Surgical Techniques for the repair of Ebsteins' Anomaly

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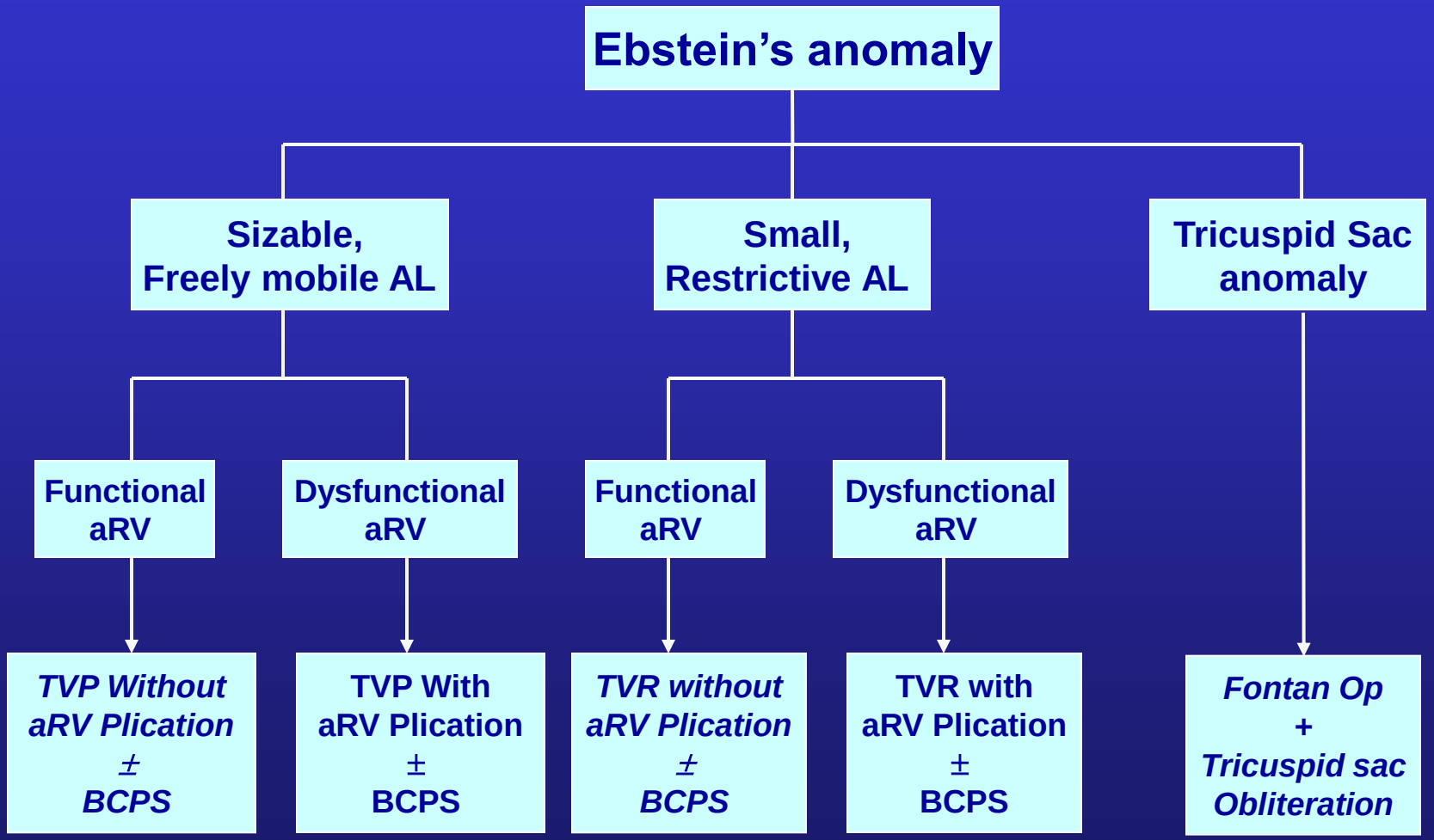
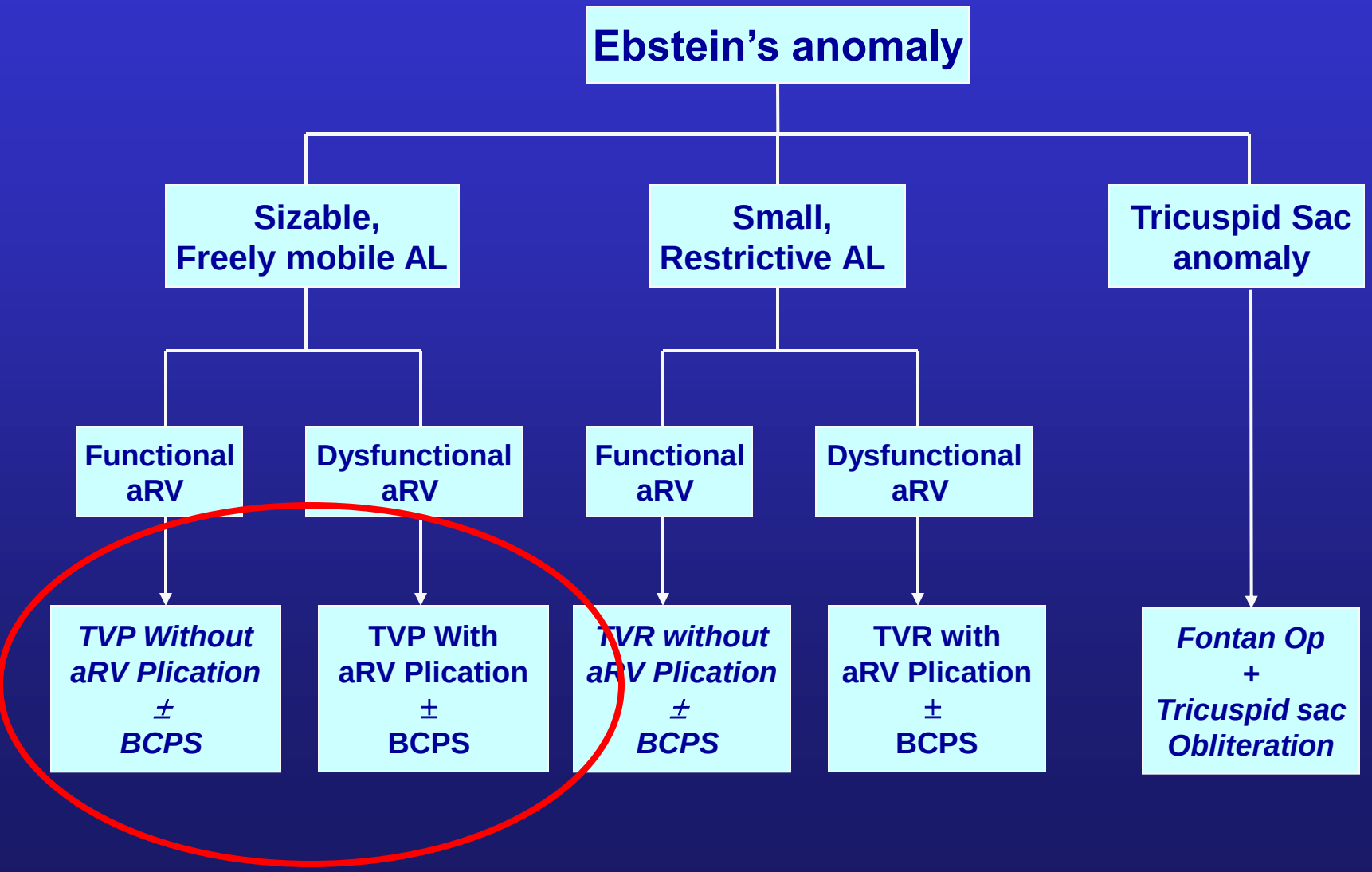
Surgical algorithm (AMC)



Surgical algorithm (AMC)



Typical scenario

- M / 45, Carpentier type B
- Functional class III
- Op finding: Hugely dilated RA
  Large, thin-walled aRV
  Small functional RV
- Operation
  TVP (cone procedure)
  → High CVP, RV dysfunction, severe TR on Echo
  → Addition of TAP without success
  → TVR with bioprosthesis
- Rocky postoperative course with RV dysfunction

Ebstein anomaly: Myths

- **TV repair is technically feasible in all Ebstein anomaly patients!**
- **Surgical outcome of each surgical technique is excellent with minimal residual TR !**
- **Excellent early surgical outcome can lead to excellent long term outcome !**

Surgical outcome of Ebstein Anomaly
(Chen et al, JTCVS 2004)

- Children's hospital of New York
- 1990-2002, 25 Patients
- Age: 2 m – 46.8 yrs (median: 8.2 yrs)
- Carpentier technique
- Residual moderate to severe TR: 13/25(52%)
- Reoperation (3) : TVR(2), Addition of BCPS (1)
- Late death: 2 patients with severe residual TR

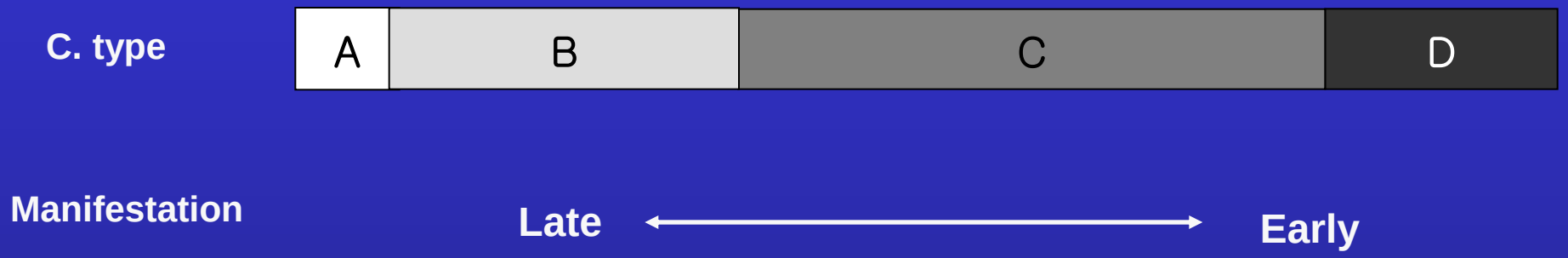
Repair of Ebstein anomaly is tricky !

- Wide spectrum of disease
- Difficult to define morphology
- Limited experience
- No standardized surgical strategy
- Various surgical techniques

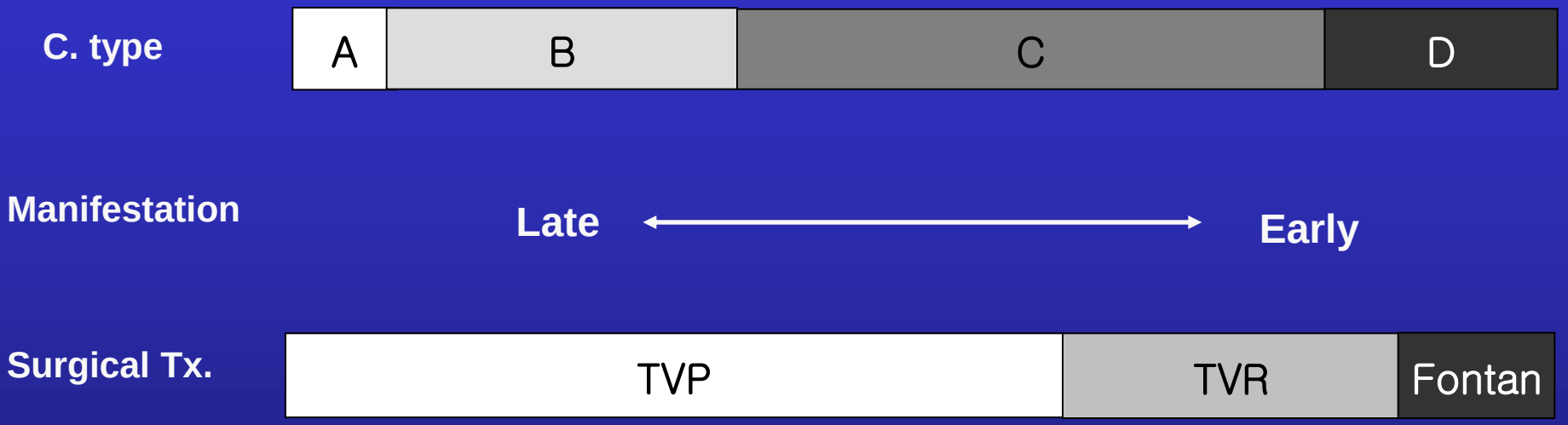
Ebstein Anomaly as a Spectrum

C. type

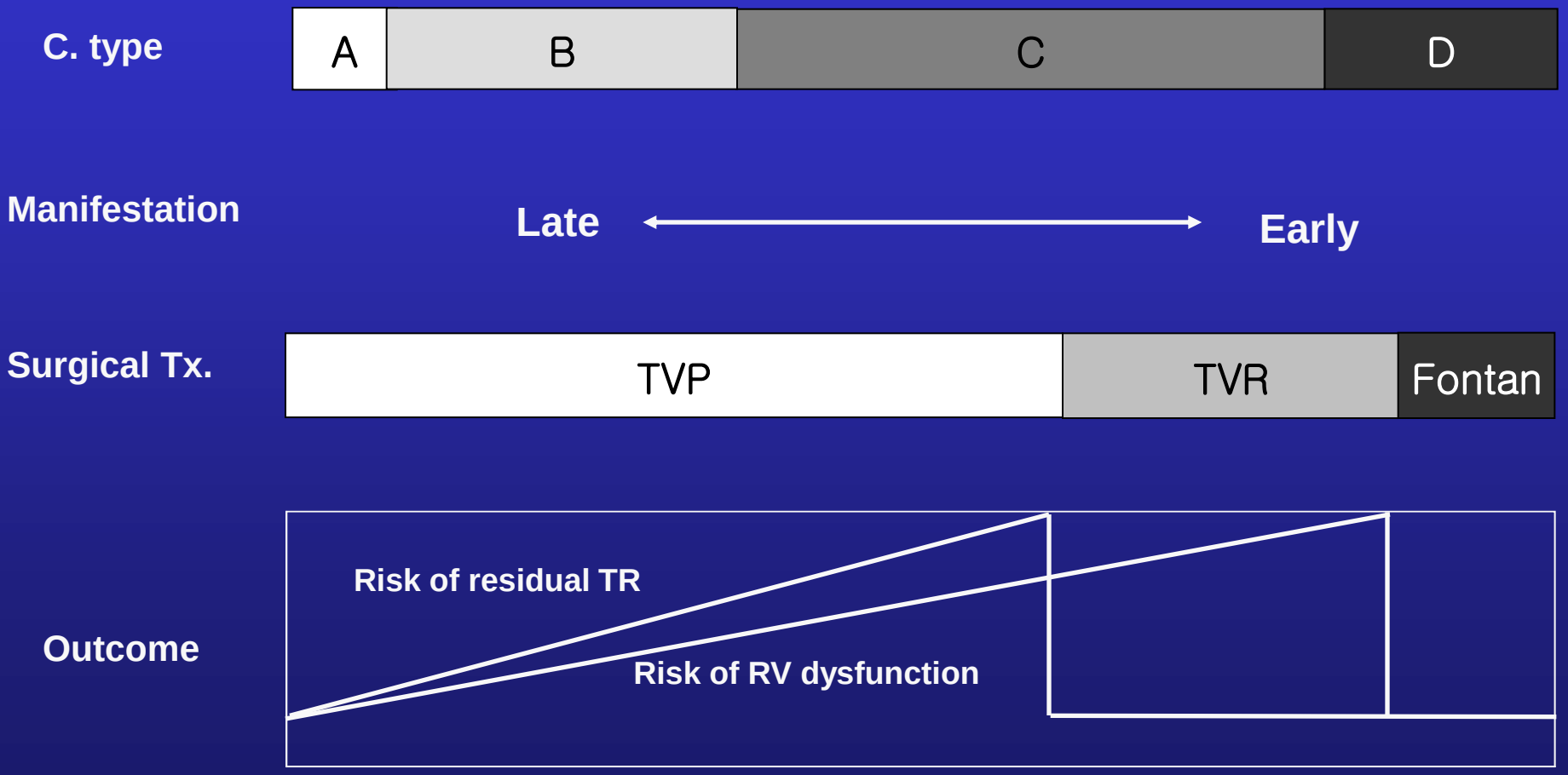
Ebstein Anomaly as a Spectrum



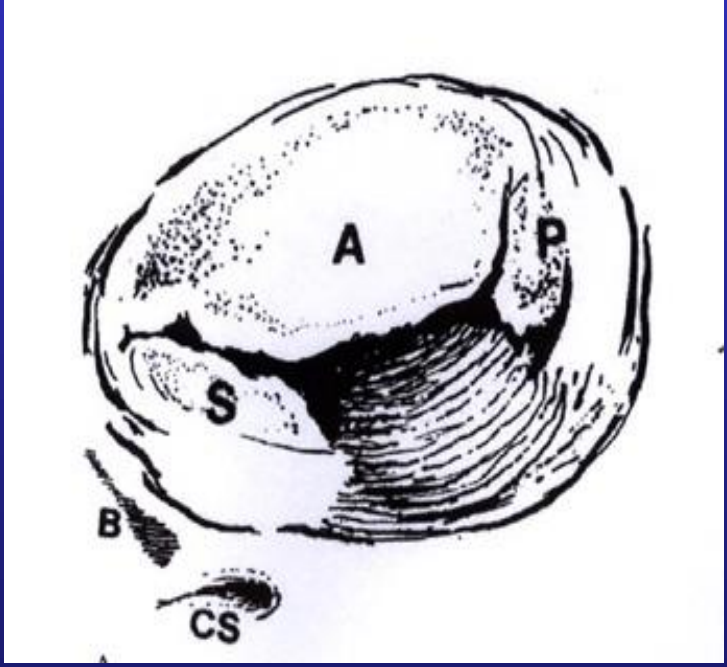
Ebstein Anomaly as a Spectrum



Ebstein Anomaly as a Spectrum



Delineation of TV leaflets



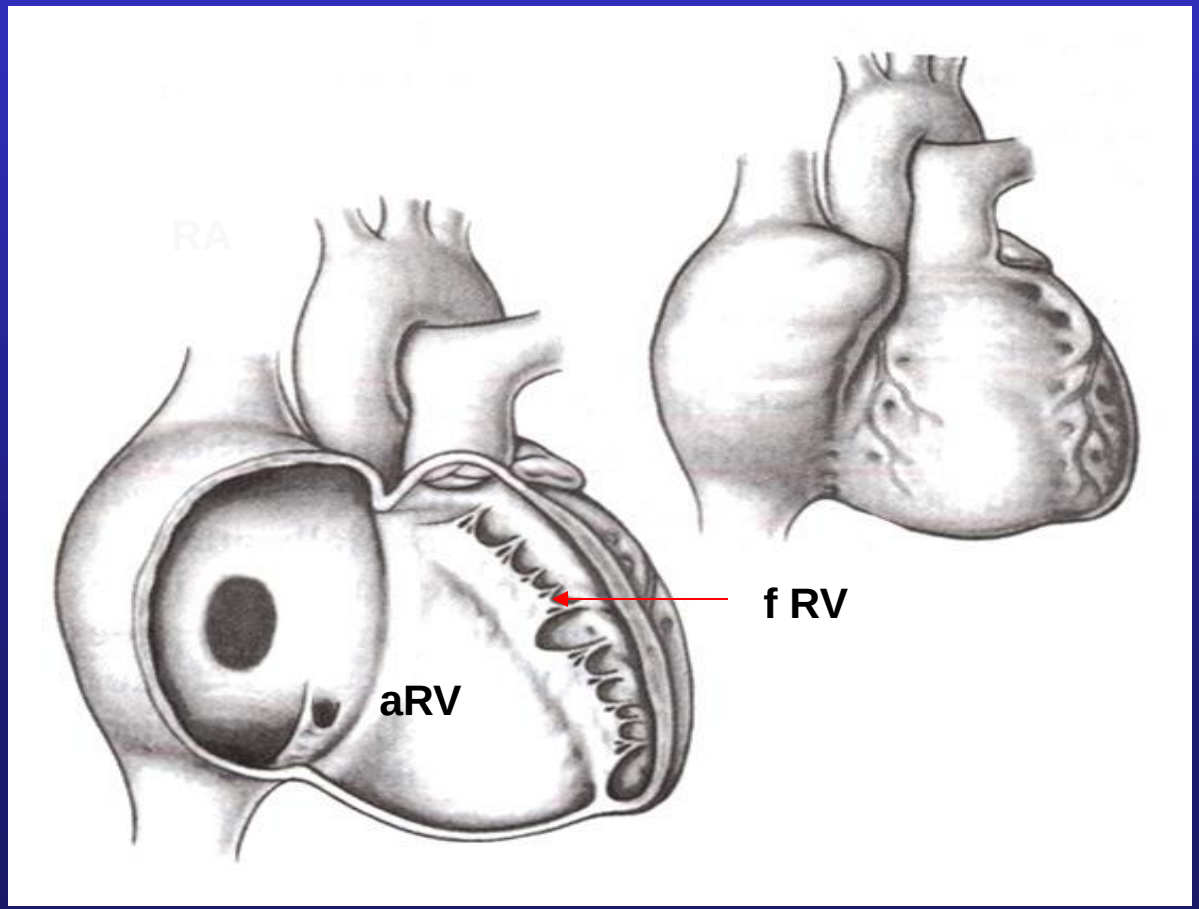
1. Commissures in systole
2. Papillary muscles

⇒ Difficult in Ebstein Anomaly

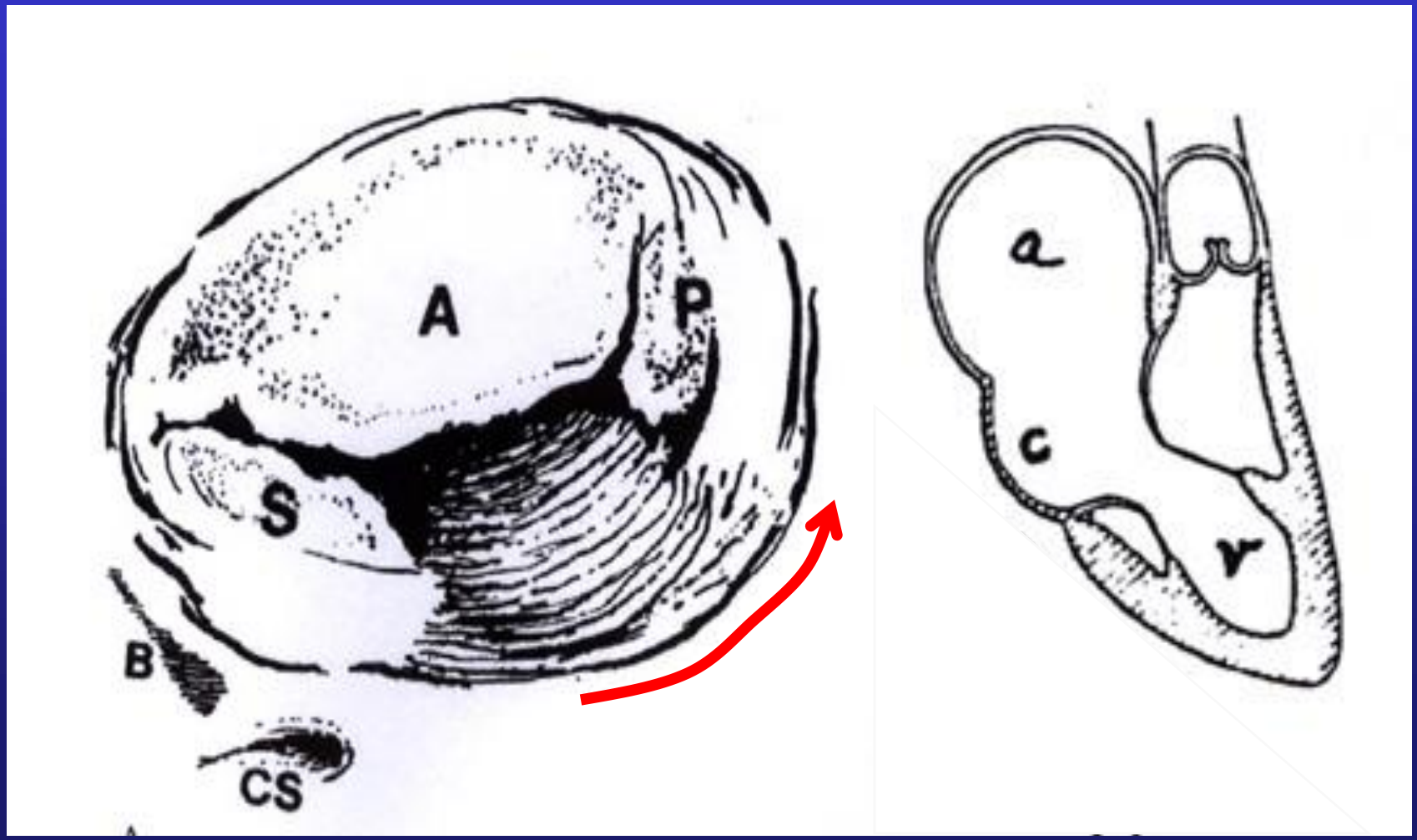
Ebstein anomaly :

Delamination failure of TV leaflets

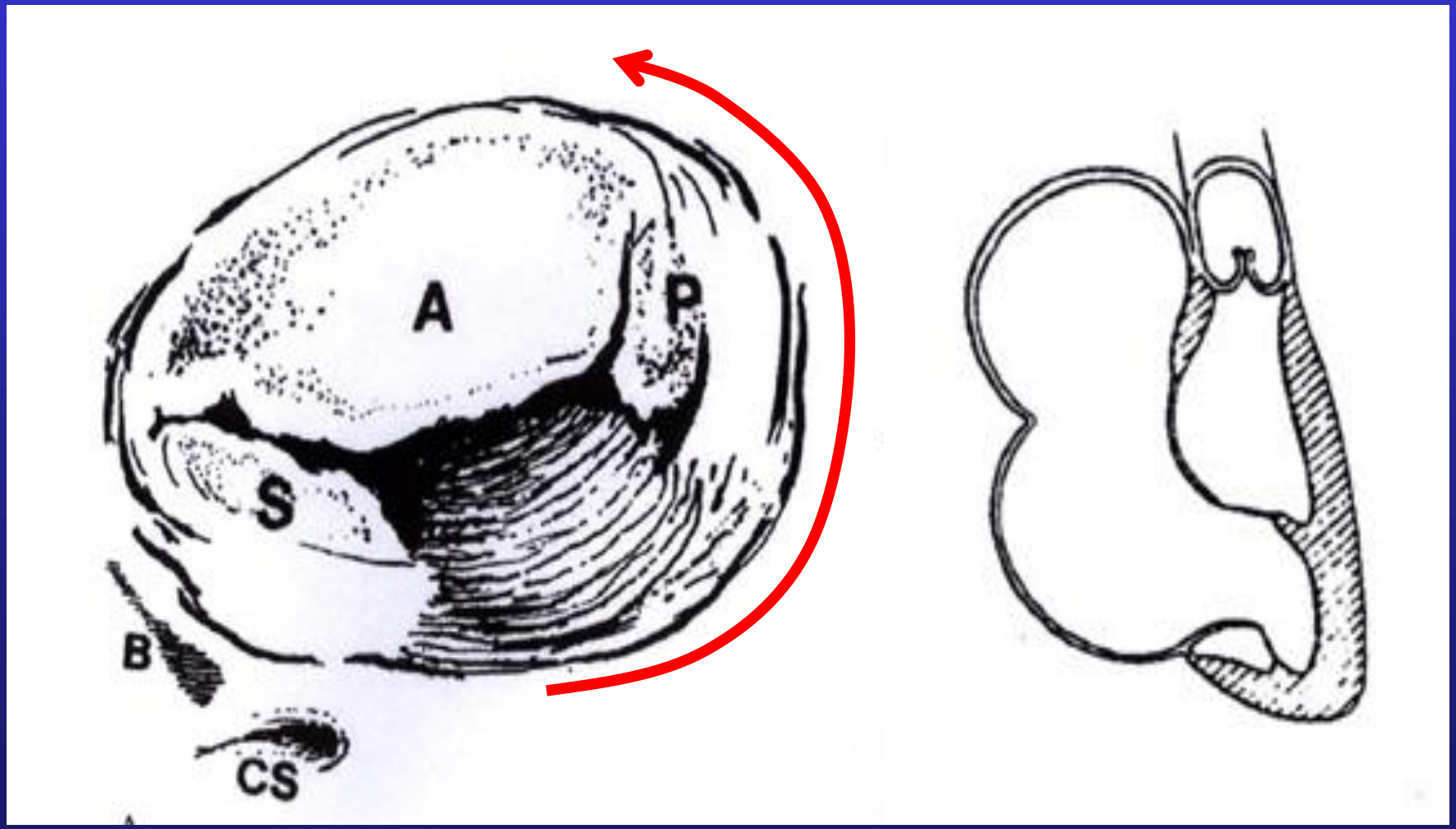
→ Downward displacement of valve mechanism



