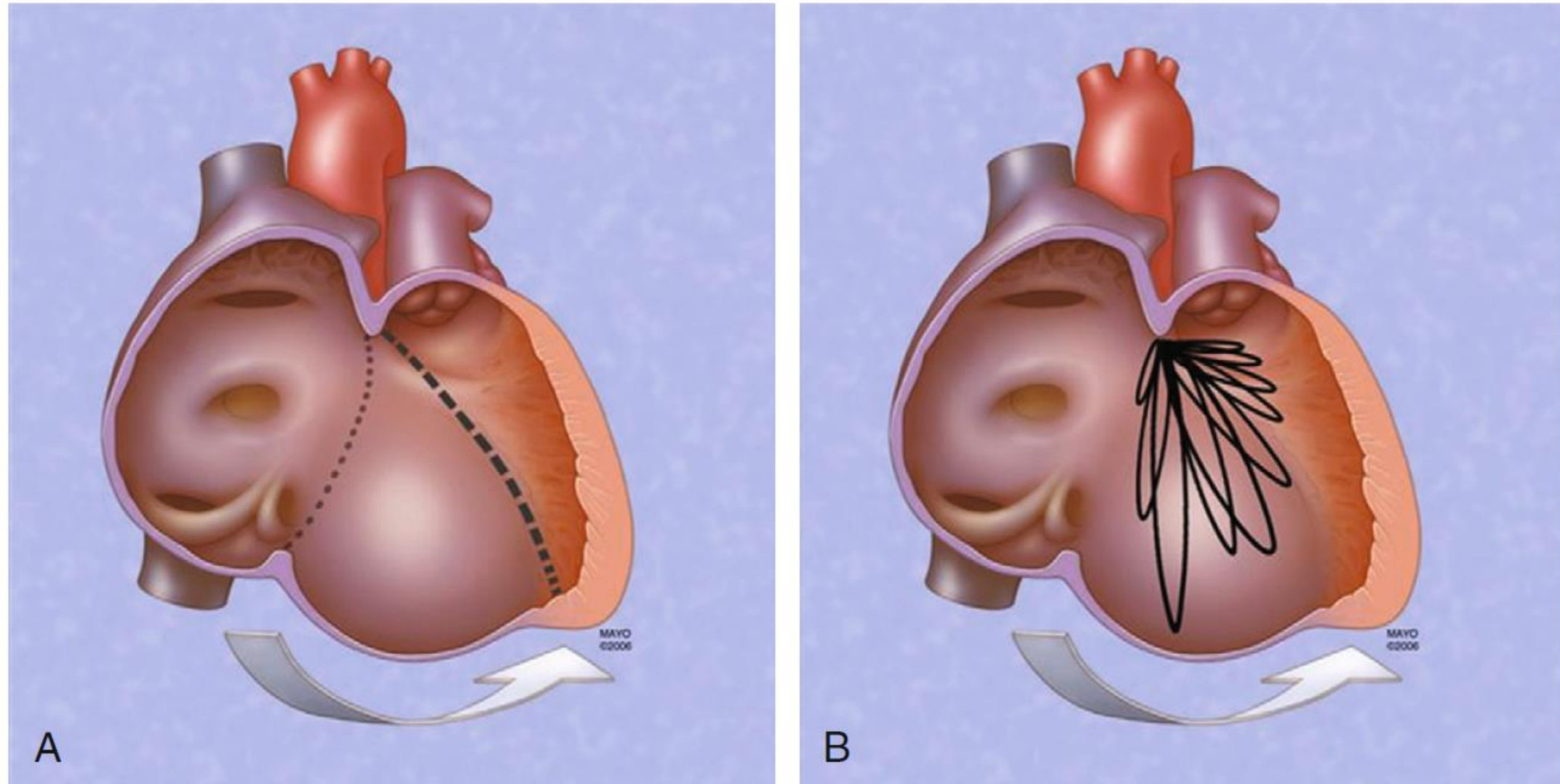


Van Mierop, Prog Card Dis, 15:67, 1972





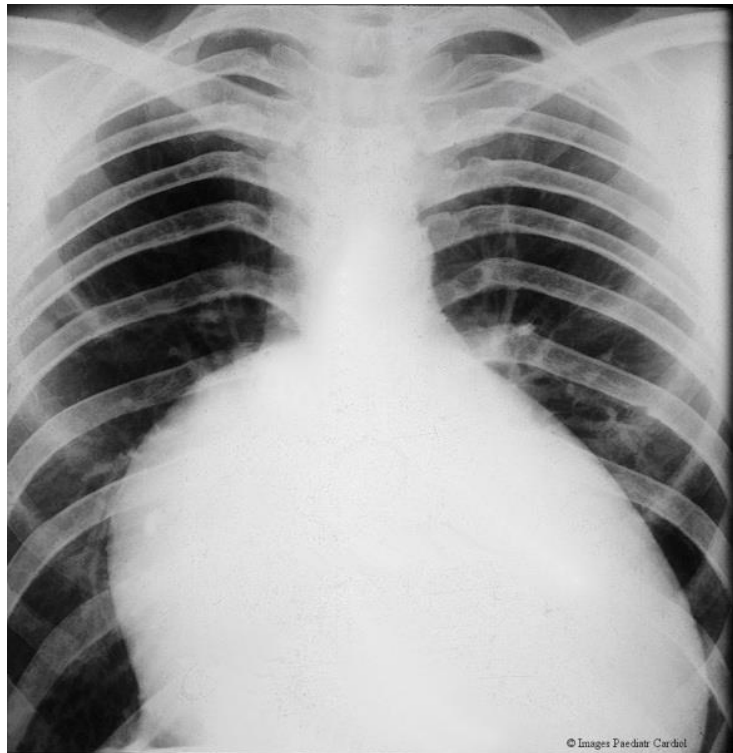
# Ebstein's Anomaly



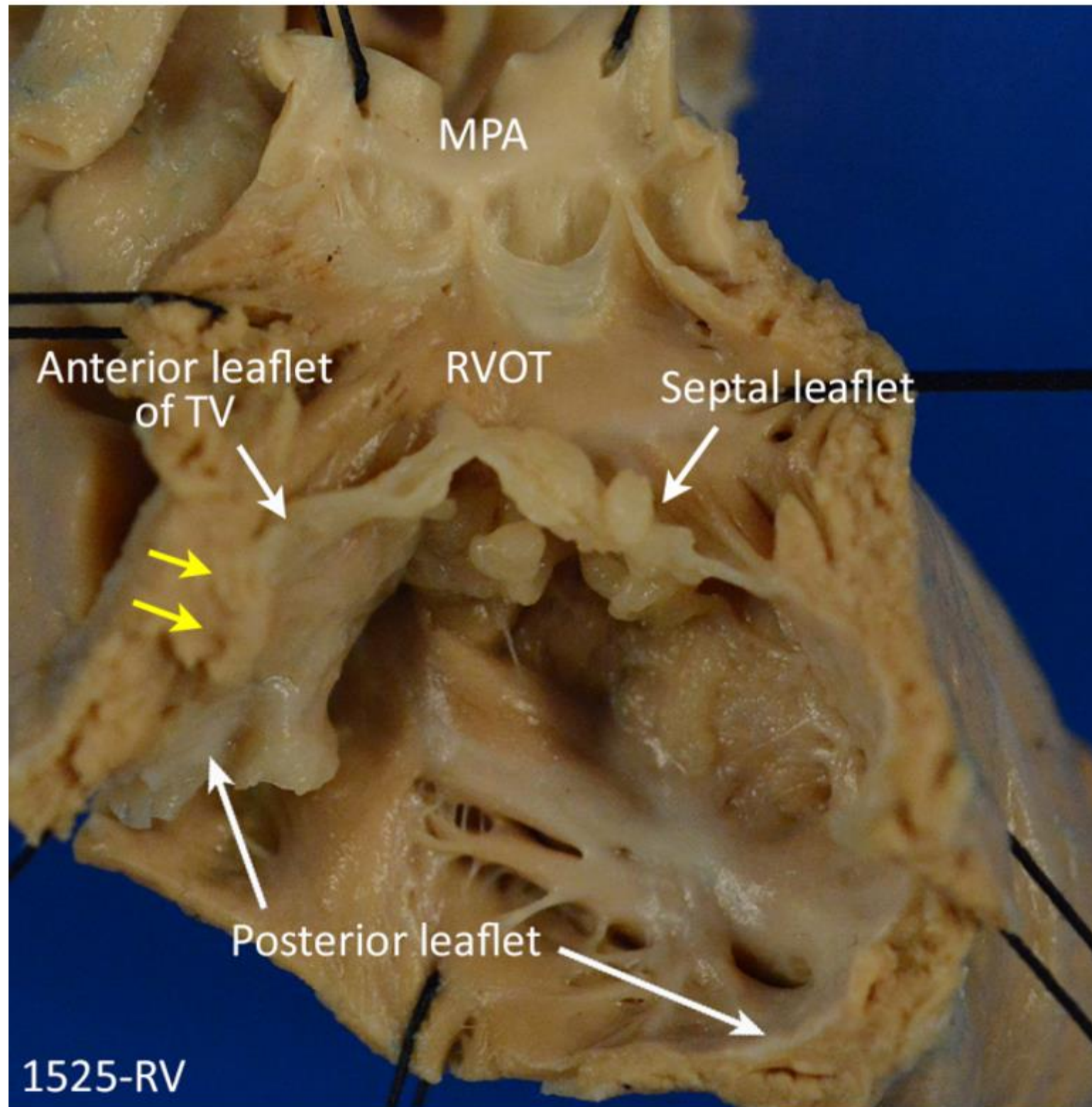
**Spectrum of disease**  
**Both in the degree of displacement**  
**the degree of leaflet dysplasia, chordal dysplasia and failed**  
**delamination**



# The Paradox of Size of the Right Ventricle



**Is the true RV too small or too big?**



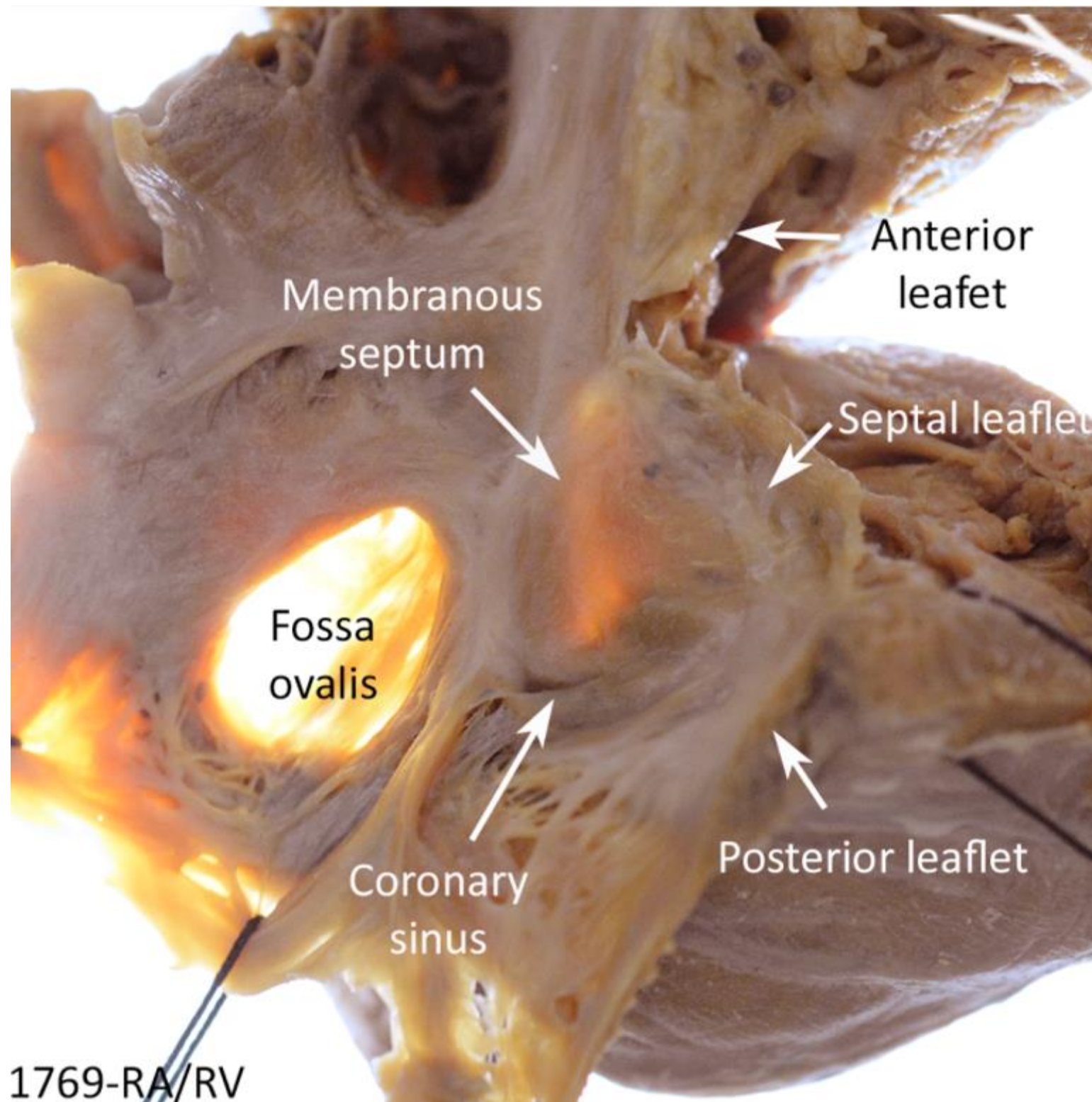
# Associated Lesions

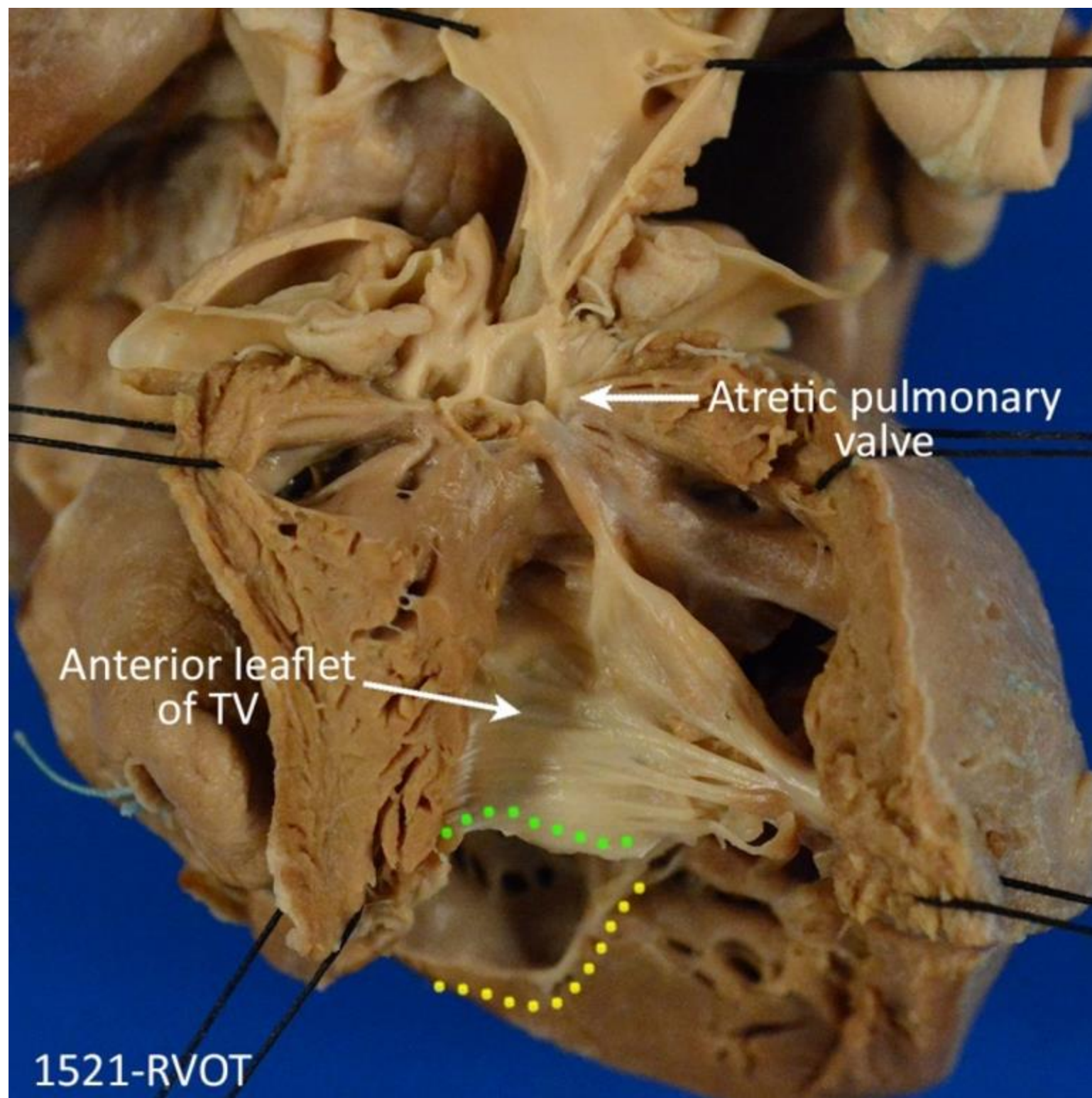
**ASD/PFO 70-80%**

**VSD Uncommon  
Can be ABOVE the septal leaflet**

**Pulmonary Atresia – can be anatomical or functional**

**Hypoplasia of Pulmonary Arteries and Lung  
hypoplasia (usually in severe neonatal presentation)**





Anterior leaflet  
of TV

Atretic pulmonary  
valve

1521-RVOT

# **Clinical Correlates: Assessing the Severity of Disease**

## **Spectrum**

**Neonatal presentation (minority 6-10%)**

**..but a common cause of hydrops and fetal loss**

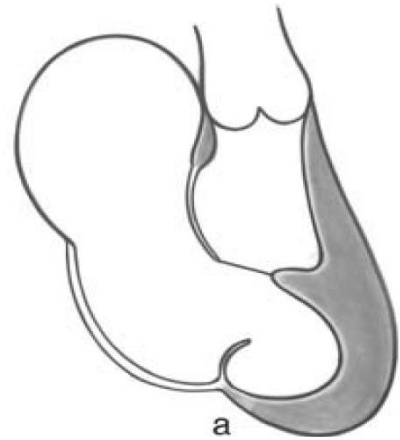
**Extreme cases – Fontan circulation**

**Majority of clinical management:  
*Elective intervention in stable adolescents/adults***

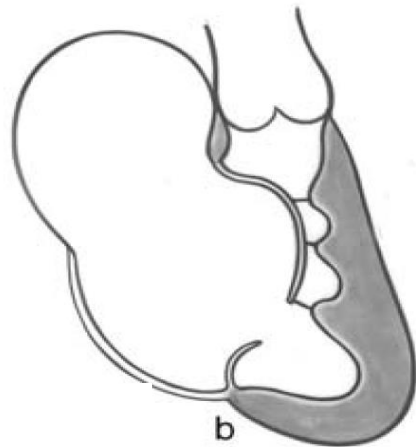
**Timing and indications for surgery**

# Carpentier Classification

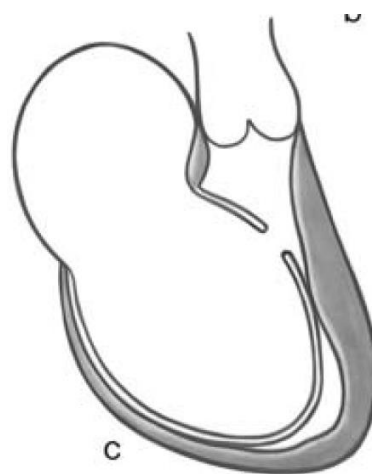
## Focused on the Anterior Leaflet



**Type a: displacement of the septal leaflet. The anterior leaflet is large, free and has well formed chordae and free edge**



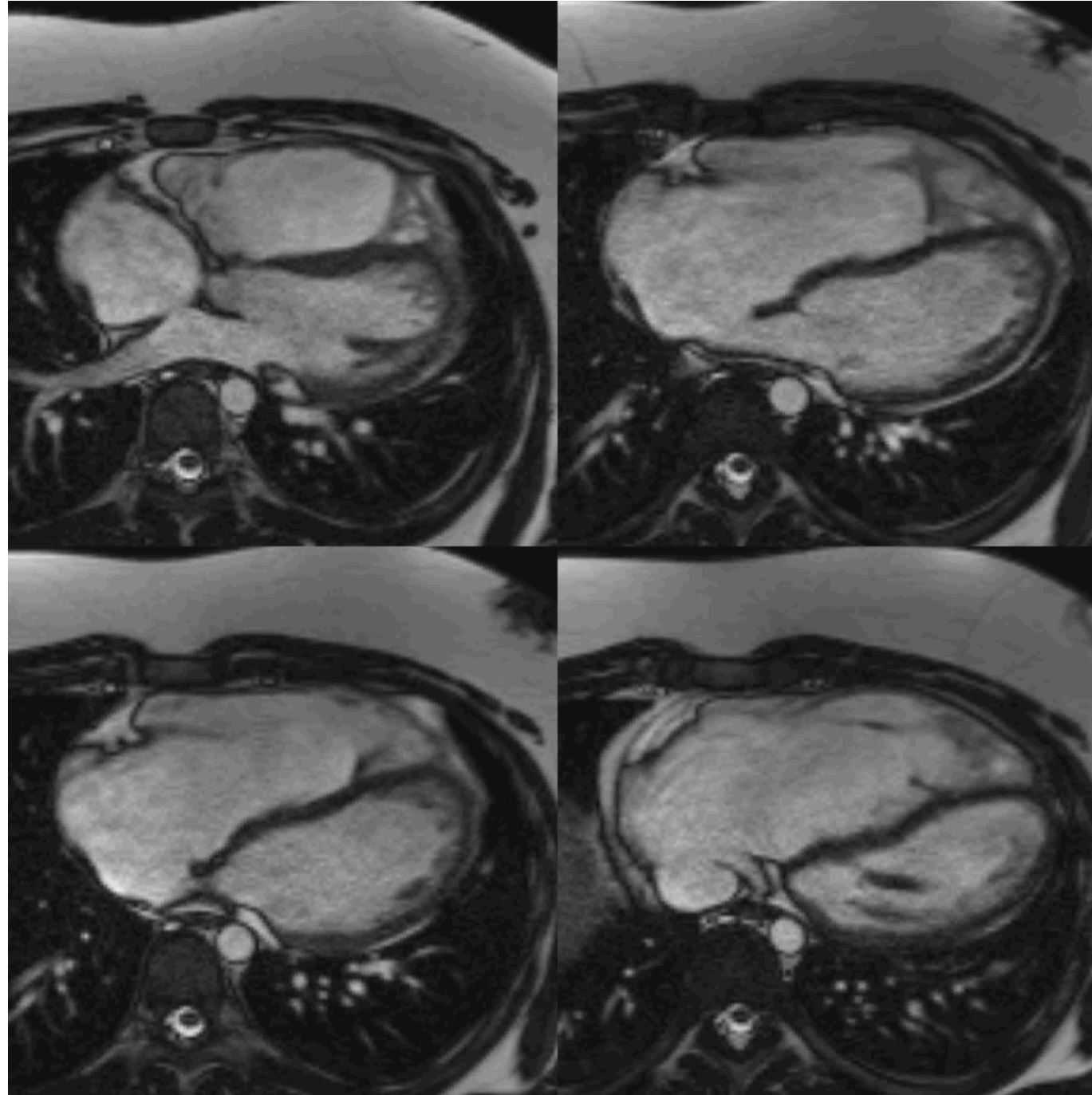
**Type b: The anterior leaflet is abnormal, restricted motion, tethering to the ventricular wall and less well defined commissures**



