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OIRPOSDRU REGIUNEA CENTRU
UNIVERSITATEA DE MEDICINĂ ŞI
FARMACIE "CAROL DAVILA"
BUCUREŞTI

AD-COR Program inovativ de formare in domeniul cardiologiei pediatrice
POSDRU/179/3.2/S/152012

Data: **19-10-2015**

MODUL TEORETIC

Valvular Disease in Children

Imputernicit: Prof. Dr. Tammam Youssef

Activitate prestata de I.R.C.C.S. POLICLINICO SAN DONATO – MILANO, ITALIA
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Beneficiar: Universitatea de Medicină și Farmacie „Carol Davila” București

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POSDRU/179/3.2/S/152012

Status of the Art: Mitral Valve Repair

Alessandro Frigiola
Tammam Youssef

IRCCS Policlinico San Donato
Milan (Italy)

Valvular Disease in Children

Preservation of the natural valve is the major goal of any heart surgery particularly in neonates and children.

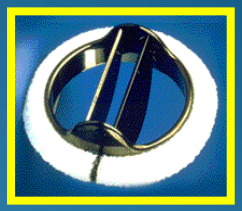
Yet, prosthetic valve replacement, considered the last therapeutic option in paediatric patients, is sometimes unavoidable.

The small size of the native valve annulus, atrium and ventricle pose problems that are unique in children.

Paediatric valve disease

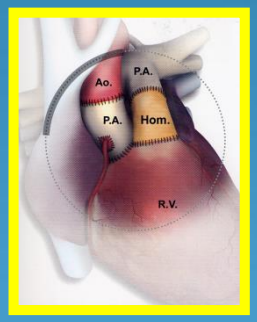
Surgical options

- Mechanical valve prosthesis



- Homografts and Bioprosthesis

- Ross Operations



- Repair

Paediatric valve disease

Surgical options

Limitations

Mechanical valves

- Lack of appropriately sized prosthesis
- Absence of the potential for growth:

  repeat replacement is needed
- Lifetime anticoaugulation is associated with difficulties with future pregnancy, thronboembolism and bleeding
- Risk of endocarditis

Paediatric valve disease

Surgical options

Limitations

Homografts and Bioprosthesis

- Do not allow for growth
- Limited durability
- Limited availability of appropriately sizes