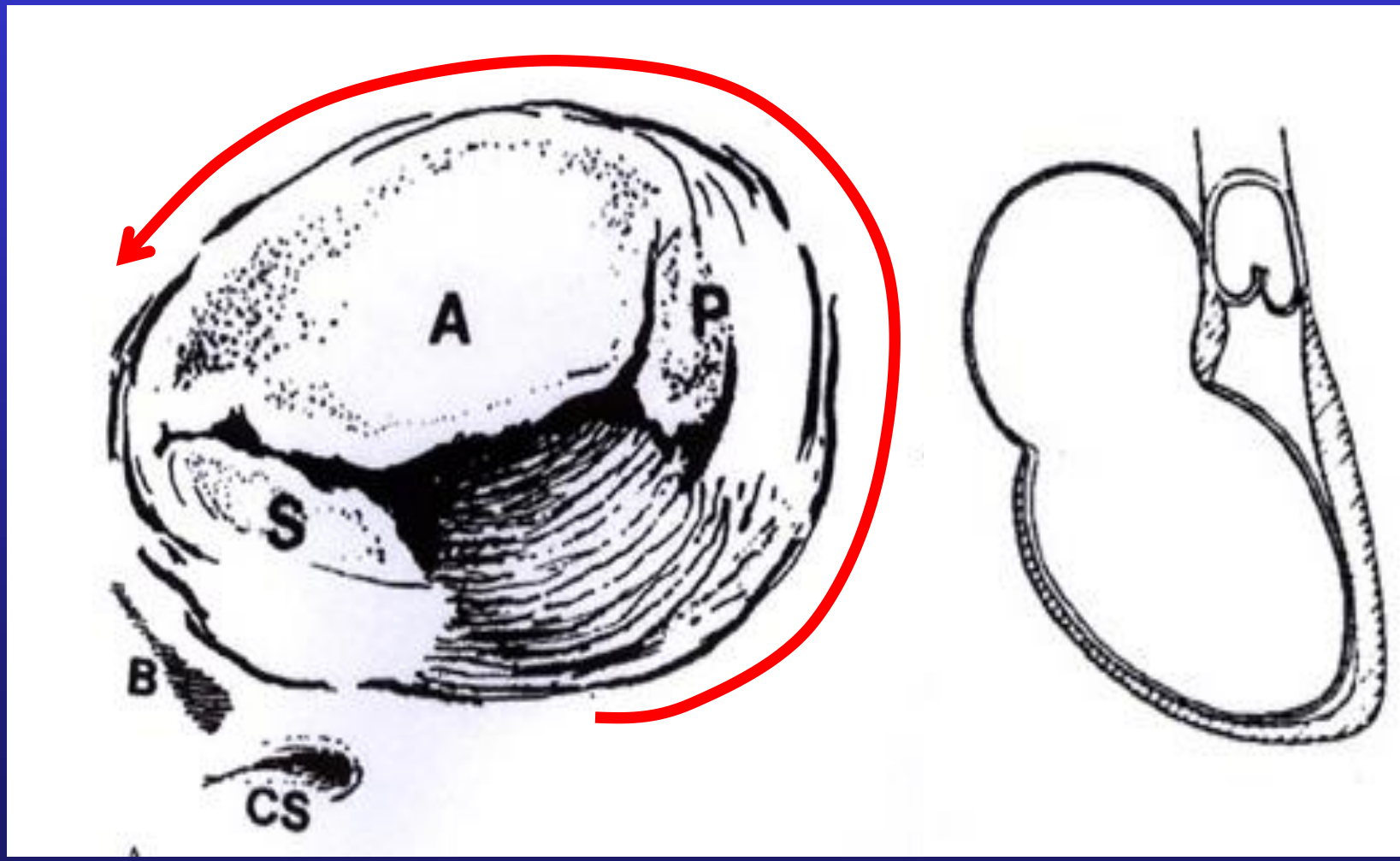
Degree of Delamination failure



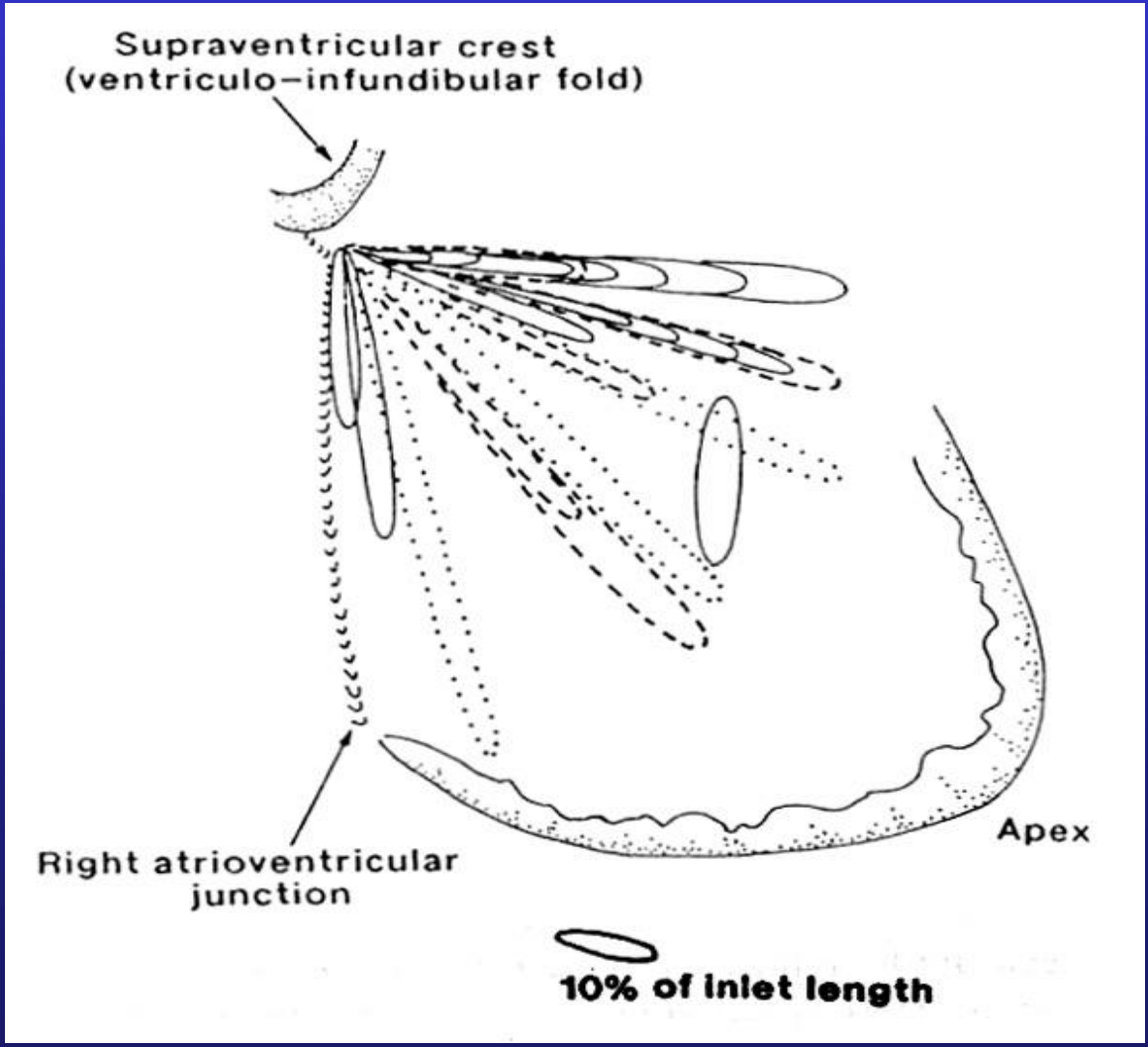
Degree of Delamination failure



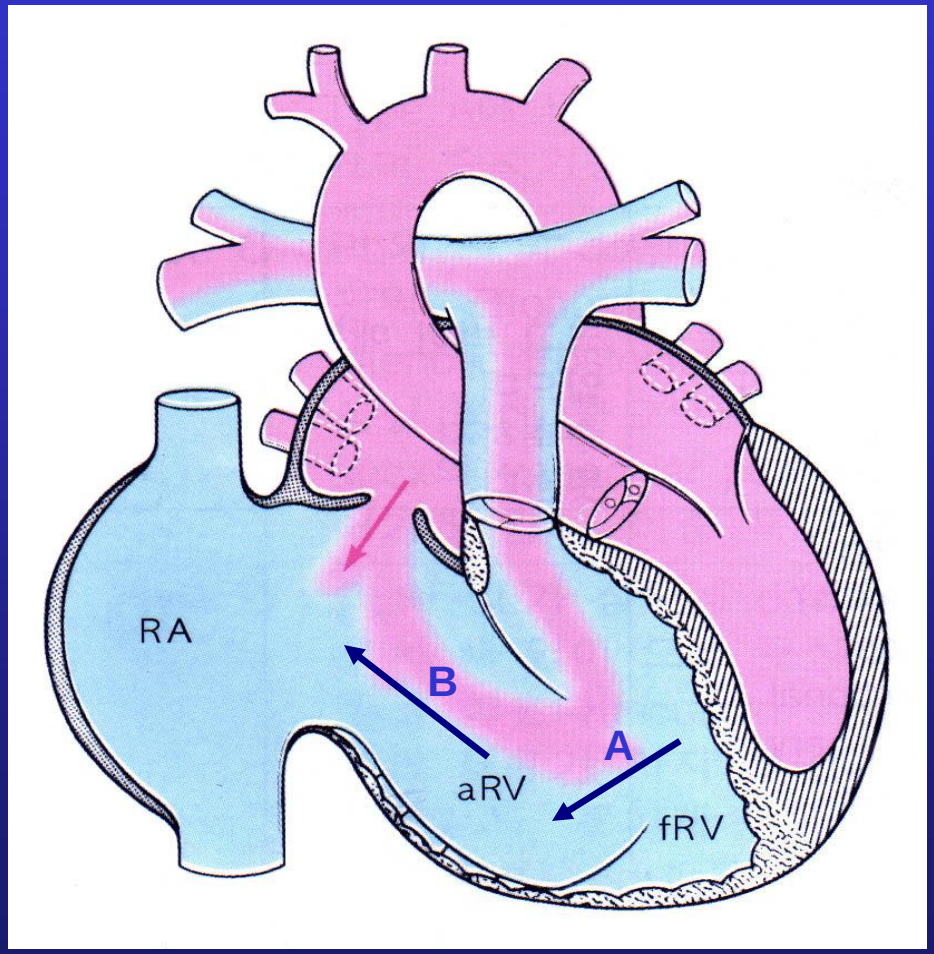
Degree of Delamination failure



Effective valve orifice in 23 specimens
(Schreiber et al, JTCVS 1999)



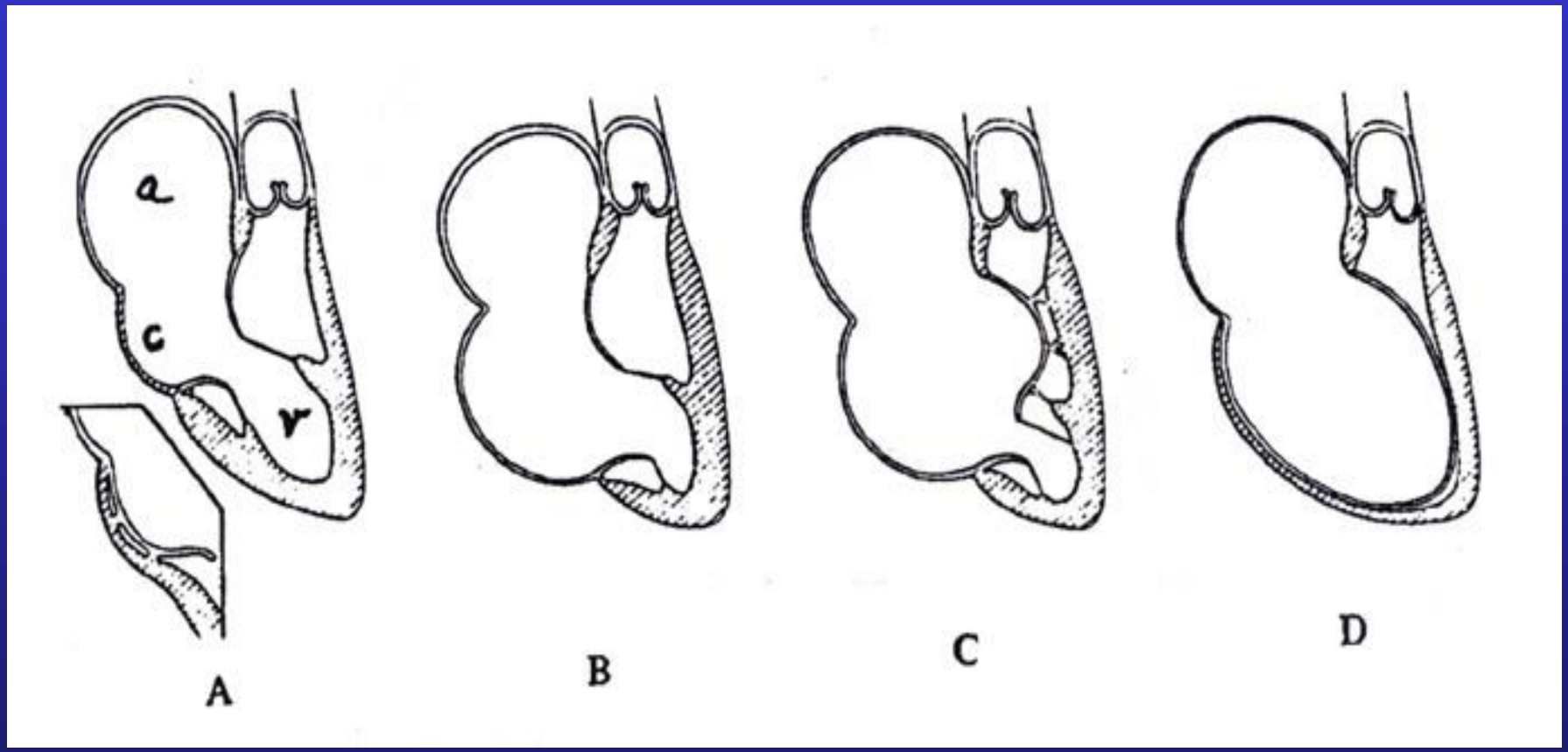
Tricuspid Regurgitation ?



A: f RV ⟶ aRV

B: aRV ⟶ RA

Carpentier Classification
(Carpenter, 1988)



Celermajer classification

(Celermajer, 1992)

$$\text{Area ratio (AR)} = \frac{\text{RA area + aRV area}}{\text{f RV area + LV area + LA area}}$$

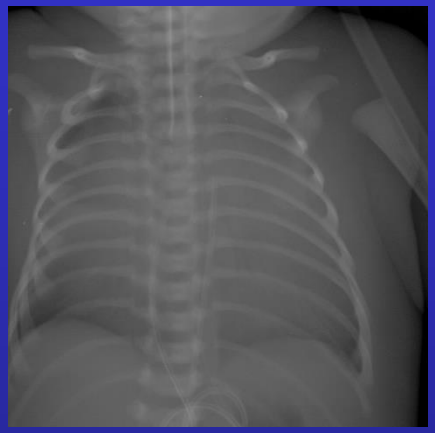
Grade I : AR < 0.5

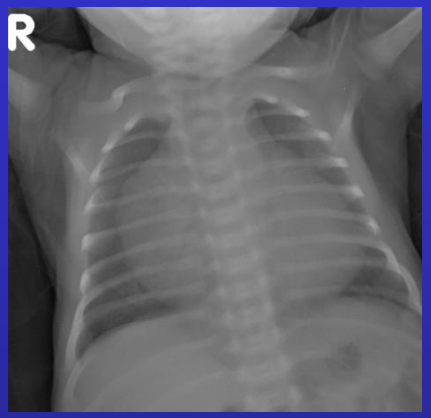
Grade II : 0.5 ≤ AR ≤ 0.99

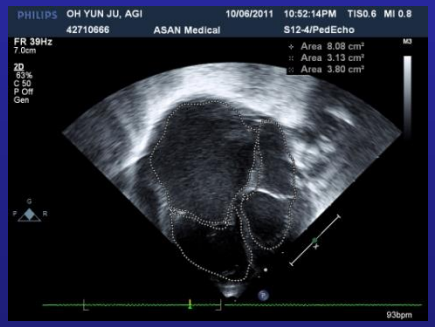
Grade III : 1.0 ≤ AR ≤ 1.49

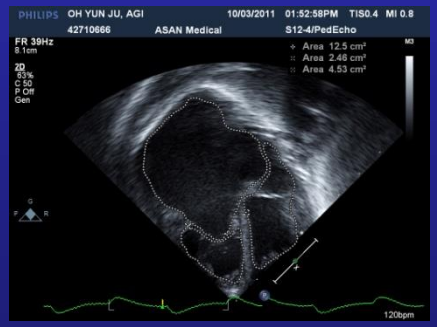
Grade IV : 1.5 ≤ AR

Celermajer index changes!









Postnatal day 0
Area idex = 1.79

Postnatal day 3
Area idex = 1.5

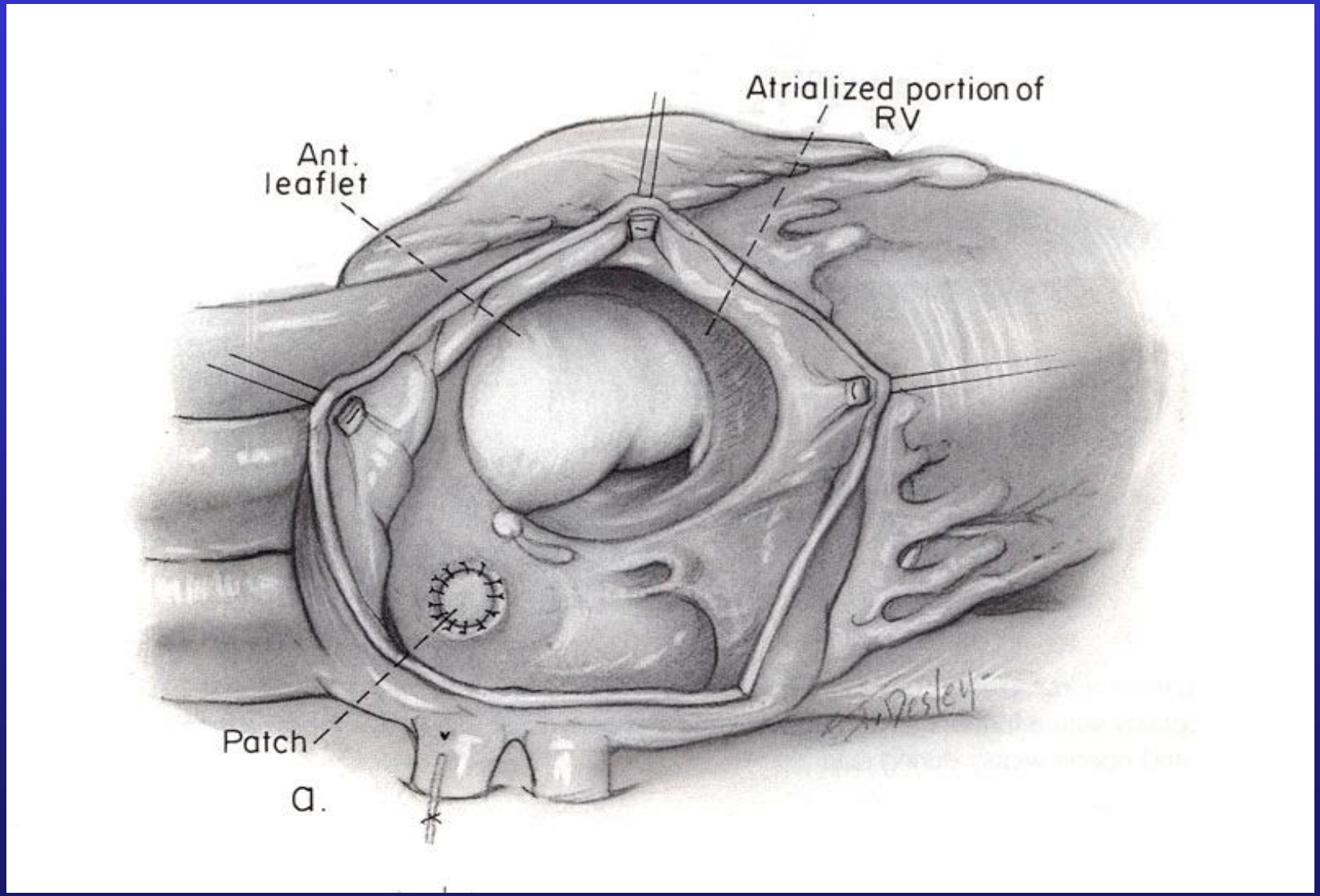
Postnatal day 21
Area idex = 0.96

Postnatal 6 m
(post 1 ½ repair)

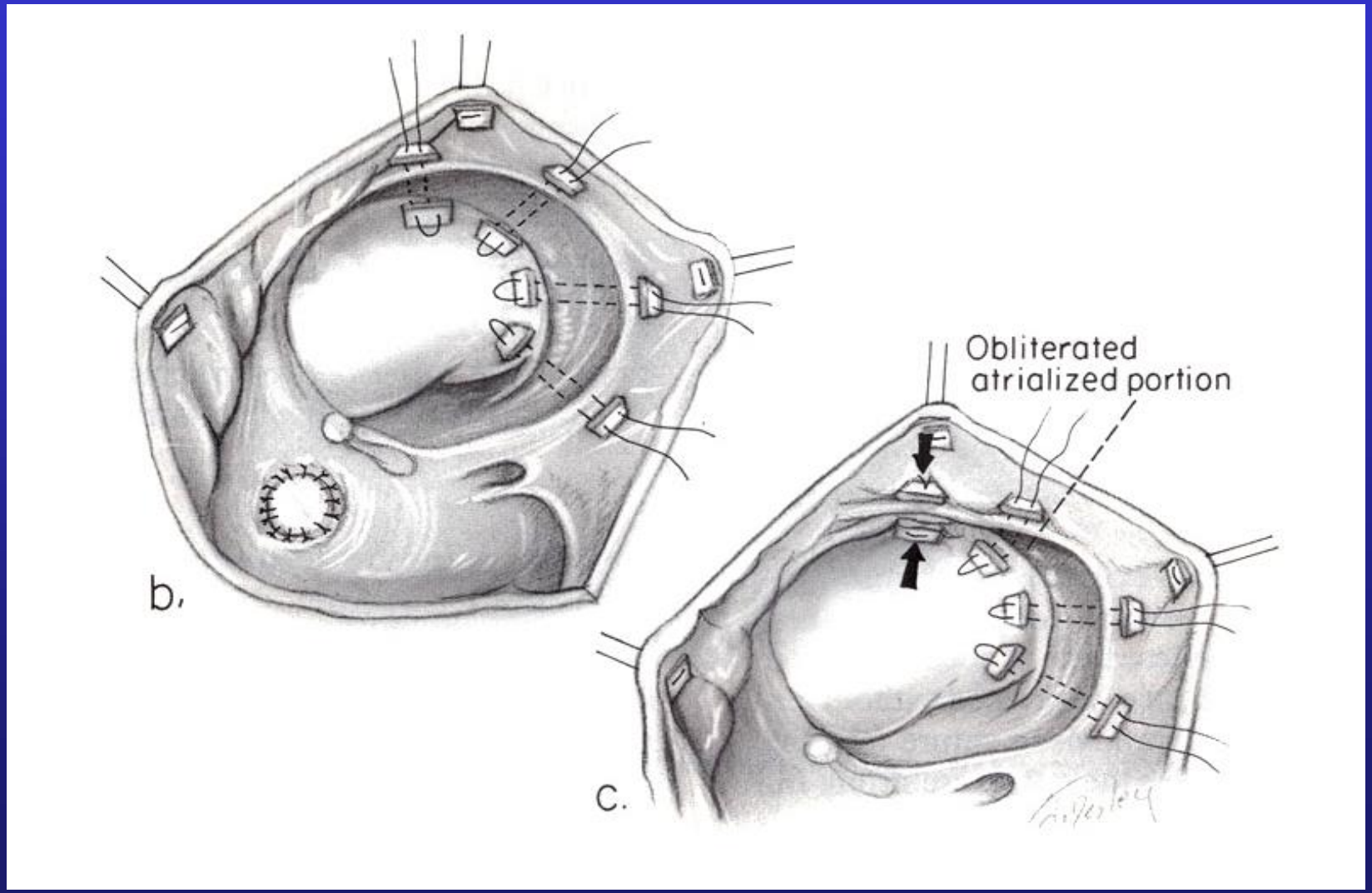
TV repair techniques

- Danielson technique
- Carpentier technique
- Cone procedure
- ‘Wu’ technique
- Hetzer technique