**Type c: The anterior leaflet inserts directly into the ventricular wall, loss of inter-chordal spaces, has attachments to the infundibulum, post leaflet fused to the wall.**

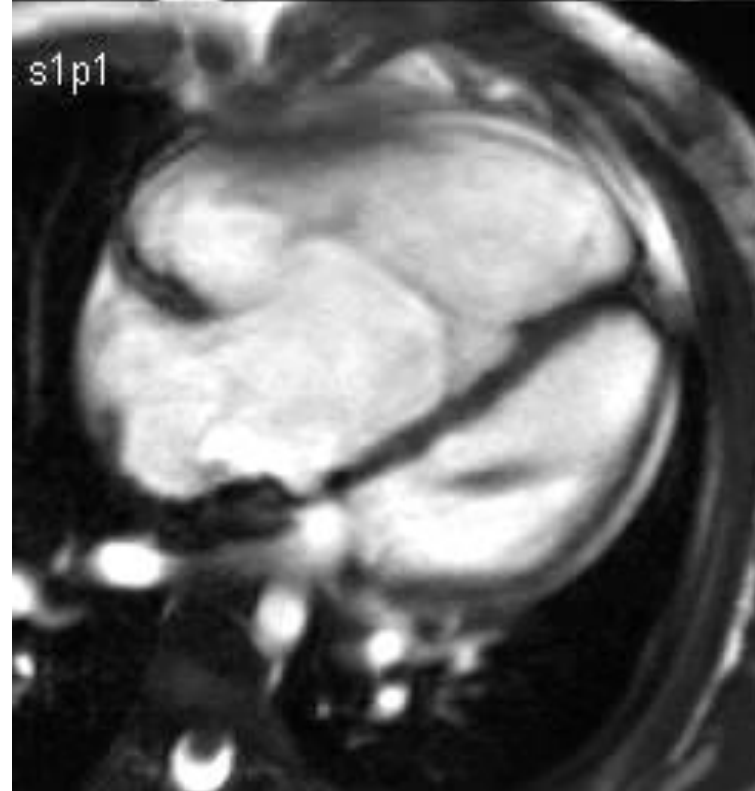
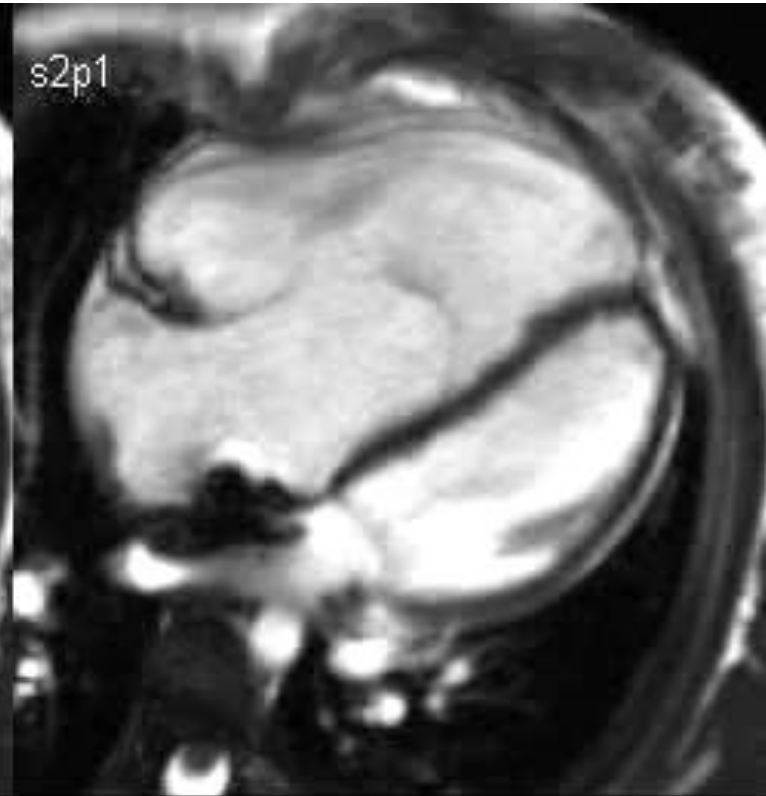
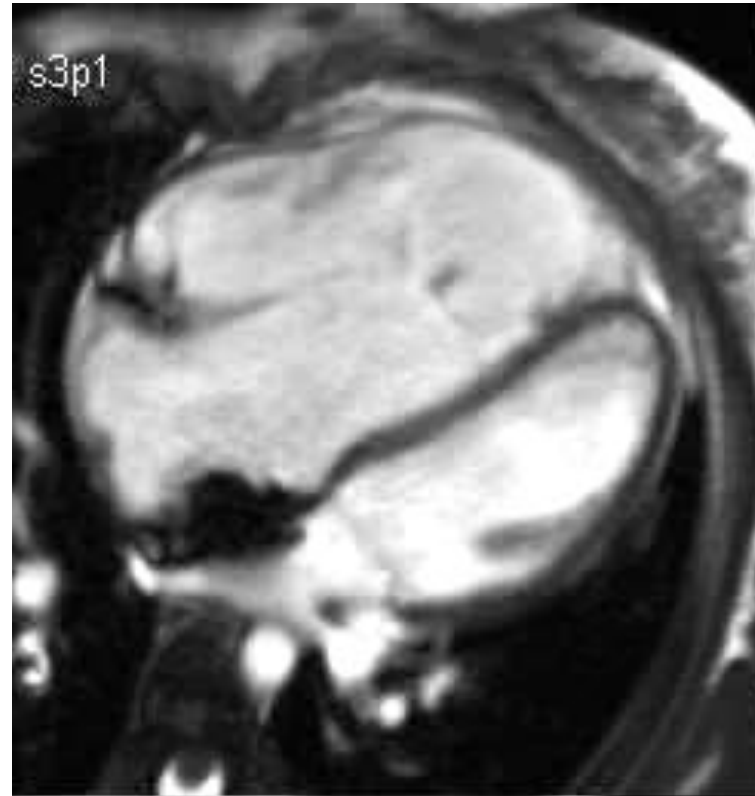
**Type d: Severe failed delamination of post and septal leaflets, ant leaflet inserted into the infundibulum, valve orifice effectively in the infundibulum**

# Type a

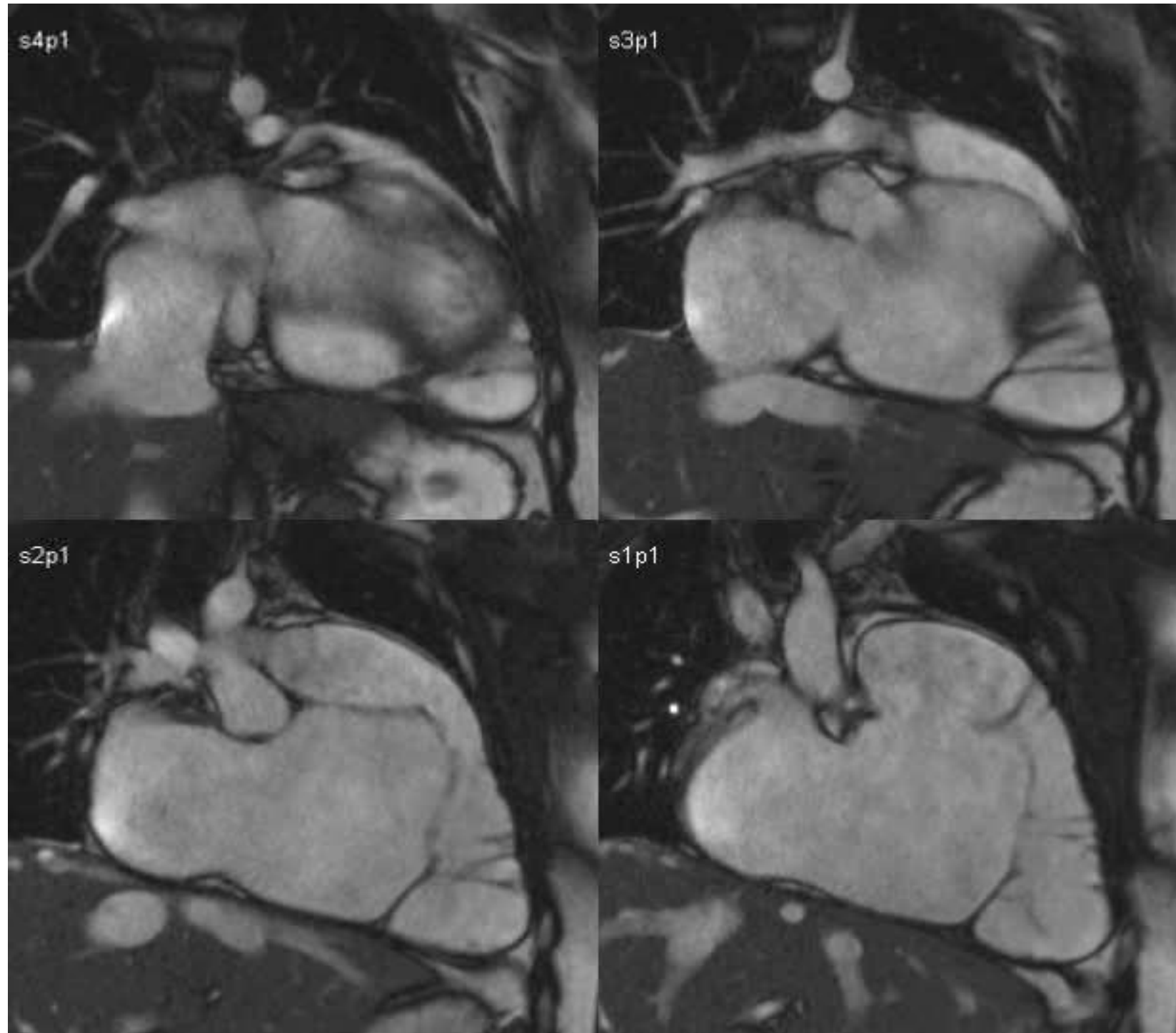




# Type b

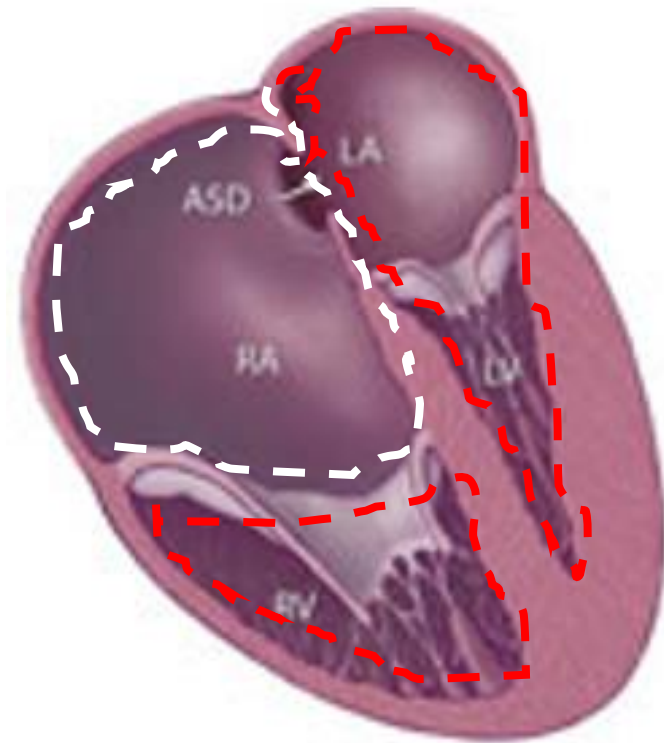


# Type c



# Celemajor Index

Based on 4-chamber echo view  
Now outdated



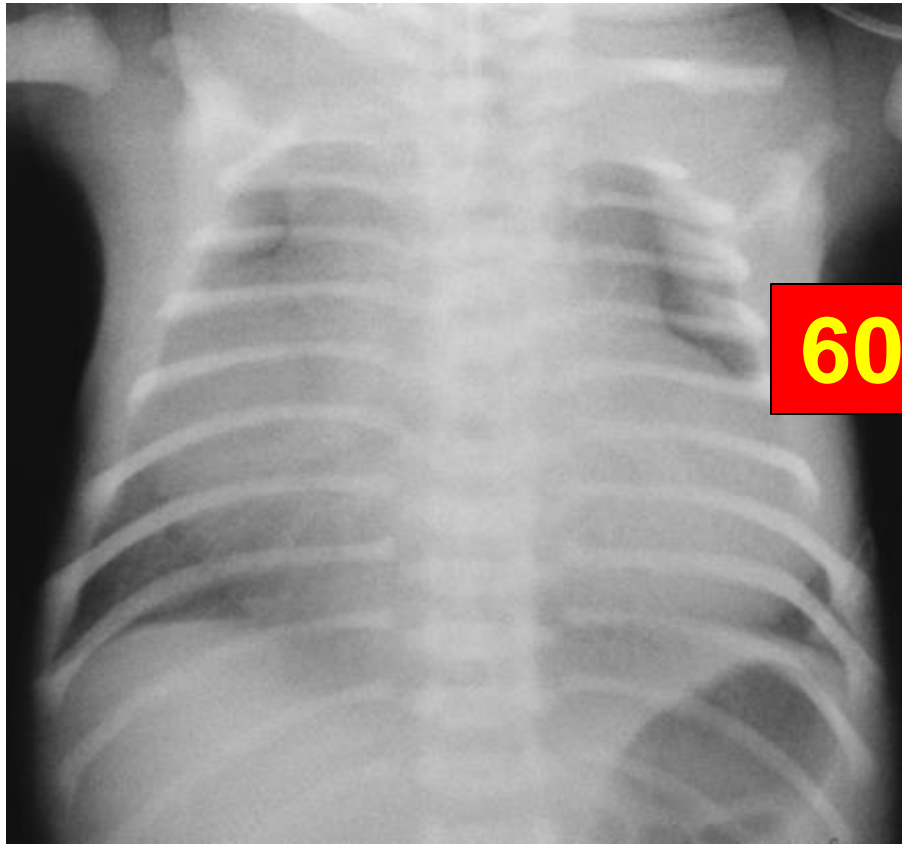
$$CI = \frac{RA + \text{Atrialised RV}}{LA + LV + \text{true RV}}$$

Index	Grade	Observed Mortality
< 0.5	1	5%
0.5 – 1.0	2	10%
1.0-1.5	3	45% *
> 1.5	4	100%

\* If cyanosed mortality 100%

JACC 23: 170, 1994

# Neonatal Ebstein



**Spectrum of disease**

**60% need no intervention (94% survival)**

**Neonates account for only 8.7% of all presentations  
But have the highest operative risk:  
75% in Pan-European Study**

**Neonatal Ebstein**

**Dependent on PGE<sub>1</sub>?**

**No**

**Yes**

**Reason for failure to wean from PGE<sub>1</sub>?**

**Cyanosis**

**Cyanosis and CCF**

**TV repairable TV not repairab**

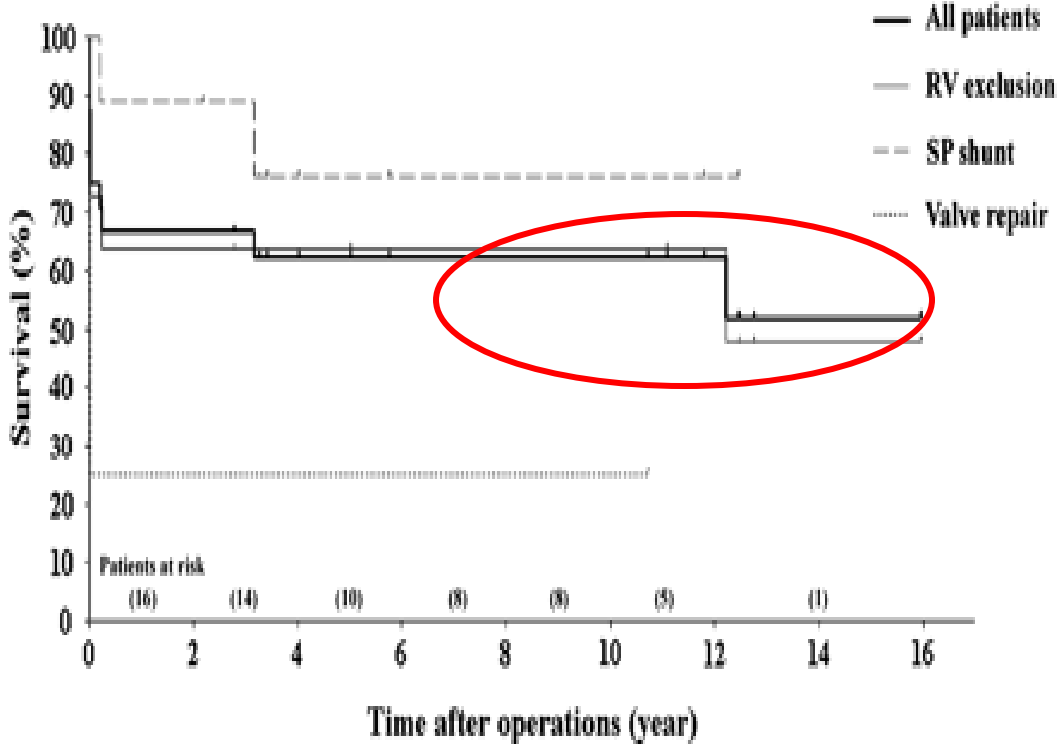
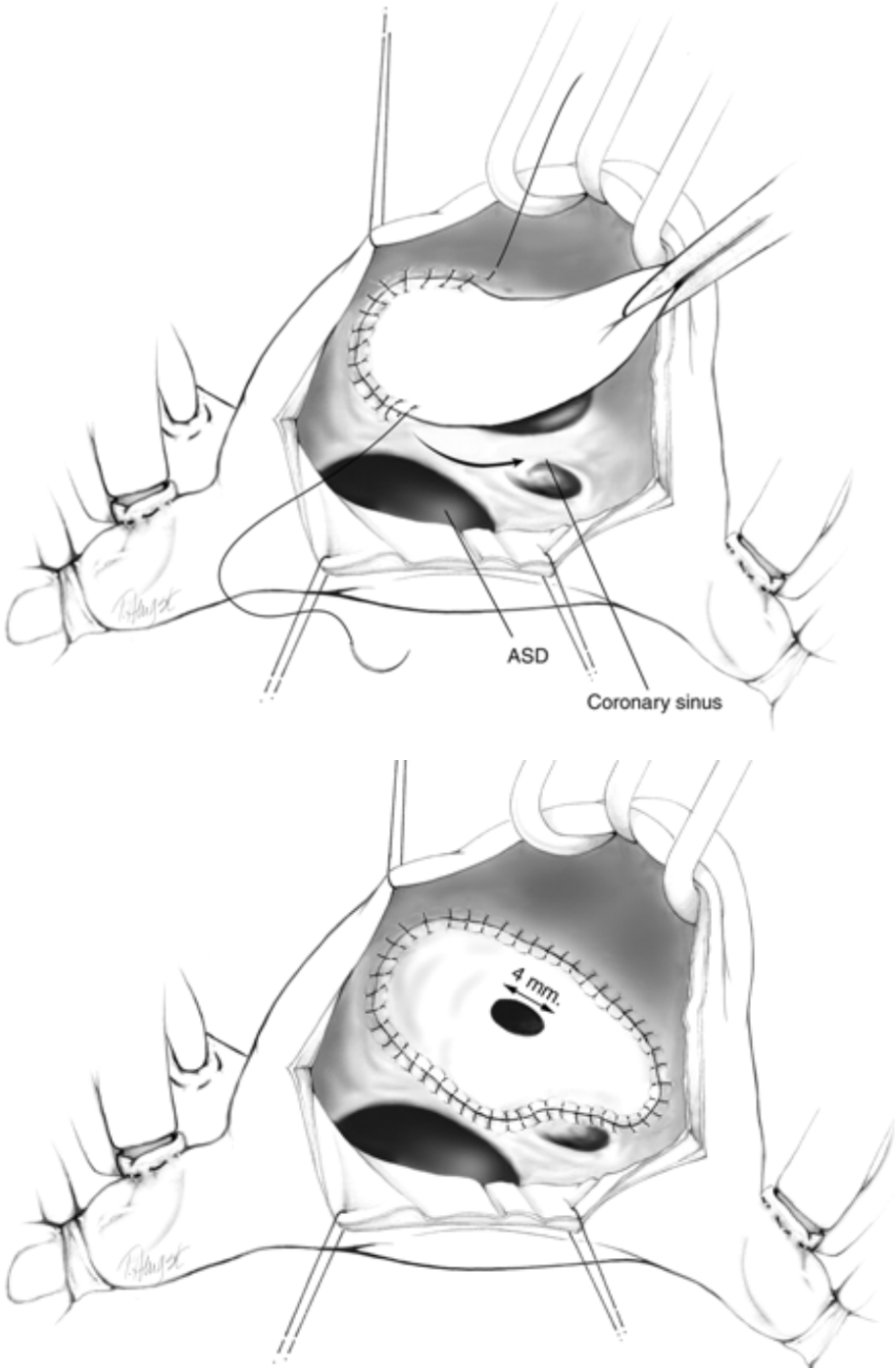
**No Intervention**