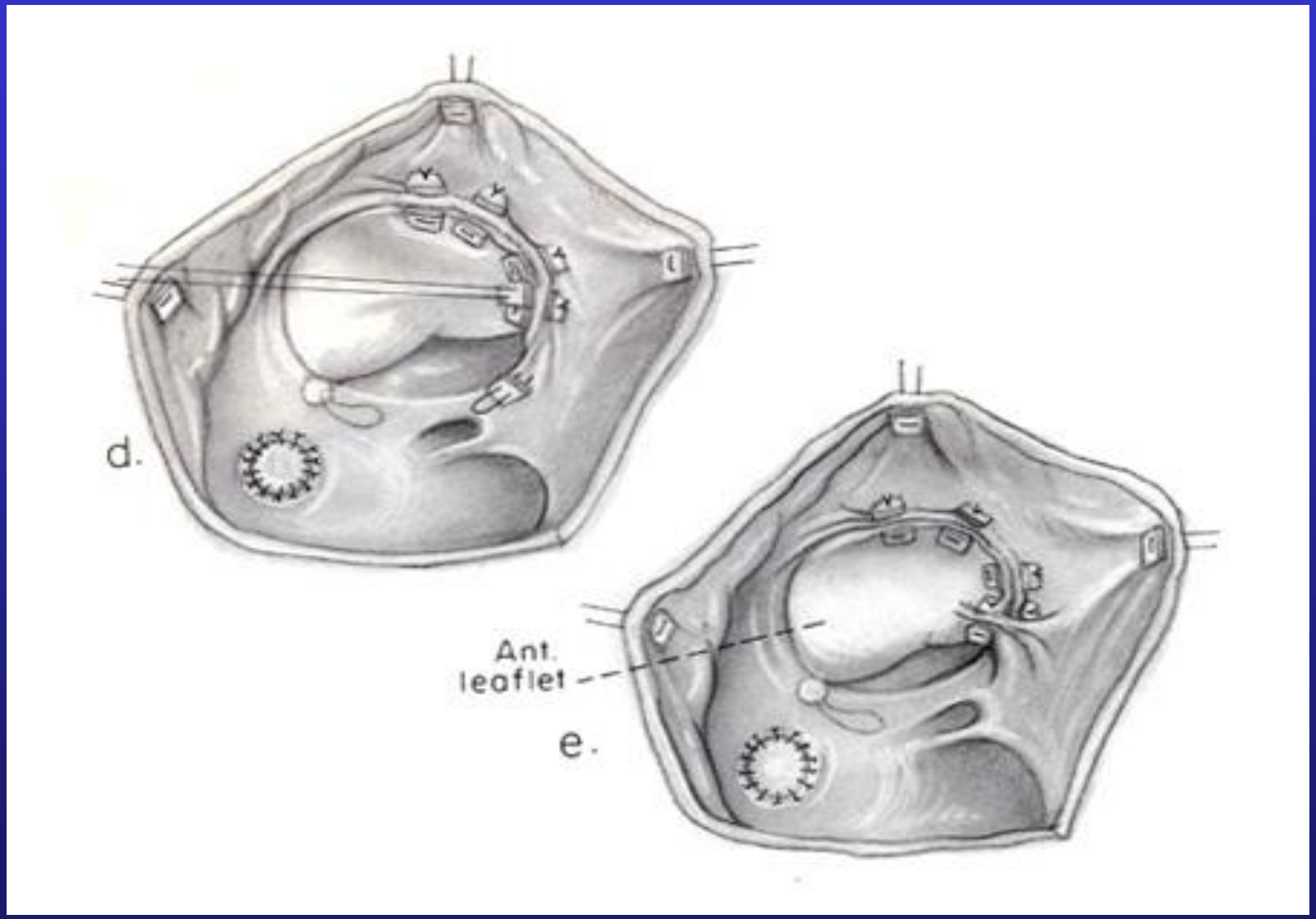
Danielson Repair



Danielson Repair



Danielson Repair

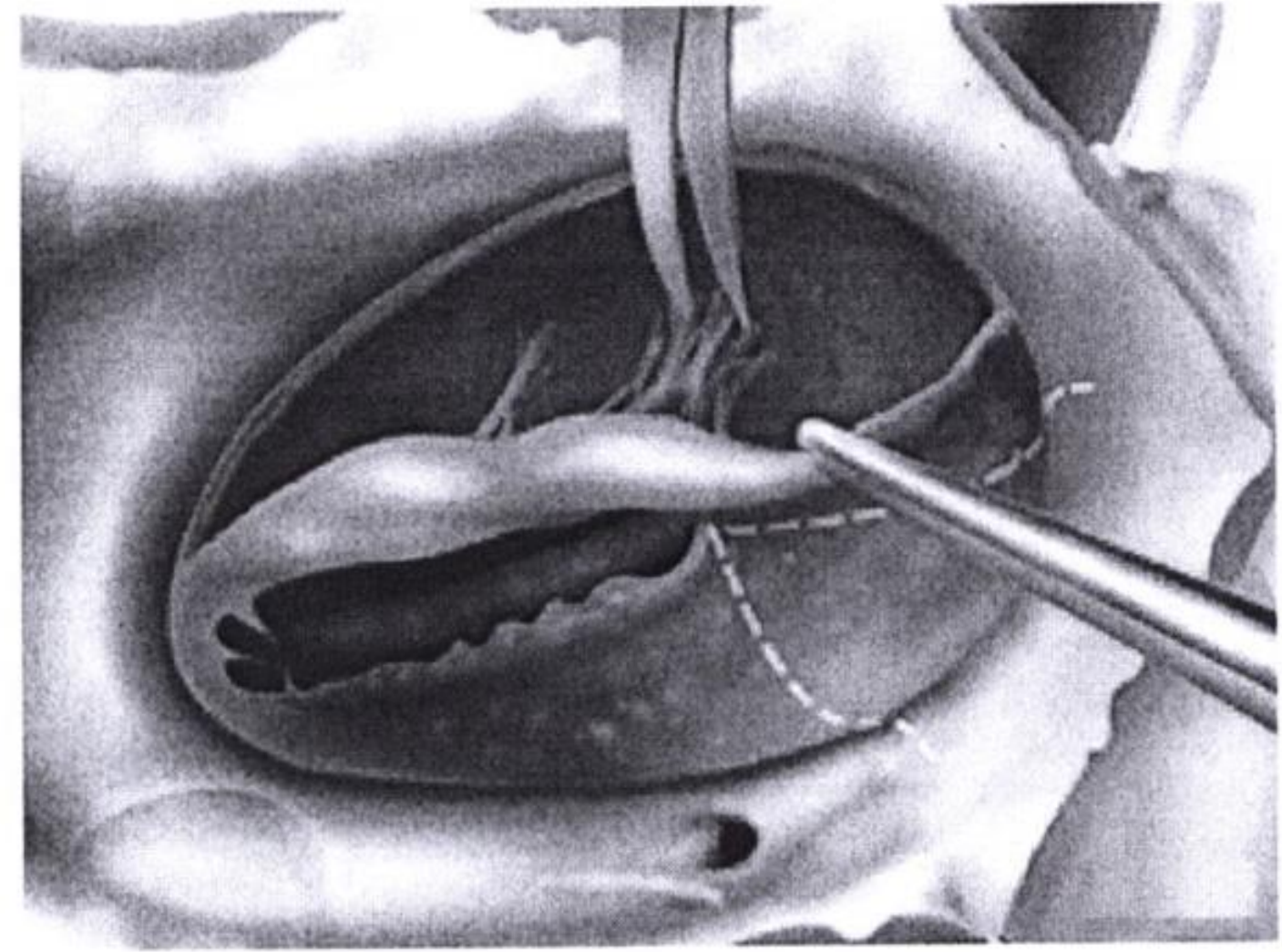


Operative series of Ebstein Anomaly

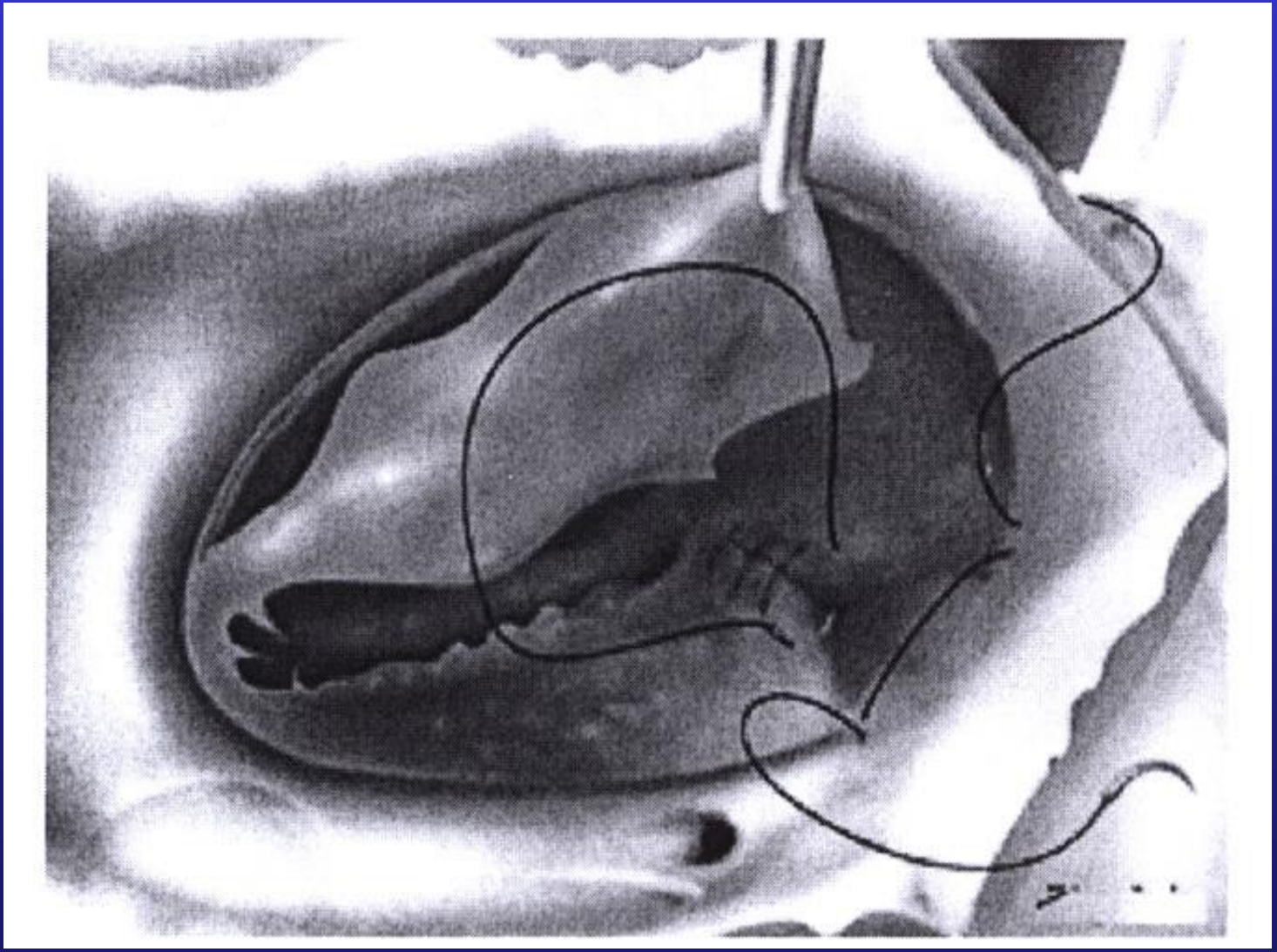
Source	Pt. No.	Age (median)	TVP (%)	Op.Mortality
Lilehei, 1967	8	6-18 yr (12)	25%	50%
Hardy and Roe,1969	6	5-41 yr (22)	100%	17%
Westaby, 1982	24	3-55 yr (20)	4%	17%
*Carpentier, 1988	14	9-51 yr (26.9)	93%	14%
Quagebeur, 1991	10	4-44 yr (22)	100%	0%
Starnes, 1991	5	1-9 d (5)	0%	0%
†Danielson, 1992	189	1-64 yr (19.1)	58.2%	6.3%
Hetzer, 1998	19	2-54 yr (21)	100%	5%
†Kiziltan, 1998	158	1-70 yr (19)	0%	6%
Marianeschi, 1998	10	2.4-31 yr (9)	60%	0%
Vargas, 1998	7	7-16 yr (12)	100%	0%
Chauvaud, 2000	142	1-65 yr (25)	97%	10%
Chauvad, 2003	191	1-65 yr (24.5)	98%	9%

* Clinical reports from the ‘Hospital Broussais † Clinical reports from ‘Mayo Clinic’

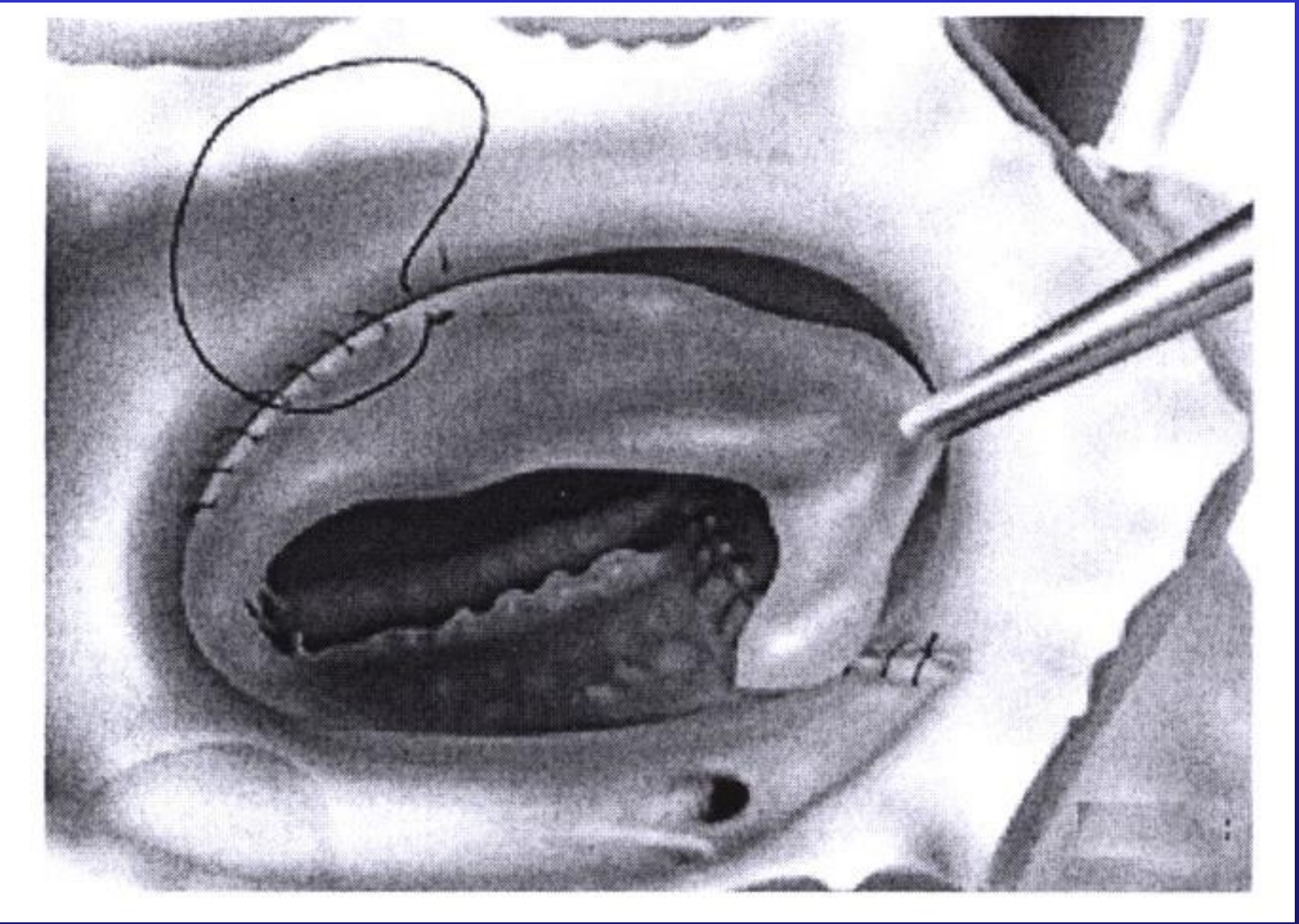
Carpentier repair (1)



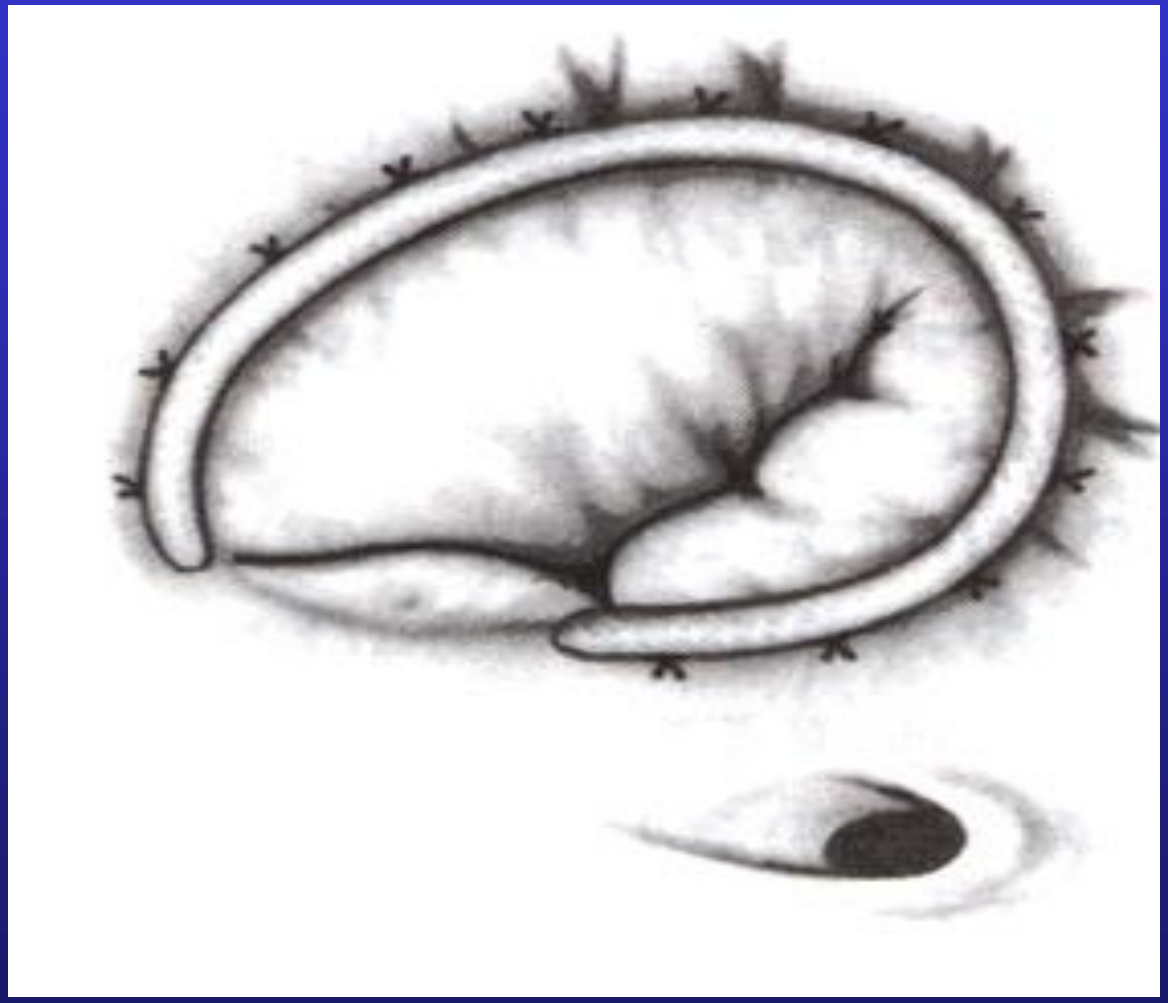
Carpentier repair (2)



Carpentier repair (3)

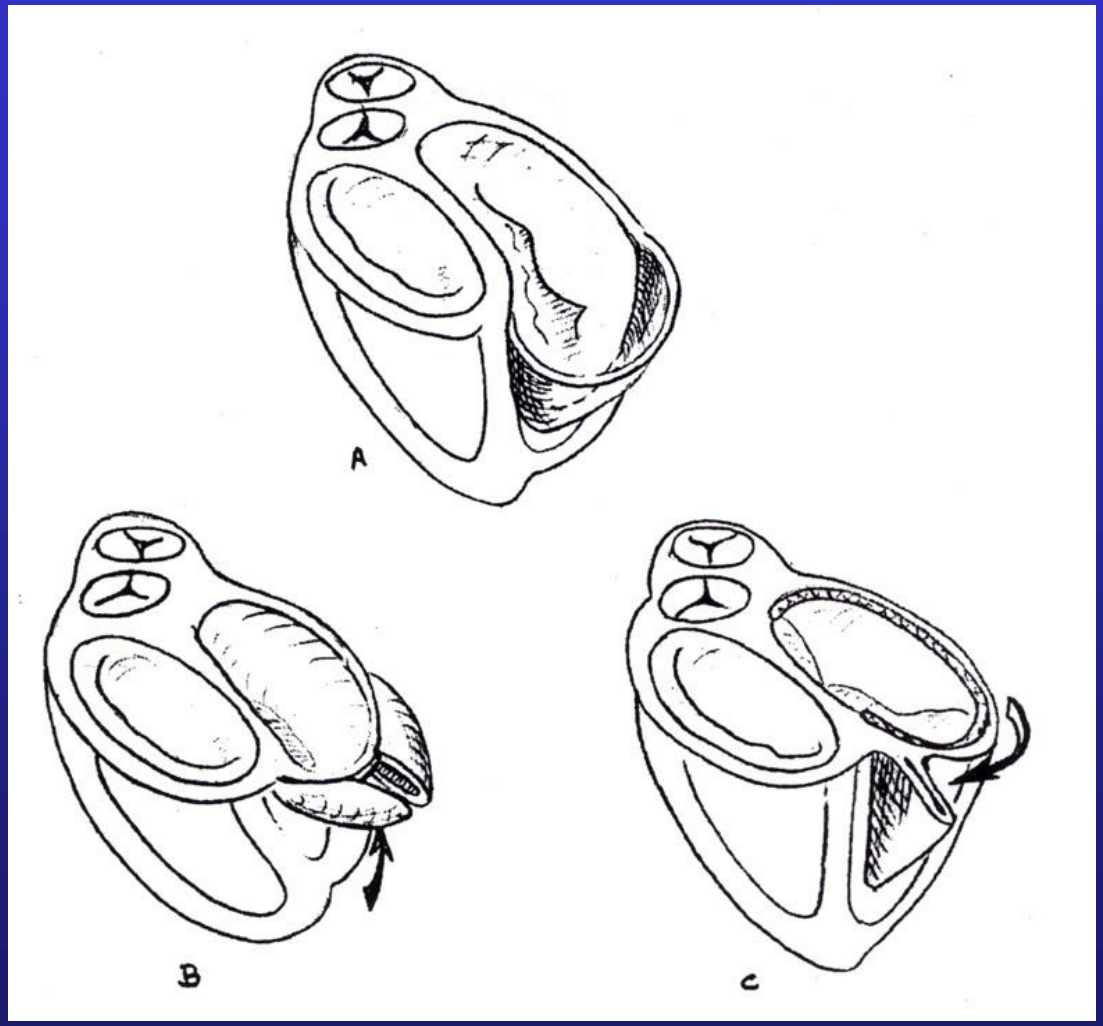


Carpentier repair (4)