**BT Shunt**

**TV repair  
BT shunt**

**RV Exclusion  
BT Shunt**

# Ebstein – RV Exclusion

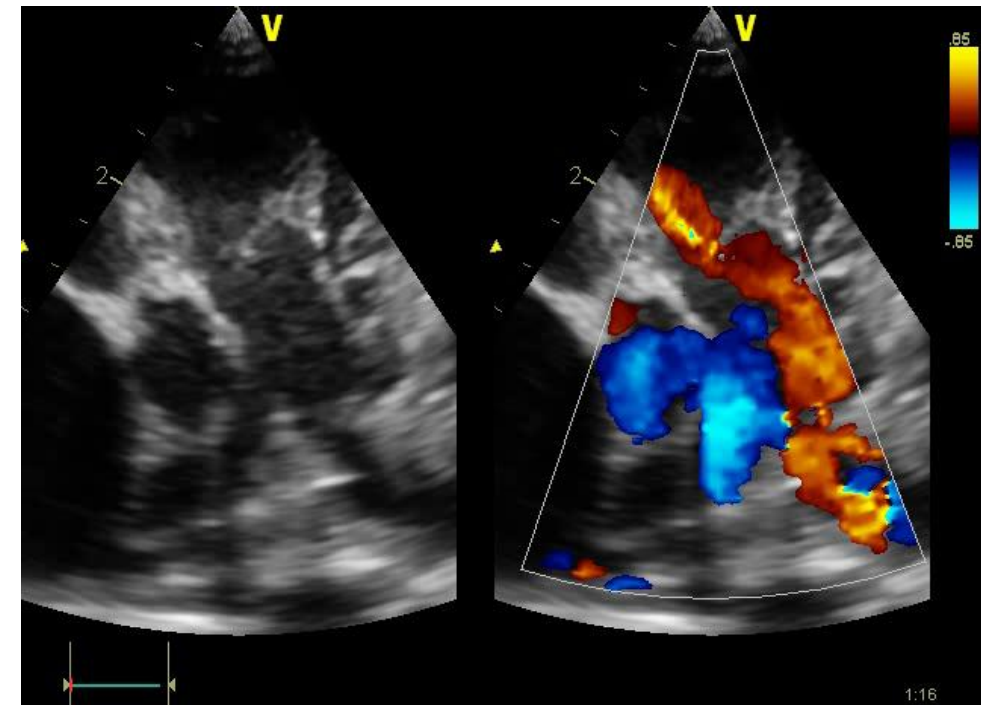
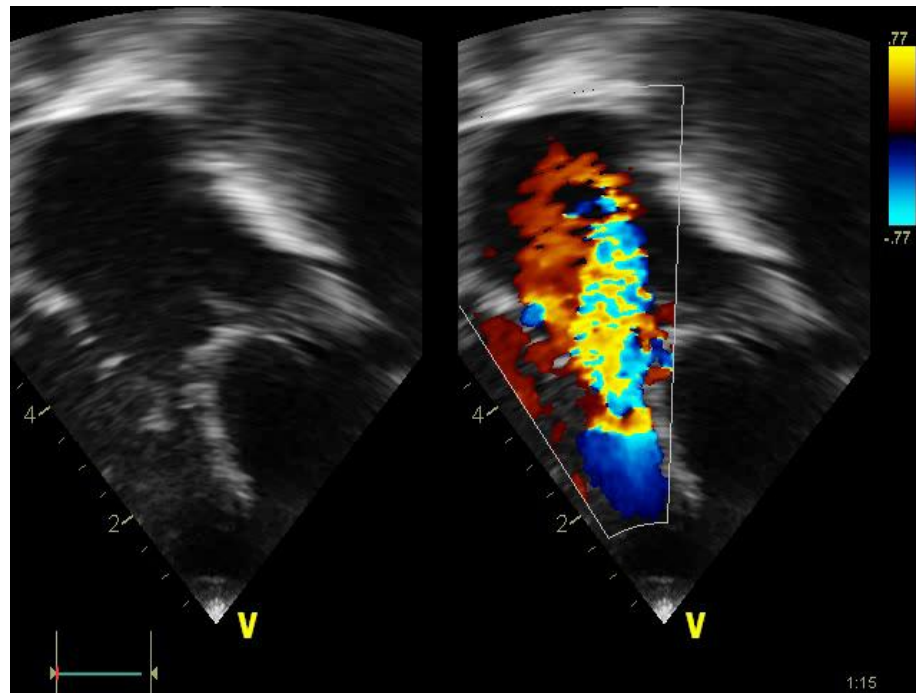
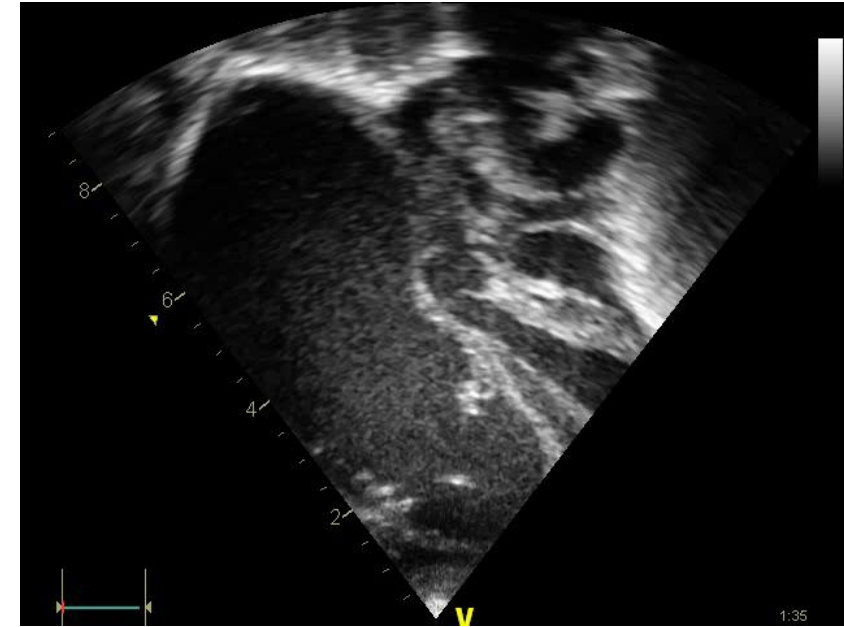
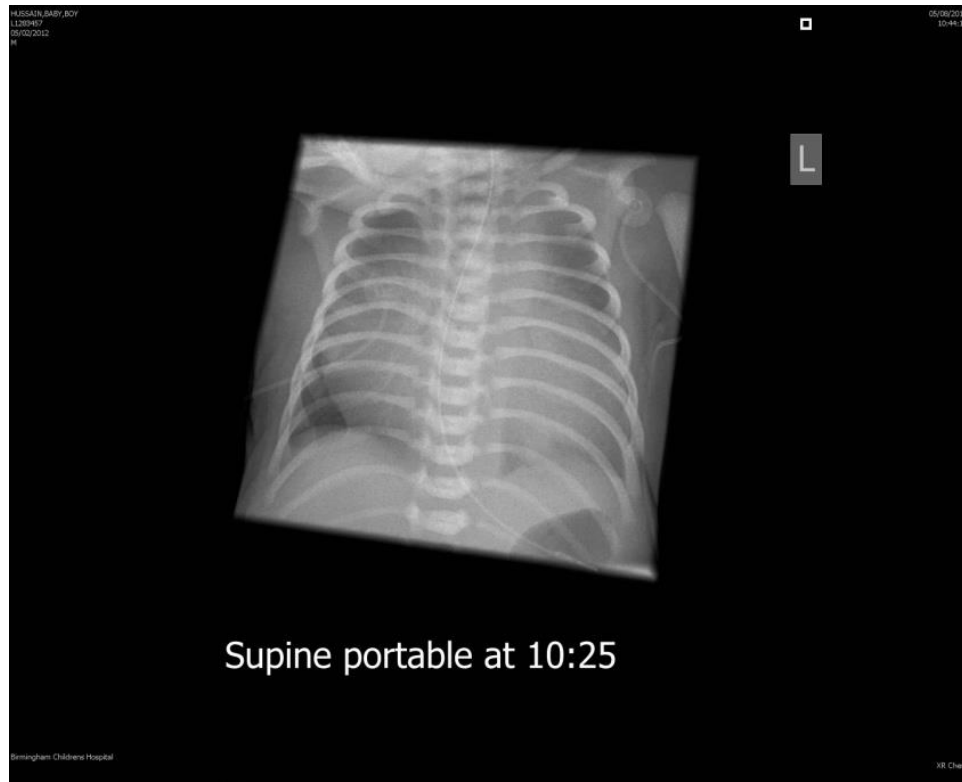


**Ann Arbor**

JTCVS 139: 254 2010

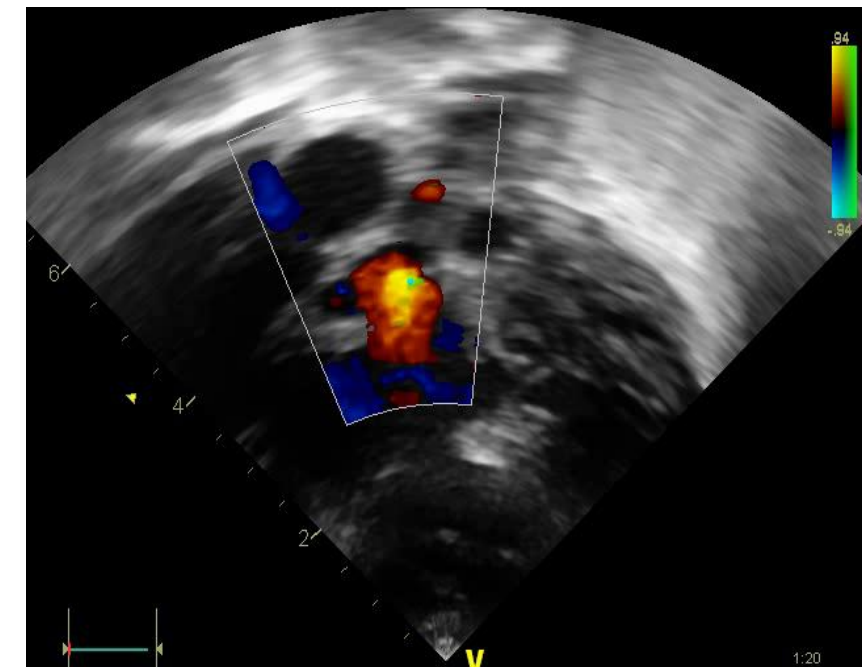
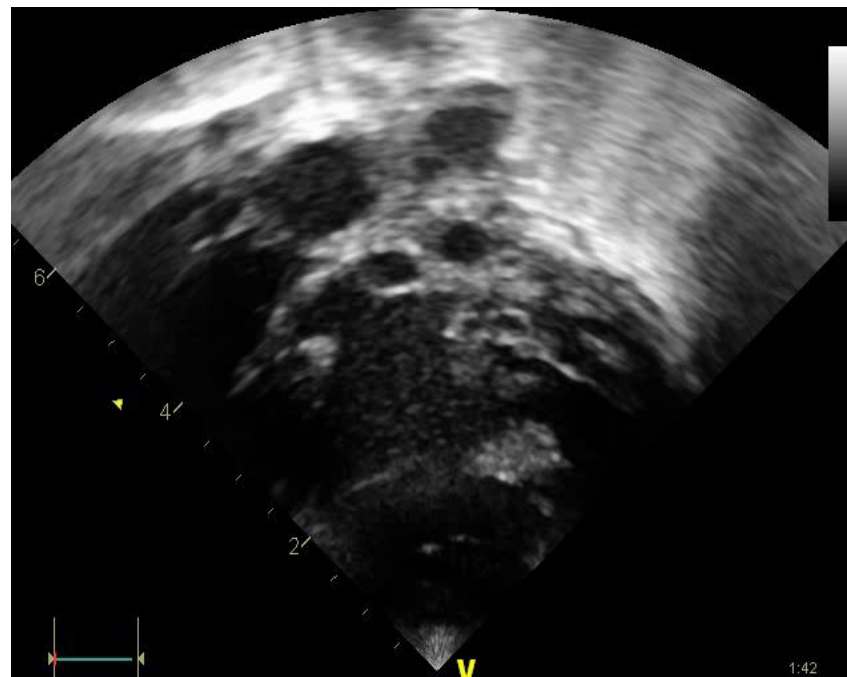
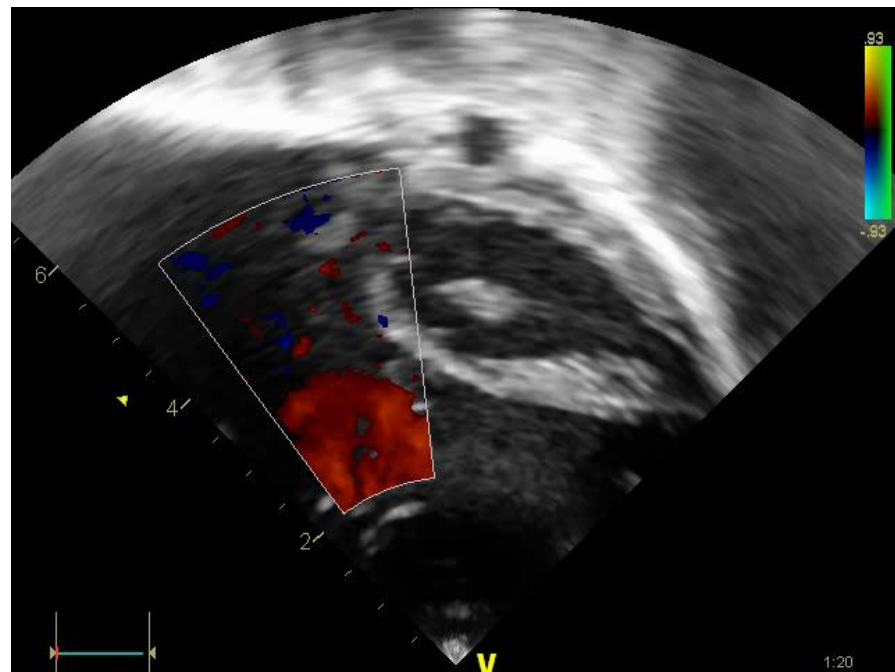
JTCVS 132: 1285, 2005 Vaughan Starnes

# Newborn Ebstein - Ventilated



# PGE<sub>1</sub> Stopped

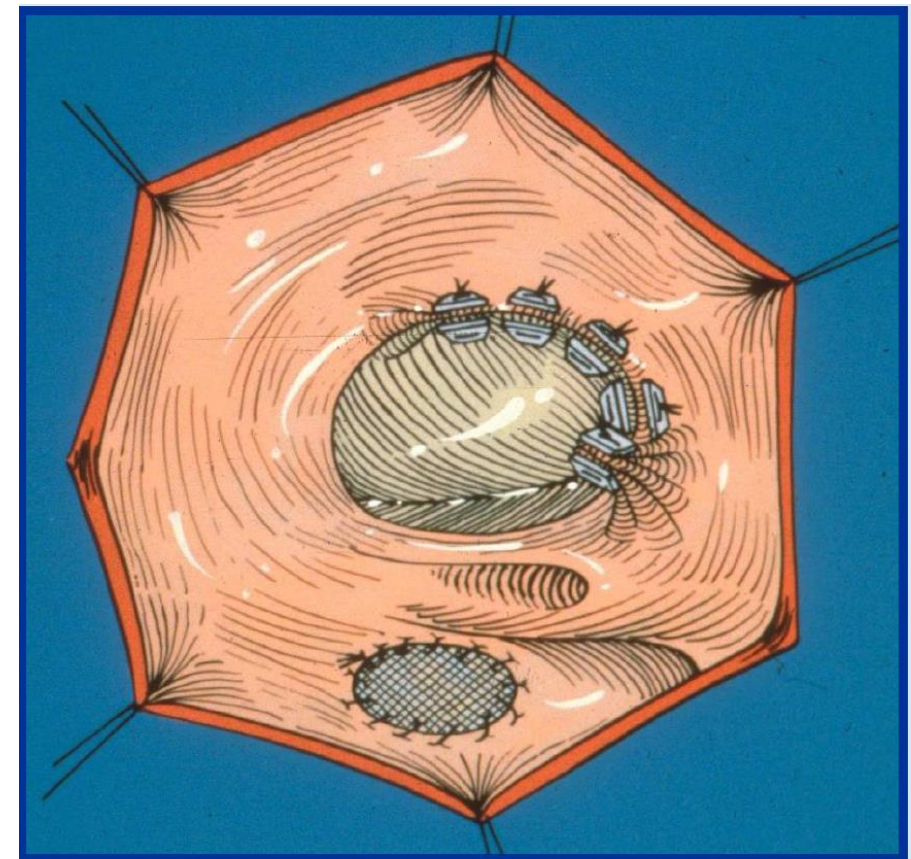
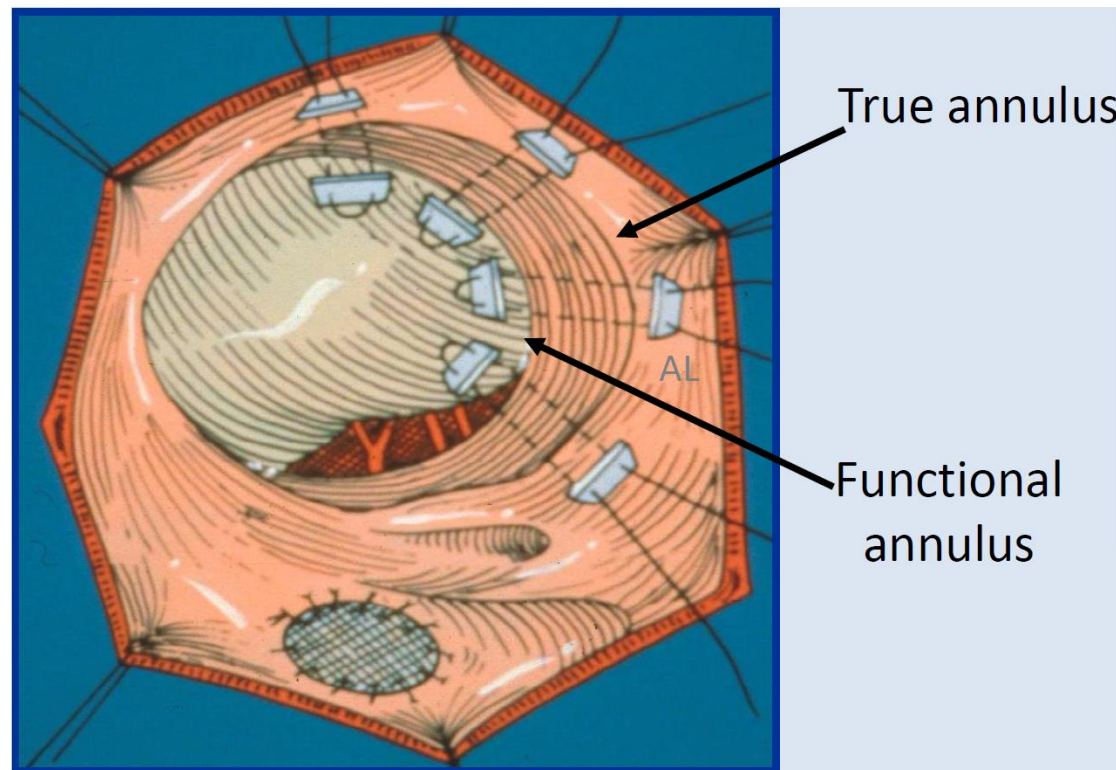
## Echo Five days later





# Traditional Repair Techniques

## Danielson Repair 1970s (Mayo)

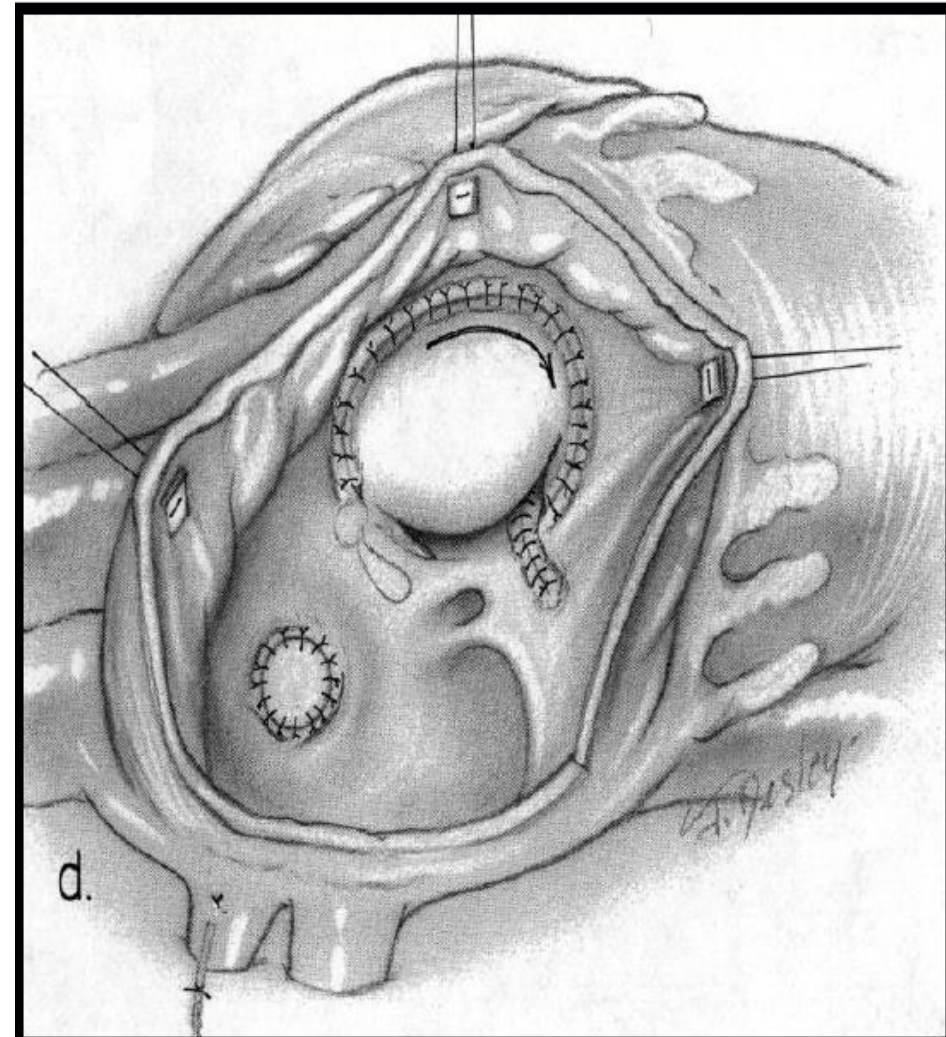
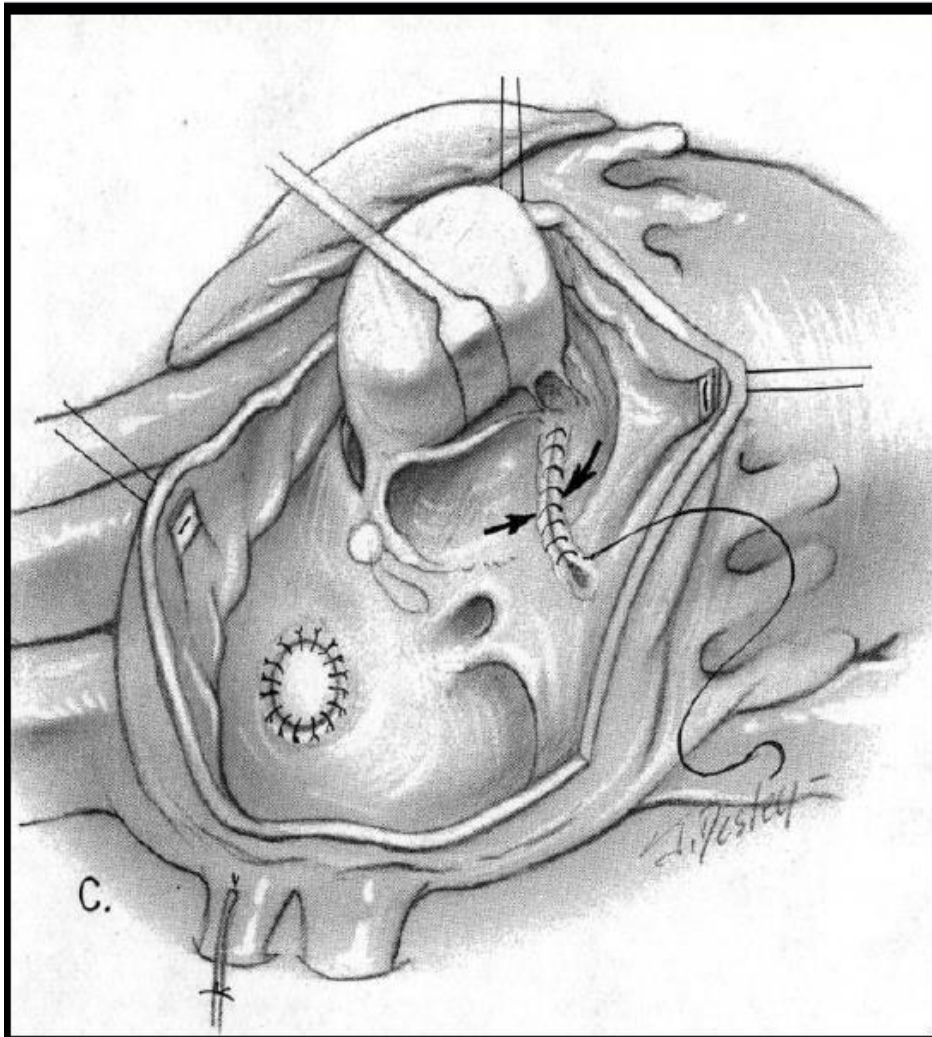


Posterior annuloplasty

**Bring the leaflets up to the level of the AV junction  
Longitudinal plication  
Plus Annuloplasty**

# Traditional Repair Techniques

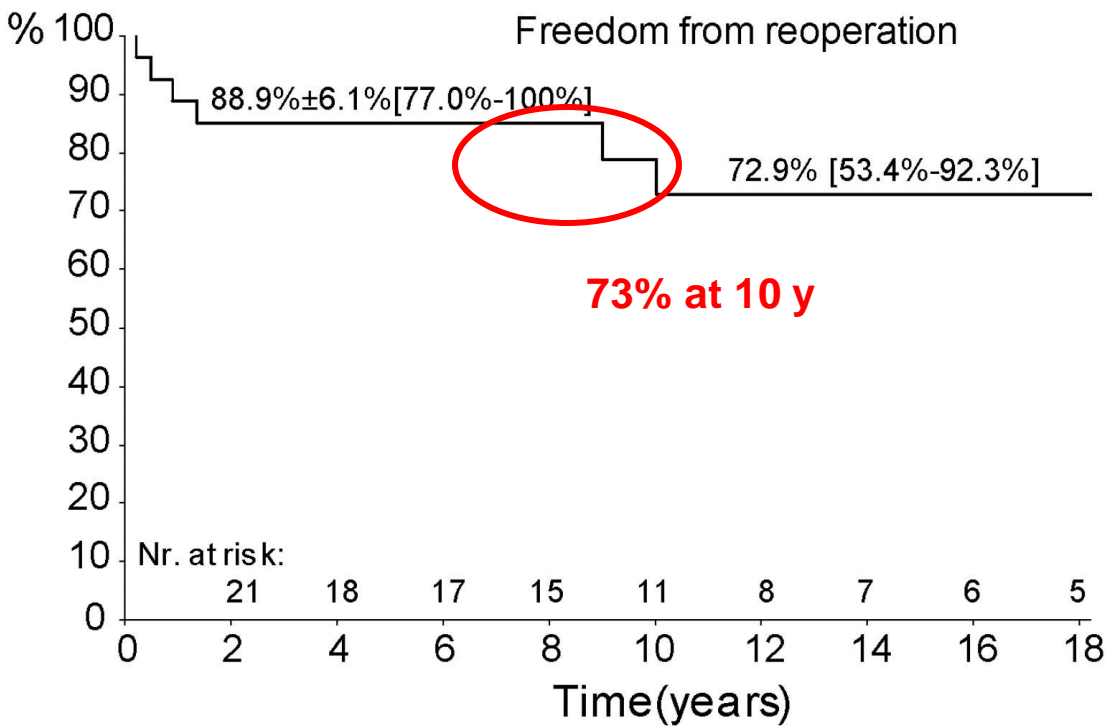
## Carpentier Repair 1980s



**Transverse Plication**  
**Rotation of the Anterior Leaflet**

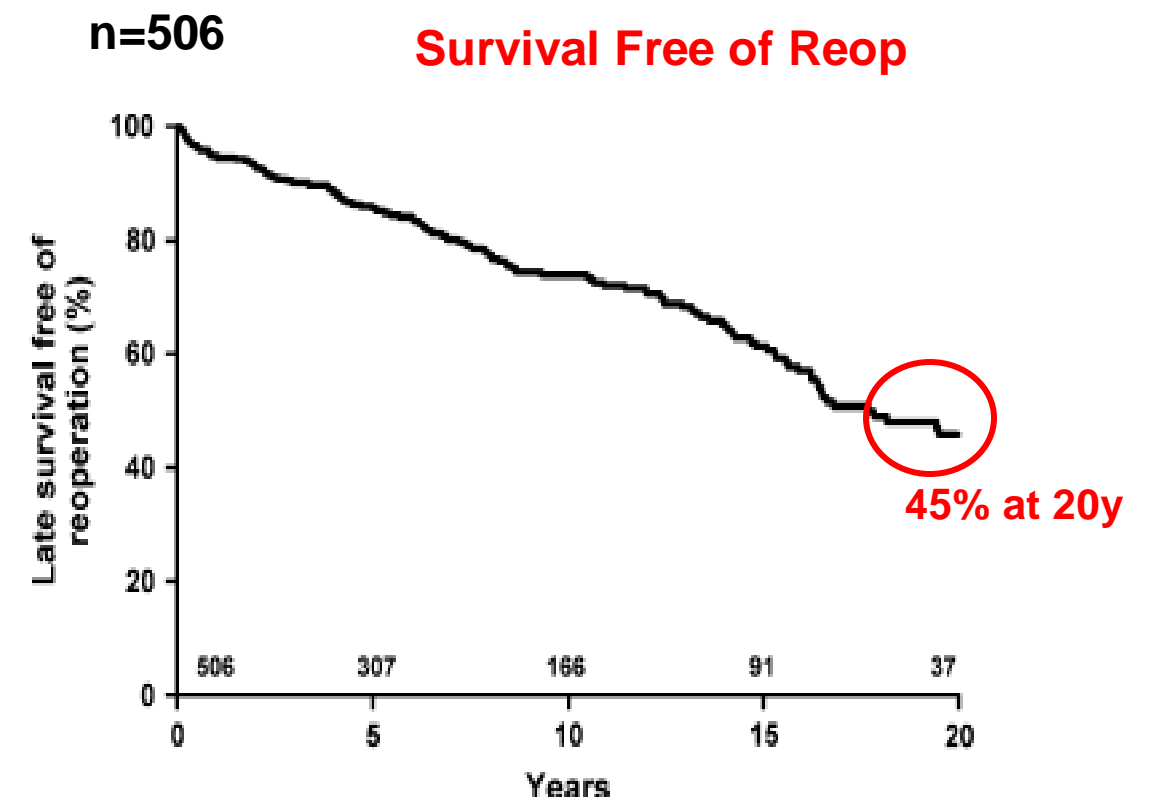
# Repair Outcomes

## Rotterdam Carpentier Technique



EJCTS 34:48, 2008

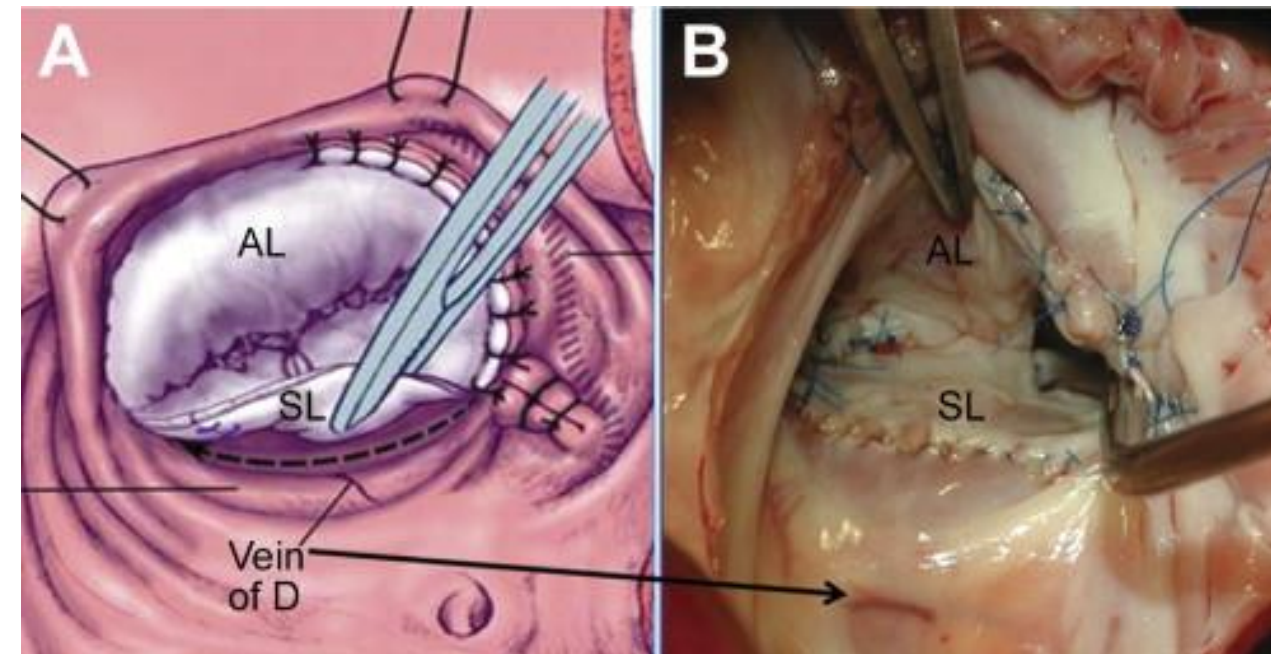
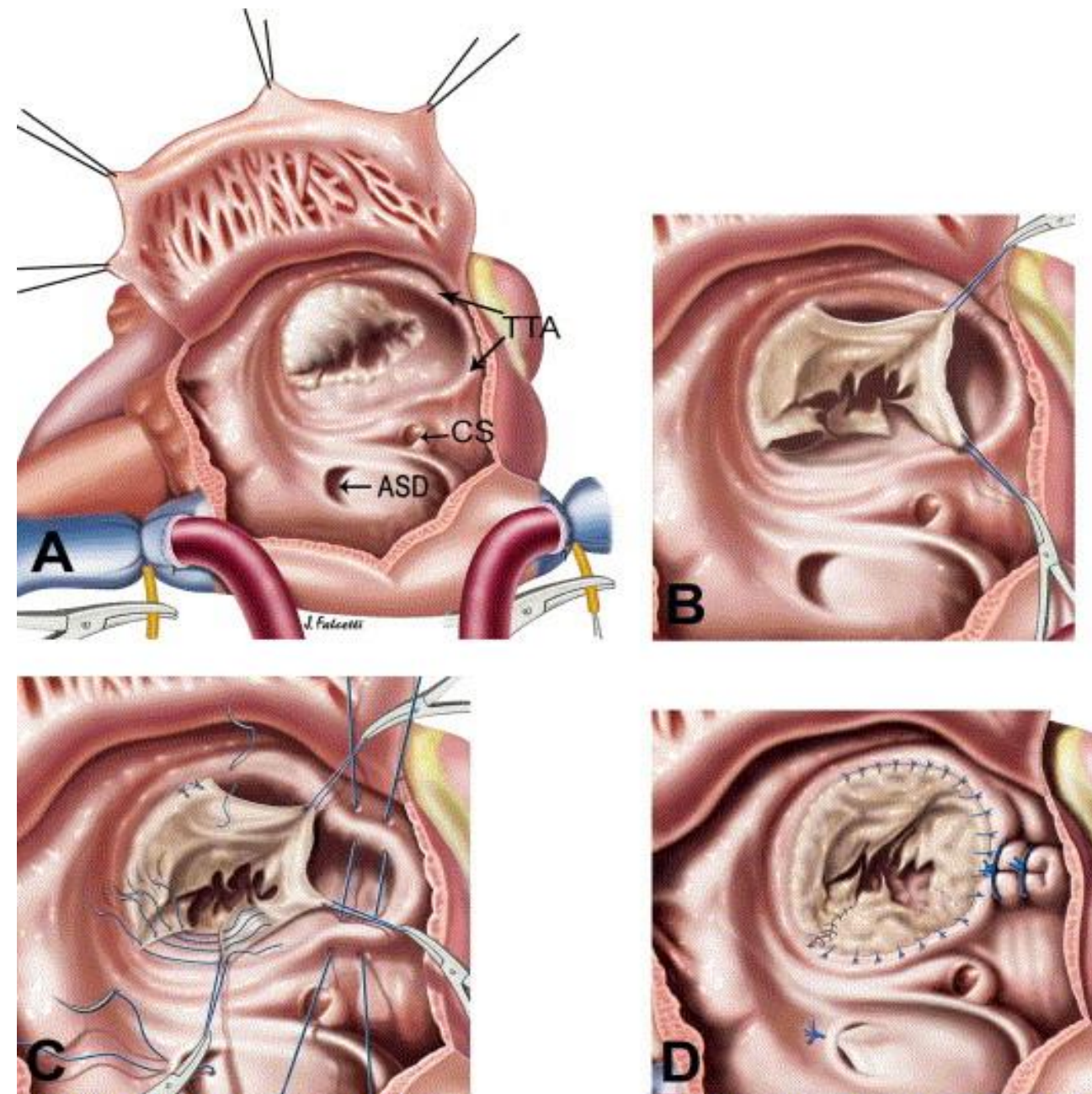
## Mayo Experience – the Danielson repair



JTCVS 135: 1220, 2008

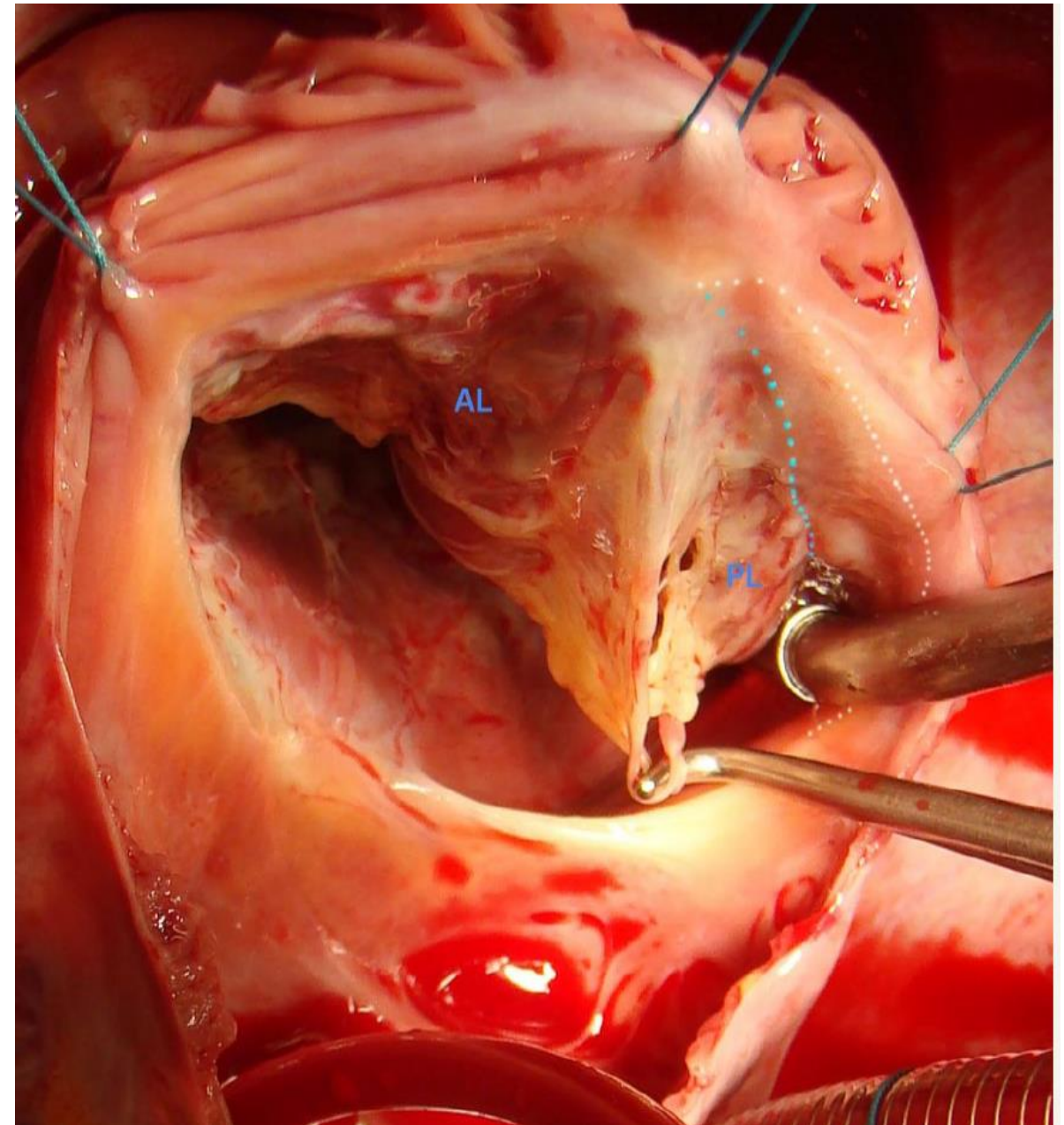
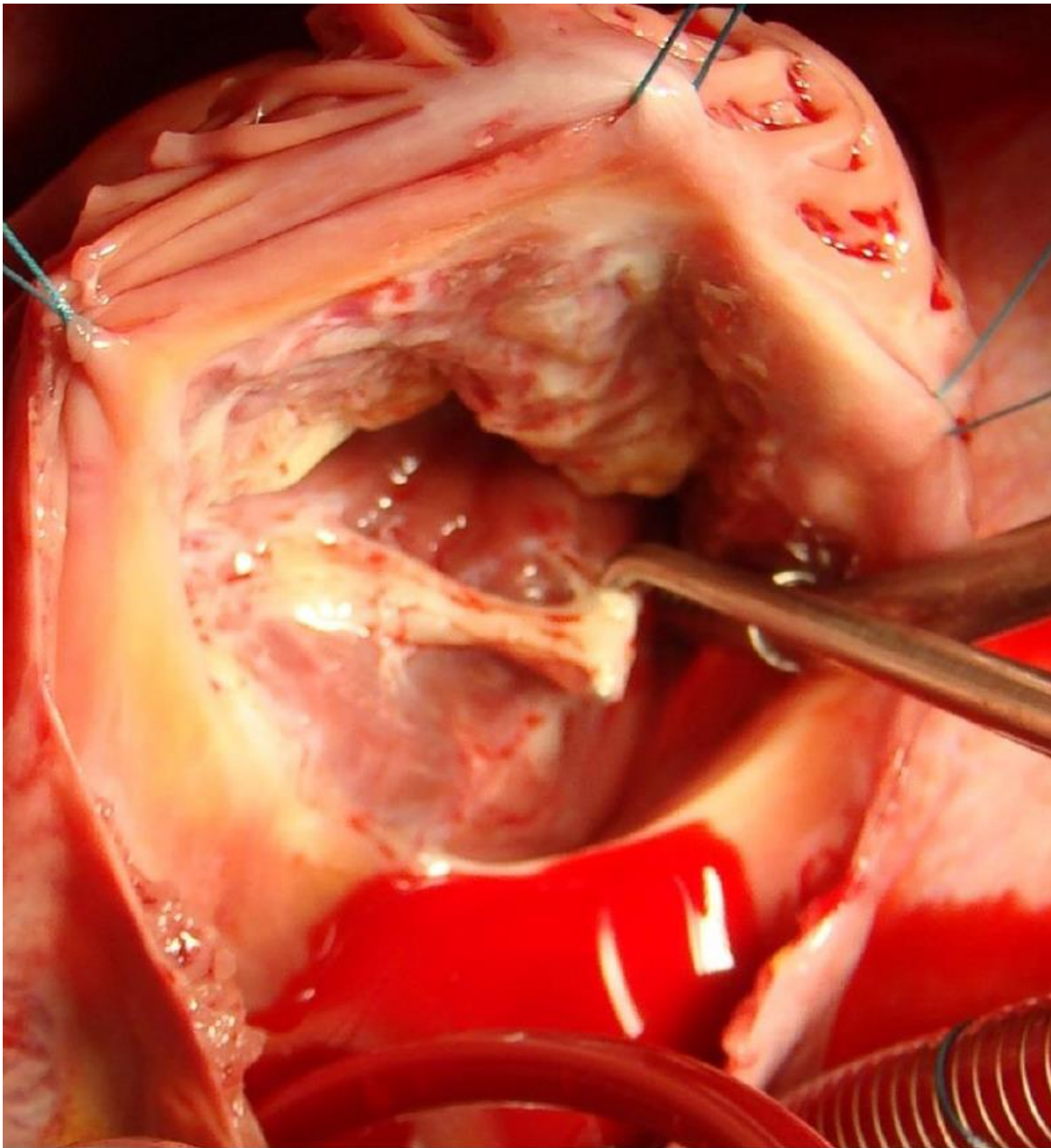
# The Cone Repair

Da Silva  
Dearani

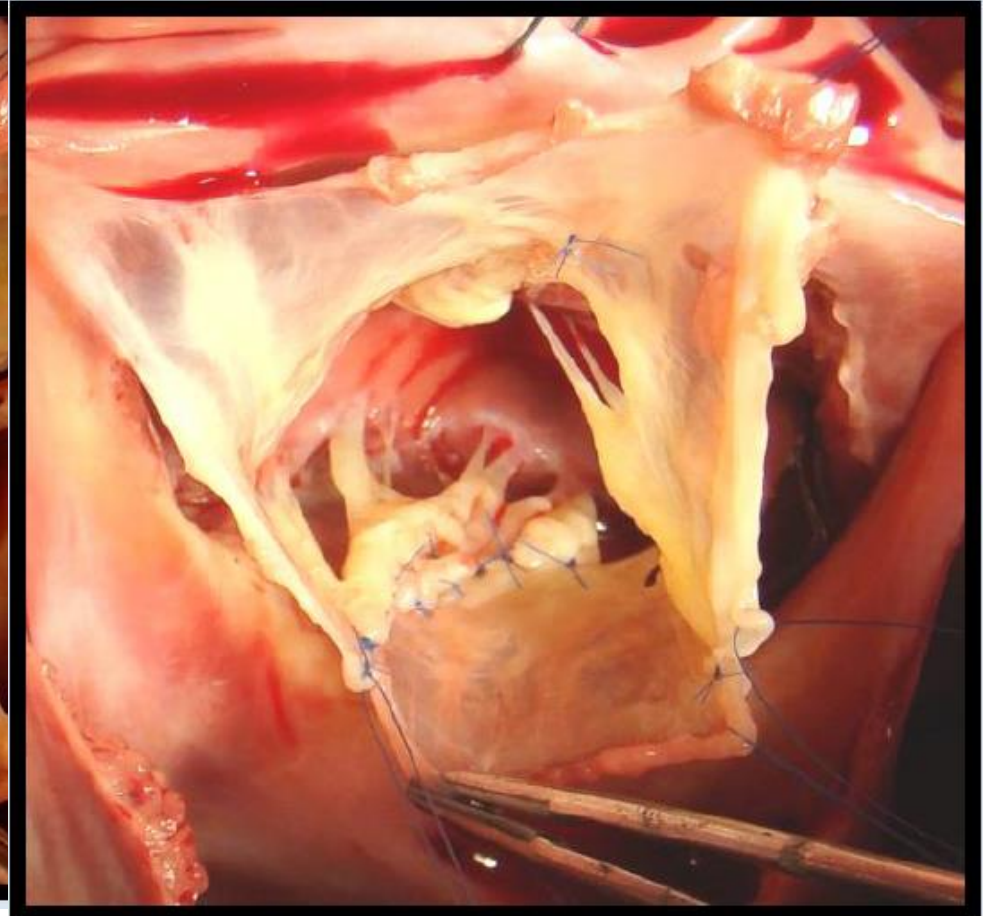
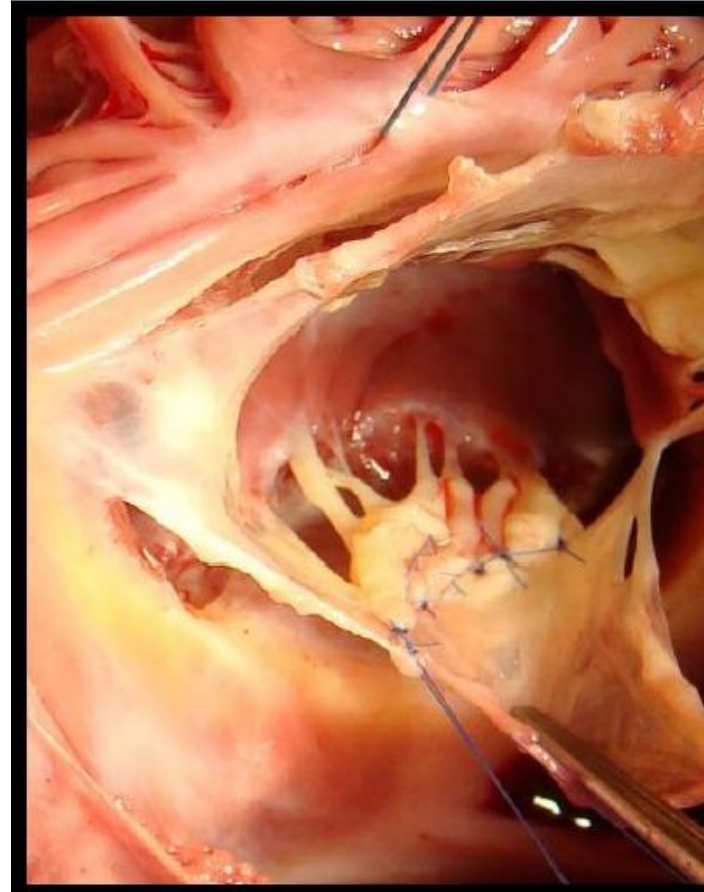
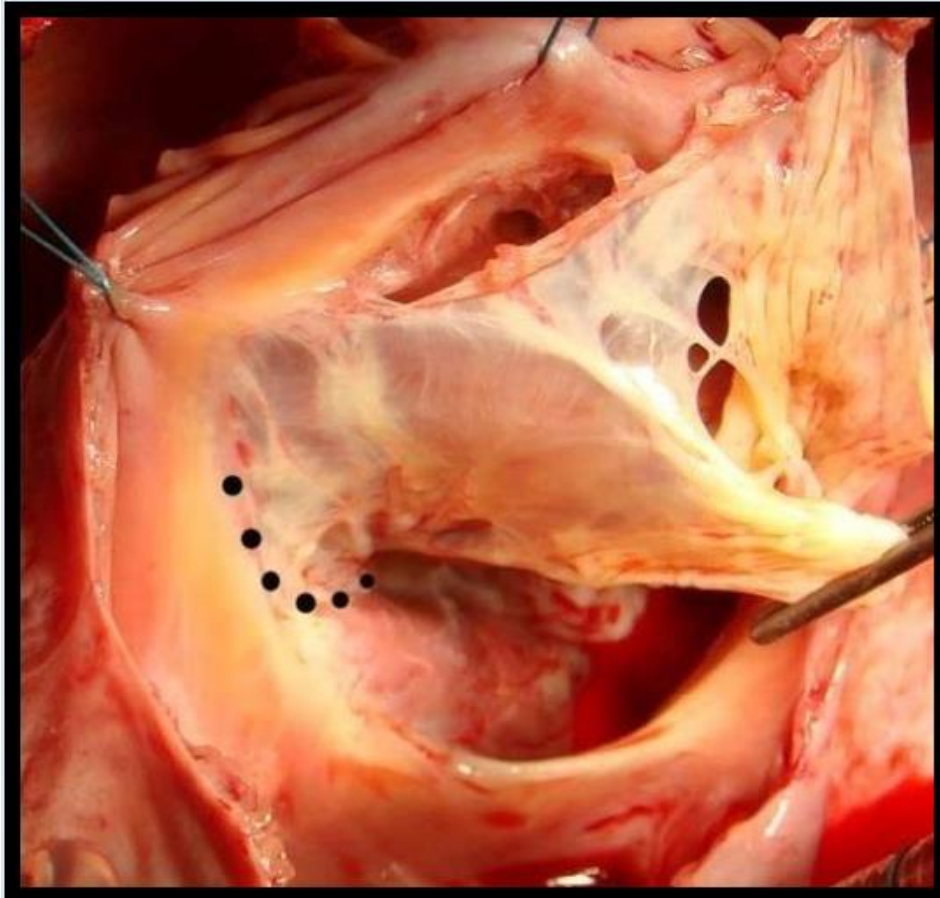