Advantages of vertical plication of aRV

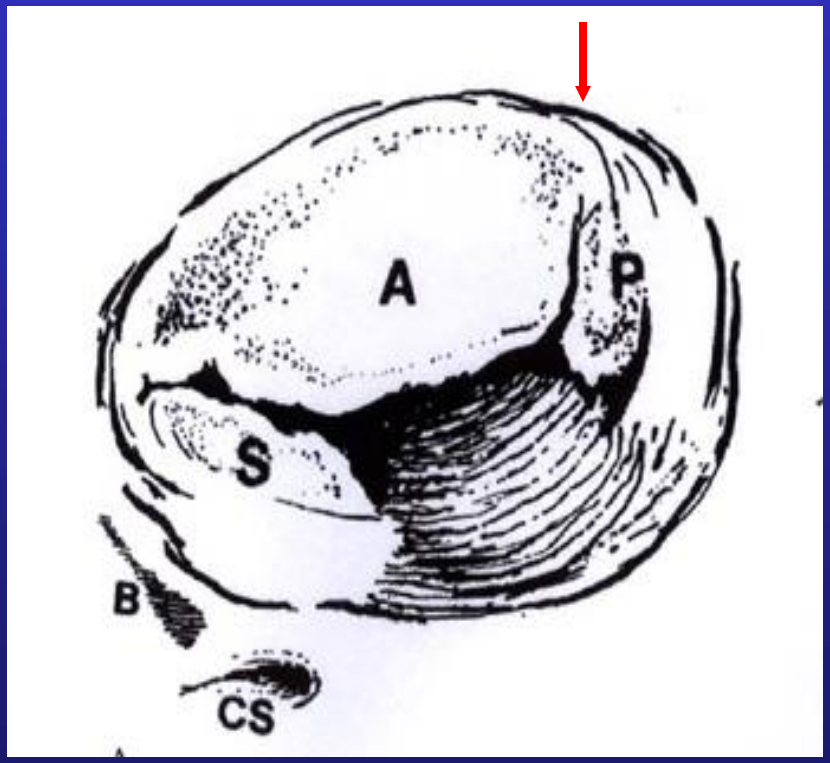
Transverse (Horizontal) Plication



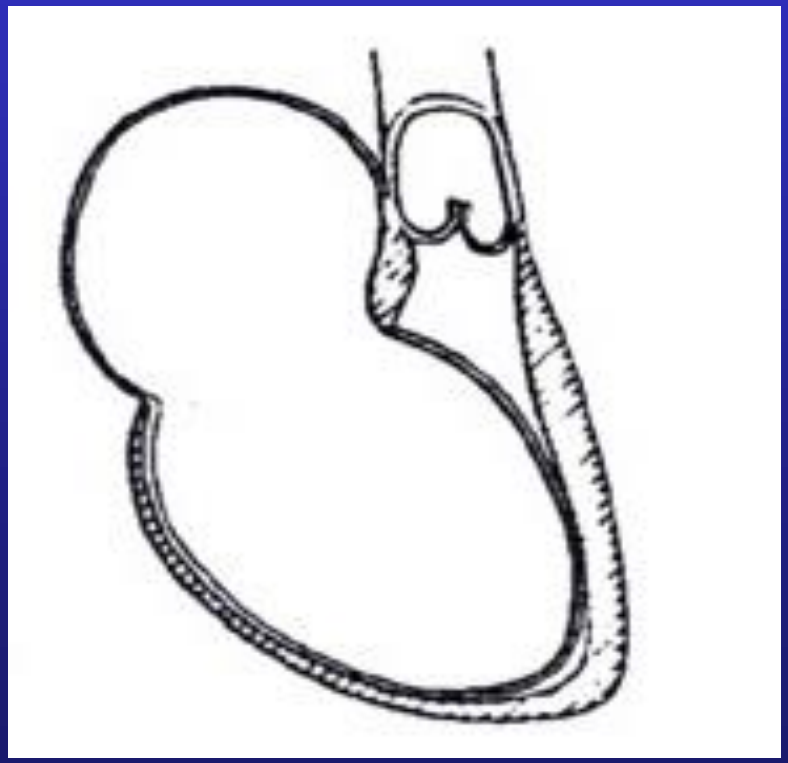
Vertical (Longitudianl) Plication

Limitations of Carpentier technique

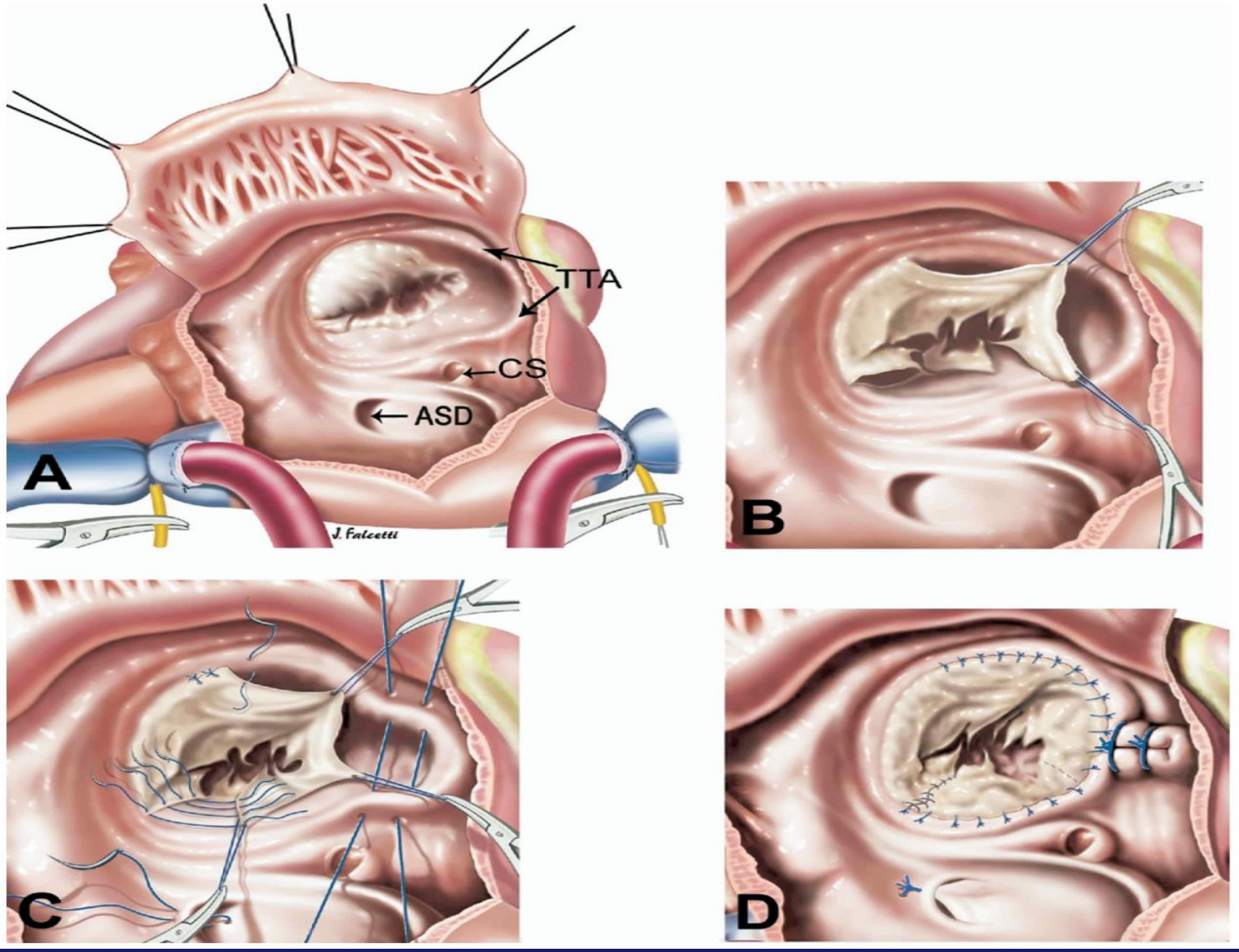
1. Tethering of AP leaflets



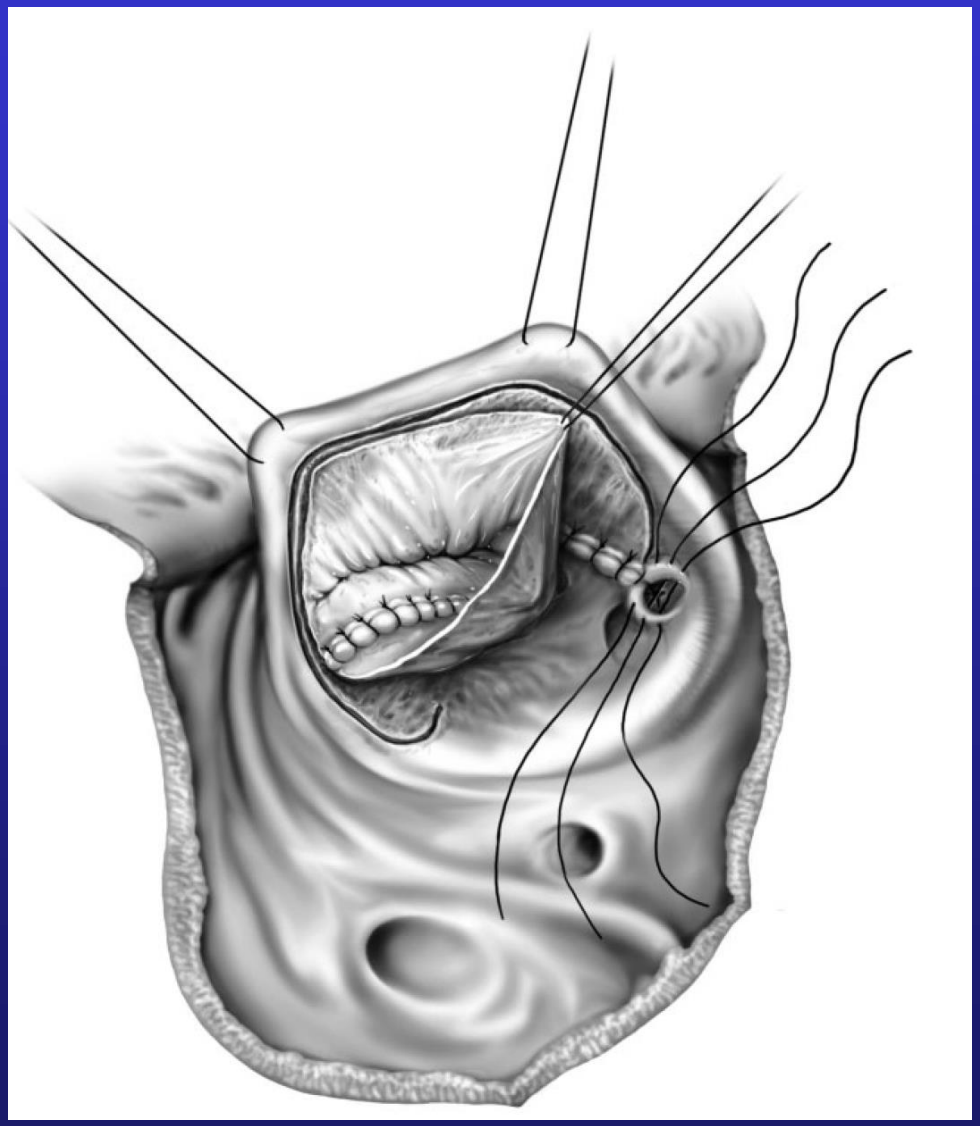
2. Tricuspid sac anomaly

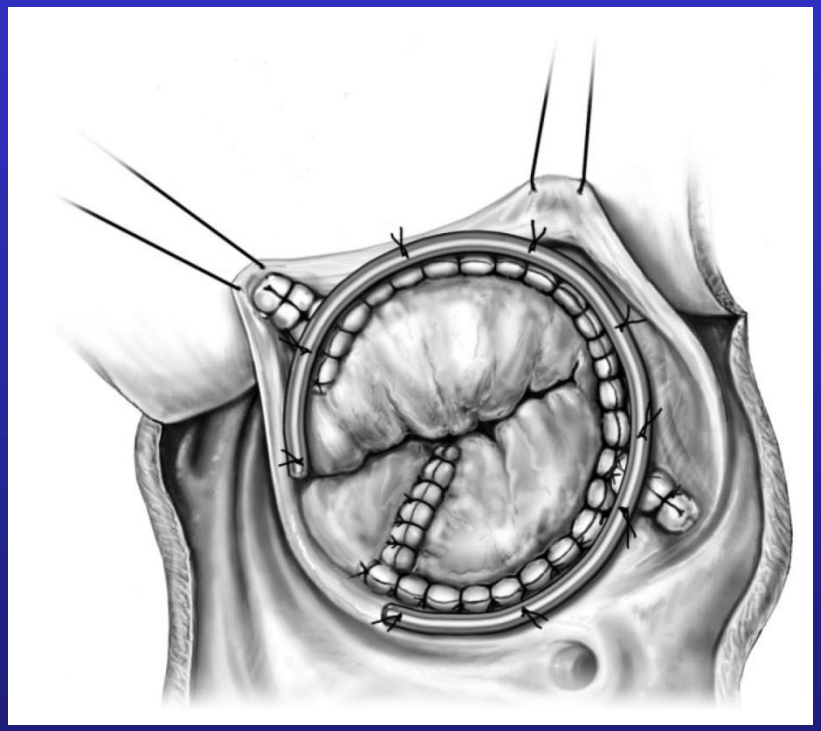


Cone procedure

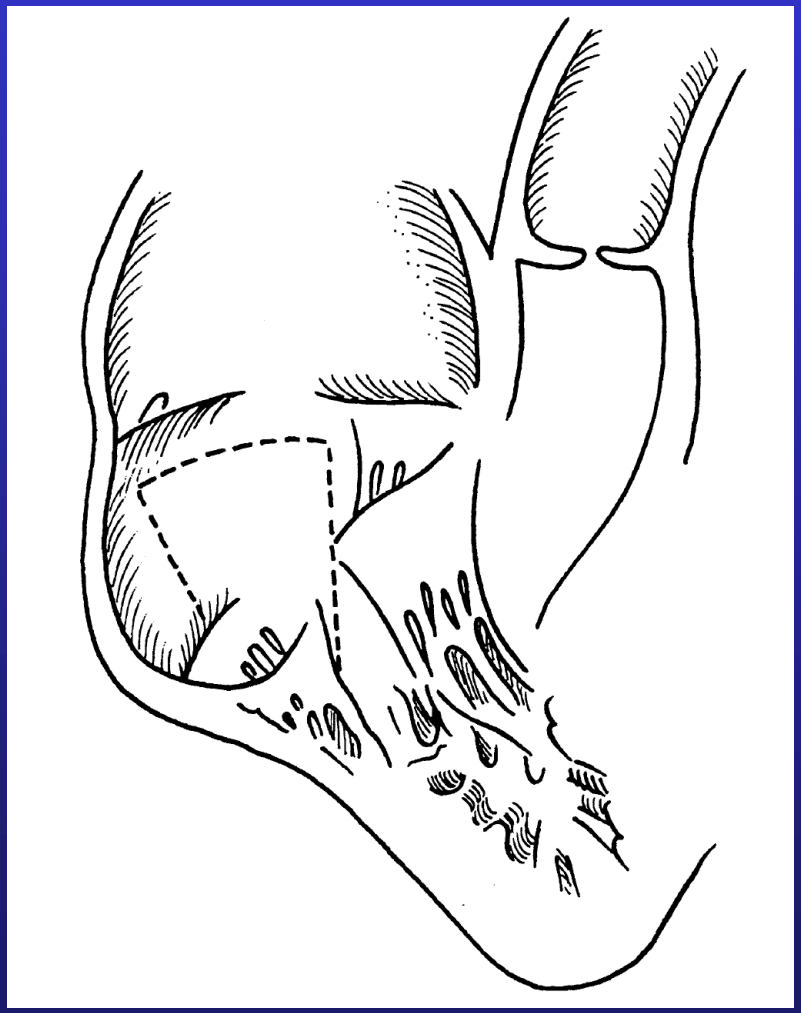


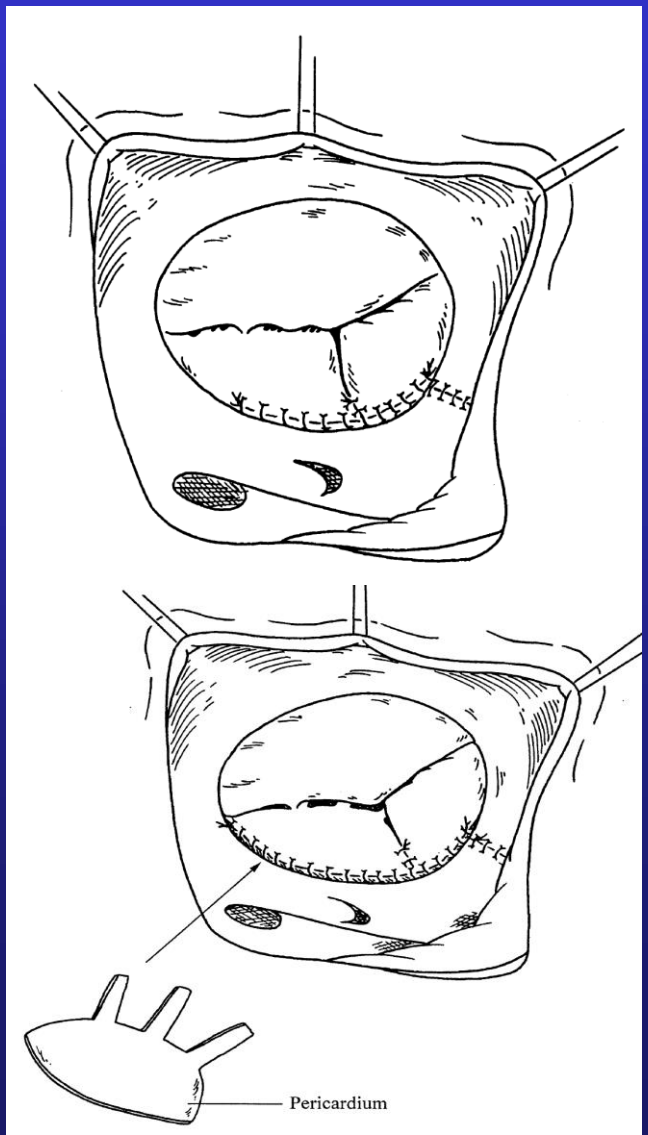
Cone procedure





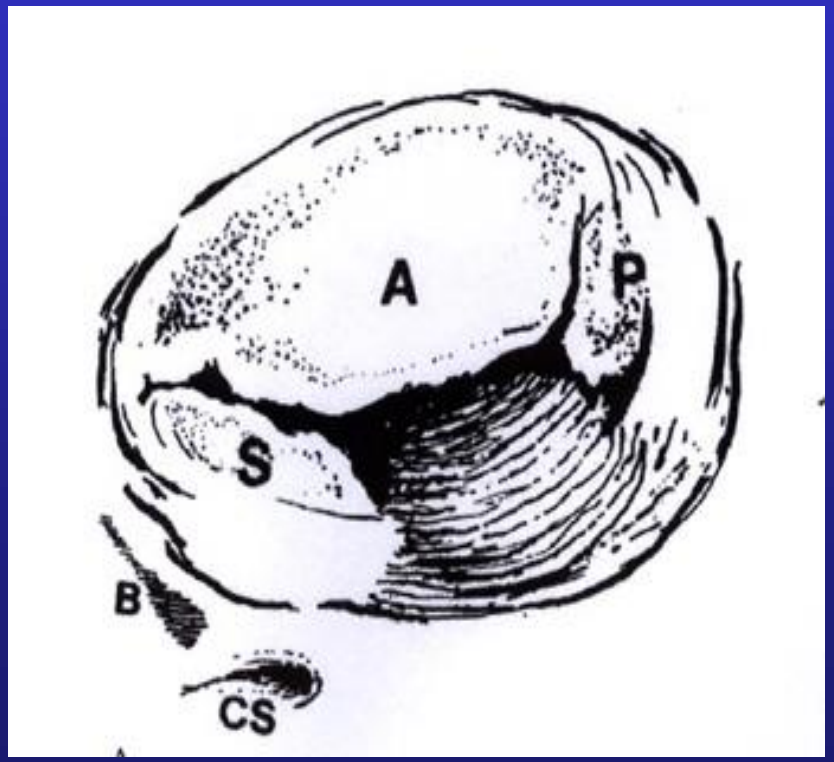
Wu technique
(Wu et al, ATS 2004)



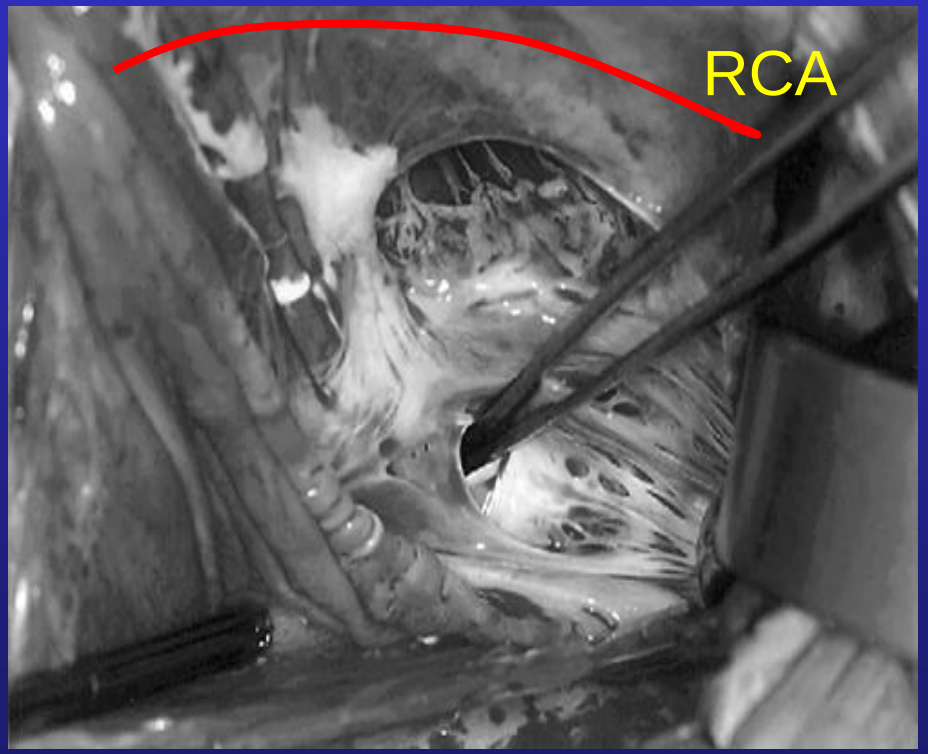


Wu technique
(Wu et al, ATS 2004)

Ebstein's anomaly



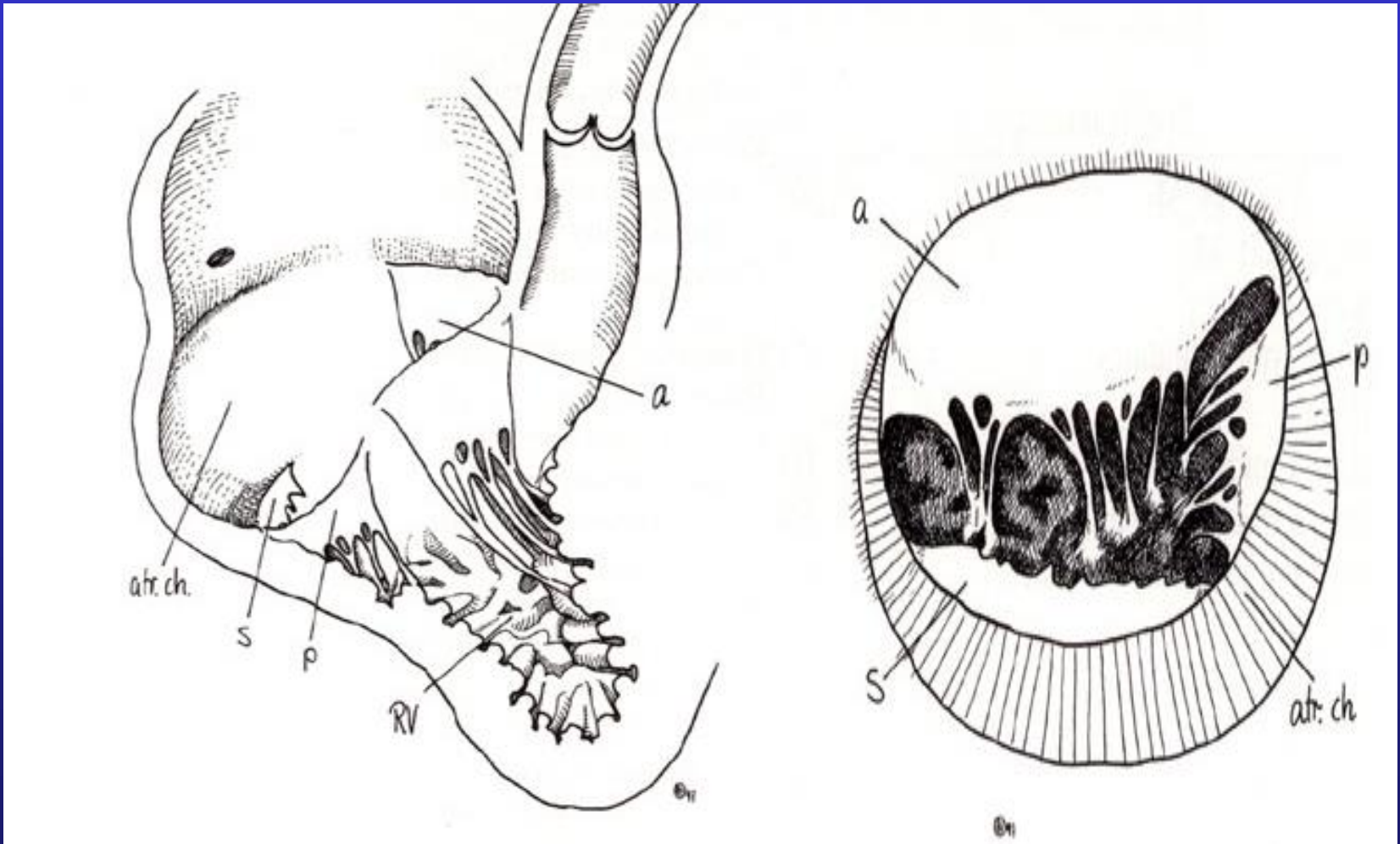
Giant RA



Hetzer repair

- No plication of aRV
- Incorporation of aRV to f RV
- Allowance of multiple trial and error
- Drawbacks
  1. Requires well developed AL
  2. Applicable to mild disease
  3. Risk of heart block

Hetzer repair
(Hetzer et al, JTCVS1998)



Hetzer repair
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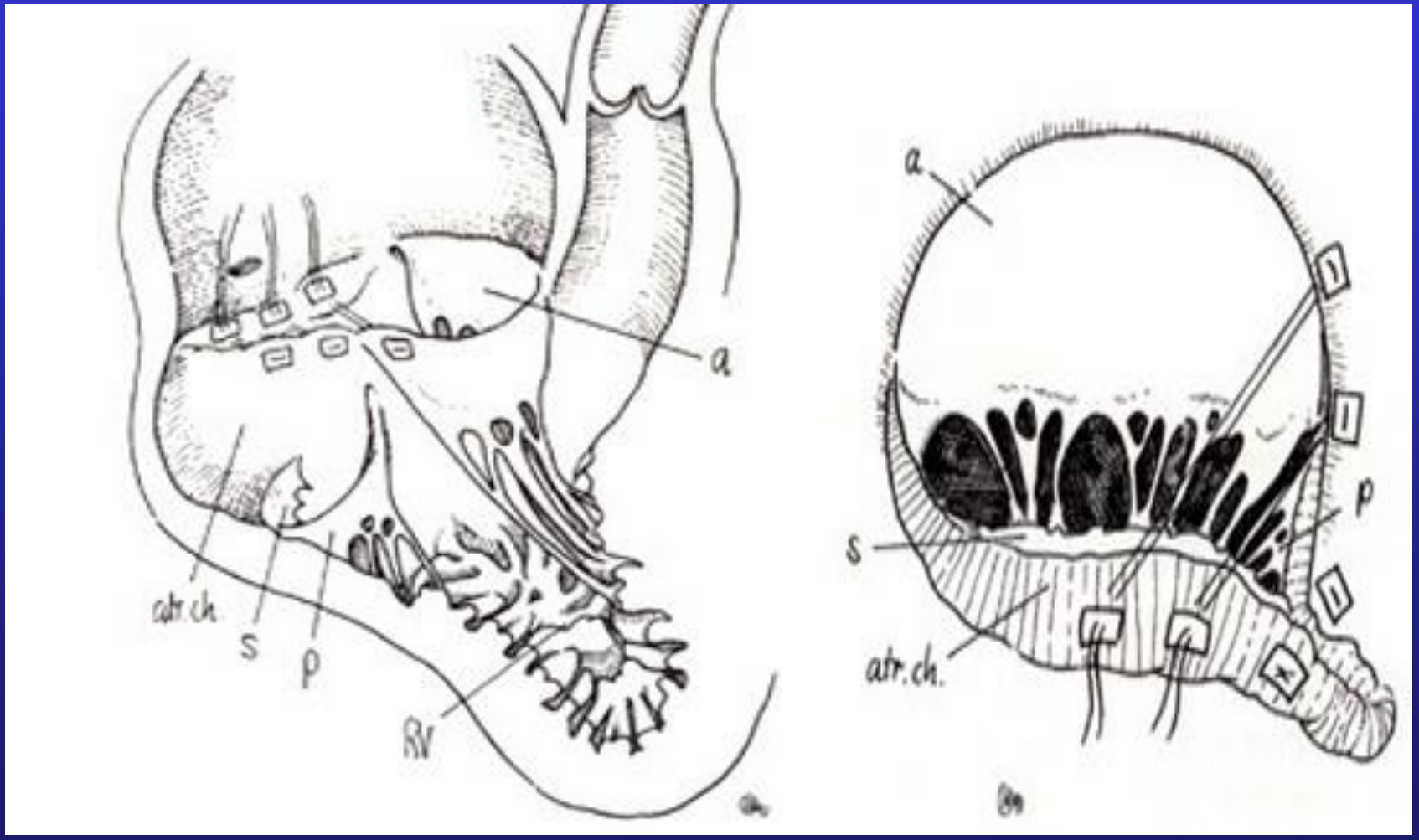

Hetzer repair
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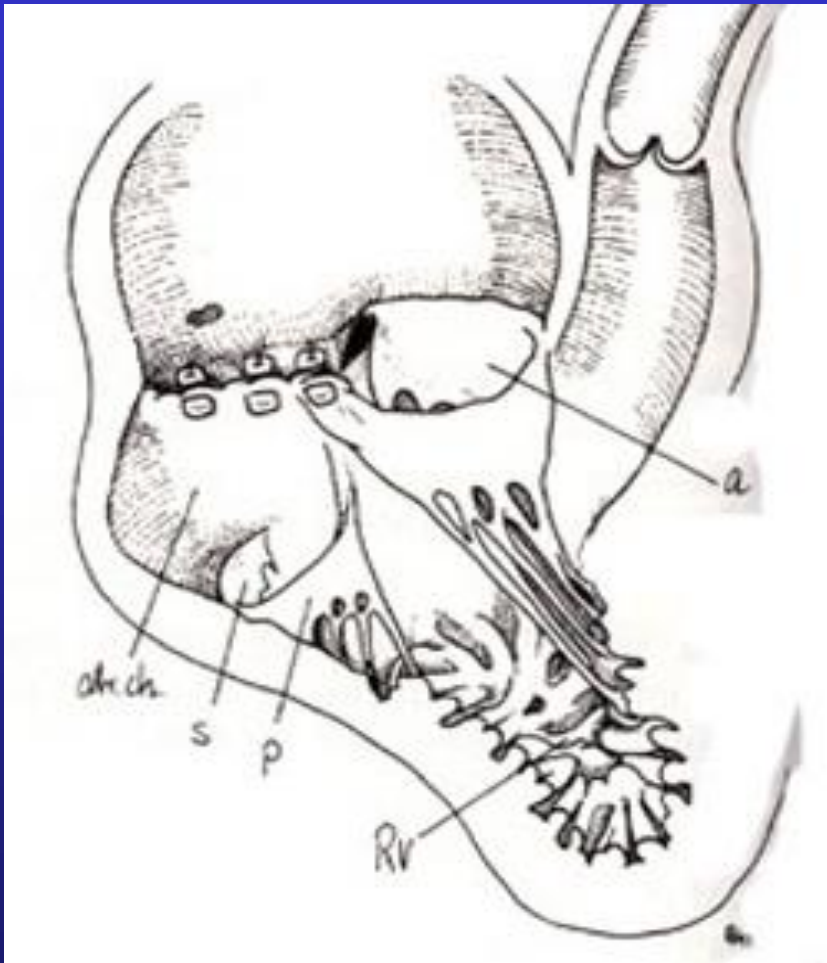

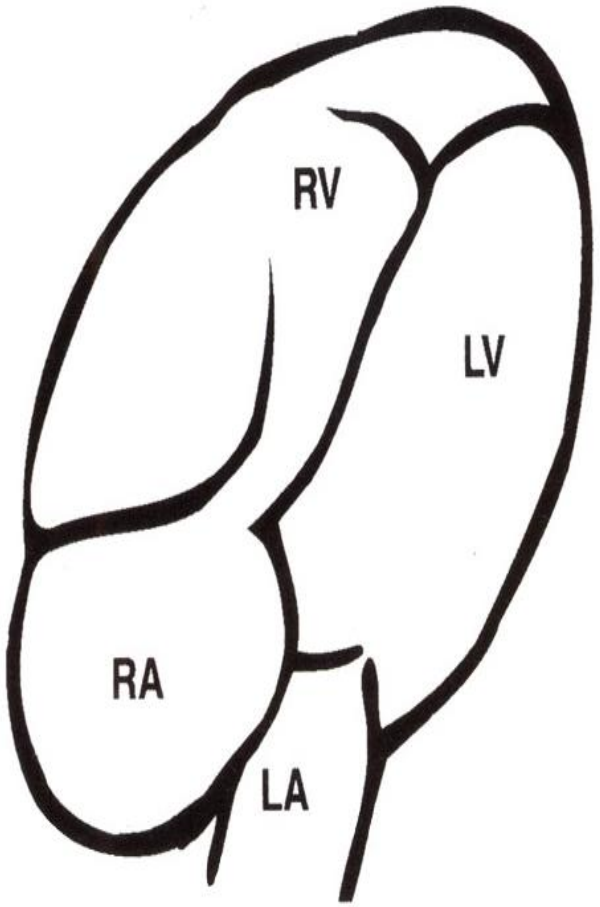

Hetzer repair
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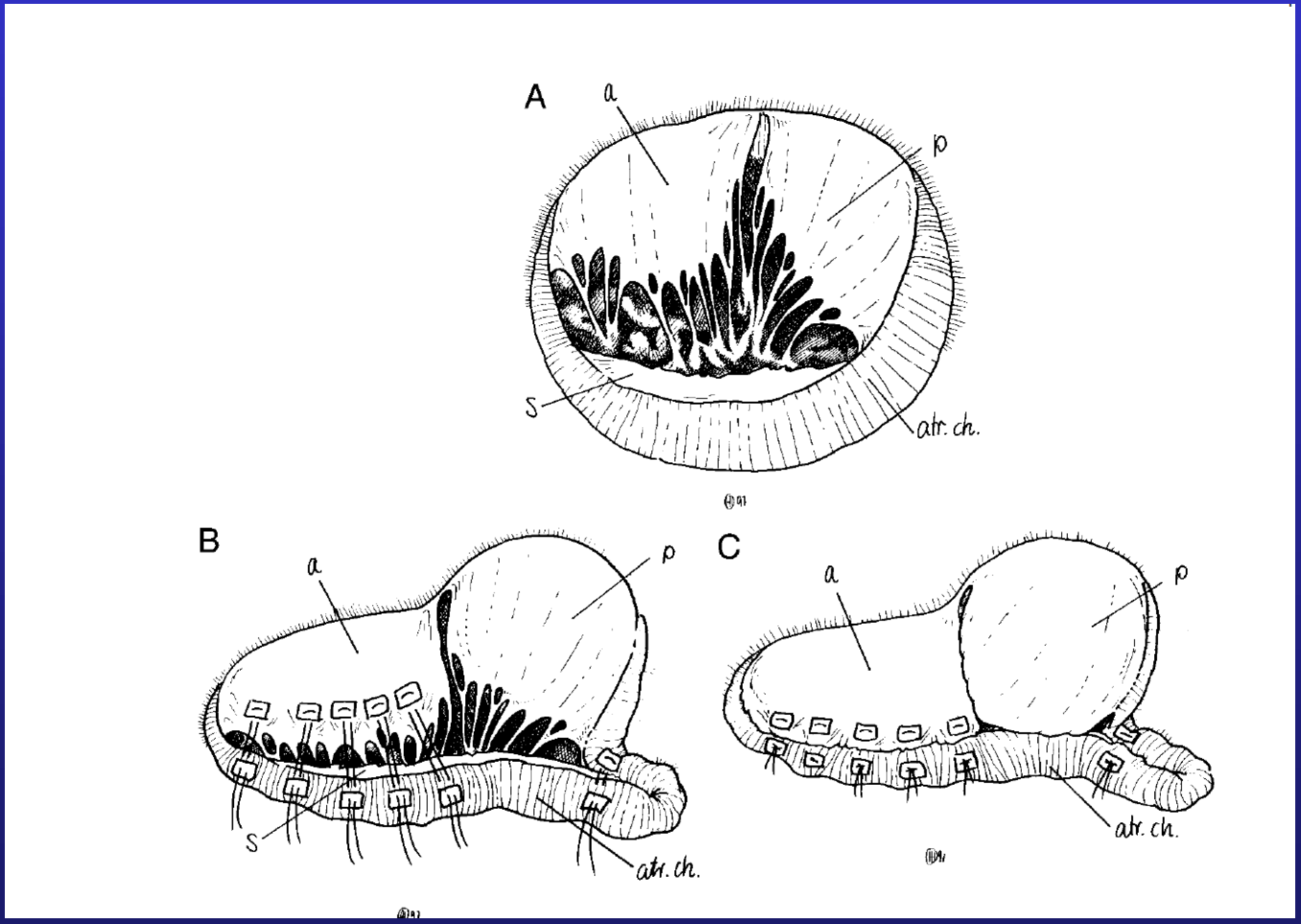
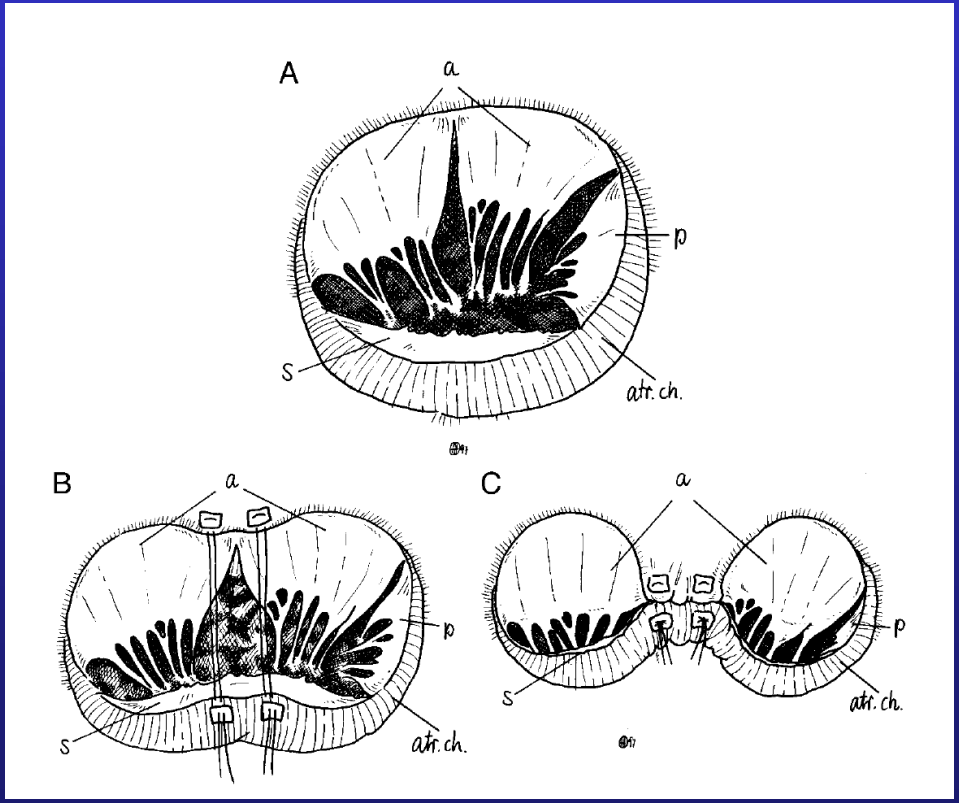
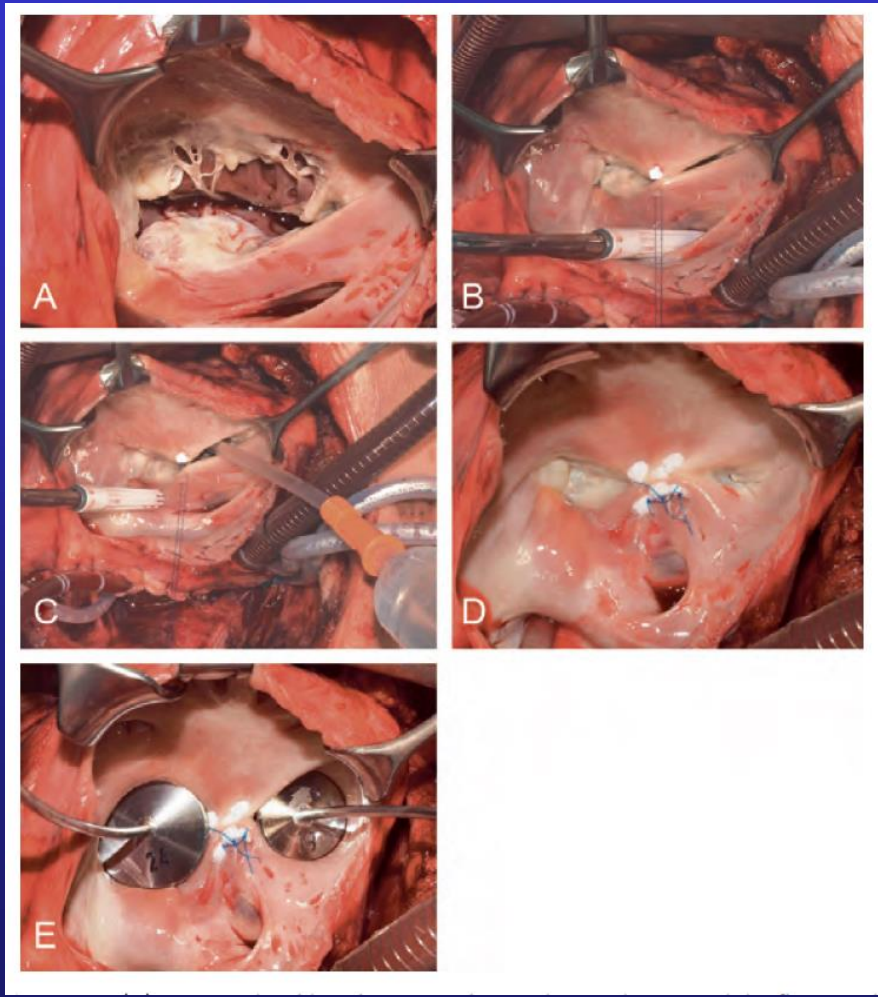