**Mobilises the whole valve apparatus**  
**Creates a 360° 'cone' of valve tissue**  
**Brings the whole valve back to the AV junction**  
**Annuloplasty and transverse plication**

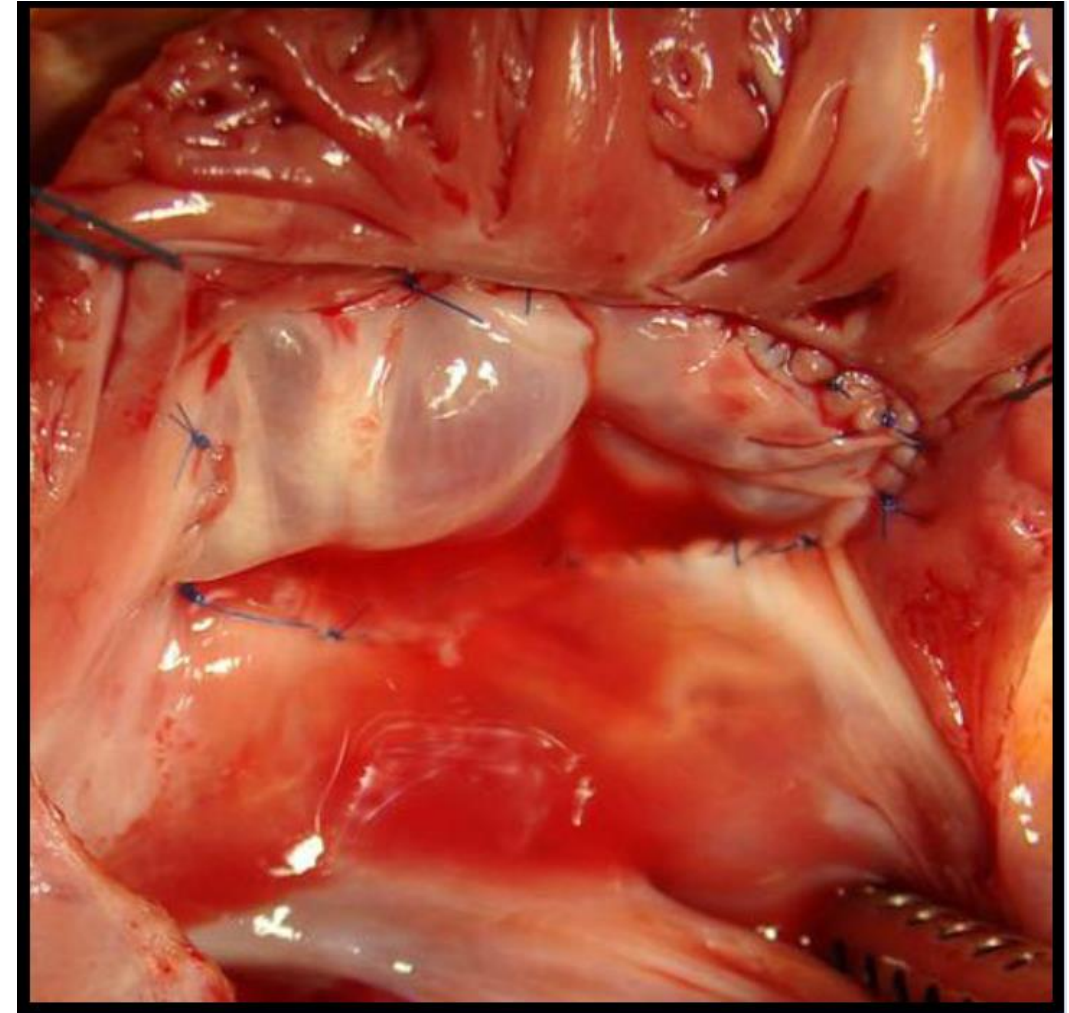
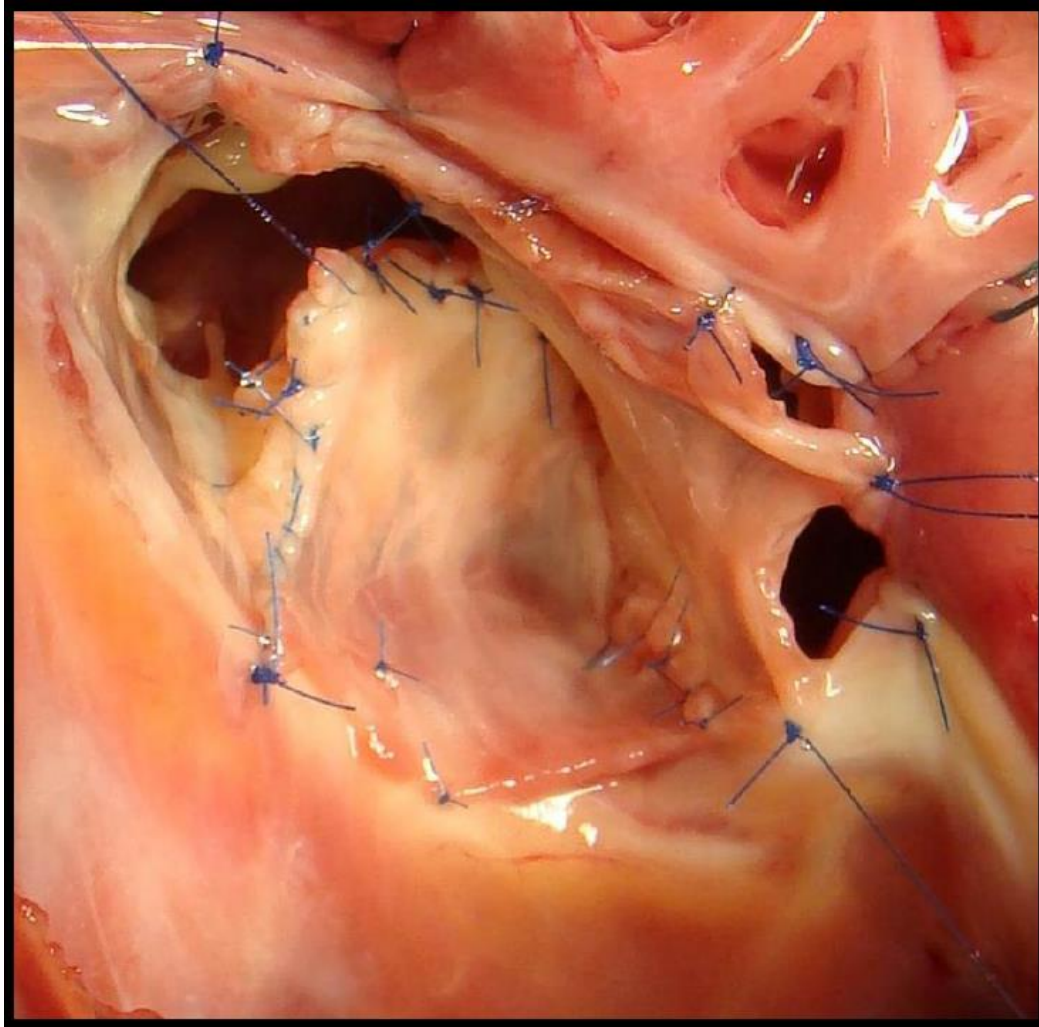
# The Cone Repair



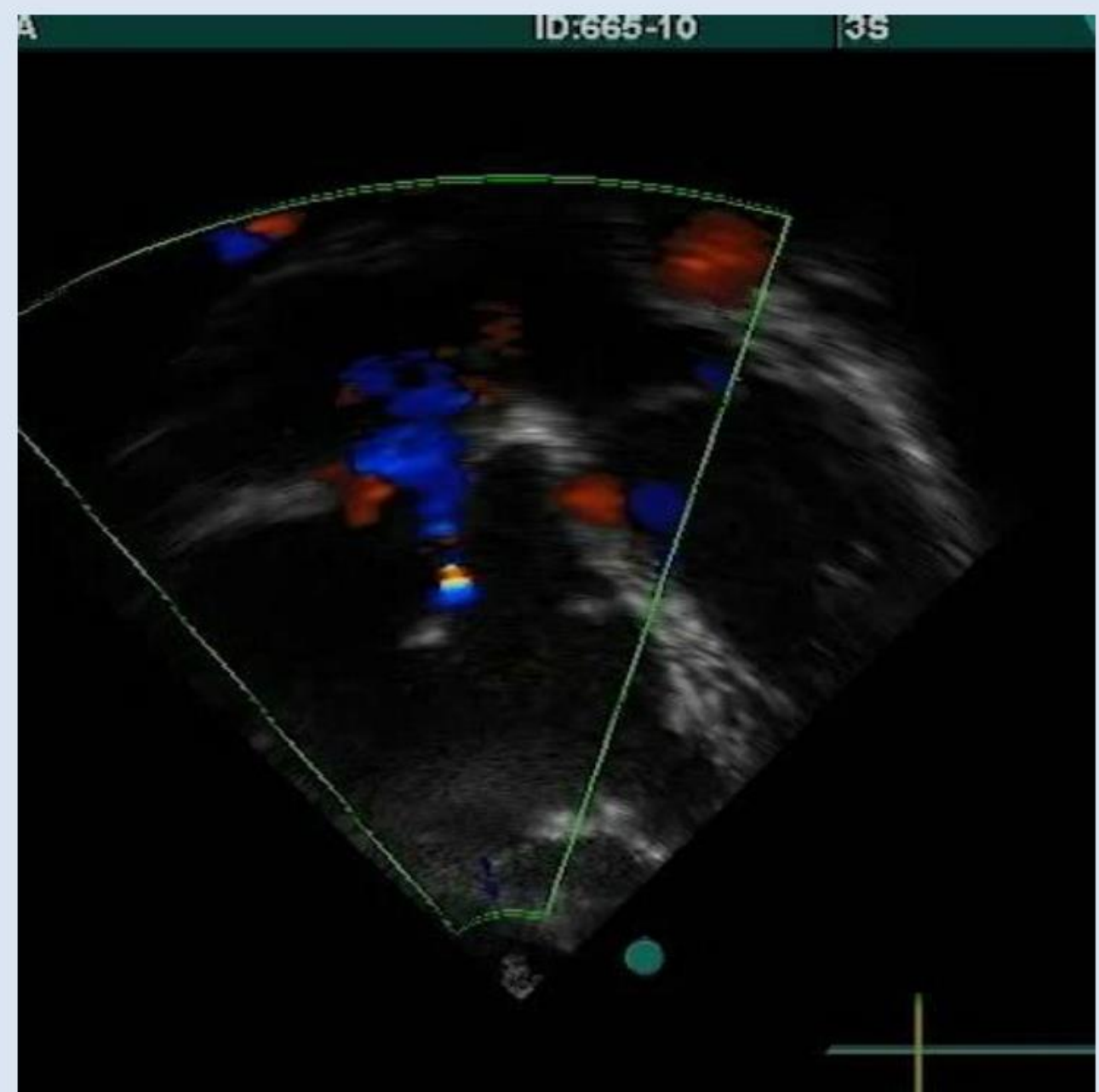
# The Cone Repair



# The Cone Repair



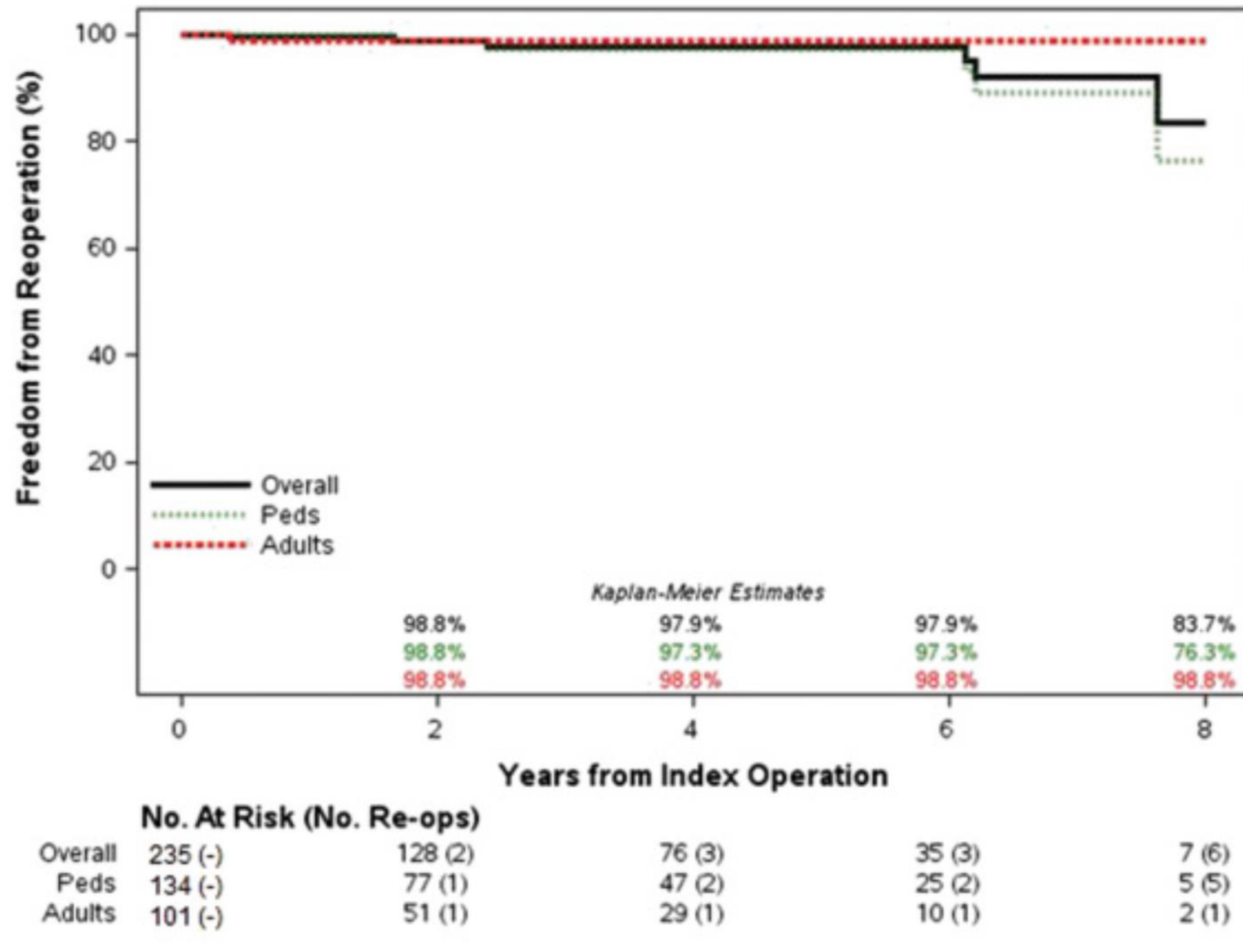
# Echo Post- Cone Repair





# The Cone Repair

Mayo n=235



# Mayo Conclusions

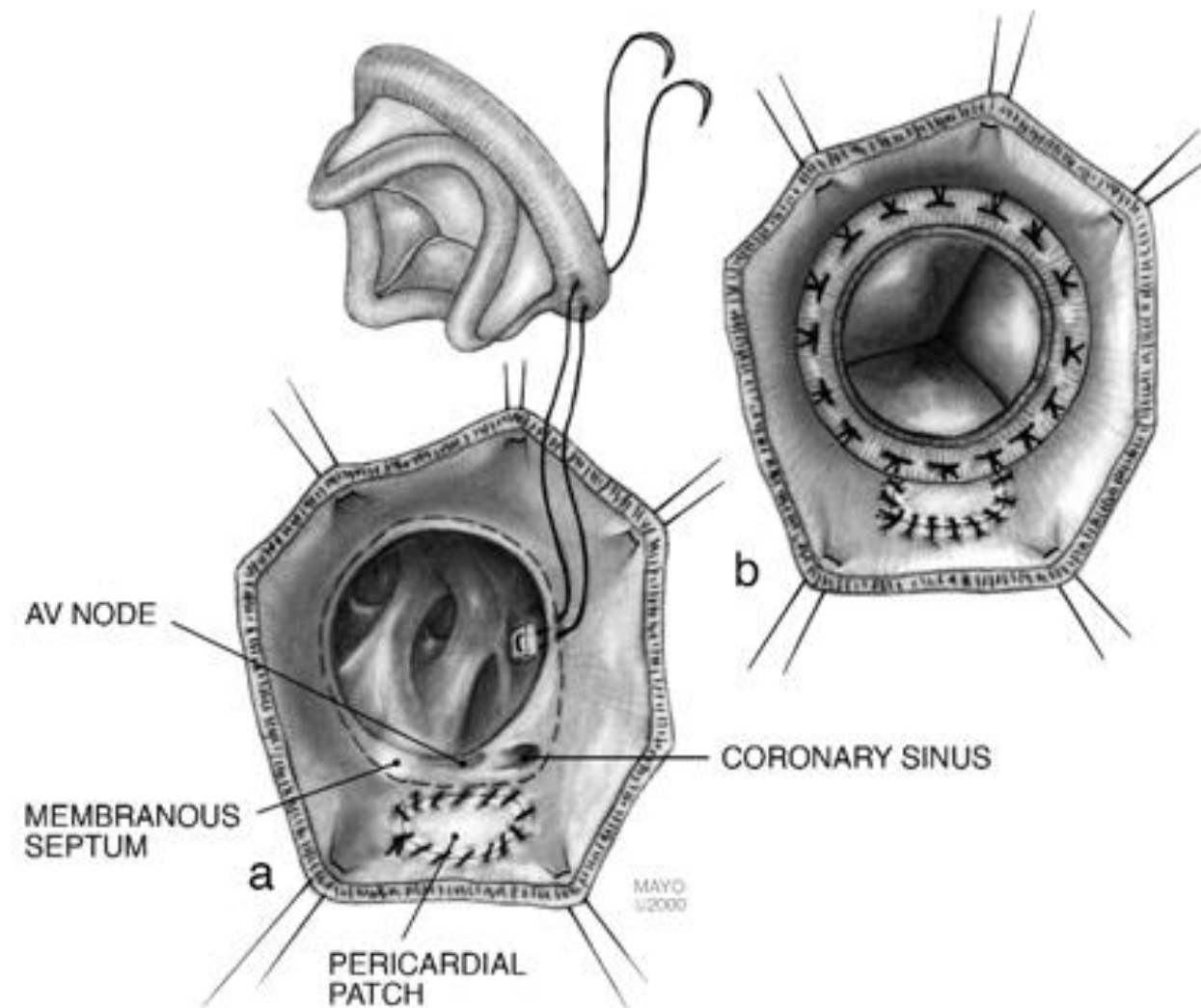
**Avoid attempting Repair if:**

- . Severe degrees of displacement: failure to see TV tissue on the apical 4-chamber view**
  - . Severe TR with multiple jets**
  - . Extensive failed delamination of anterior leaflet**
  - . Severely reduced RV function**
    - . +/- impaired LV function**



**“to replace the valve is not a failure”**  
**“Cannot underestimate the sequelae of leaving residual TR in a failing RV”**