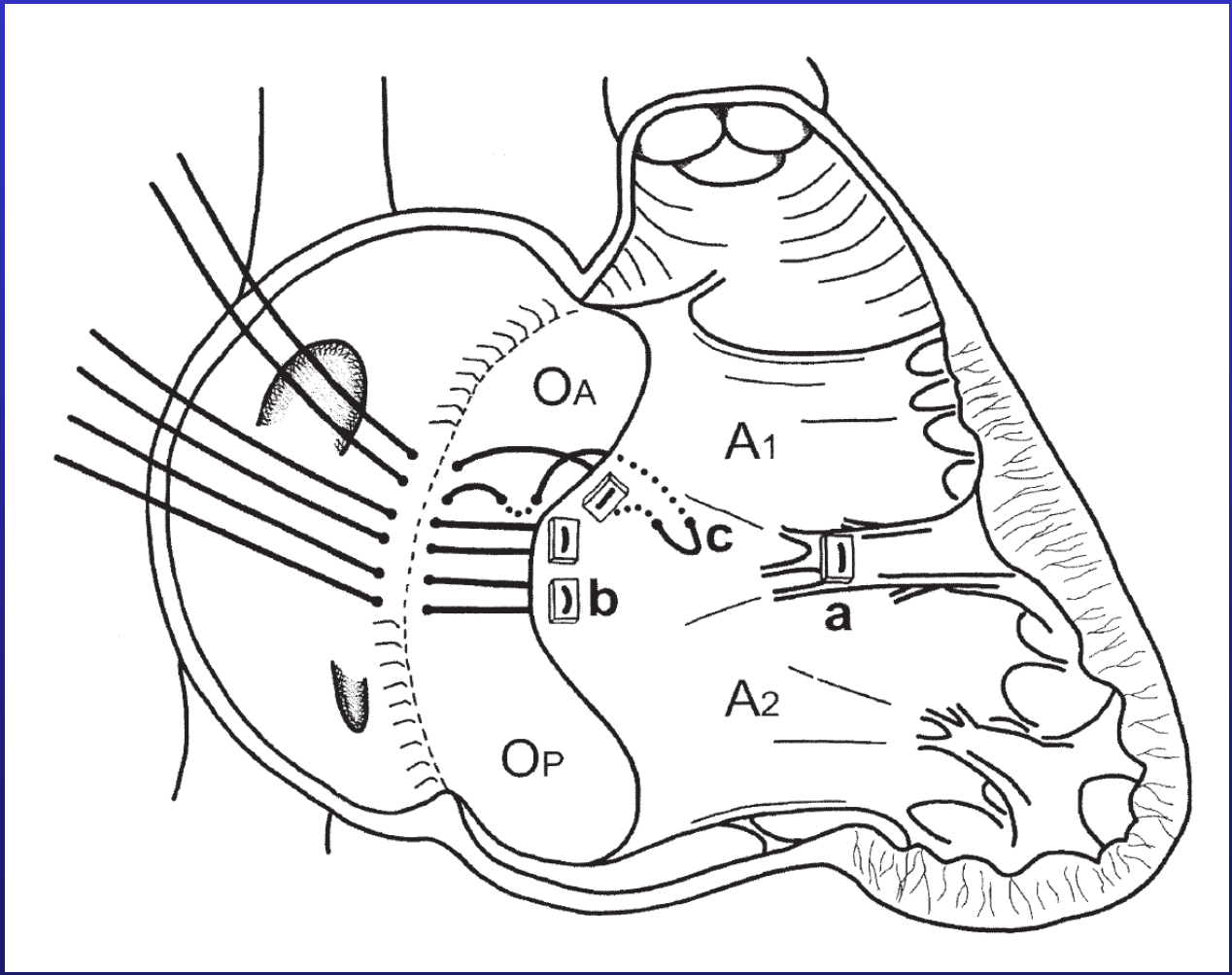
Hetzer repair
(Hetzer et al, JTCVS1998)



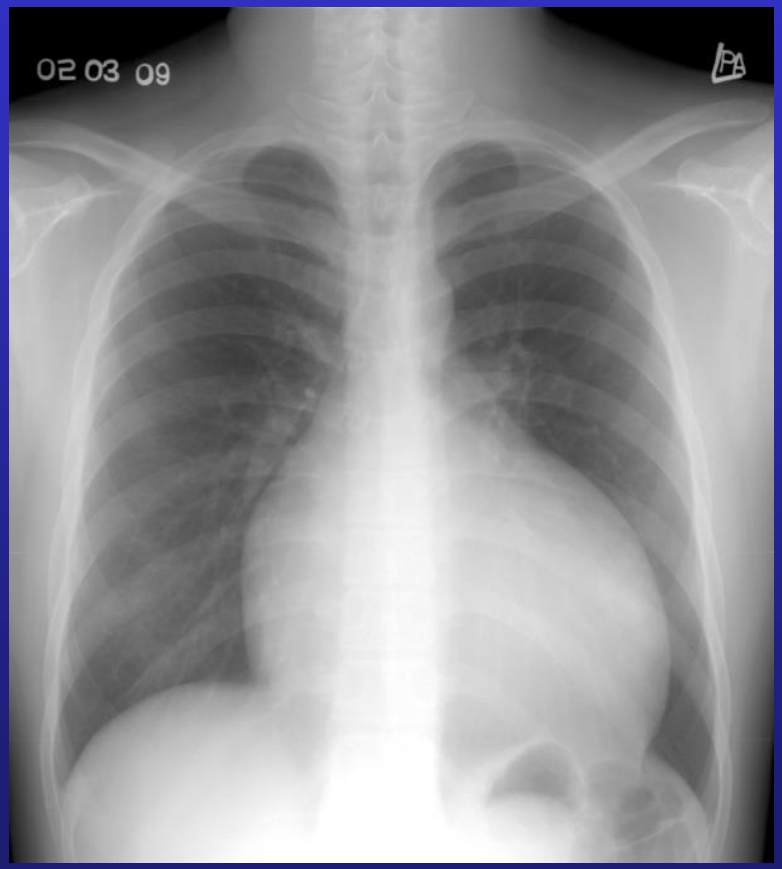


Hetzer repair

-Sebening stitch-



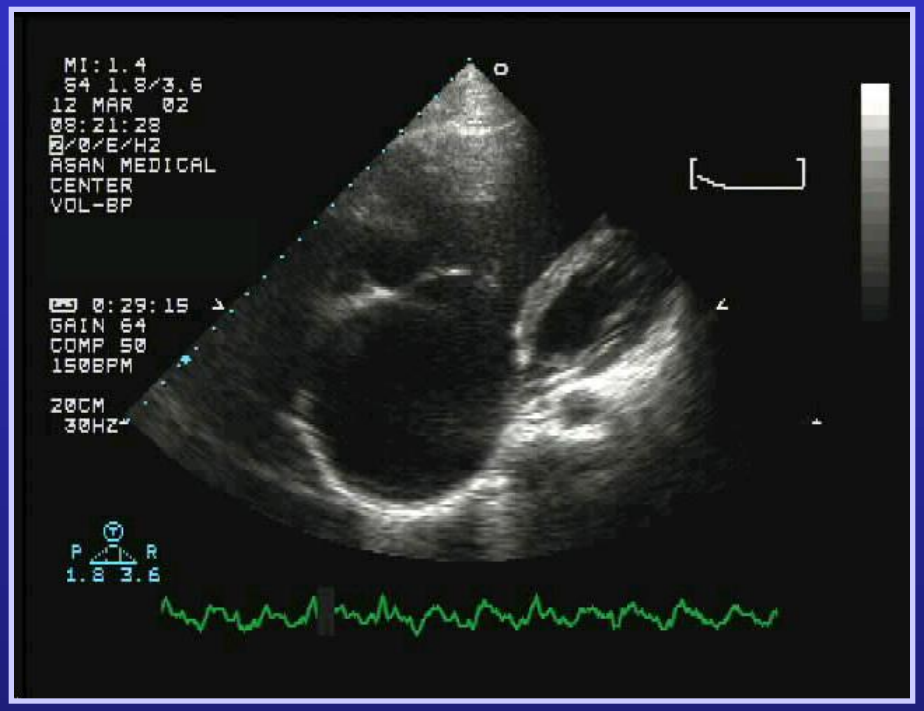
21-year-old man with Ebstein's anomaly

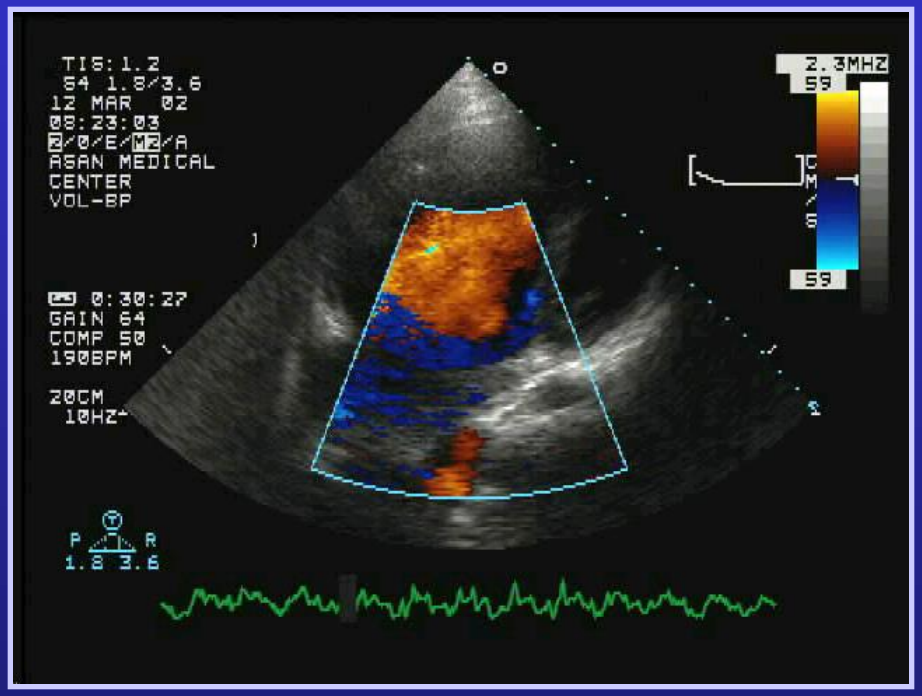


Pre-op chest PA

Functional class III
Severe TR: TR jet area 25 cm^2
Association of Atrial flutter

aRV function after Hetzer repair

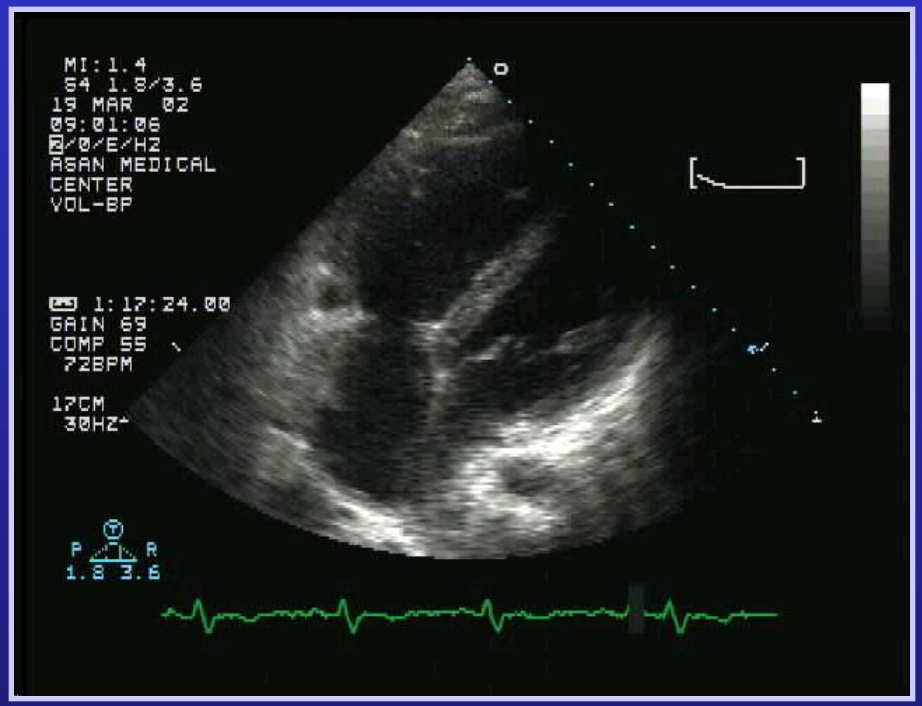


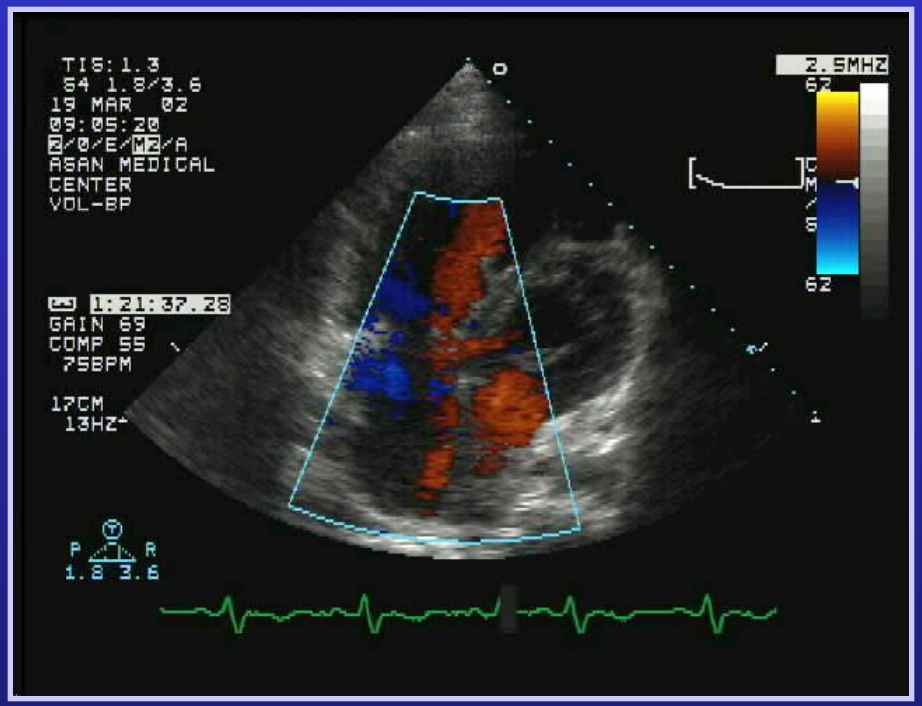


M/21, Ebstein anomaly (A)

Pre-operative

aRV function after Hetzer repair

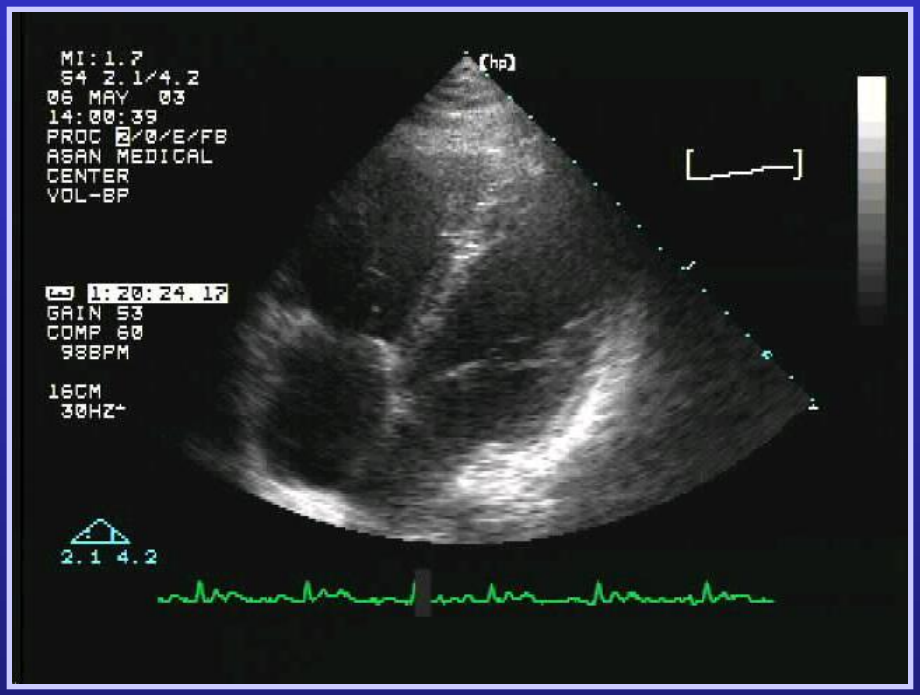


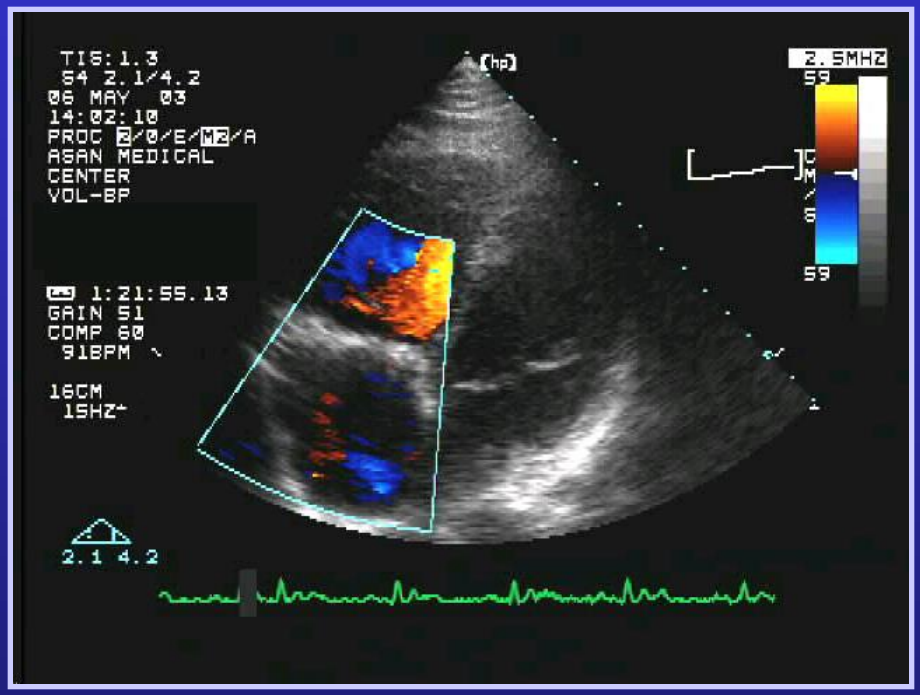


M/21, Ebstein anomaly (A)

Immediate Post-operative

aRV function after Hetzer repair

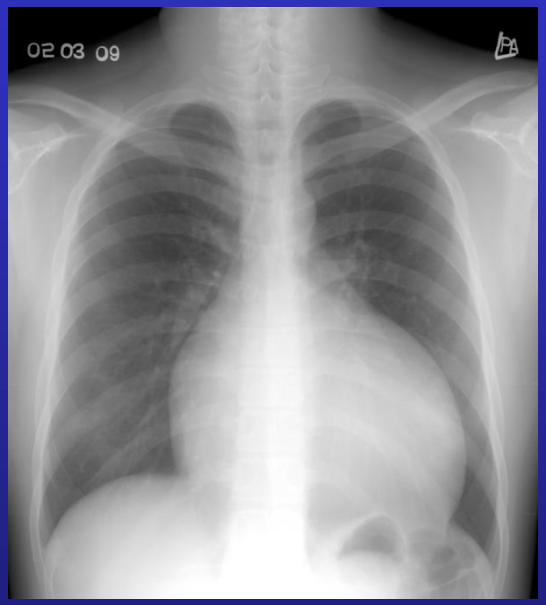




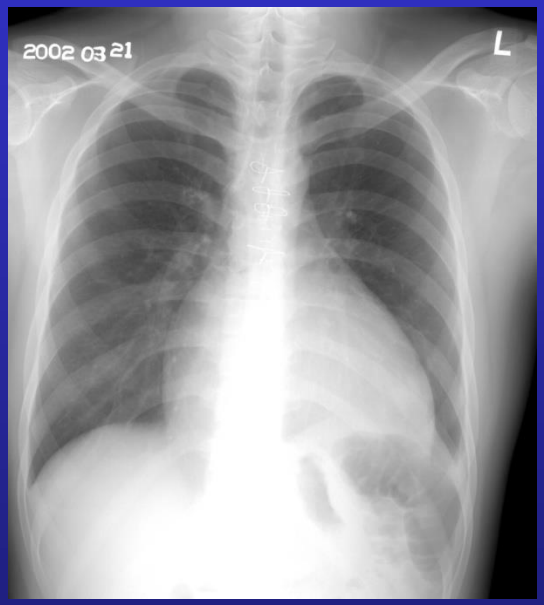
M/21, Ebstein anomaly (A)

Post-operative 14 months

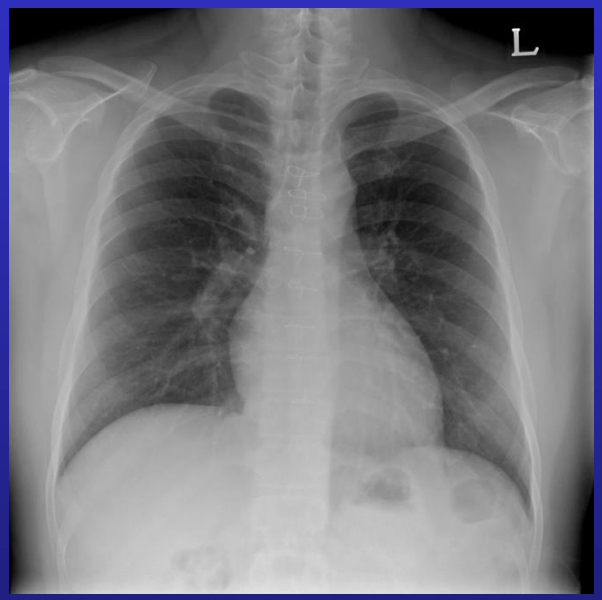
21-year-old man with Ebstein's anomaly



Pre-op

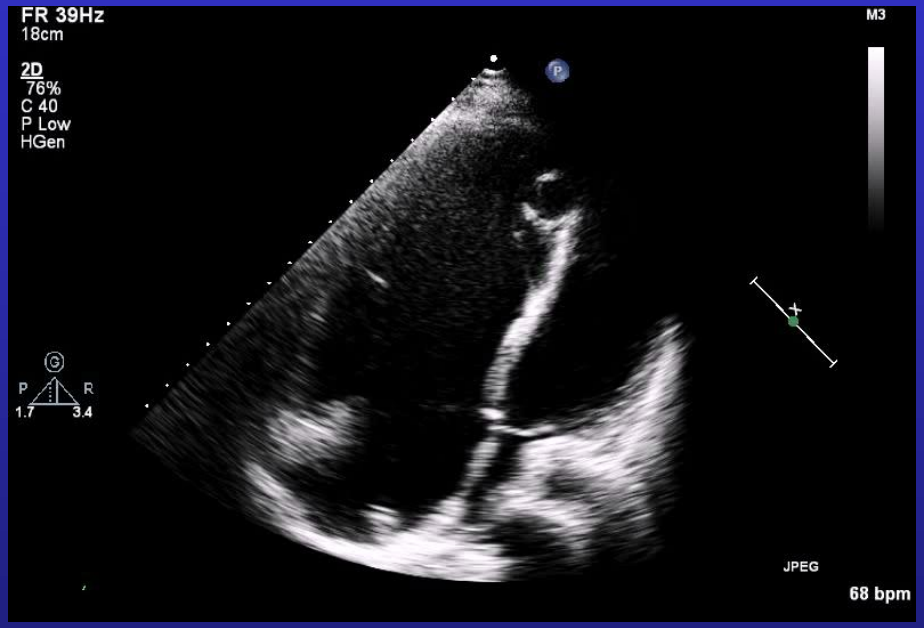


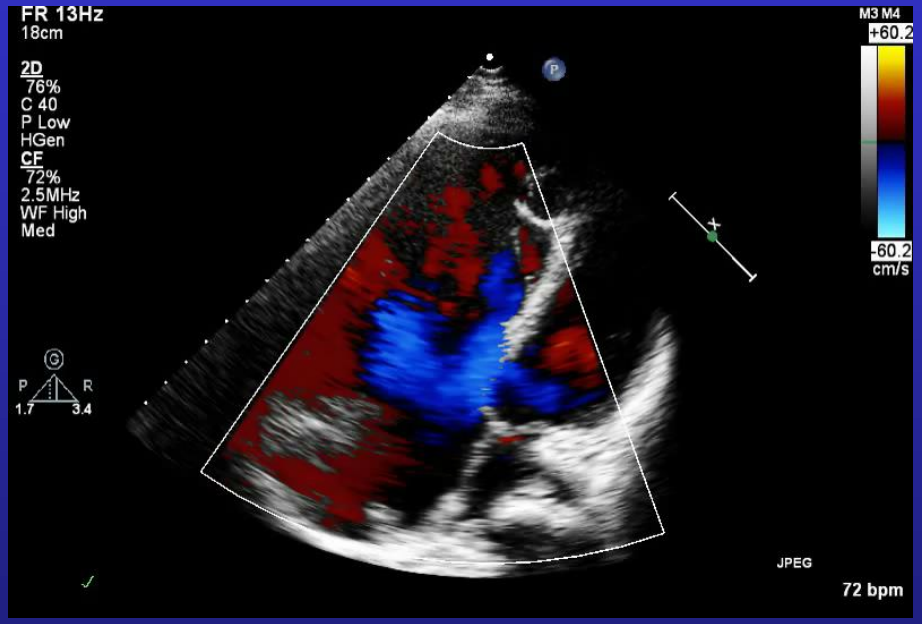
Immediate Post-op



Post-op 9 years

aRV function after Hetzer repair

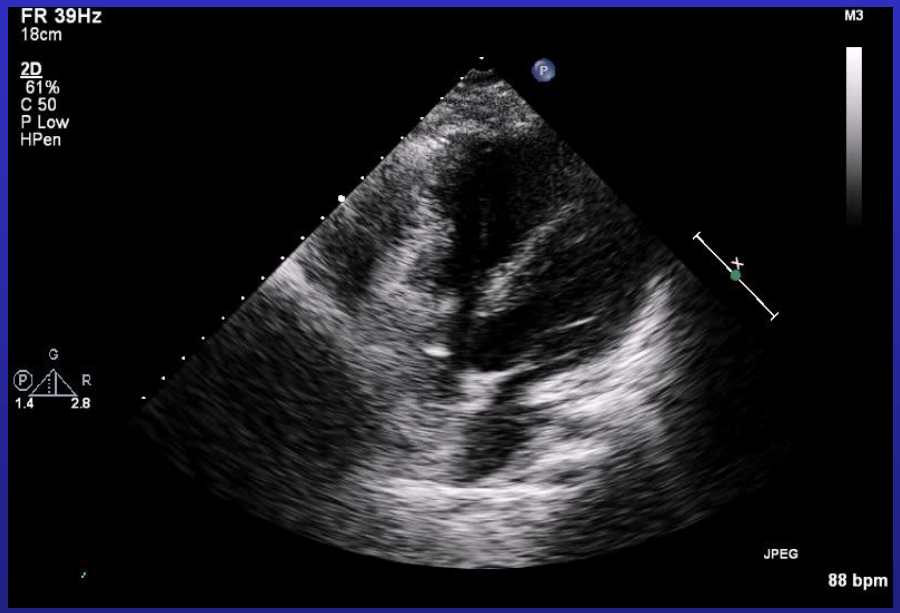


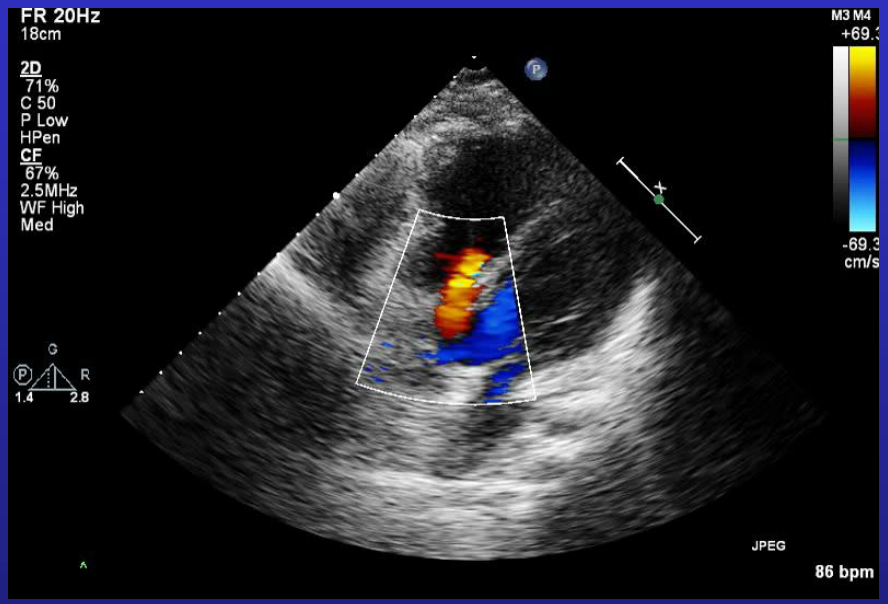


F/25, Ebstein anomaly (B)