# Tricususpid Valve Replacement

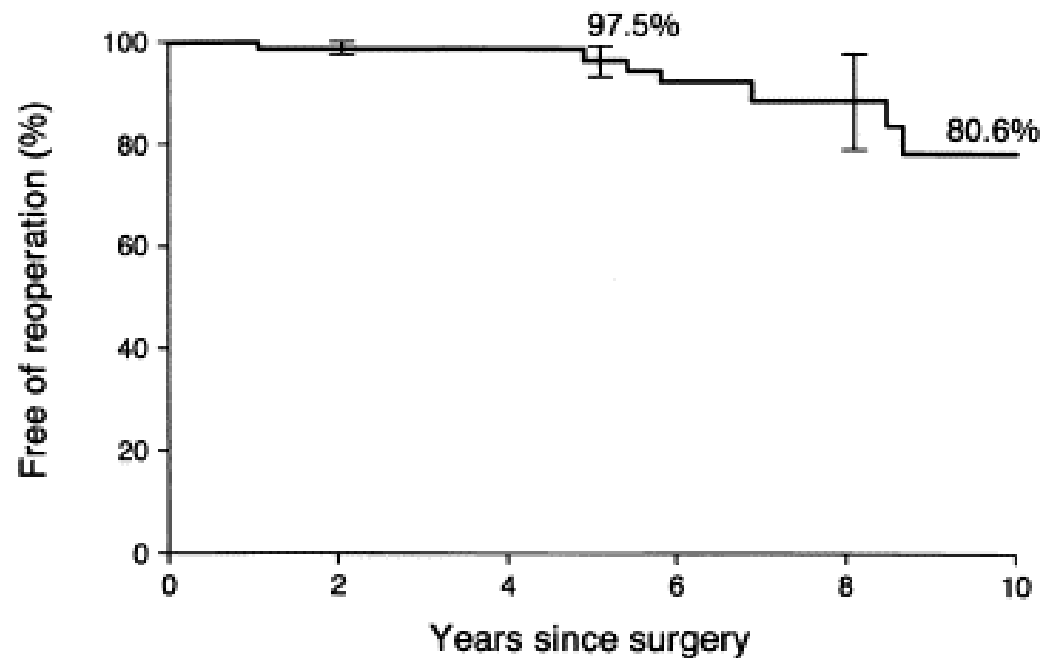


Place 'supra-annular' to avoid Conduction tissue and RCA

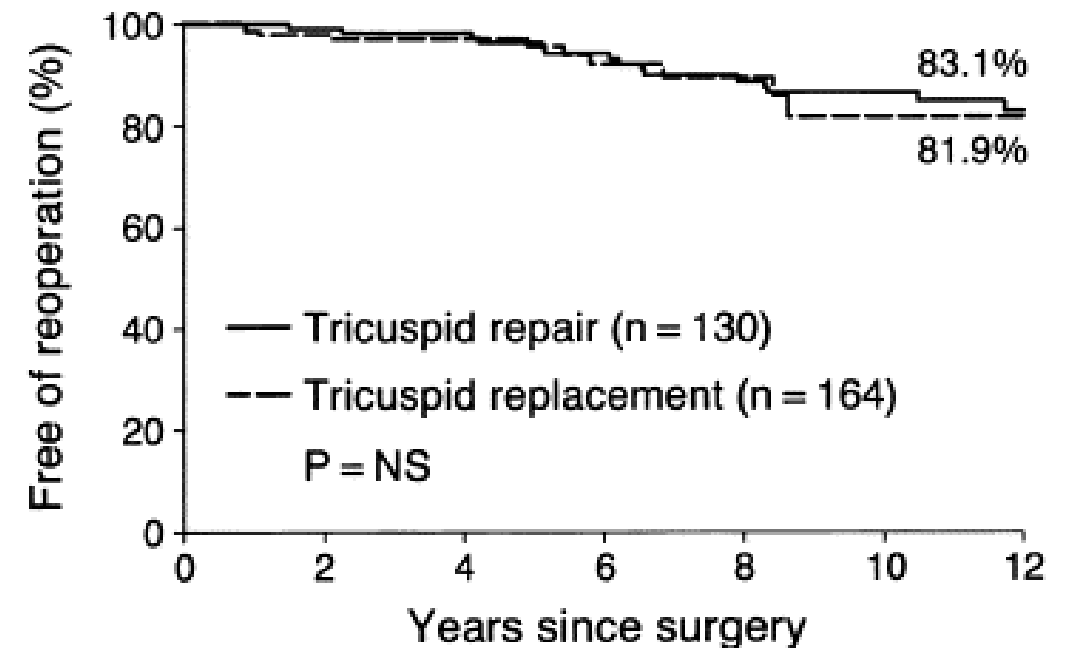
# Performance of the Bioprosthetic TVR

n=164

## Freedom from Reoperation



## Comparison with Repair



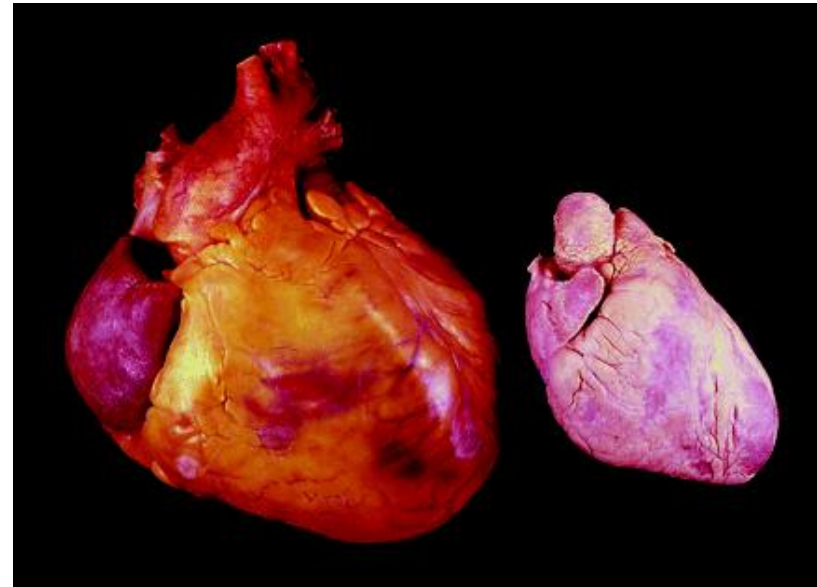
Freedom from replacement was  $97.5\% \pm 1.9\%$  at 5 years and  $80.6\% \pm 7.6\%$  at 10 and 15 years.

Included All age-groups:

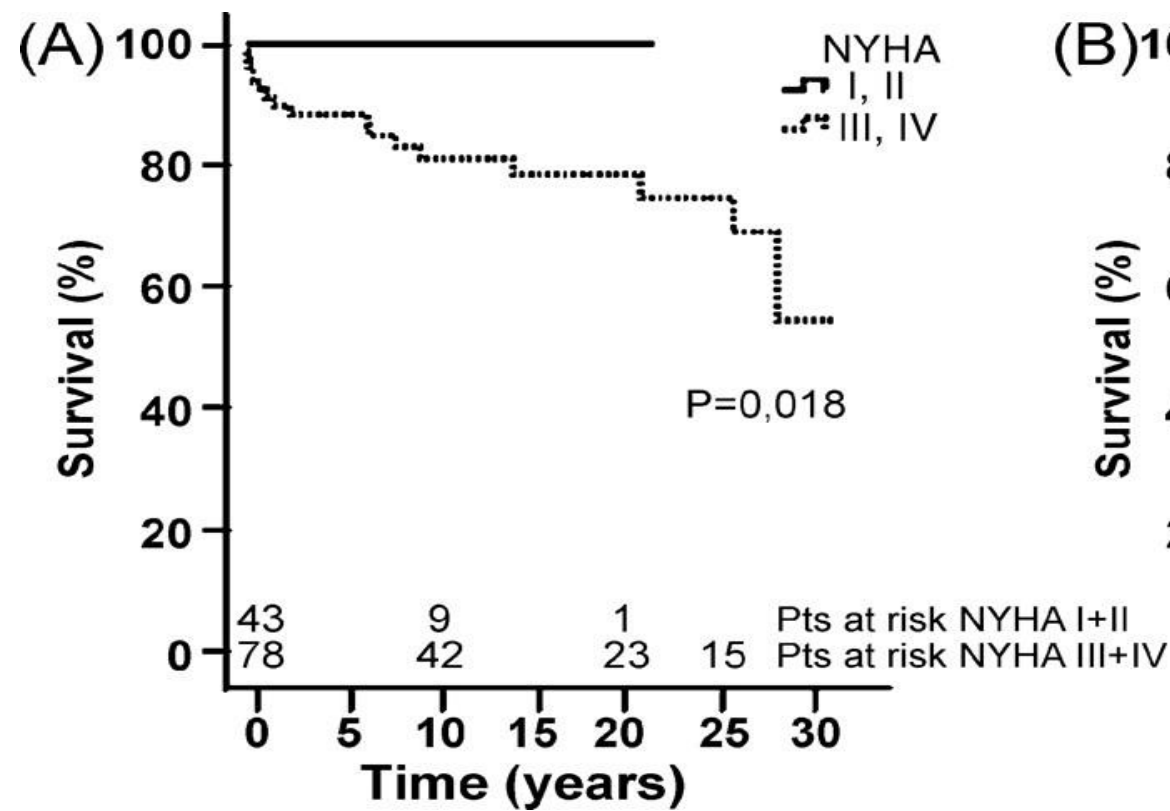
If limit to adults only: the performance was even better with 94.6% freedom reop at 10

y

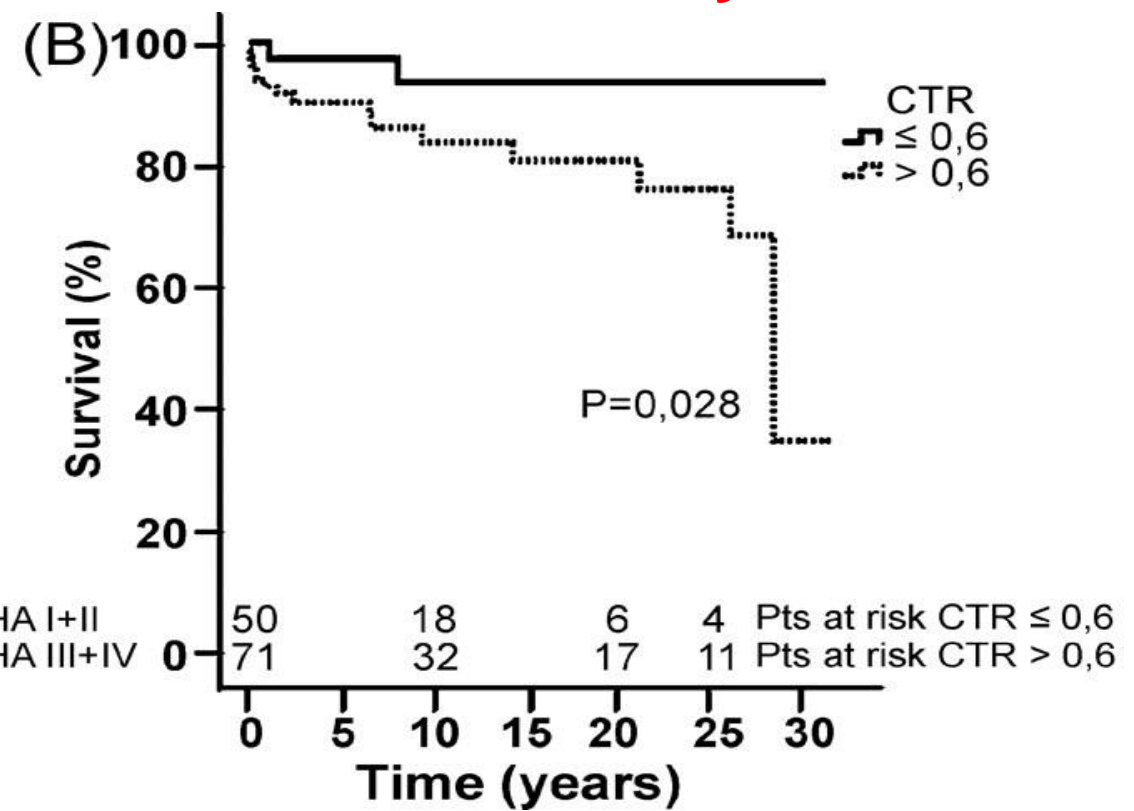
# Influence of Functional Class and RV Size on Outcome



**Survival by NYHA**



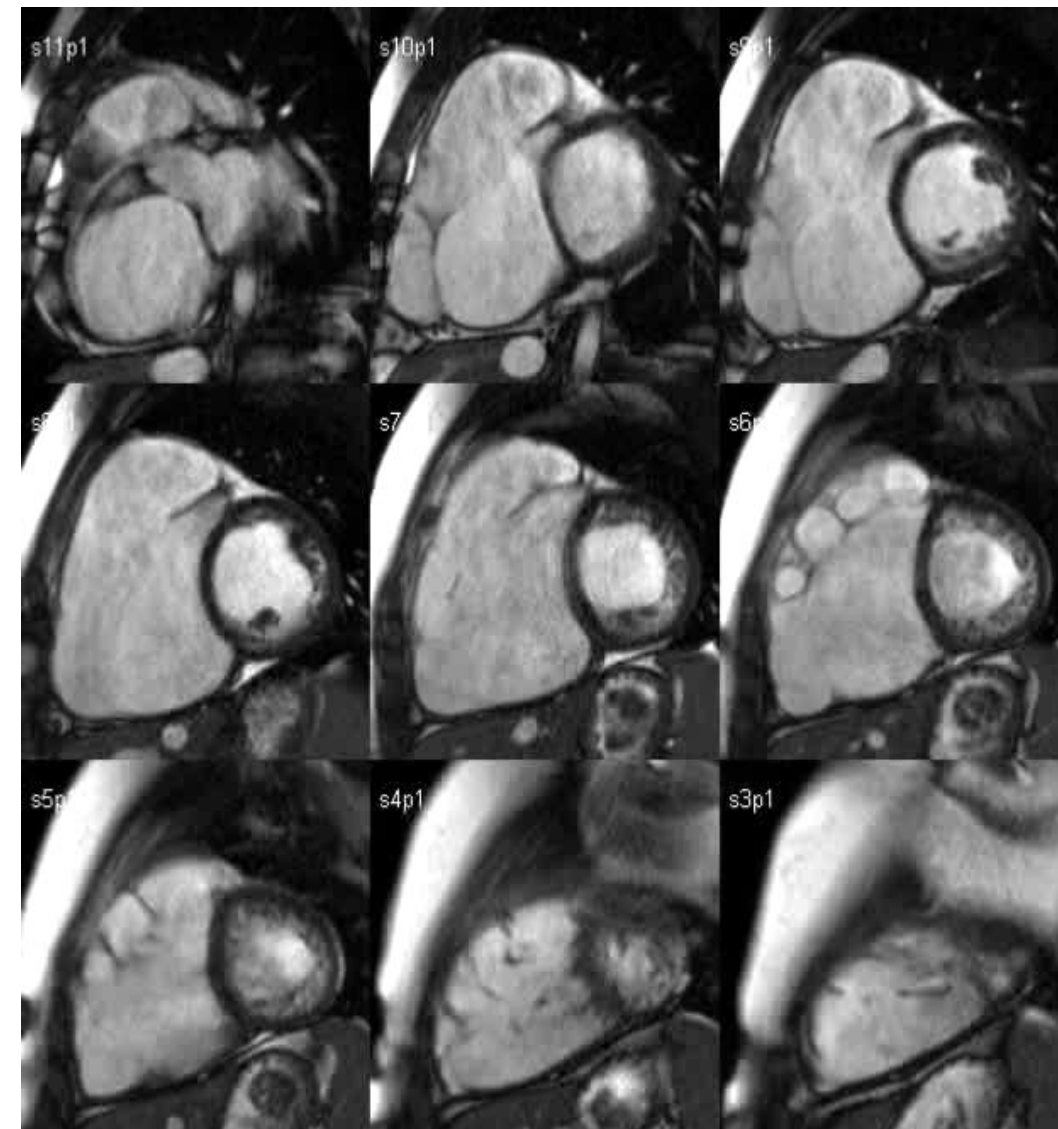
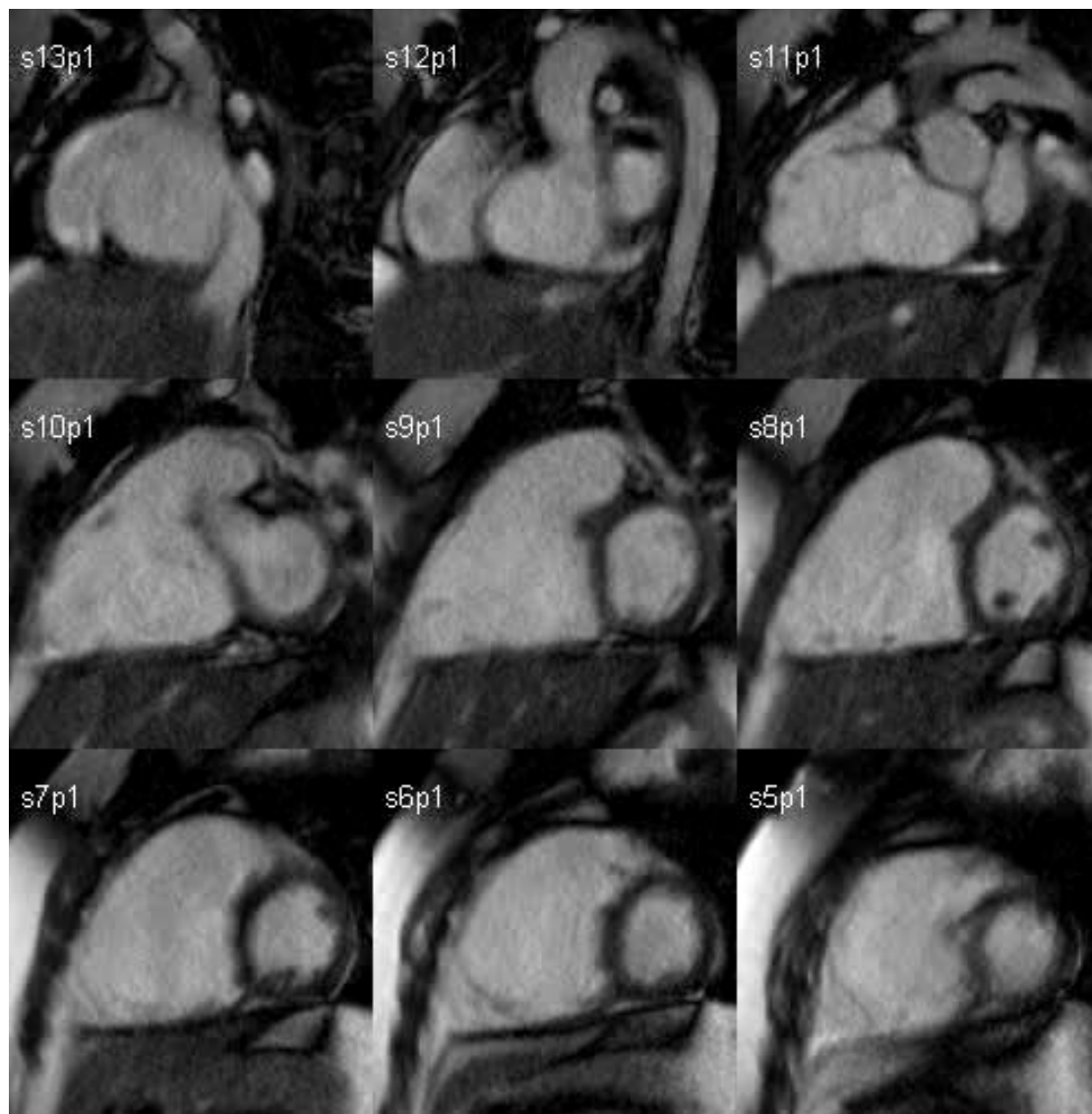
**Survival by RV Size**



# Ventricular Interaction and LV function

Being recognised as increasingly important

**Septal dyskinesia and reduction in LVEF are independent risk factors for death.**



# Summary

**This is a defect that involves the whole right ventricle,  
remember the spiralling motion**

**Spectrum of morphology, focus on:**

***the extent of failed delamination***

***the morphology of the anterior leaflet***

***the insertion of the leaflets and the tricuspid orifice***

**Associated lesions:**

**ASD/PFO**

**Pulmonary atresia**

**Careful echo and MRI assessment guide repair**

**Repair techniques depend on quality of the leaflets, RV size and function**

**Outside of the neonatal period, it's the big RV you worry about, not the small ones.**





**The outcomes of operations for 539 patients with  
Ebstein anomaly**

Morgan L. Brown, MD,<sup>a</sup> Joseph A. Dearani, MD,<sup>a</sup> Gordon K. Danielson, MD,<sup>a</sup> Frank Cetta, MD,<sup>b</sup> Heidi M. Connolly, MD,<sup>c</sup>  
Carole A. Warnes, MD,<sup>c</sup> Zhuo Li, MS,<sup>d</sup> David O. Hodge, MS,<sup>d</sup> and David J. Driscoll, MD,<sup>b</sup> for the Mayo Clinic Congenital  
Heart Center

**Comparison of Repair vs Replacement**

**No difference in early mortality (5% vs 6%)**

**No difference in 1 y survival (93% vs 91%)**

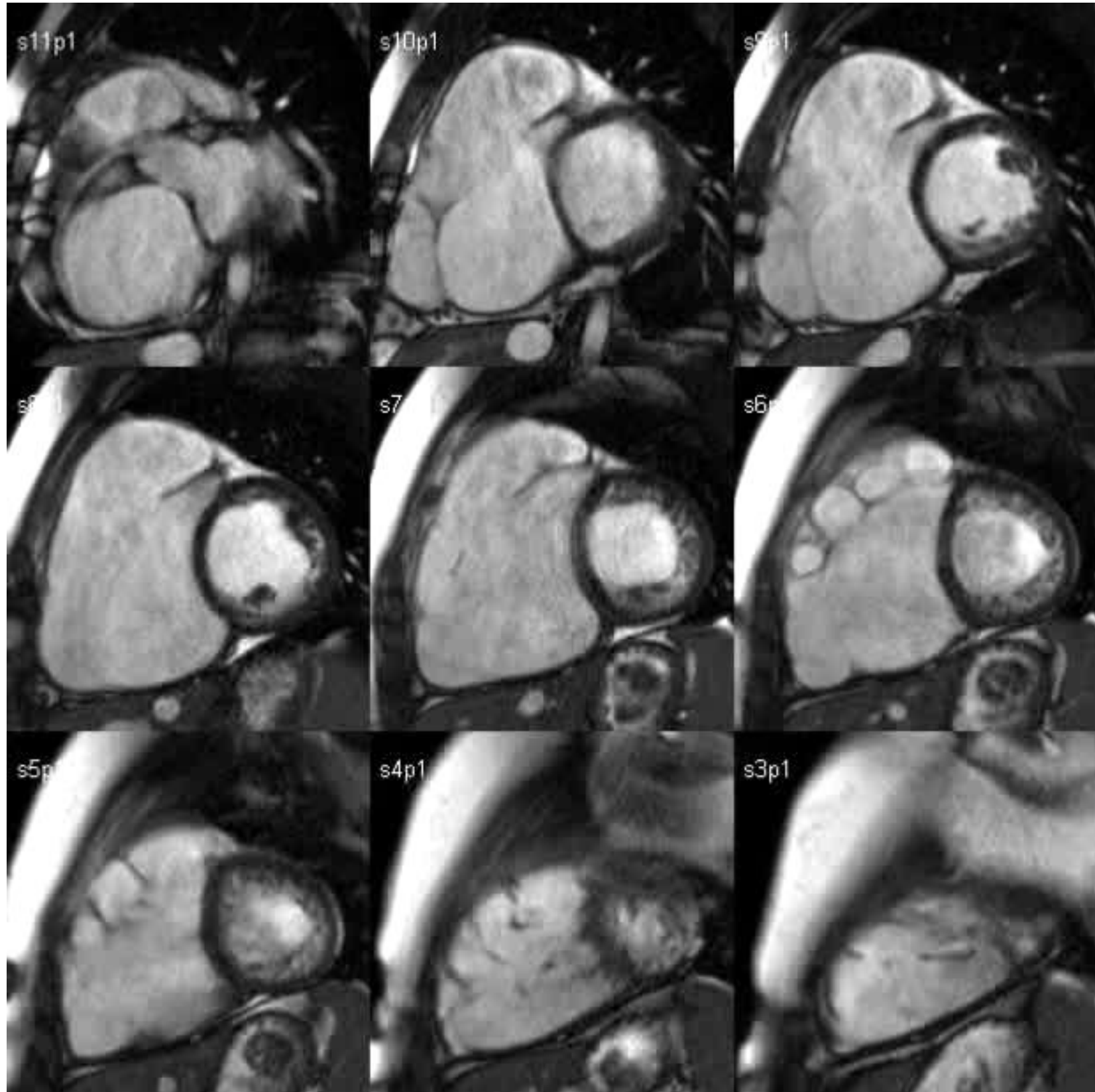
**No difference in 10y survival (88% vs 83%)**

**No difference in 20 y survival (76% vs 68%)**

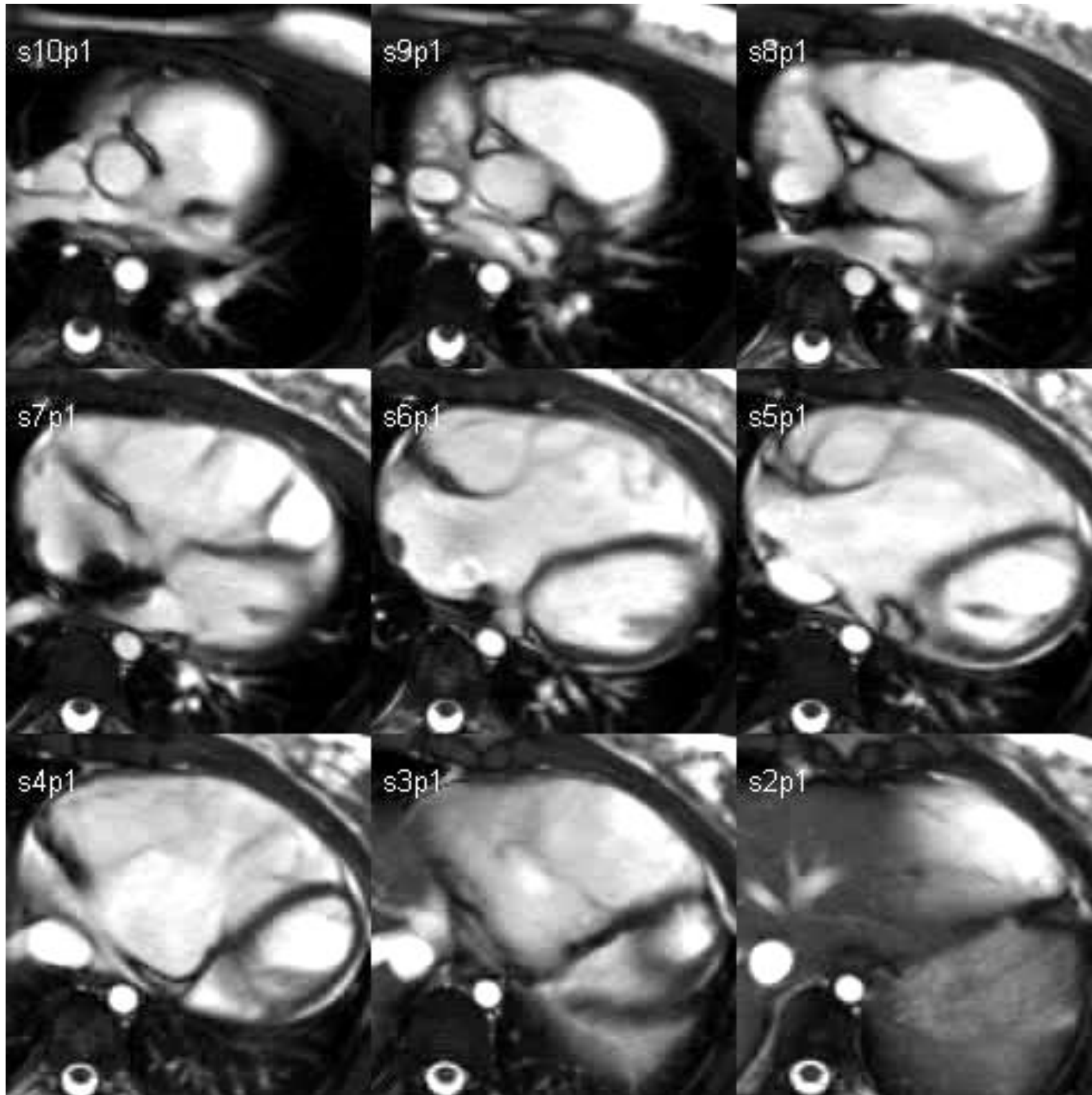
**Munich: No difference in 1 y, 5y 10y, 20y survival between repairs and replacements**

**EJCTS 37:186, 2010**

**Both Studies:  
Only significant risk factor for survival was mod-severe RV  
dysfunction at time of surgery**

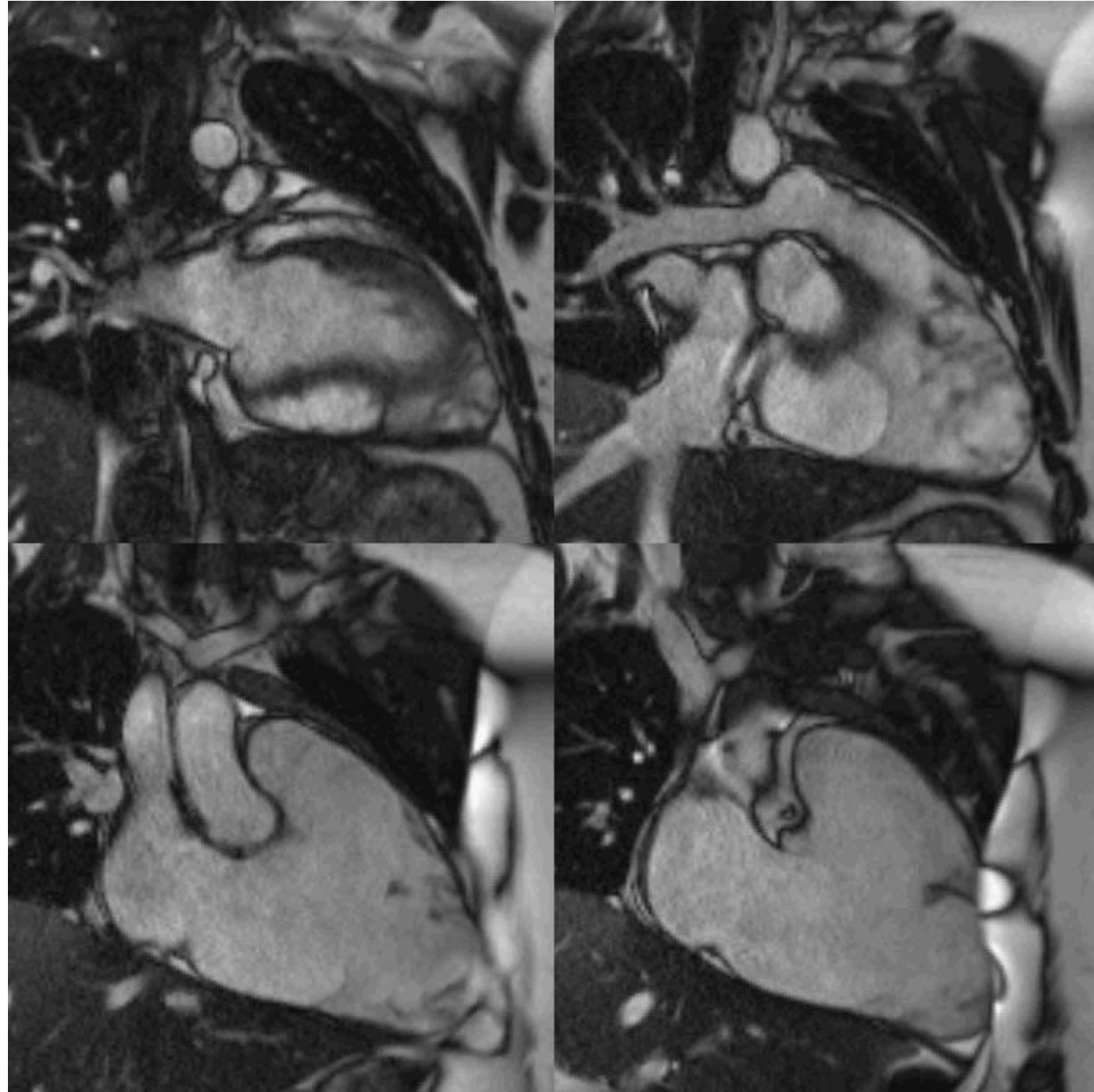


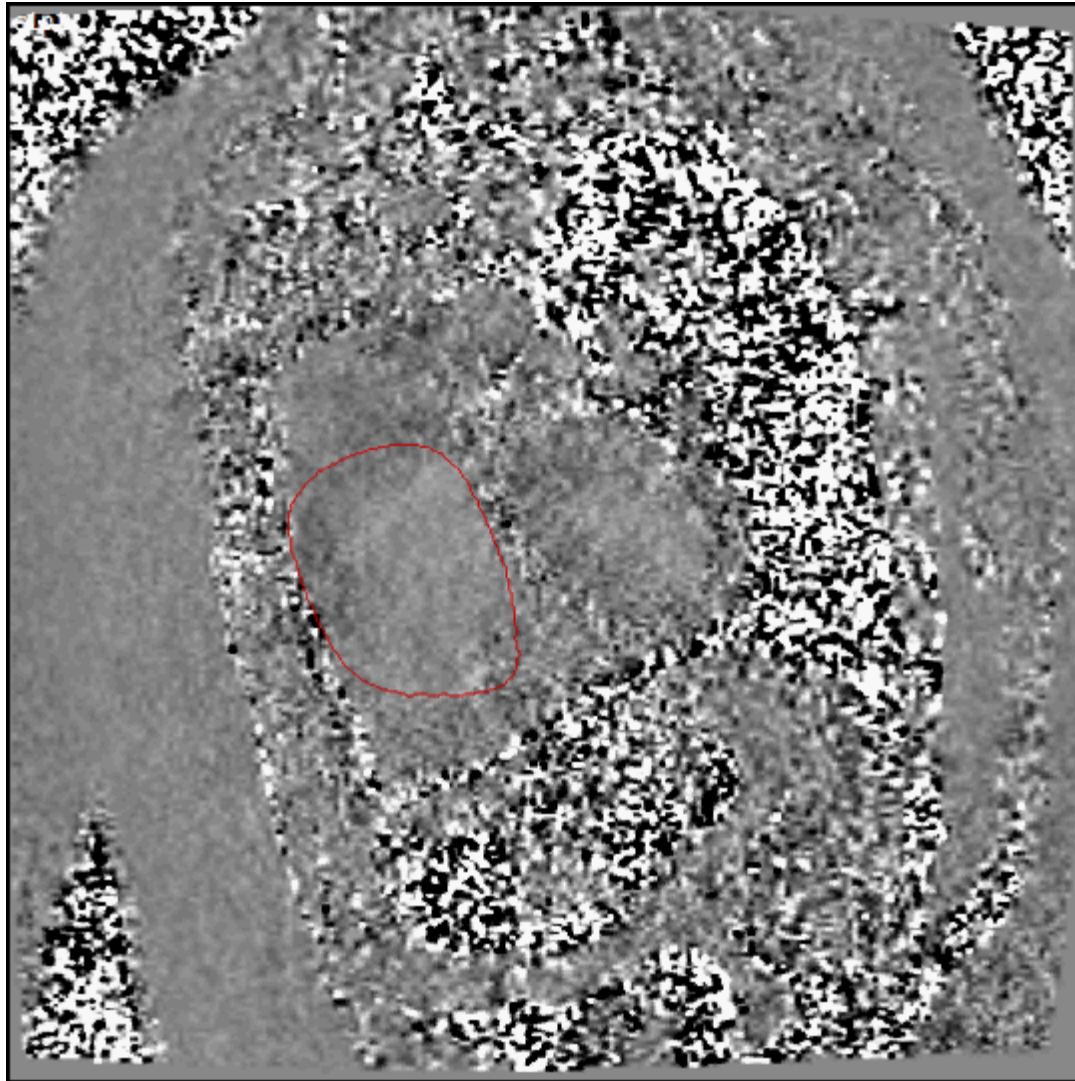
## Ebstein S/P ASD device closure



Ebstein 2266758

13Y





### LV Volumetry

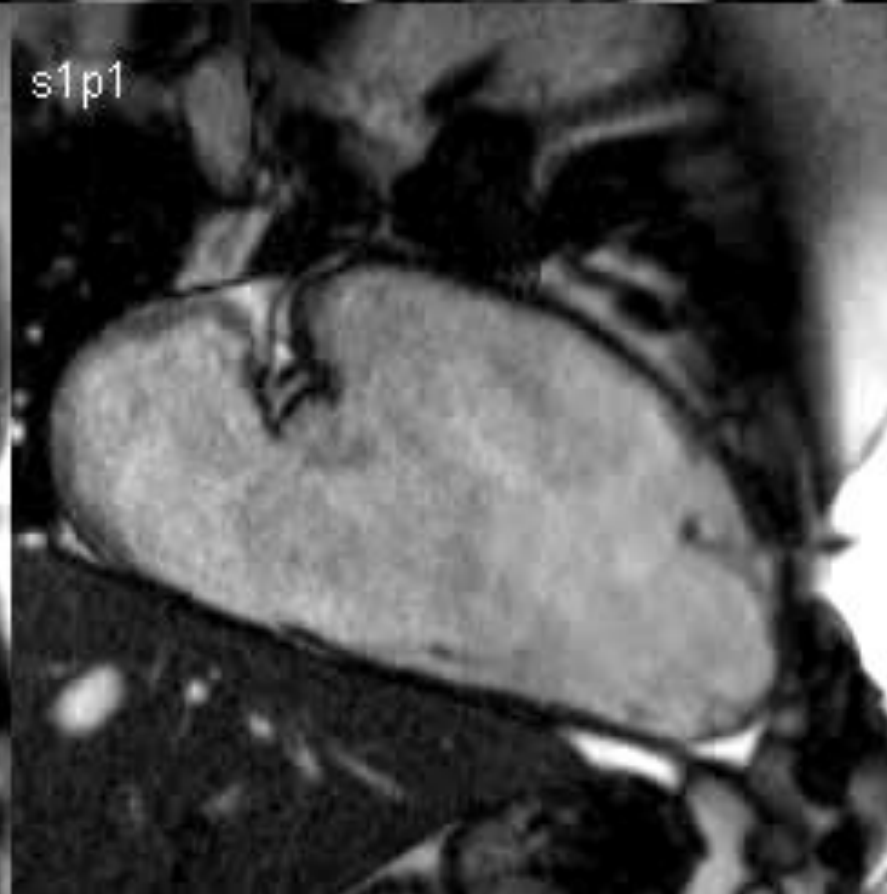
ED mass	67.42 g	ED Mass/BSA	31.94 g/m <sup>2</sup>
EDV	176.00 ml	EDV/BSA	83.38 ml/m <sup>2</sup> (56-108)
ESV	91.29 ml	ESV/BSA	<b>43.25</b> ml/m <sup>2</sup> (14-42)
SV	84.71 ml	SV/BSA	40.13 ml/m <sup>2</sup> (36-71)
EF	<b>48.13</b> % (55-76)		
CO	5.28 l/min	CO/BSA	2.50 l/(min*m <sup>2</sup> ) (2-7)

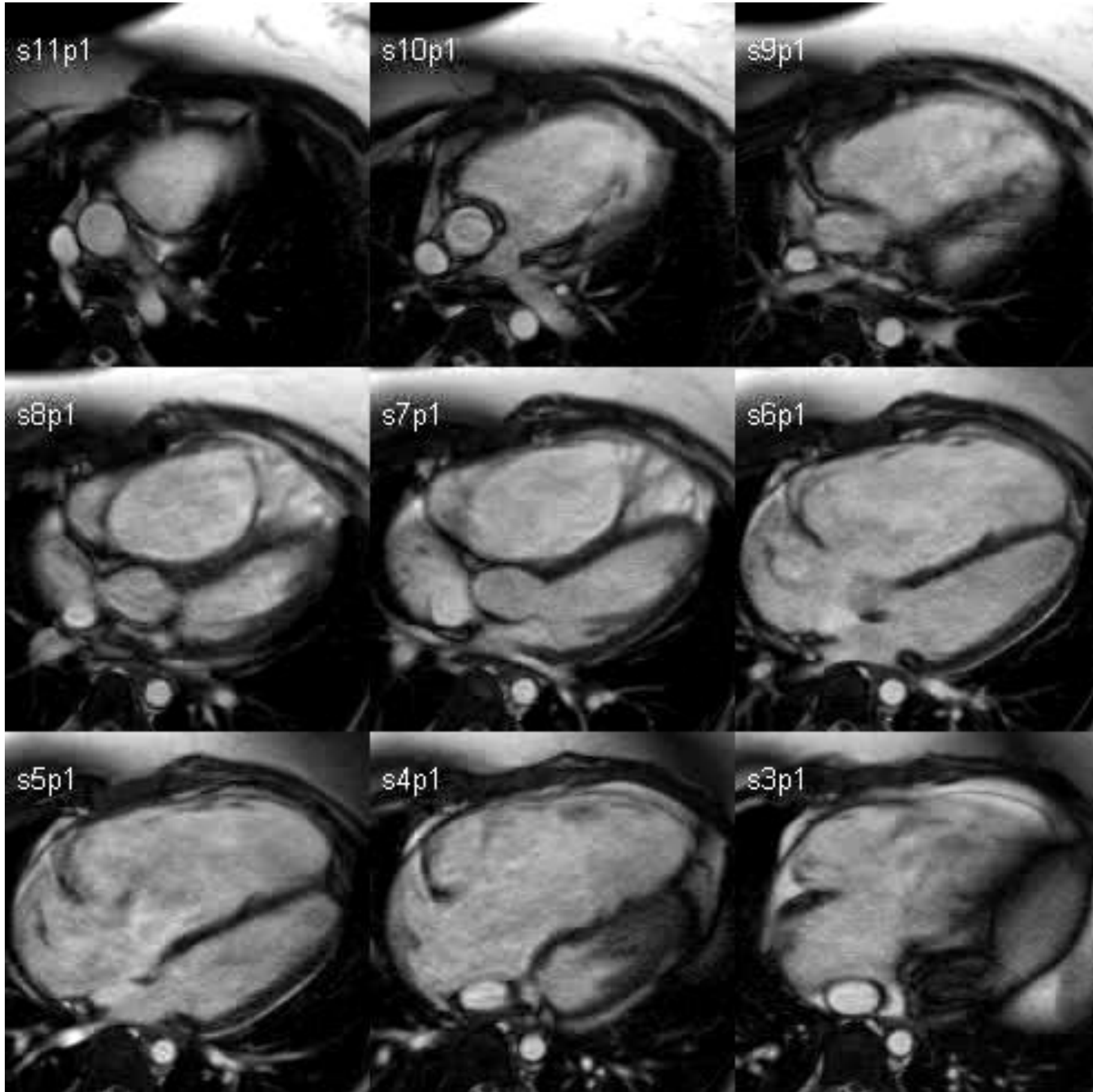
### RV Volumetry

ED mass	0.00 g	ED Mass/BSA	0.00 g/m <sup>2</sup>
EDV	421.42 ml	EDV/BSA	<b>199.65</b> ml/m <sup>2</sup> (57-109)
ESV	227.55 ml	ESV/BSA	<b>107.80</b> ml/m <sup>2</sup> (19-44)
SV	193.87 ml	SV/BSA	<b>91.84</b> ml/m <sup>2</sup> (34-69)
EF	<b>46.00</b> % (53-71)		
CO	12.08 l/min	CO/BSA	5.72 l/(min*m <sup>2</sup> )