Pre-operative

aRV function after Hetzer repair



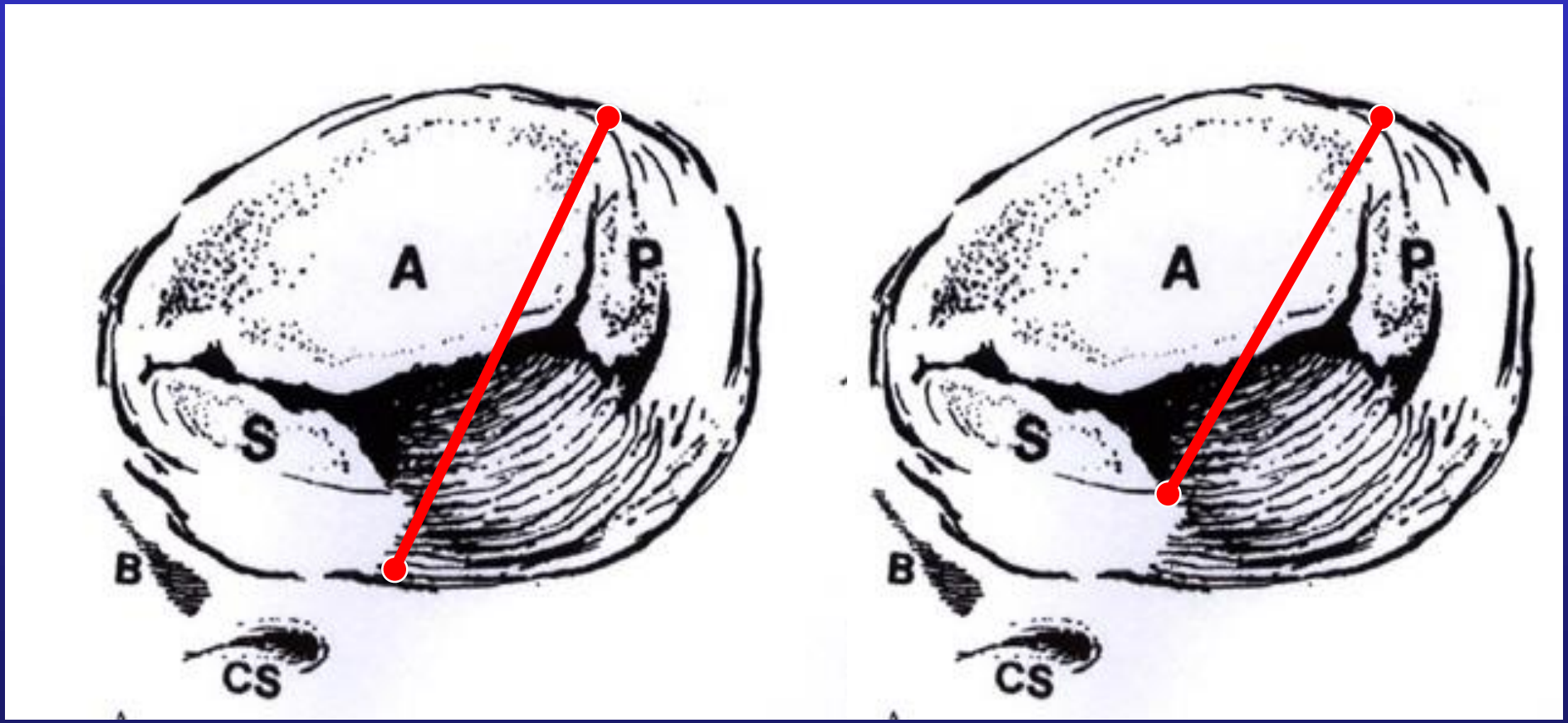


F/25, Ebstein anomaly (B)

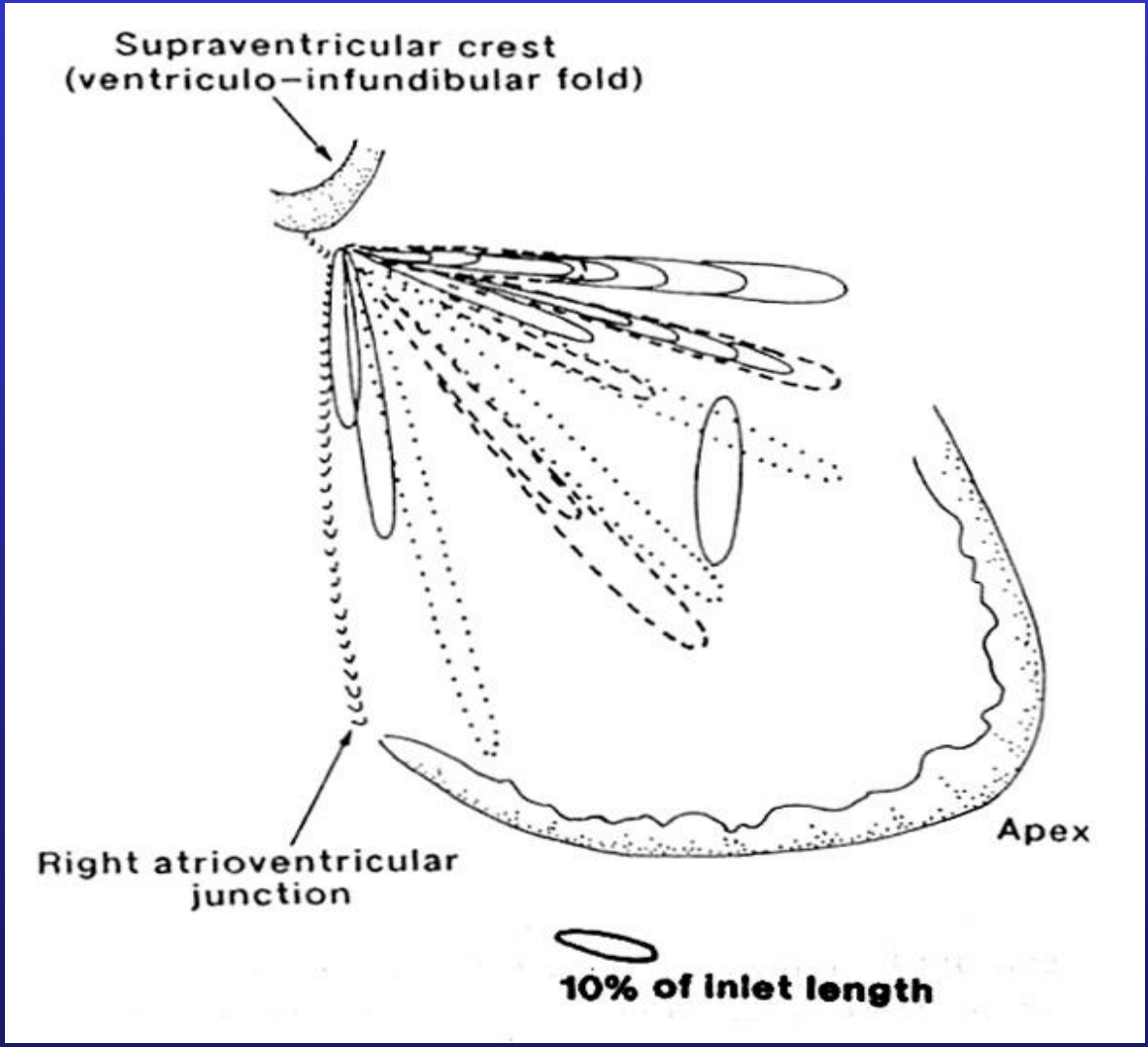
Immediate postoperative

AMC experience of Hetzer repair

- Original Hetzer technique
- Modified Hetzer technique



Effective valve orifice in 23 specimens
(Schreiber et al, JTCVS 1999)



AMC experience of Hetzer Technique

- Duration: 2002-2013
- Patient No. : 27
- Male / Female : 7 / 20
- Age at operation: 1 m – 57 y (median: 29 y)
- Carpentier type: A (7), B (13), C (3), unknown (4)
- Pre-op desaturation (<95%): 7/27
- Associated lesion

  AF-Af (7), AVNRT (1), WPW (1), ASD (9), RPA atresia (1)

AMC experience of Hetzer Technique

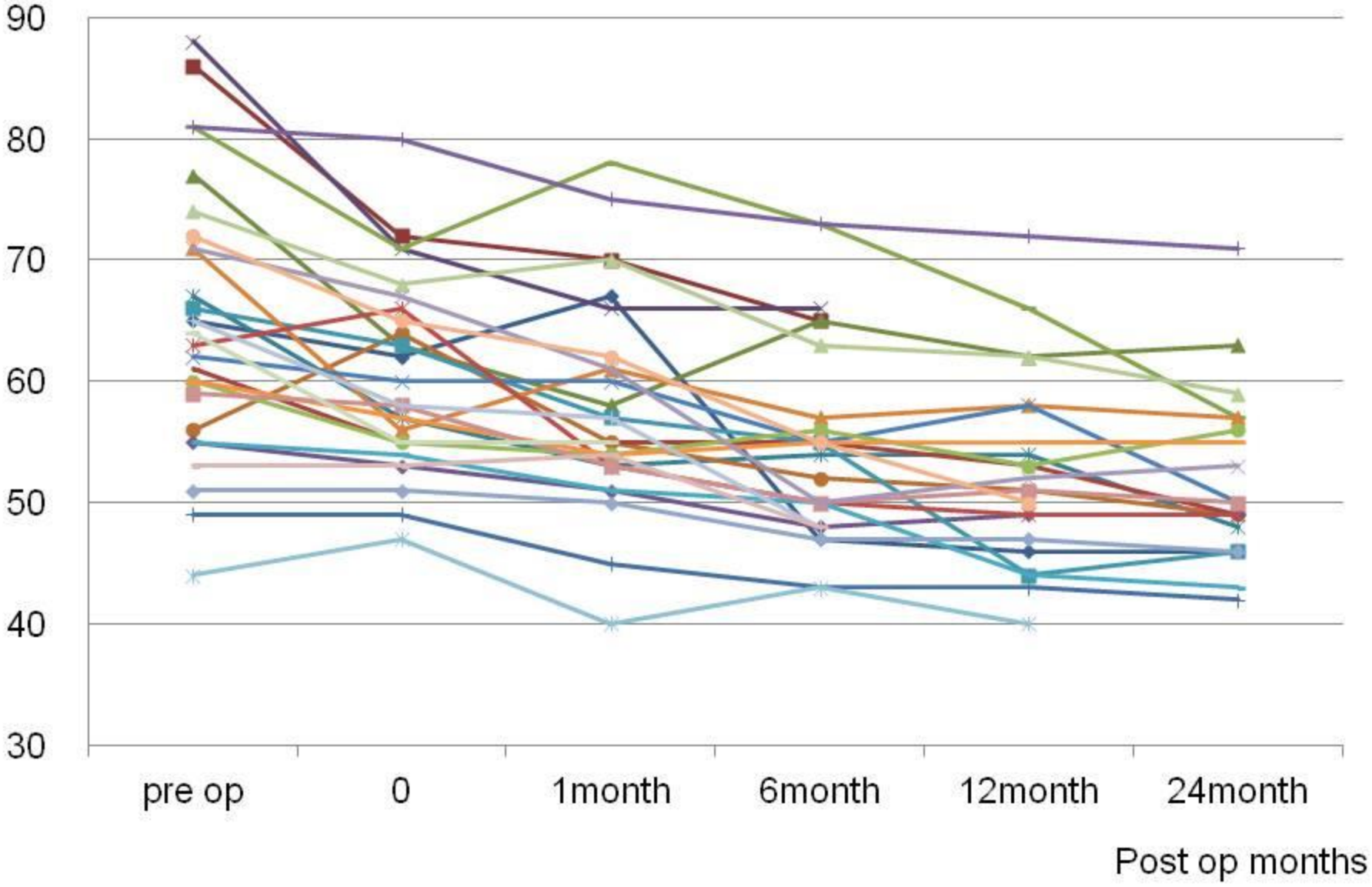
- Hetzer technique: Original (6), Modified (21)
- Associated Procedure

  BCS (23), Arrhythmia Op (8), ASD closure (9)
- CPB / ACC : 119 min / 54 min
- aRV plication: 1/27
- aRV Obliteration: 2/27
- RA reduction: 11/27
- Reoperation: 1/27

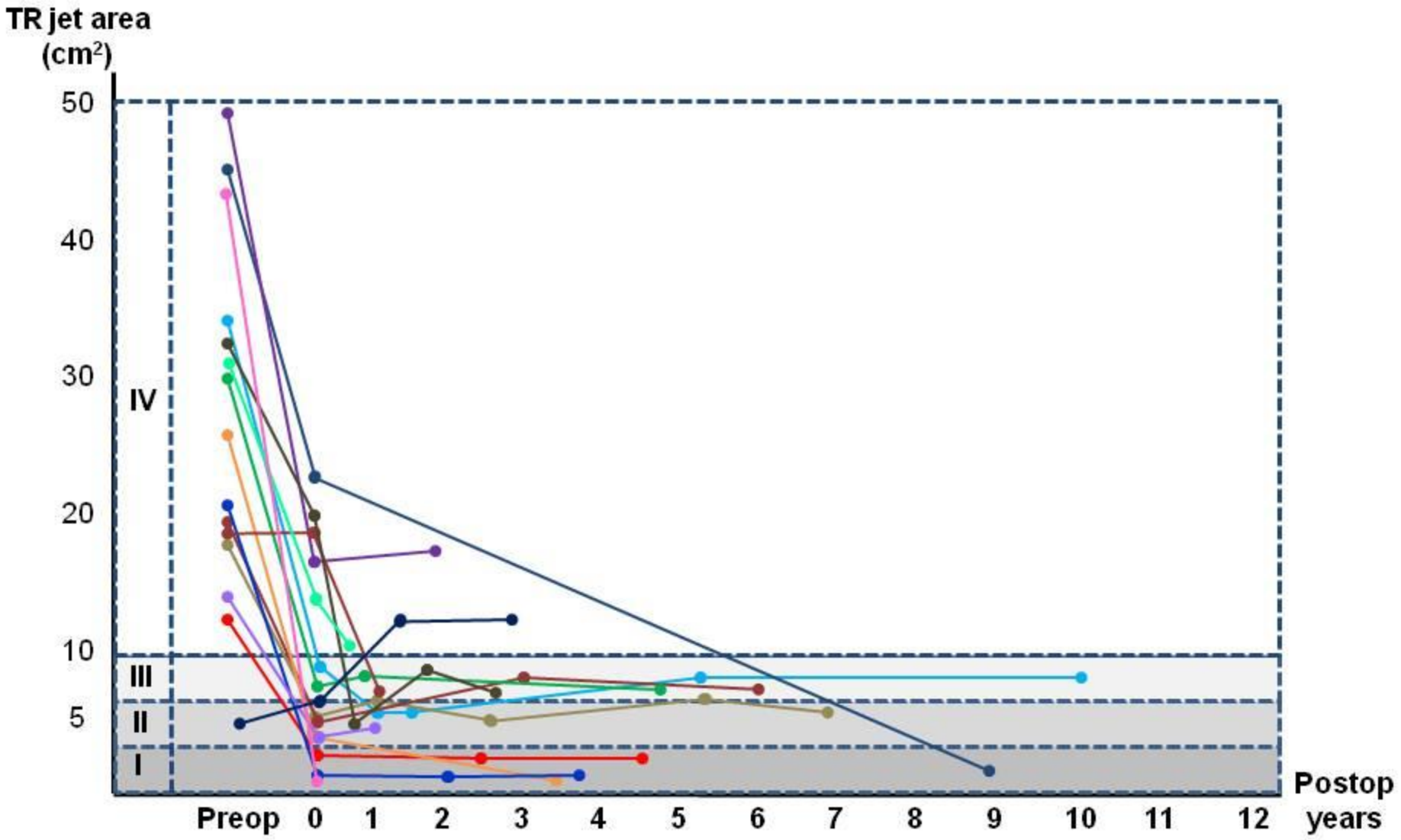
AMC experience of Hetzer Technique

- ICU stay / Hospital stay: 2.6 day / 12.6 day
- No early or late death
- F/U: 1 m – 124.8 m (median: 55 m)
- NYHA functional class $\leq$ II: 22/27
- TR on Immediate post-op TTE
  - < II (18/27)
  - III (6/27)
  - IV (3/27) : *TR jet area 46 cm^2 $\rightarrow$ 22 cm^2*

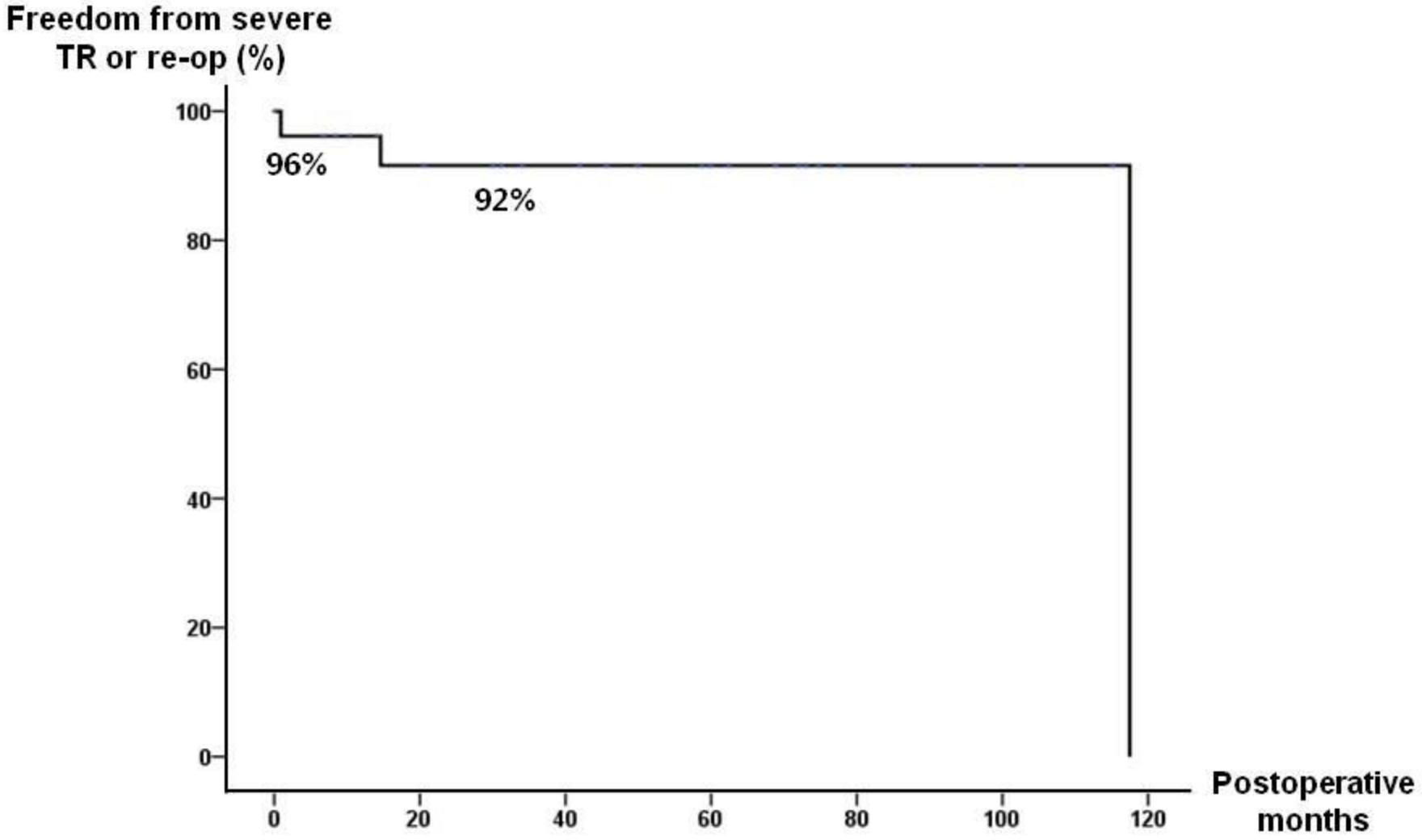
CT ratio on Chest x-ray



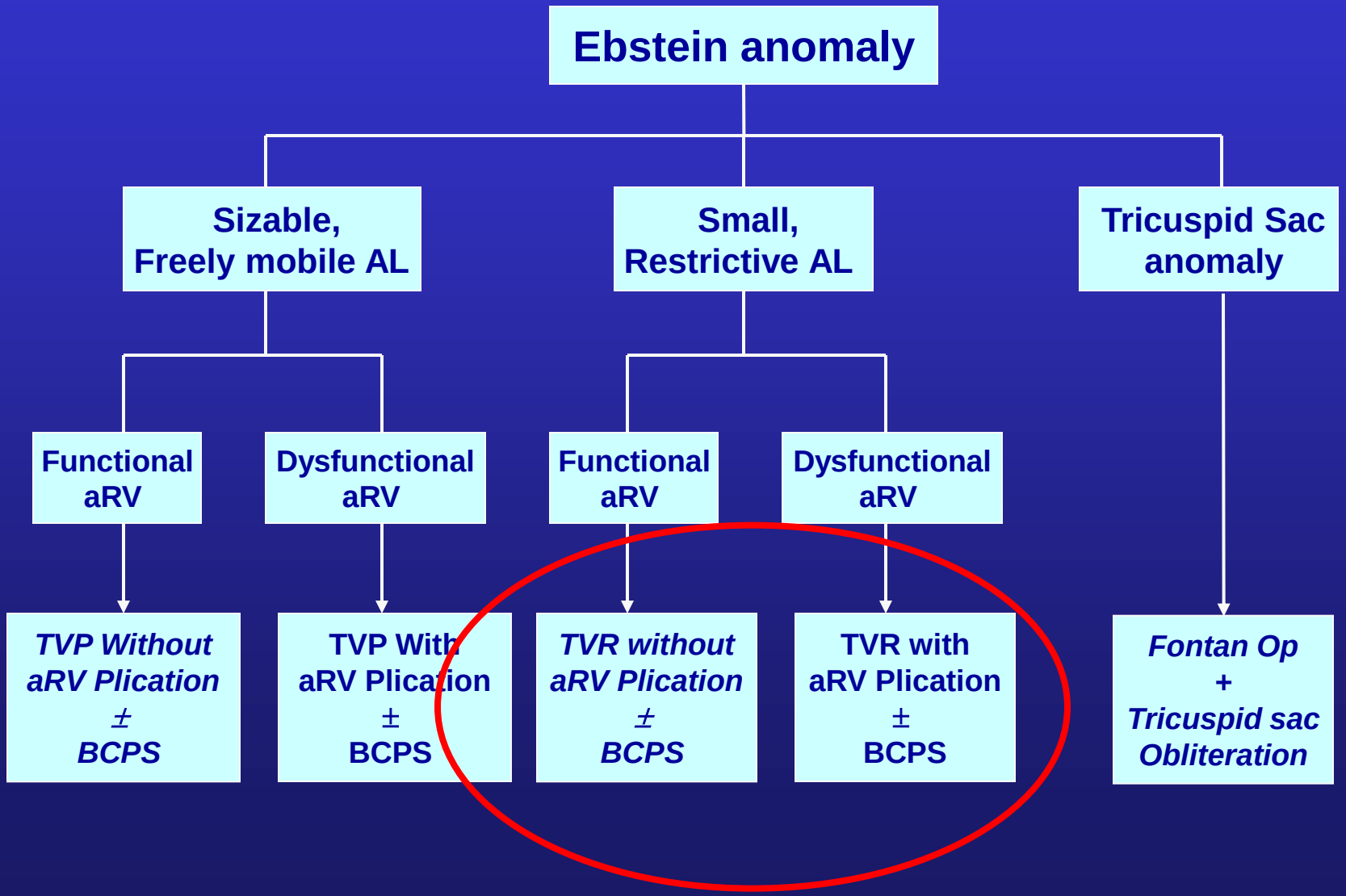
Postoperative changes in TR



Freedom of significant TR or re-operation



Surgical algorithm (AMC)



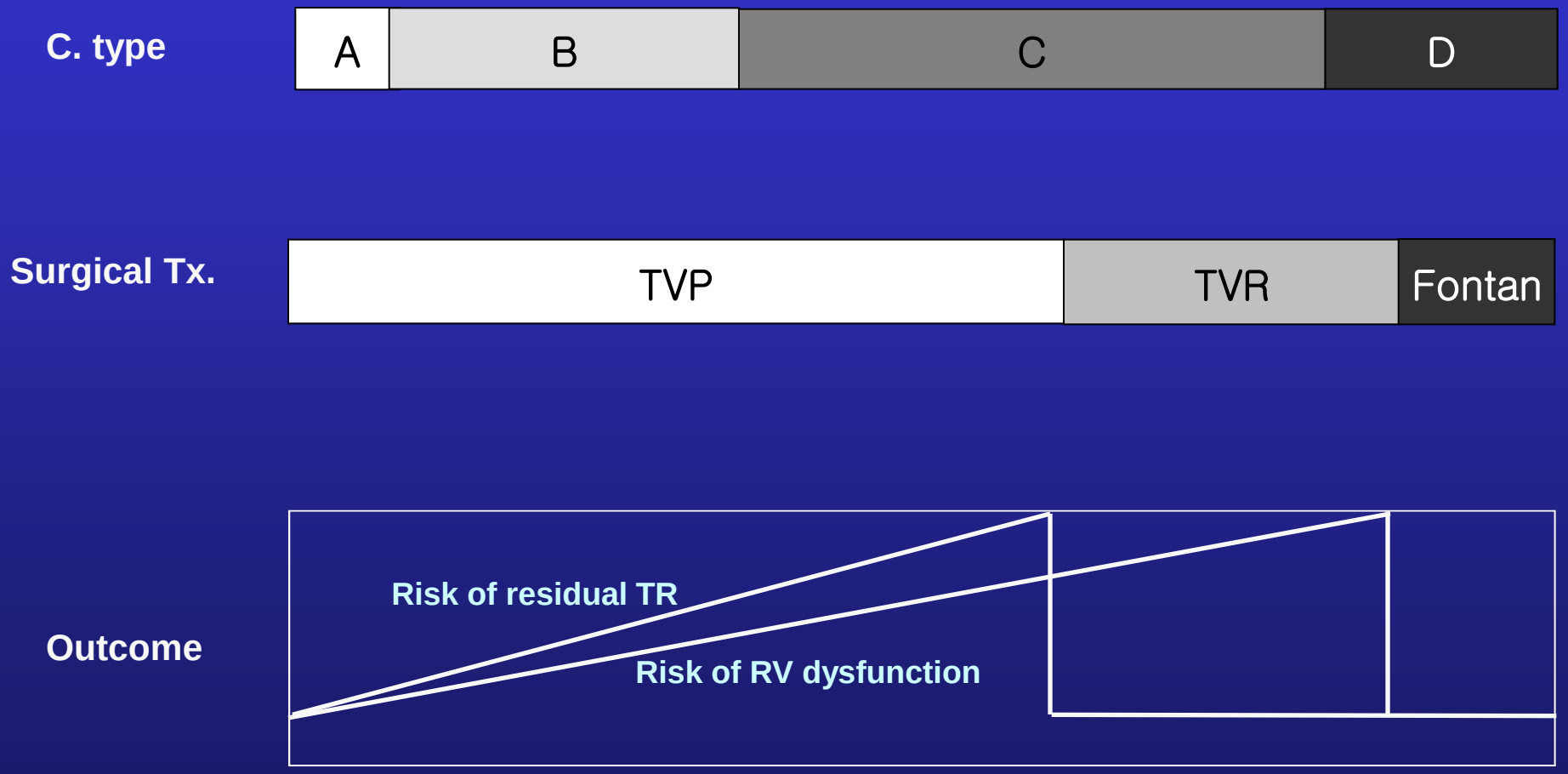
TVR in Ebstein anomaly

- **20 – 80% of entire cohort**
- **Excellent long term outcome of porcine bioprosthetic valve (Mayo Clinic)**
- **Issues in surgical technique:**

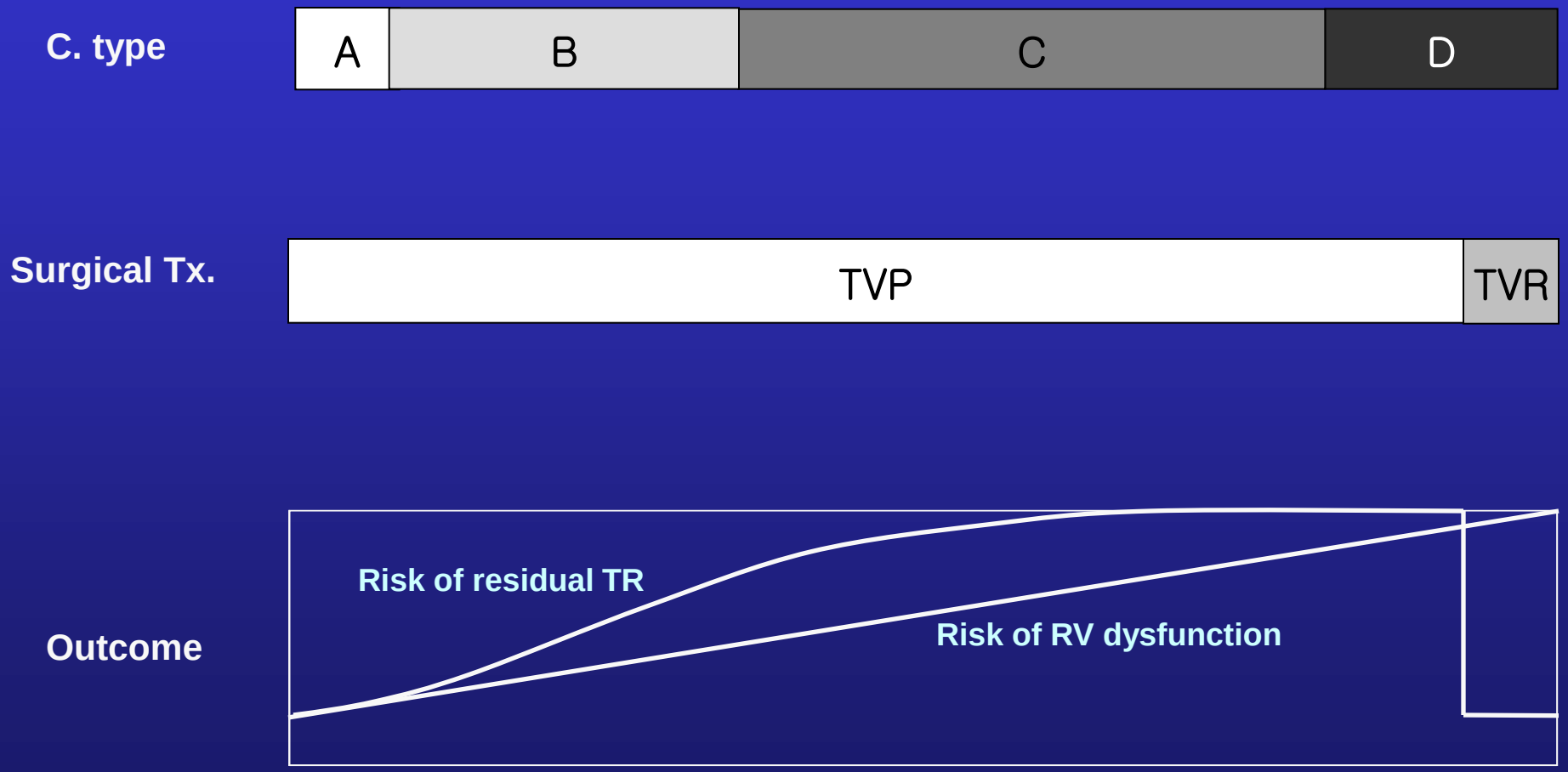
  1) above coronary sinus vs. under coronary sinus

  2) Combined procedure: plication of aRV

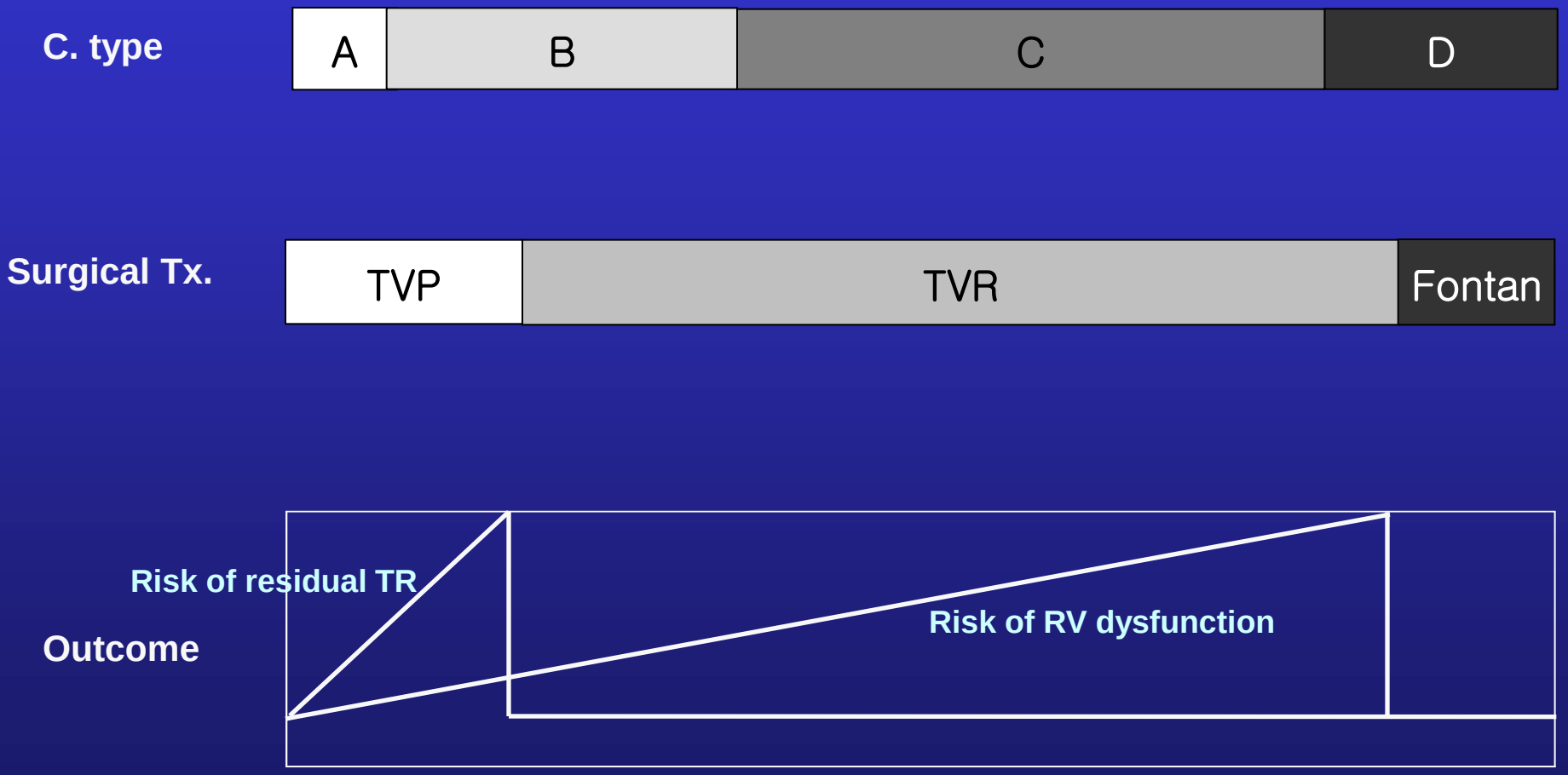
Ebstein Anomaly as a Spectrum



Overly aggressive TVP



Over-reliance on TVR

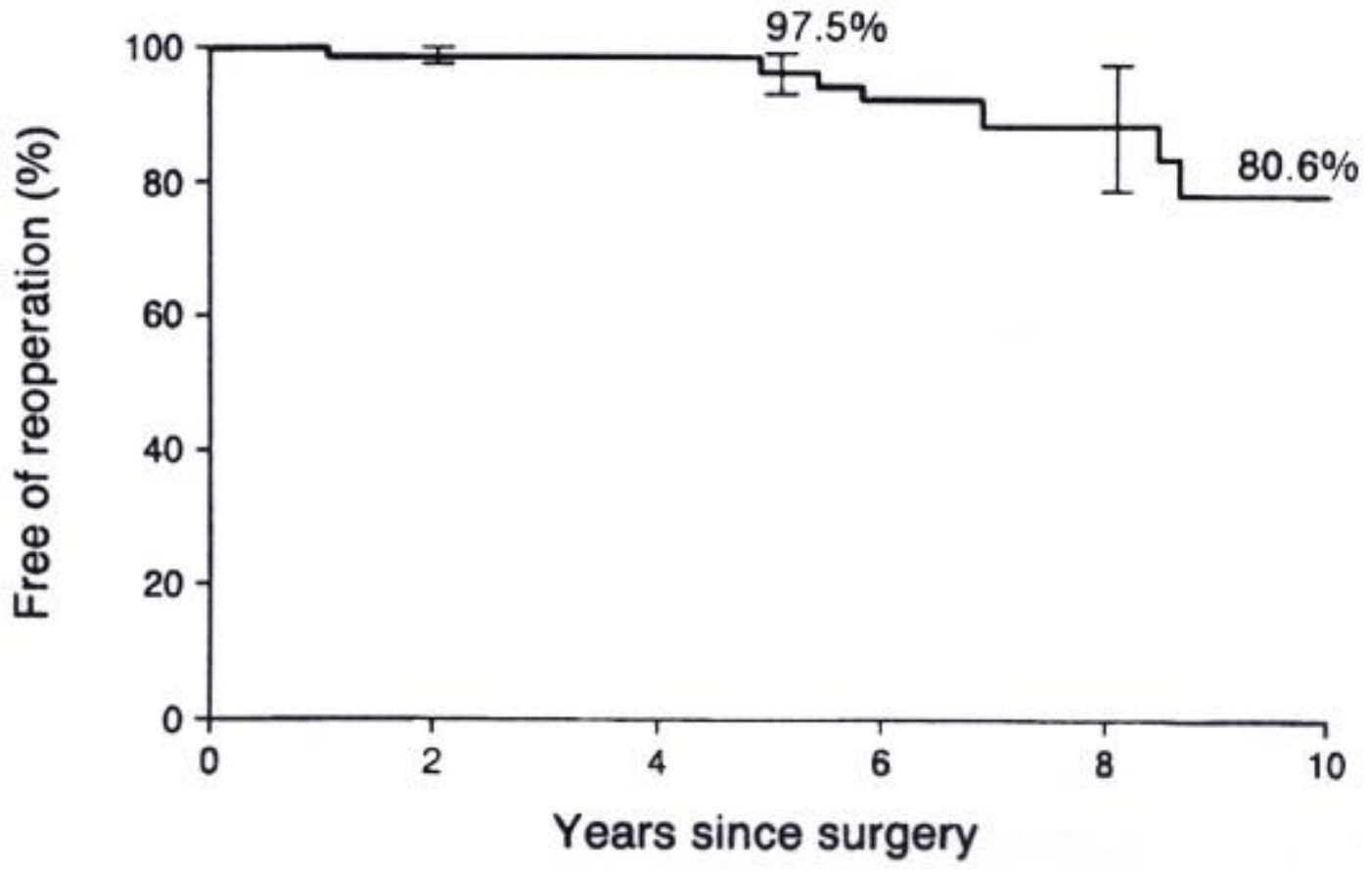


Operative series of Ebstein Anomaly

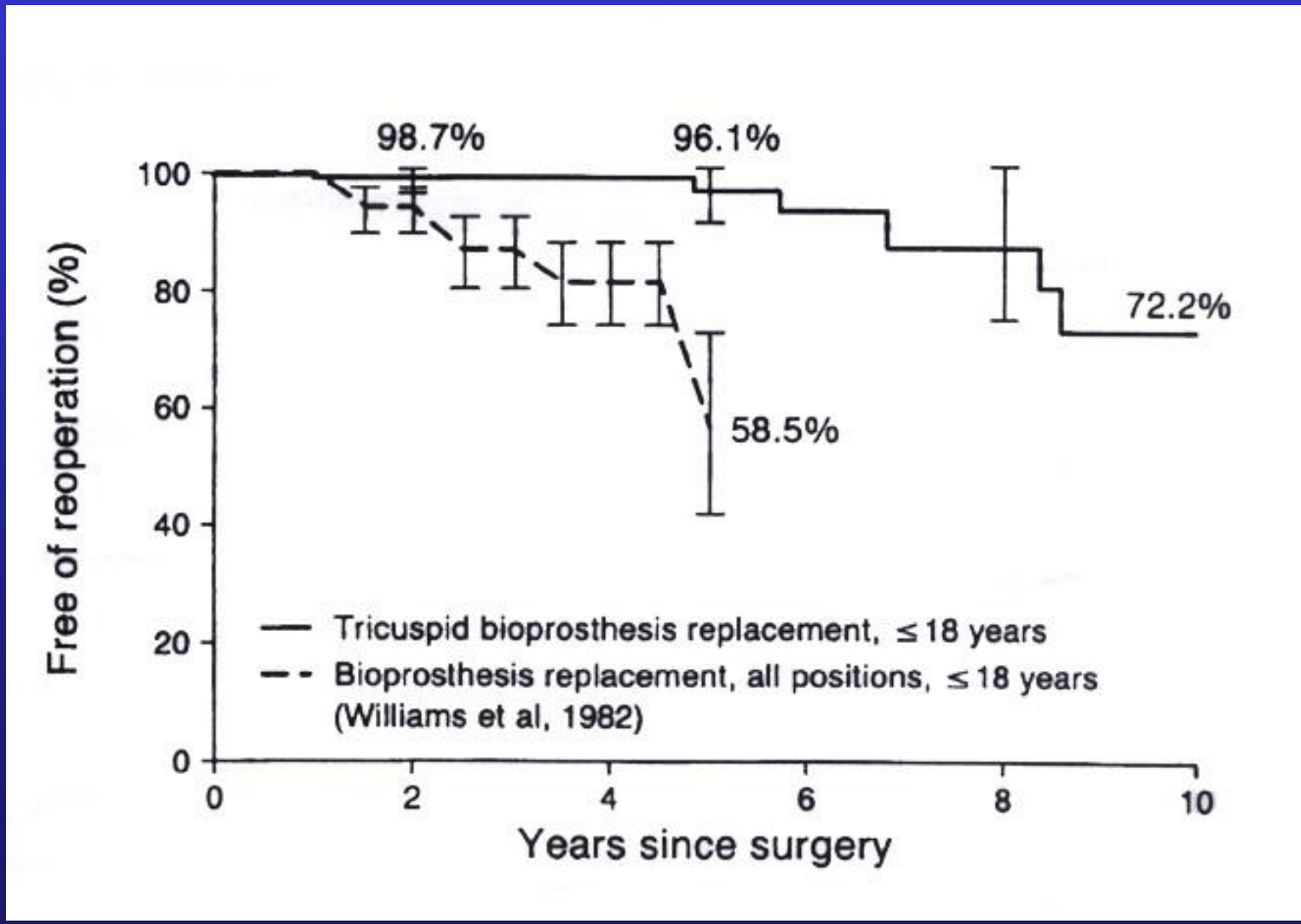
Source	Pt. No.	Age (median)	TVP (%)	Op.Mortality
Lilehei, 1967	8	6-18 yr (12)	25%	50%
Hardy and Roe,1969	6	5-41 yr (22)	100%	17%
Westaby, 1982	24	3-55 yr (20)	4%	17%
*Carpentier, 1988	14	9-51 yr (26.9)	93%	14%
Quagebeur, 1991	10	4-44 yr (22)	100%	0%
Starnes, 1991	5	1-9 d (5)	0%	0%
†Danielson, 1992	189	1-64 yr (19.1)	58.2%	6.3%
Hetzer, 1998	19	2-54 yr (21)	100%	5%
†Kiziltan, 1998	158	1-70 yr (19)	0%	6%
Marianeschi, 1998	10	2.4-31 yr (9)	60%	0%
Vargas, 1998	7	7-16 yr (12)	100%	0%
Chauvaud, 2000	142	1-65 yr (25)	97%	10%
Chauvad, 2003	191	1-65 yr (24.5)	98%	9%

* Clinical reports from the ‘Hospital Broussais † Clinical reports from ‘Mayo Clinic’

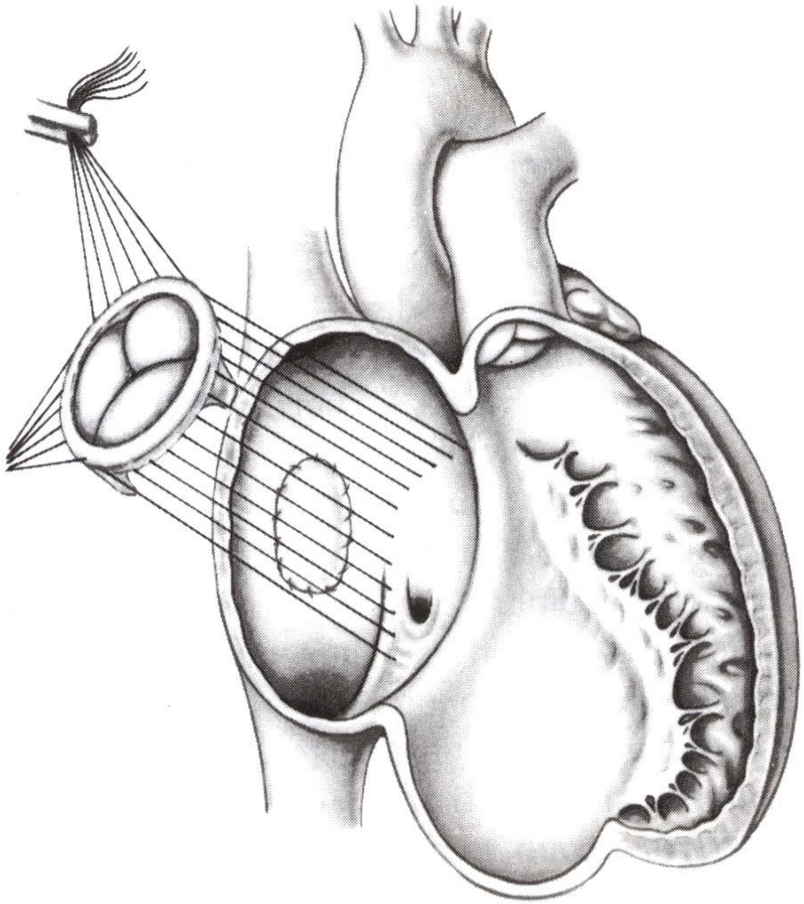
TVR with Bioprosthesis
(Mayo clinic, 1998)

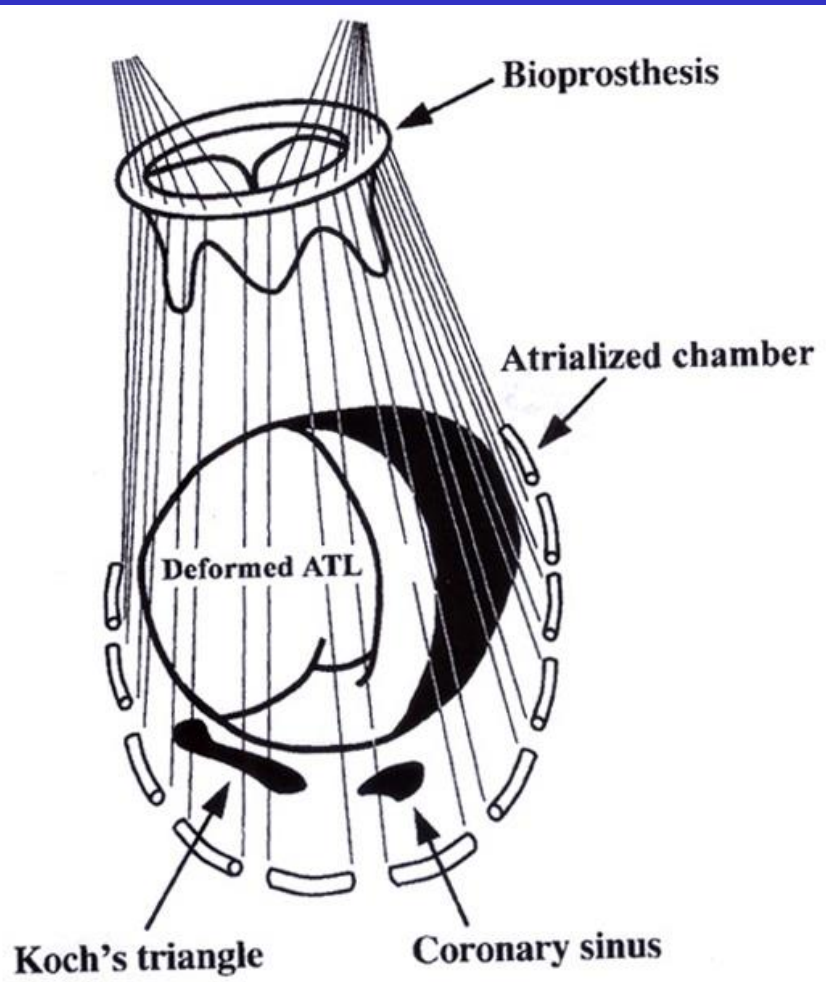


TVR with Bioprosthesis in young age
(Mayo clinic, 1998)



TVR in Ebstein anomaly





1 ½ repair in Ebstein anomaly
-Theoretical advantages-

- Increased capability of valve repair
- Decreased early and late mortality
- Decreased risk of re-operation
- Better functional class

Operative series of Ebstein Anomaly

Source	Pt. No.	Age (median)	TVP (%)	Op.Mortality
Lilehei, 1967	8	6-18 yr (12)	25%	50%
Hardy and Roe,1969	6	5-41 yr (22)	100%	17%
Westaby, 1982	24	3-55 yr (20)	4%	17%
*Carpentier, 1988	14	9-51 yr (26.9)	93%	14%
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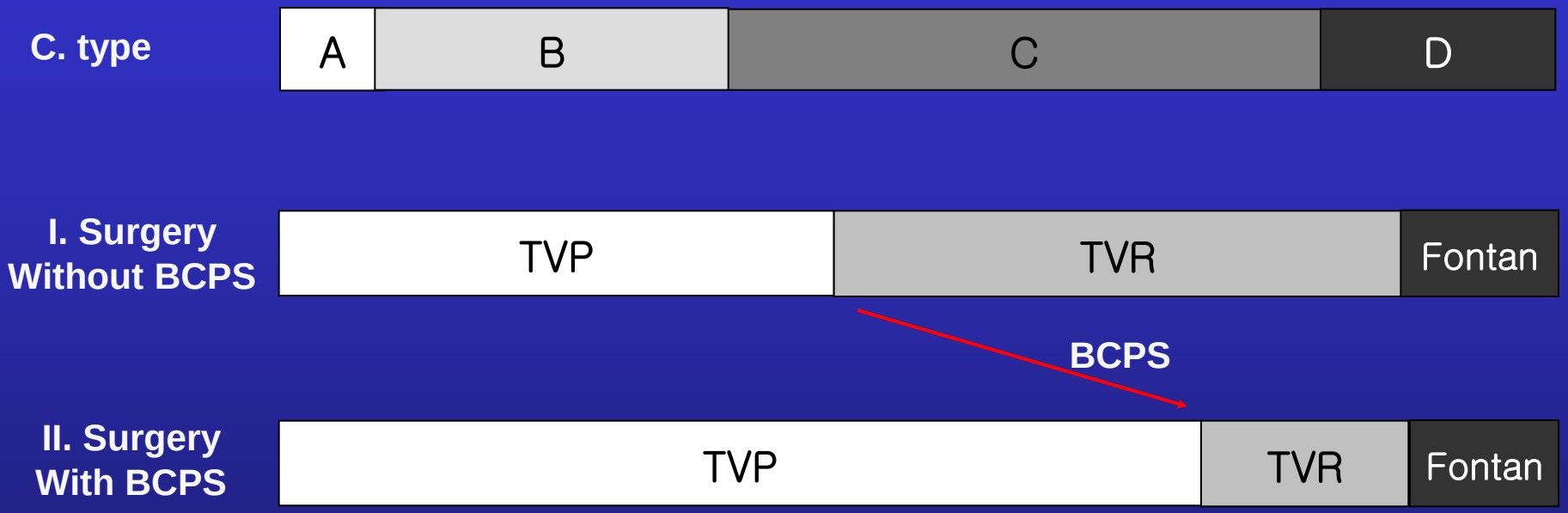
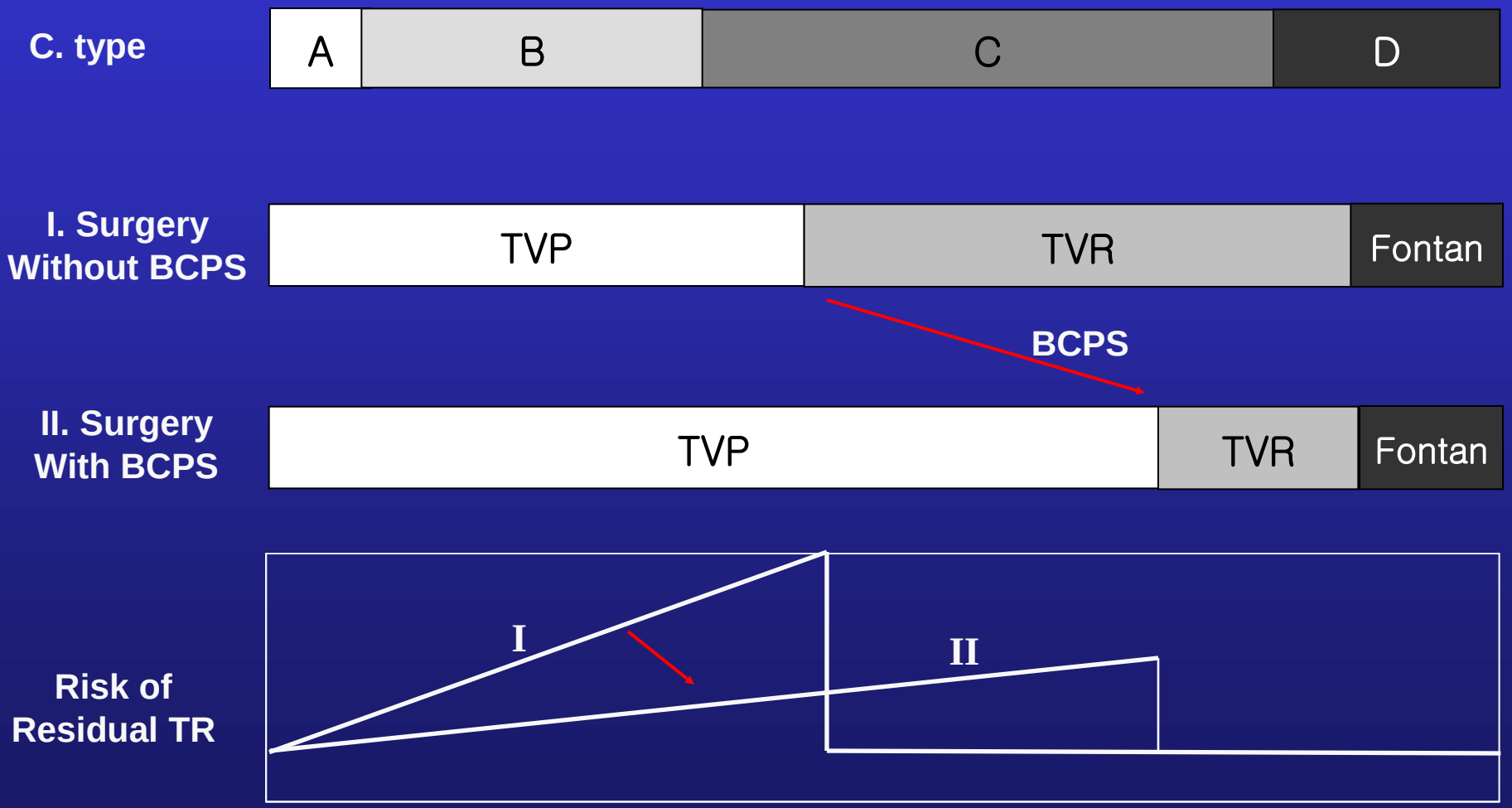
* Clinical reports from the 'Hospital Broussais † Clinical reports from 'Mayo Clinic'