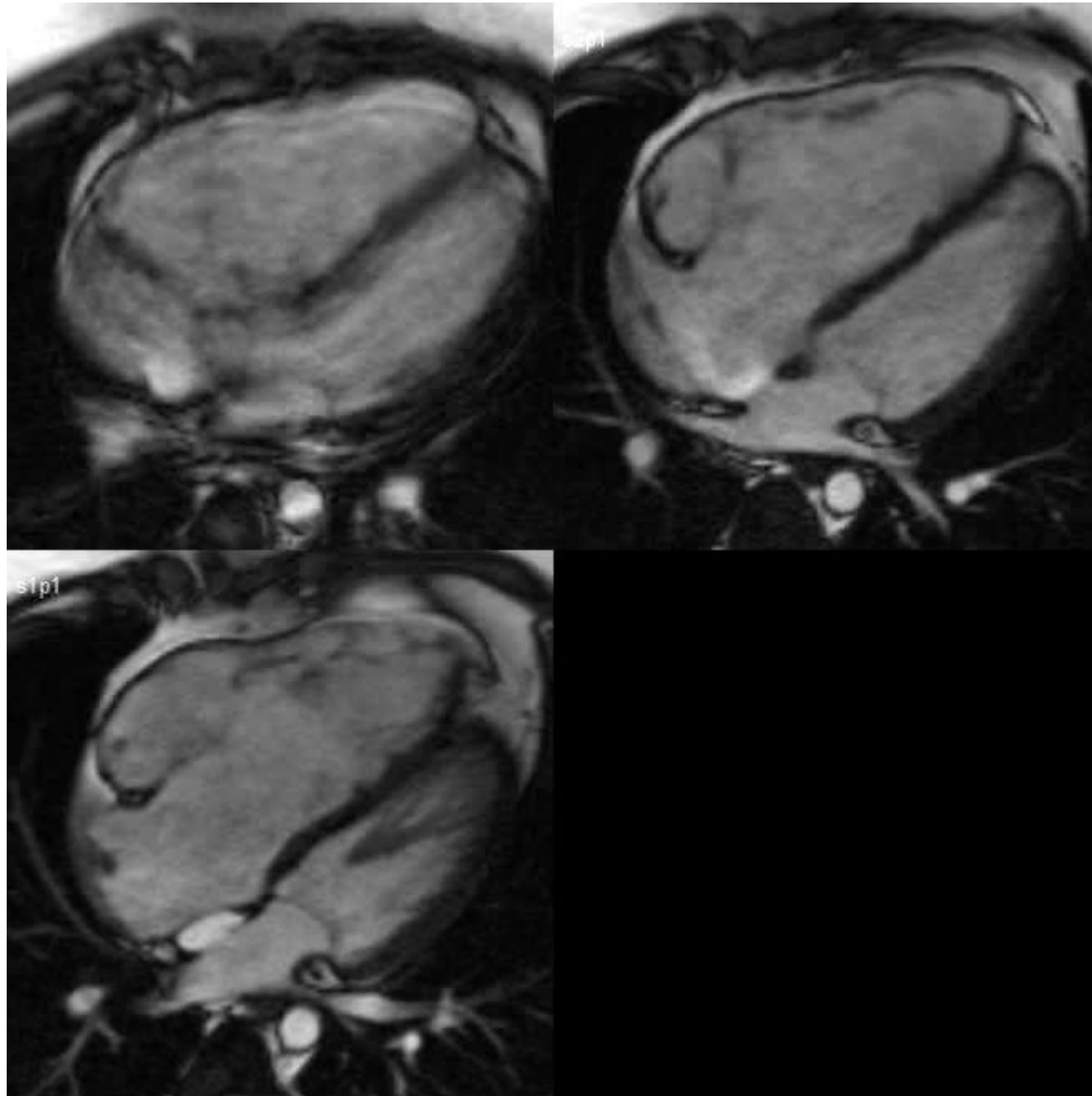
TV DYSPLASIA 5708188



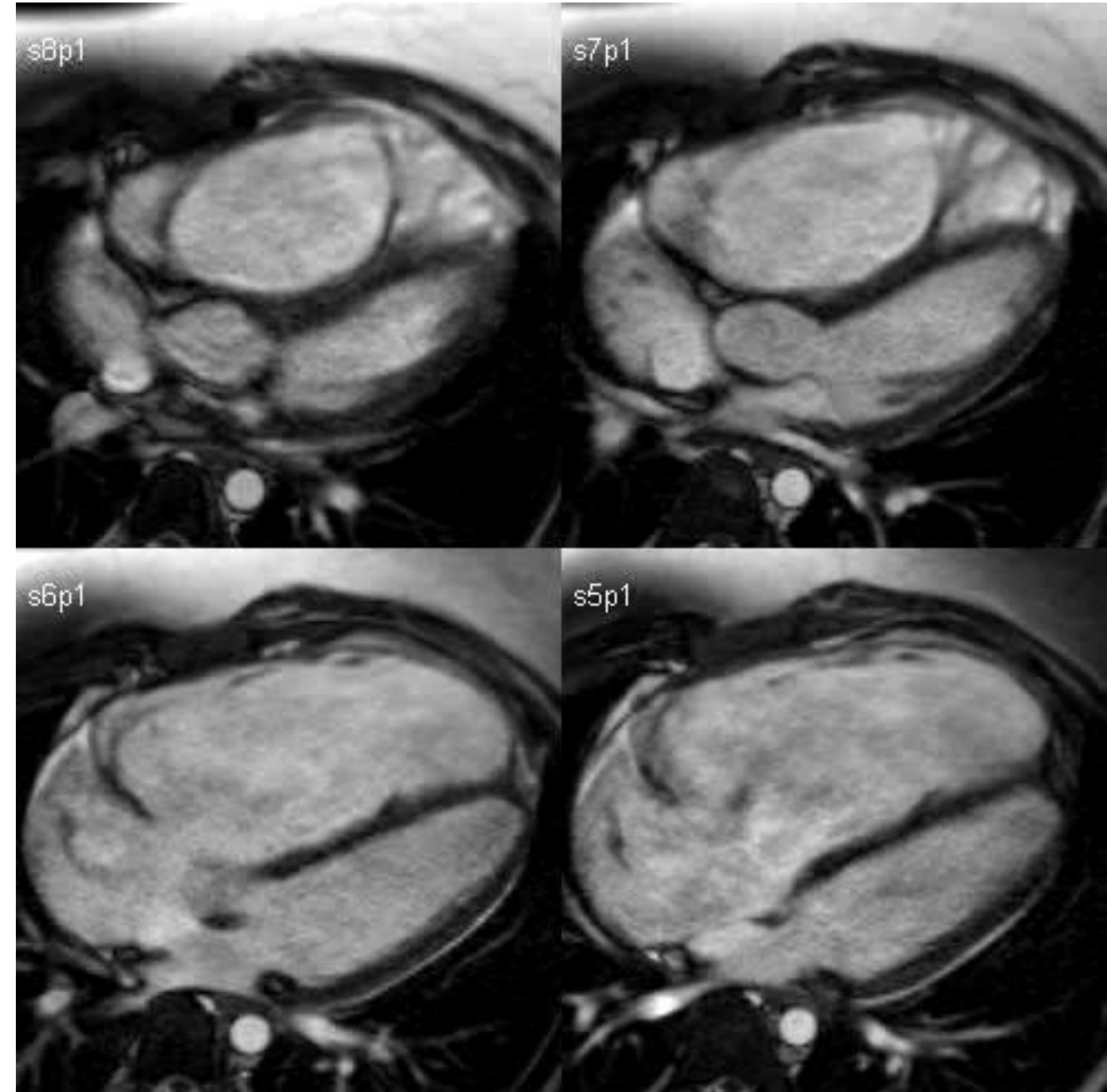




12Y8M

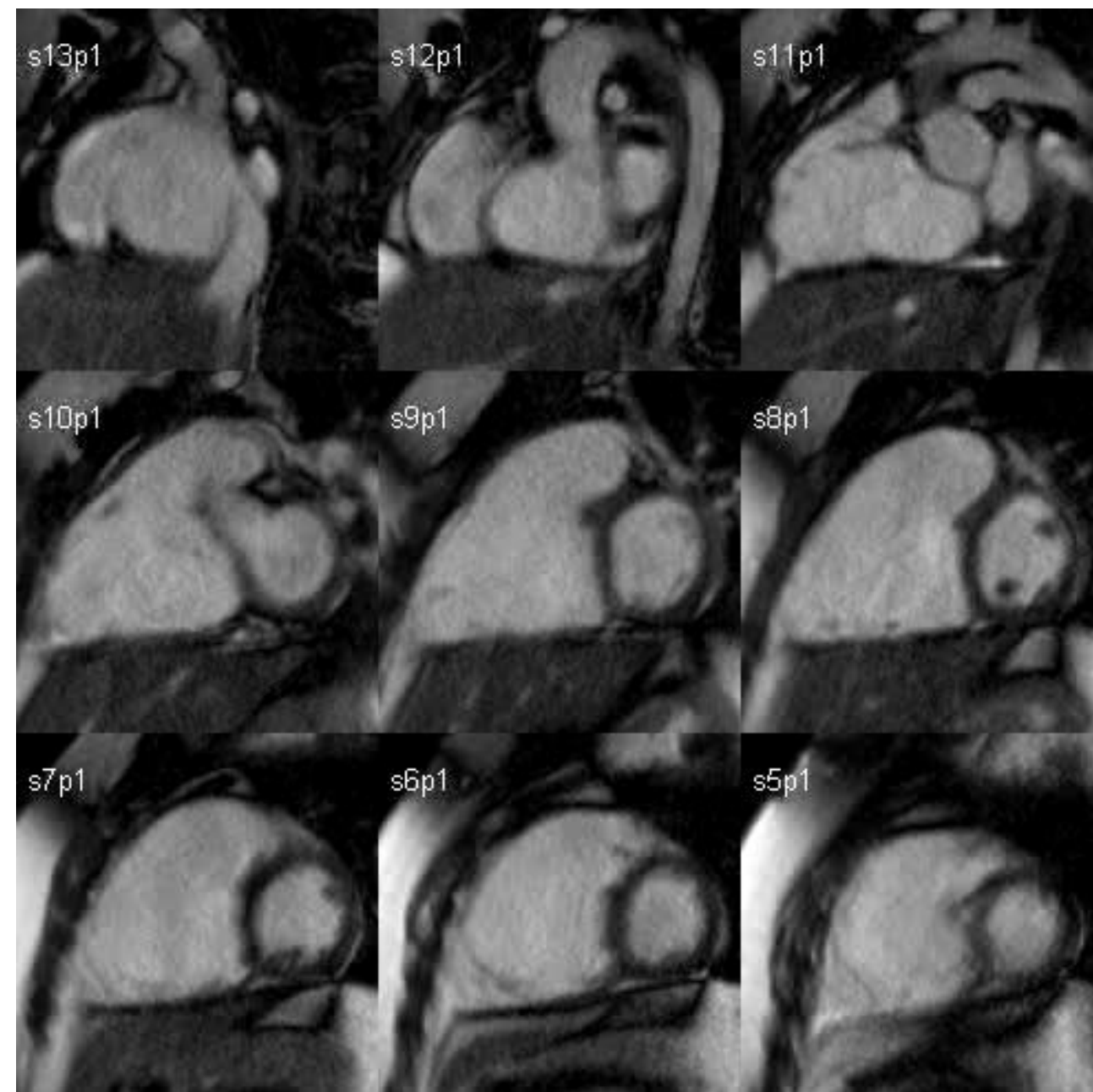
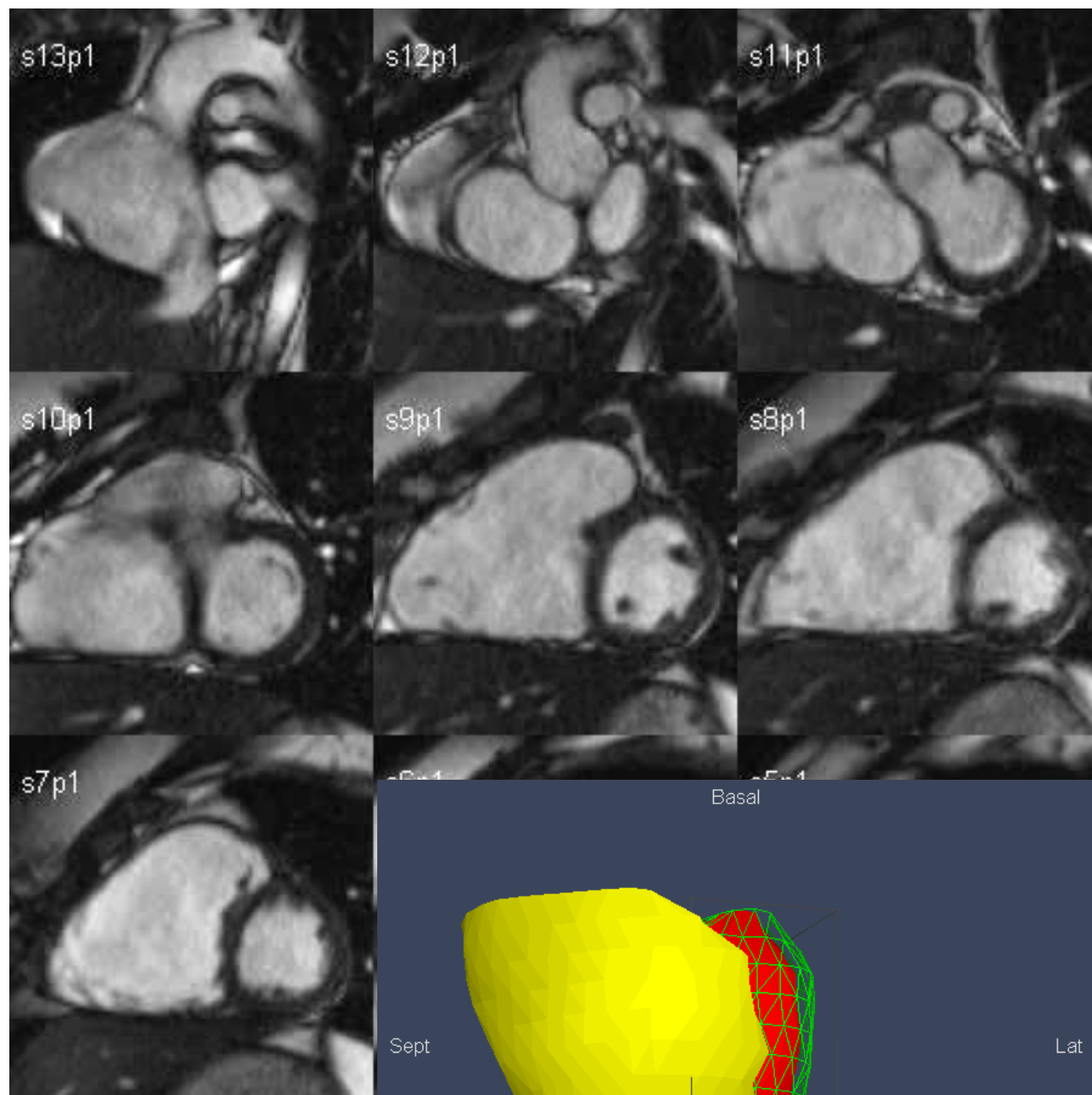


16Y



12Y8M

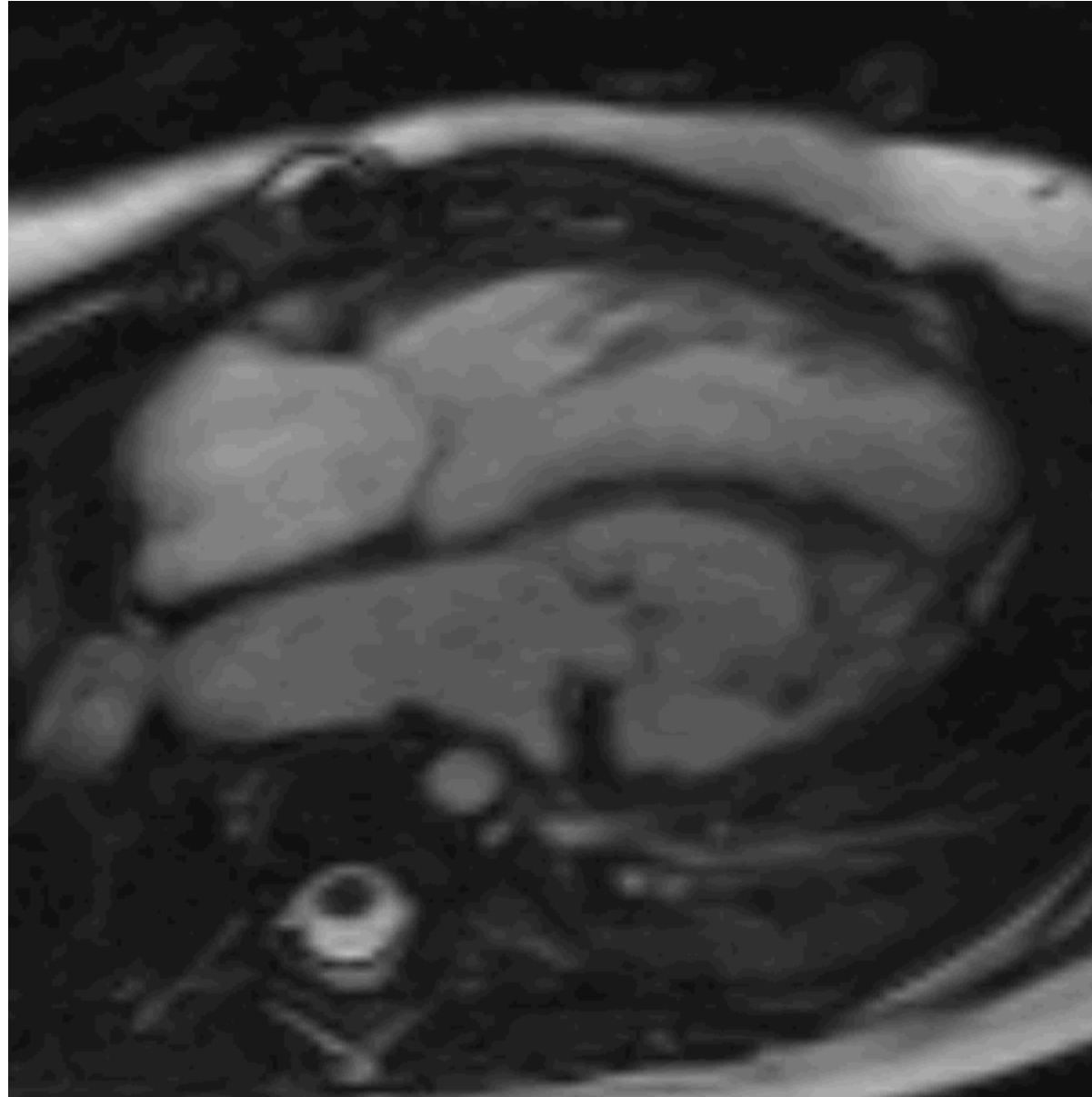
16Y

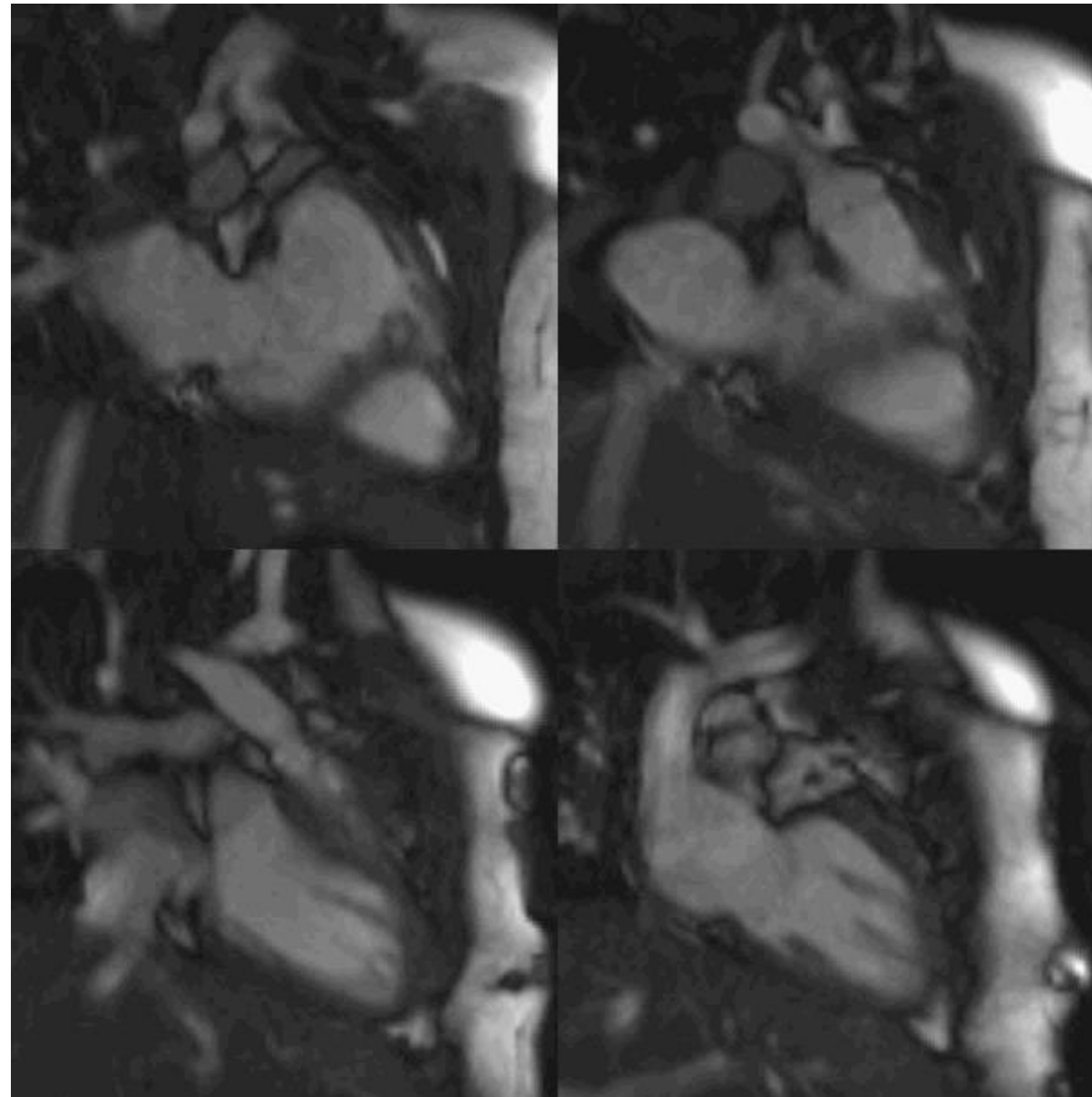


	2010/08/03 (12Y8M)	2012/12/17 (15Y)
Body parameters	163-77.4-1.83	170-102-2.12
Heart rate	100 bpm	90 bpm
RVEDVI (ml/m <sup>2</sup> )	160	239
RVESVI (ml/m <sup>2</sup> )	90	138
RVSVI (ml/m <sup>2</sup> )	70	101
RVEF (%)	44	42
LVEDVI (ml/m <sup>2</sup> )	51	64
LVESVI (ml/m <sup>2</sup> )	22	30
LVSVI (ml/m <sup>2</sup> )	29	34
LVEF (%)	54	53
Cardiac index (L/min/m <sup>2</sup> )	2.46	2.20
Right atrial volume	-	101
Estimated TR	-	5.4 L/min/m <sup>2</sup> (66%)

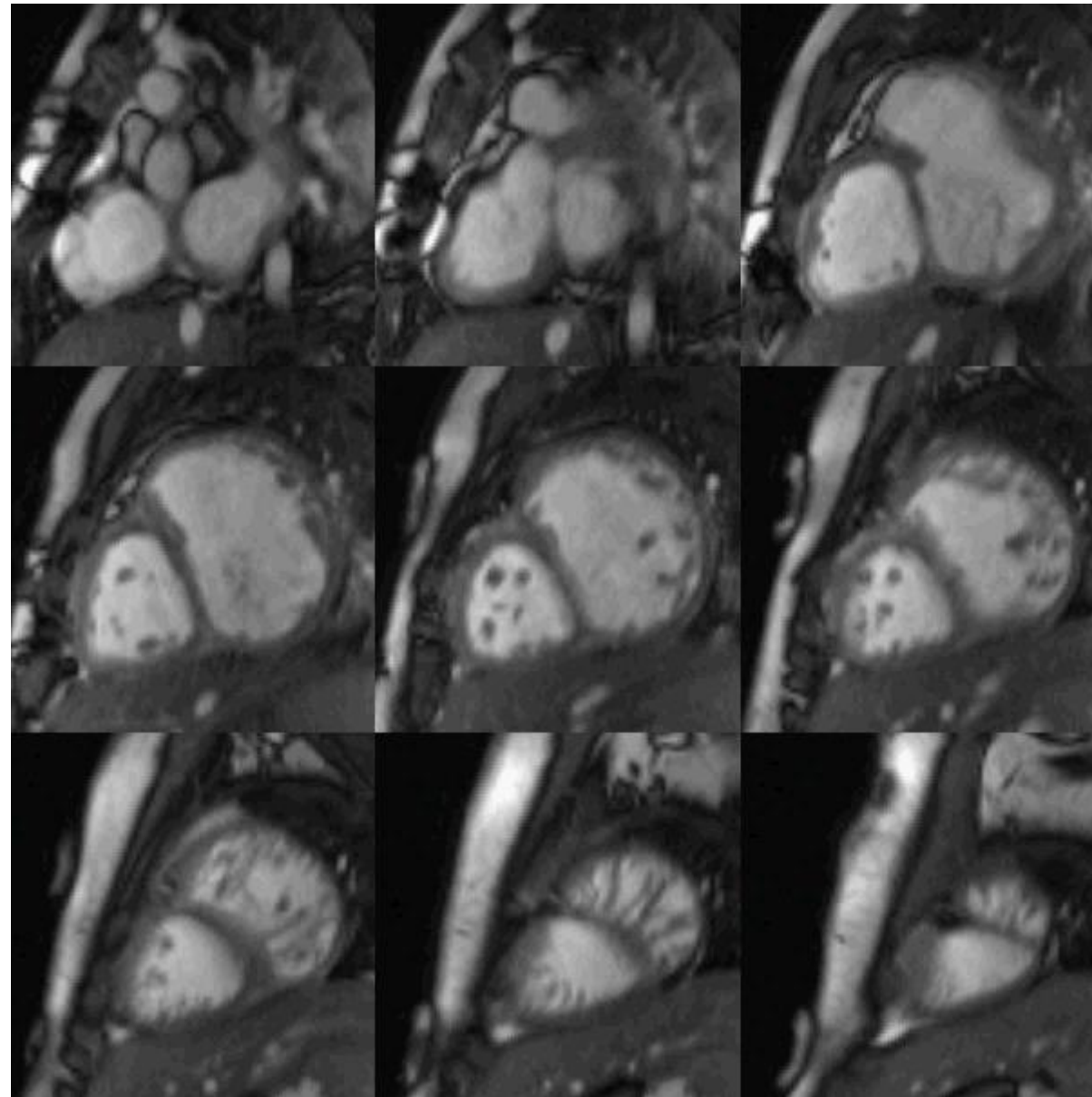
CCTGA Ebstein S/P PAB  
2745239

2y2m









# Tricuspid Repair for Ebstein's: Danielson

## Predictive Value of Preoperative Echocardiography

The echocardiographer attempted to predict the likelihood of successful surgical valve repair in 284 cases.

- Sensitivity was 59%,
- Specificity was 92%,
- Positive predictive value was 65%,
- Negative predictive value was 90%.

“Favorable echocardiographic criteria for TV repair include both *the valve leaflet location and morphology* and *the papillary muscle location and attachments*. Valves that have **severe leaflet displacement into the RV apex** or those anteriorly rotated into the RVOT are generally not suitable for the traditional monocusp repair.”