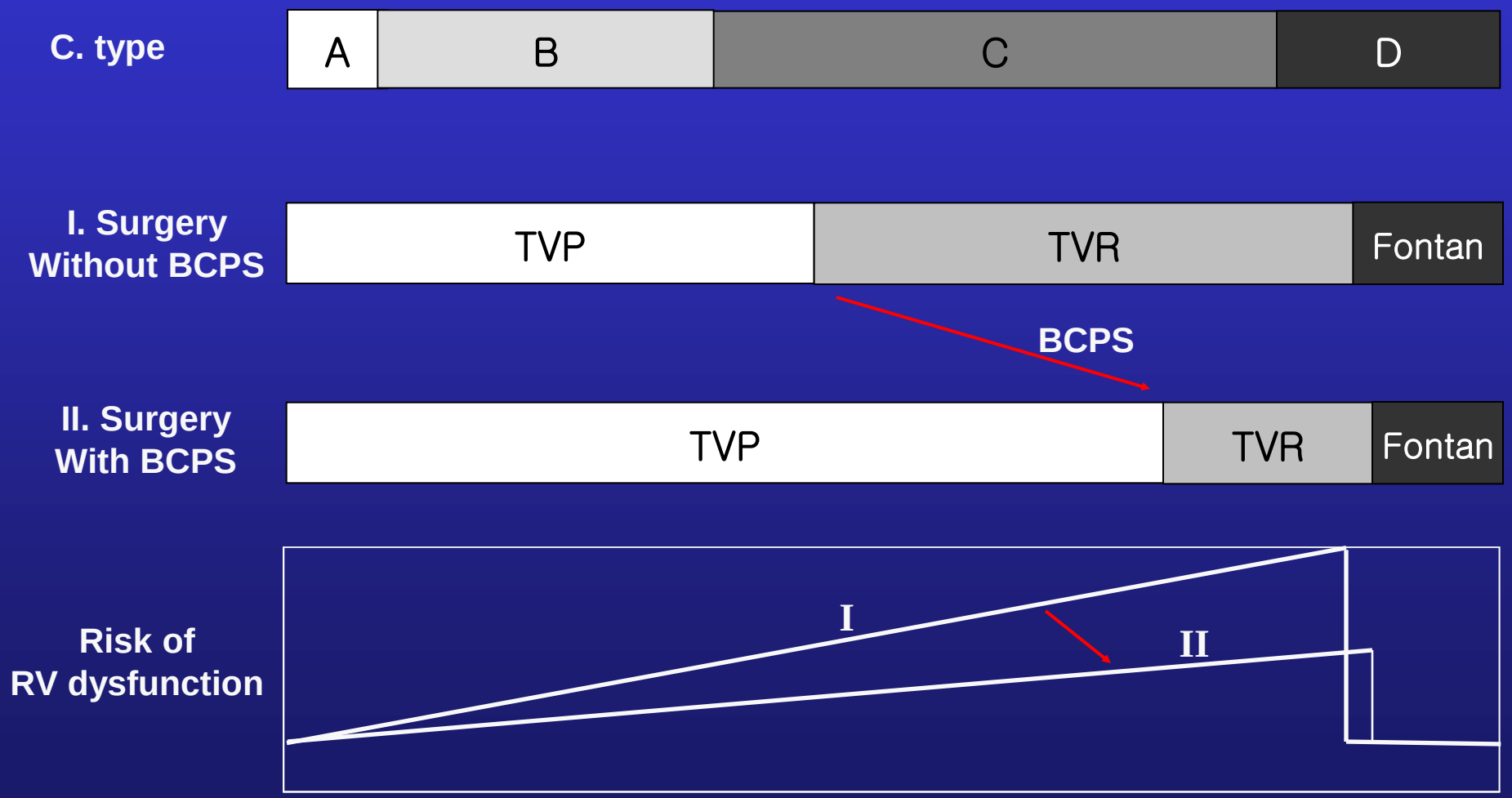
1 ½ repair in Ebstein Anomaly



1 ½ repair in Ebstein Anomaly



1 ½ repair in Ebstein Anomaly



One and a Half Ventricle Repair in Adults: Postoperative Hemodynamic Assessment Using Phase-Contrast Magnetic Resonance Imaging

Jin Woo Chung, MD, Hyun Woo Goo, MD, Yu-Mi Im, MS Hong-Ju Shin, MD, Won Kyoung Jhang, MD, Jae-Kon Ko, MD, and Tae-Jin Yun, MD, PhD

Division of Pediatric Cardiac Surgery, Department of Radiology, and Division of Pediatric Cardiology, Asan Medical Center, University of Ulsan, Seoul, Republic of Korea

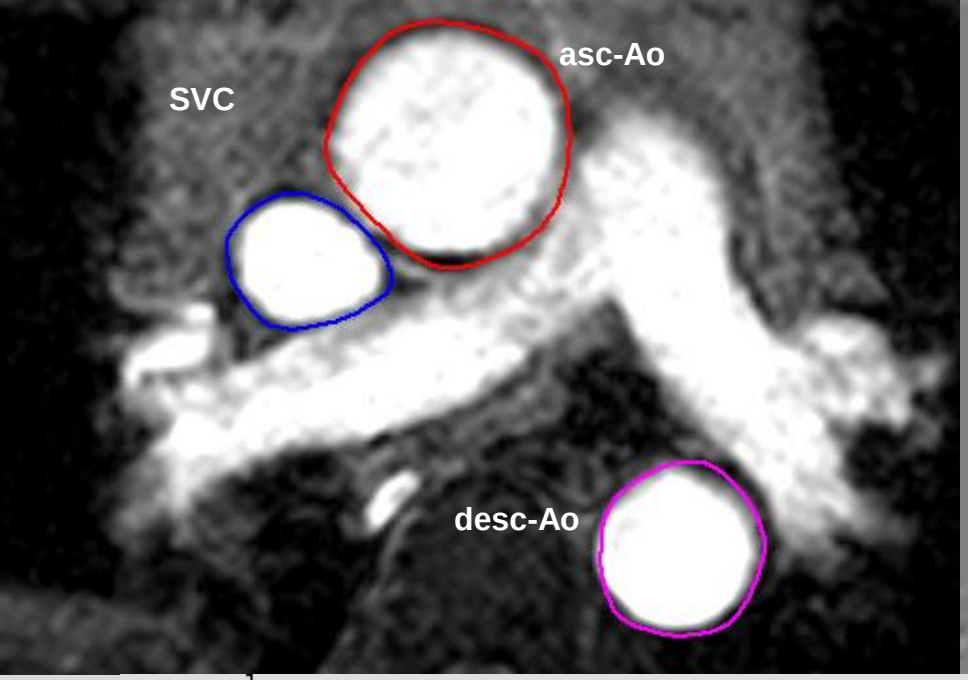
Background. One and a half ventricle repair (1½ repair) strategy has been used for patients with a hypoplastic or dysfunctional right ventricle (RV), or both. We sought to assess the postoperative hemodynamics of 1½ repair using phase-contrast magnetic resonance imaging (PC-MRI).

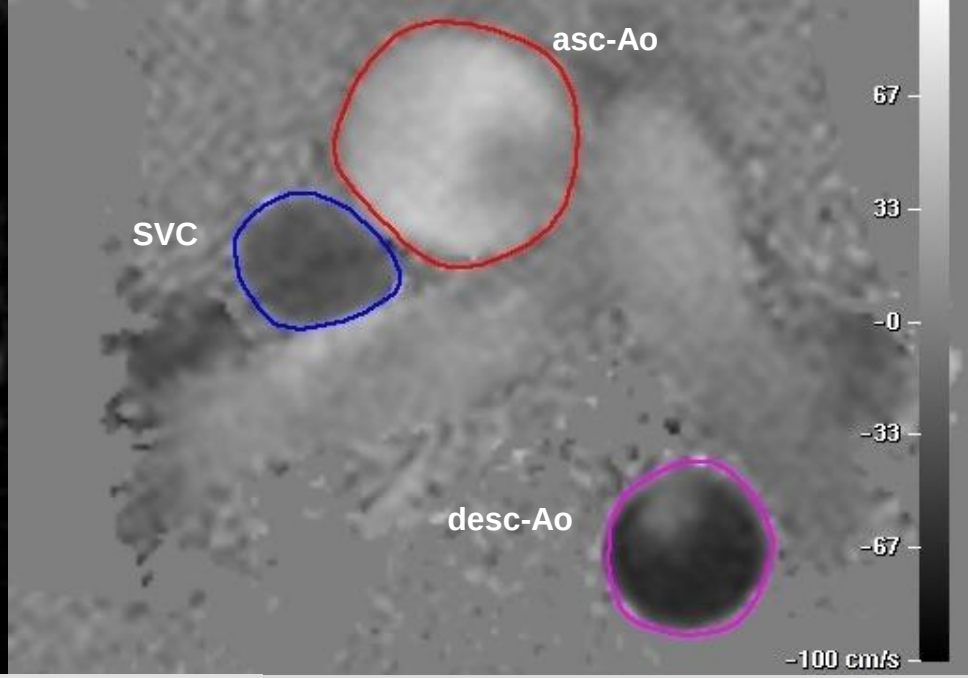
Methods. Ten adults, 9 with Ebstein's anomaly and 1 with tricuspid stenosis, underwent 1½ repair (median age at operation, 42.4 years). The azygos vein was left open in all patients on 1½ repair to prevent severe postoperative central venous hypertension. Postoperative PC-MRI studies were performed to measure blood flow to the ascending aorta ($Q_{Asc\text{-}Ao}$), the main pulmonary artery (Q_{MPA}), the superior vena cava (SVC) (Q_{SVC}), and the branch pulmonary arteries and veins. From these values, blood flow to the upper compartment of the body (Q_{UC}), right ventricular volume unloading effect ($Q_{MPA}/Q_{Asc\text{-}Ao}$), proportion of blood flow to the upper compartment of the body ($Q_{UC}/Q_{Asc\text{-}Ao}$), and venous return to arterial forward flow ratio of the upper compartment of the body (Q_{SVC}/Q_{UC}) were calculated. Two patients also underwent preoperative PC-MRI.

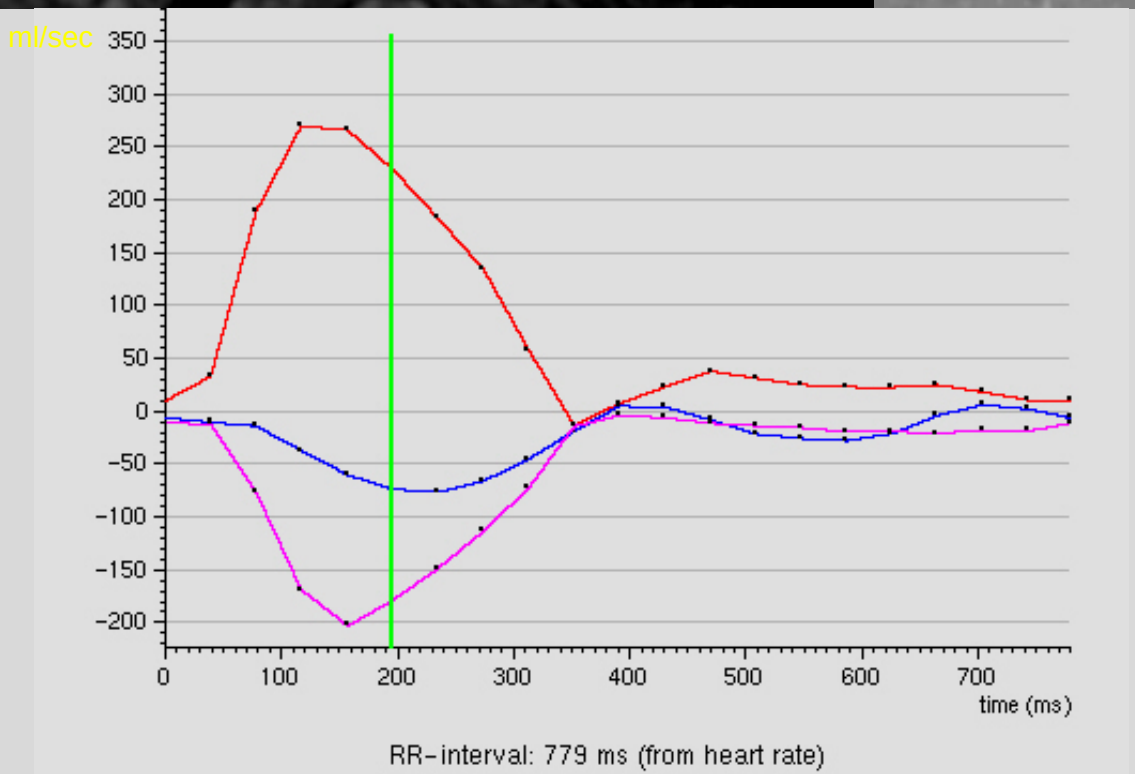
Results. On PC-MRI, $Q_{MPA}/Q_{Asc\text{-}Ao}$, $Q_{UC}/Q_{Asc\text{-}Ao}$, and Q_{SVC}/Q_{UC} were 0.58 to 0.84 (median, 0.67), 0.19 to 0.36 (median, 0.27), and 0.47 to 1.57 (median, 0.93, lower than 1.0 in 7 patients), respectively. In 2 patients who had preoperative and postoperative PC-MRI, $Q_{UC}/Q_{Asc\text{-}Ao}$ decreased from 0.26, 0.32 to 0.21, 0.28, respectively.

Conclusions. After 1½ repair, right ventricular volume unloading was effective in all patients, but intercaval collateral veins (ie. $Q_{SVC}/Q_{UC} < 1$) appeared to develop in most of the patients. Furthermore blood flow to the upper compartment of the body appeared to diminish, presumably due to postoperative elevation of central venous pressure.

(Ann Thorac Surg 2011;92:193–9)



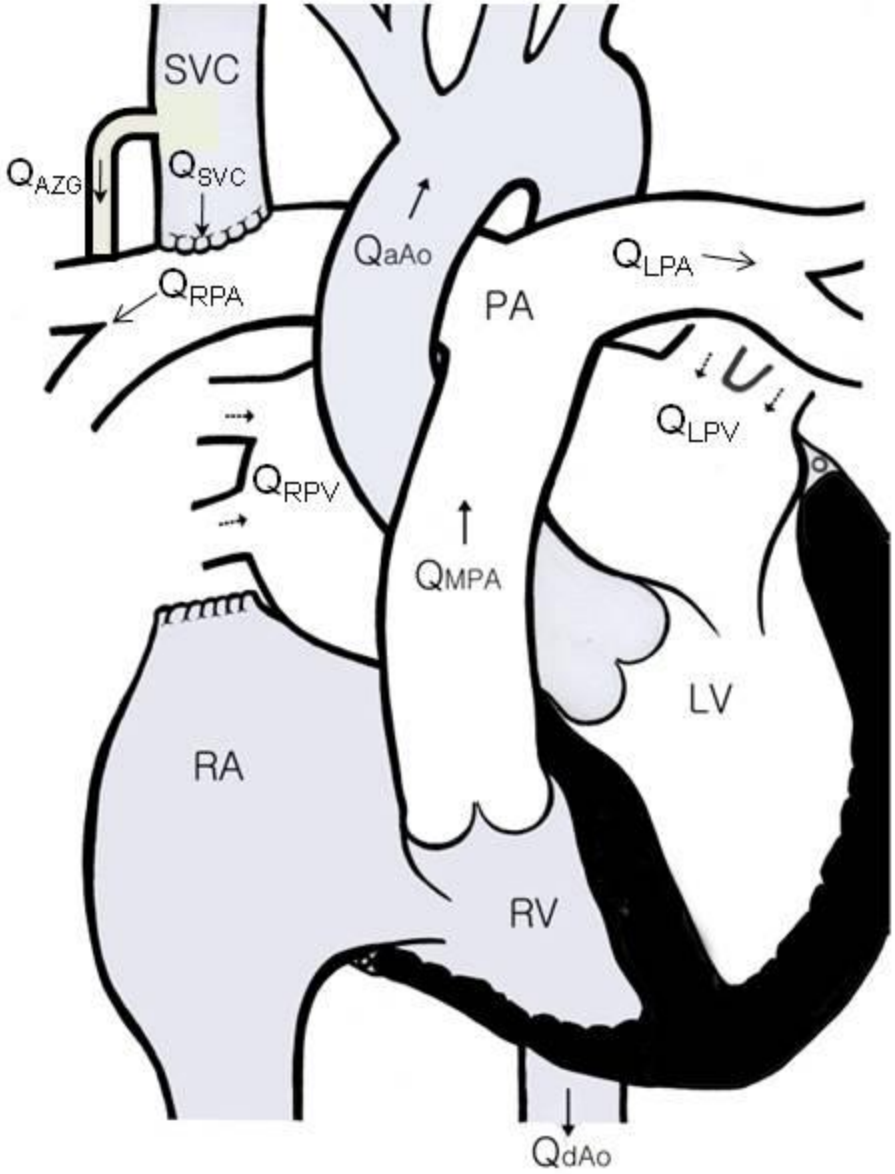




: asc-Ao

: SVC

: desc-Ao



1) RV volume unloading

Q_{MPA} / $Q_{a\text{-}Ao}$

2) Collateral flow to the lungs

$Q_{Coll} = Q_{L\text{-}Coll} + Q_{R\text{-}Coll}$

$Q_{L\text{-}Coll} = Q_{LPA} - Q_{LPV}$

$Q_{R\text{-}Coll} = Q_{RPA} - Q_{RPV}$

3) Flow to the upper body

$Q_{UC} = Q_{a\text{-}Ao} - Q_{Coll} - Q_{d\text{-}Ao}$

4) SVC-IVC Collateral flow

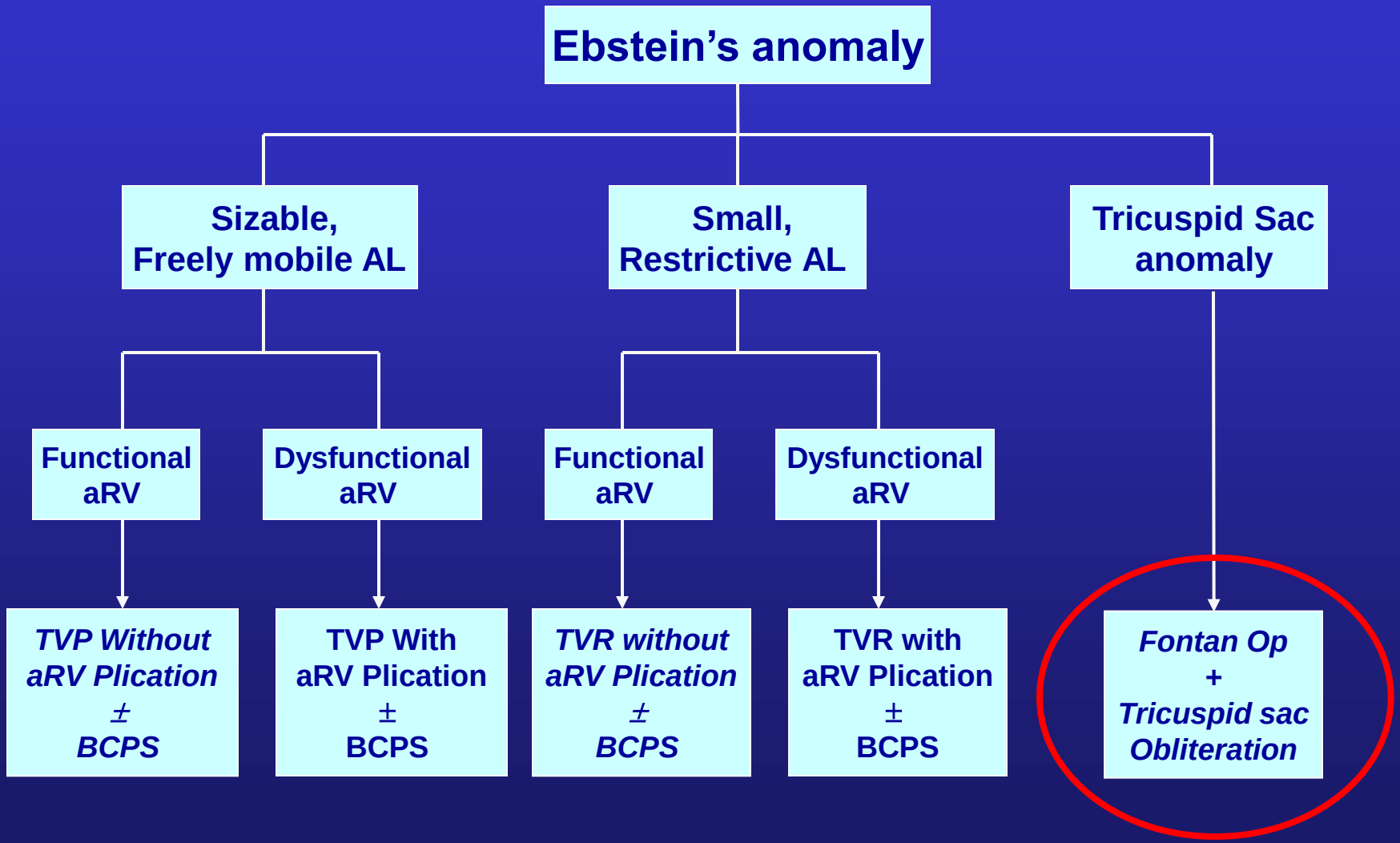
$Q_{SVC\text{-}IVC} = Q_{UC} - Q_{SVC}$

Table 3. Comparison of preoperative and postoperative PC-MRI data in patient 9 and 10 (cc/beat)

Pt		Q_{asc-Ao}	$Q_{desc-Ao}$	Q_{coll}	Q_{UC}	Q_{UC}/Q_{asc-Ao}	Q_{MPA}	Q_{MPA}/Q_{asc-Ao}
9	Pre-op	45.5	33.7	0	11.8	0.26	47.6	1.05
	Post-op	60.6	47.7	0	12.9	0.21	41	0.68
10	Pre-op	65.5	44.7	0	20.8	0.32	65.3	1.0
	Post-op	53.4	38.4	0	15	0.28	45	0.84

PC-MRI: phase-contrast velocity mapping magnetic resonance imaging; Pre-op: preoperative; Post-op: postoperative 3 months in patient 9 and 2 months in patient 10; Q_{asc-Ao}: ascending aortic blood flow; $Q_{desc-Ao}$: descending aortic blood flow; Q_{coll}: aortopulmonary collateral blood flow; Q_{UC}: blood flow to the upper compartment of the body; Q_{MPA}: blood flow of the main pulmonary artery.

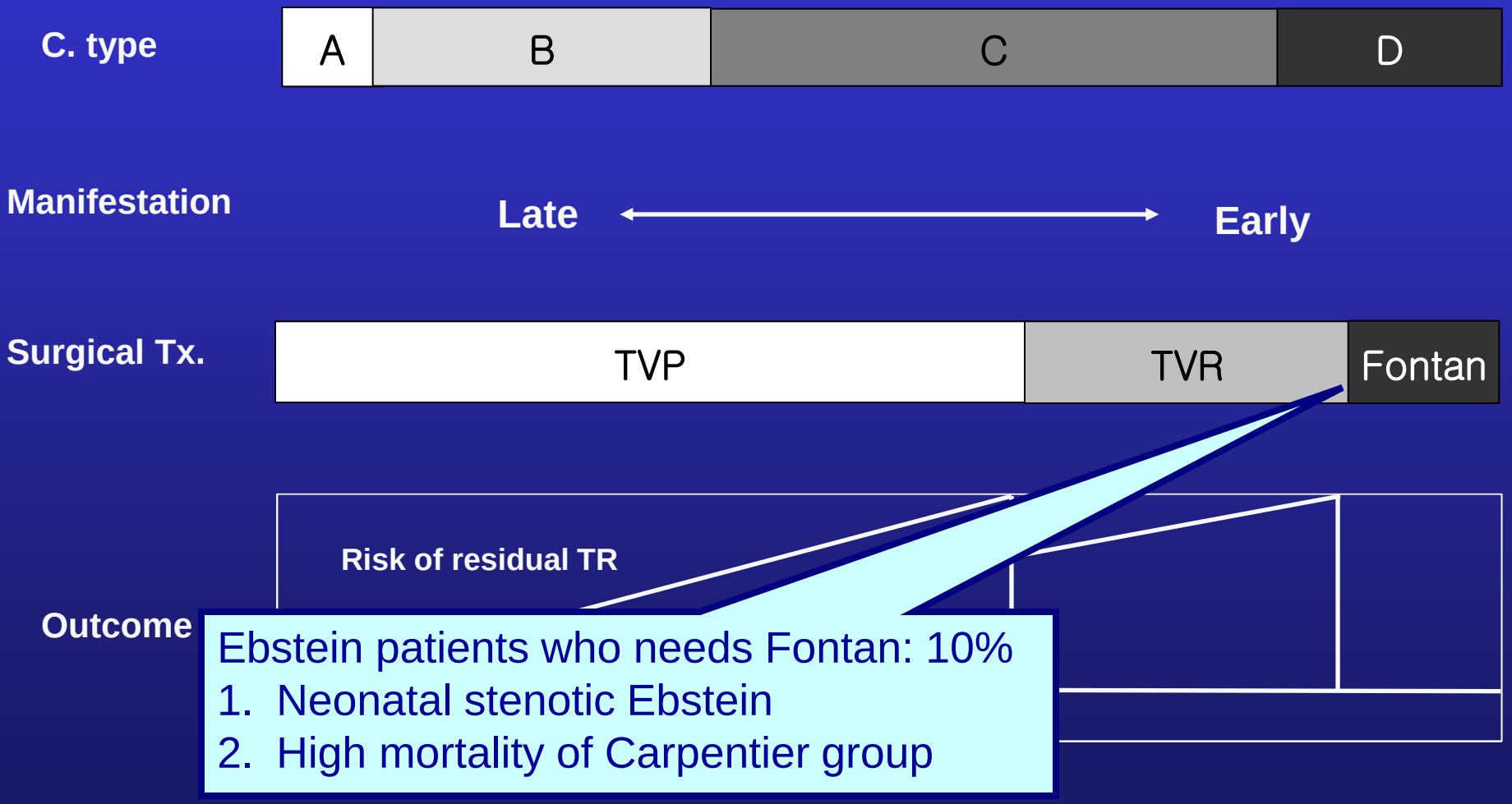
Surgical algorithm (AMC)



Fontan op in Ebstein anomaly

- 10% of entire cohort
- Role of Fontan operation in old age: ?
- Issues in surgical technique:
  1) Extracardiac Fontan
  2) Tricuspid sac obliteration

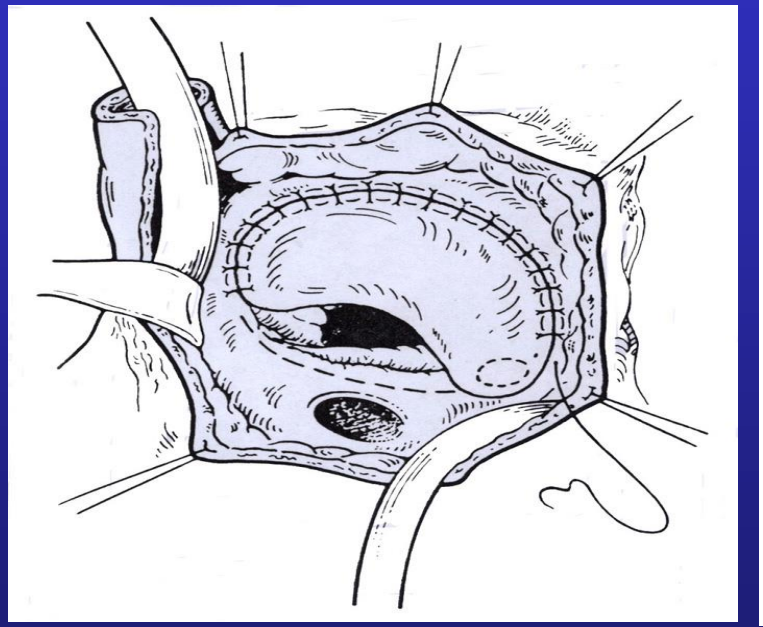
Ebstein Anomaly as a Spectrum



Ebstein patients who needs Fontan: 10%

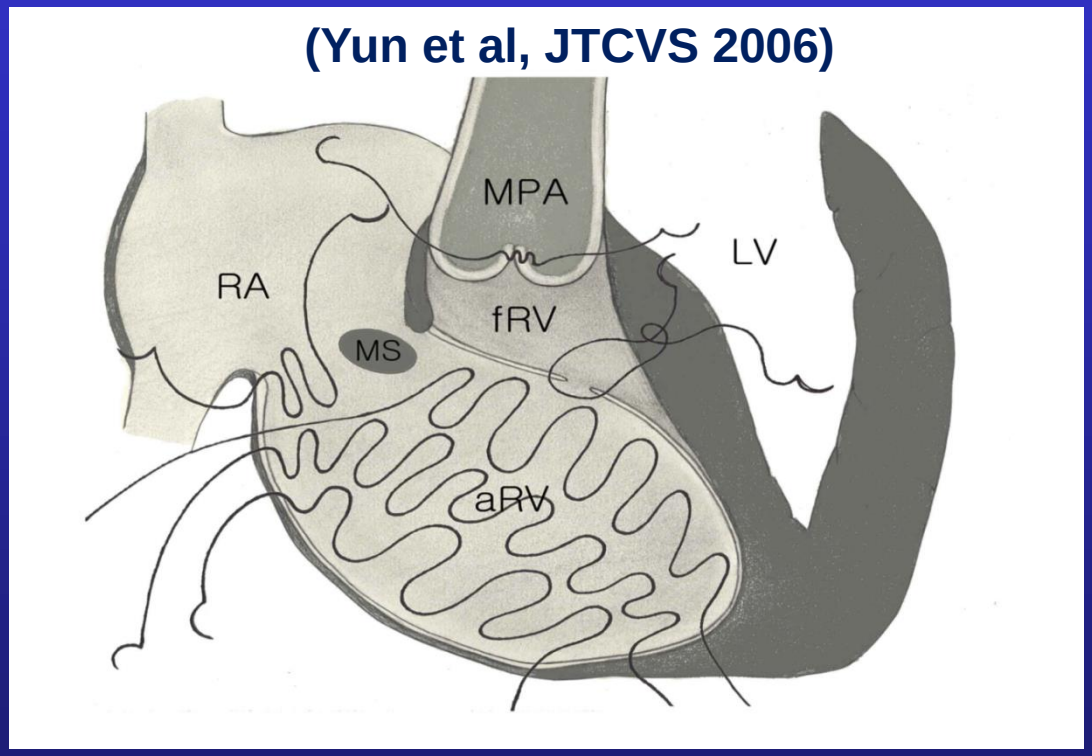
1. Neonatal stenotic Ebstein
2. High mortality of Carpentier group

Techniques of RV exclusion



Patch closure of TV annulus
(Starnes Op)

(Yun et al, JTCVS 2006)



Suture obliteration of aRV

Summary

- Fontan track: 10% of entire cohort
- TVR for Ebstein's anomaly is a risky operation, especially in patients with severe disease.
- Hetzer procedure is technically feasible, and leads to excellent longterm outcome
- Addition of BCS is beneficial upon TVP/TVR.