Paediatric valve disease

Surgical options

Limitations

Ross Operation

- **Double Valve Operation**
- **Durability**
- **Long operation**

Paediatric valve disease

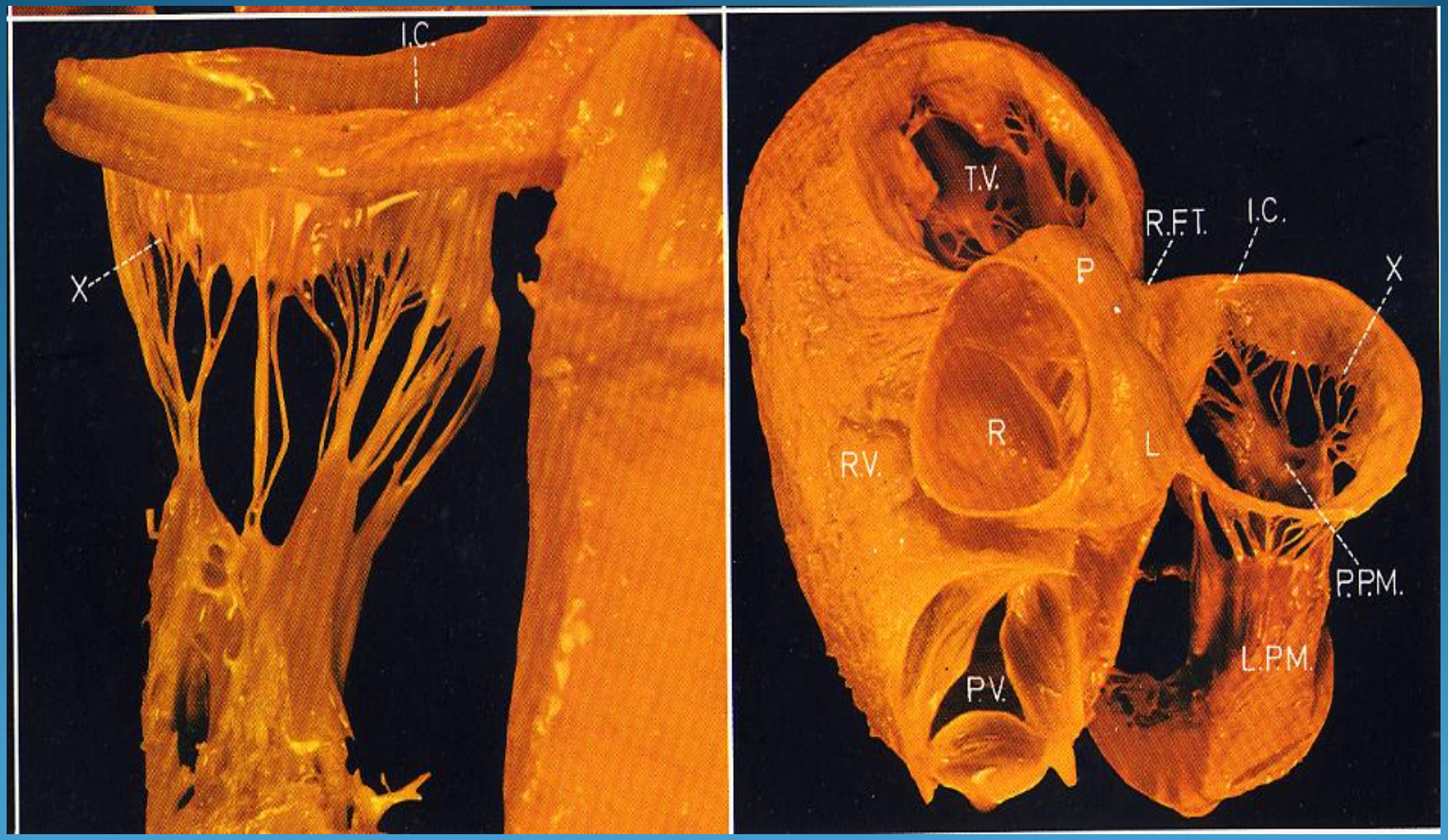
Surgical options

Limitations

Repair

- **Residual Lesions continue to progress**
- **Usually palliative solution**

Mitral Valve Disease



Mortality of Mitral valve surgery in children

Author	Year	No.of patients	Mortality
Moller J.H	2001	139	21%
Brizard C.P	2008	71	4%
James L. Monro	2001	44	Age < 5y 22% Age > 5y 4.8%
Brian K. Eble	2003	53	29%
Hunaid A. Vohra	2006	24 all <5y	30%
Alfred E. Wood	2005	45	11%
Kilian Ackermann	2007	69	13%
Wolfram Beierlein	2007	54	< 2y 42% > 2y 6%

ELSEVIER

European Journal of Cardio-thoracic Surgery 31 (2007) 860–865

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY

www.elsevier.com/locate/ejcts

Long-term follow-up after mitral valve replacement in childhood: poor event-free survival in the young child☆

Wolfram Beierlein, Vera Becker, Robert Yates, Victor Tsang, Martin Elliott, Marc de Leval, Carin van Doorn*

Cardiothoracic Unit, Great Ormond Street Hospital for Children National Health Service Trust, London, United Kingdom

Received 29 November 2006; received in revised form 22 January 2007; accepted 2 February 2007

Mortality ≤ 2 years old pts = 42%

Survival 15 years follow-up

33% ≤ 2 years old pts

75% older pts

Mortality > 2 years old pts = 6%

Freedom from redo 15 years follow-up

0% for prostheses <23mm

83% for prostheses >23mm

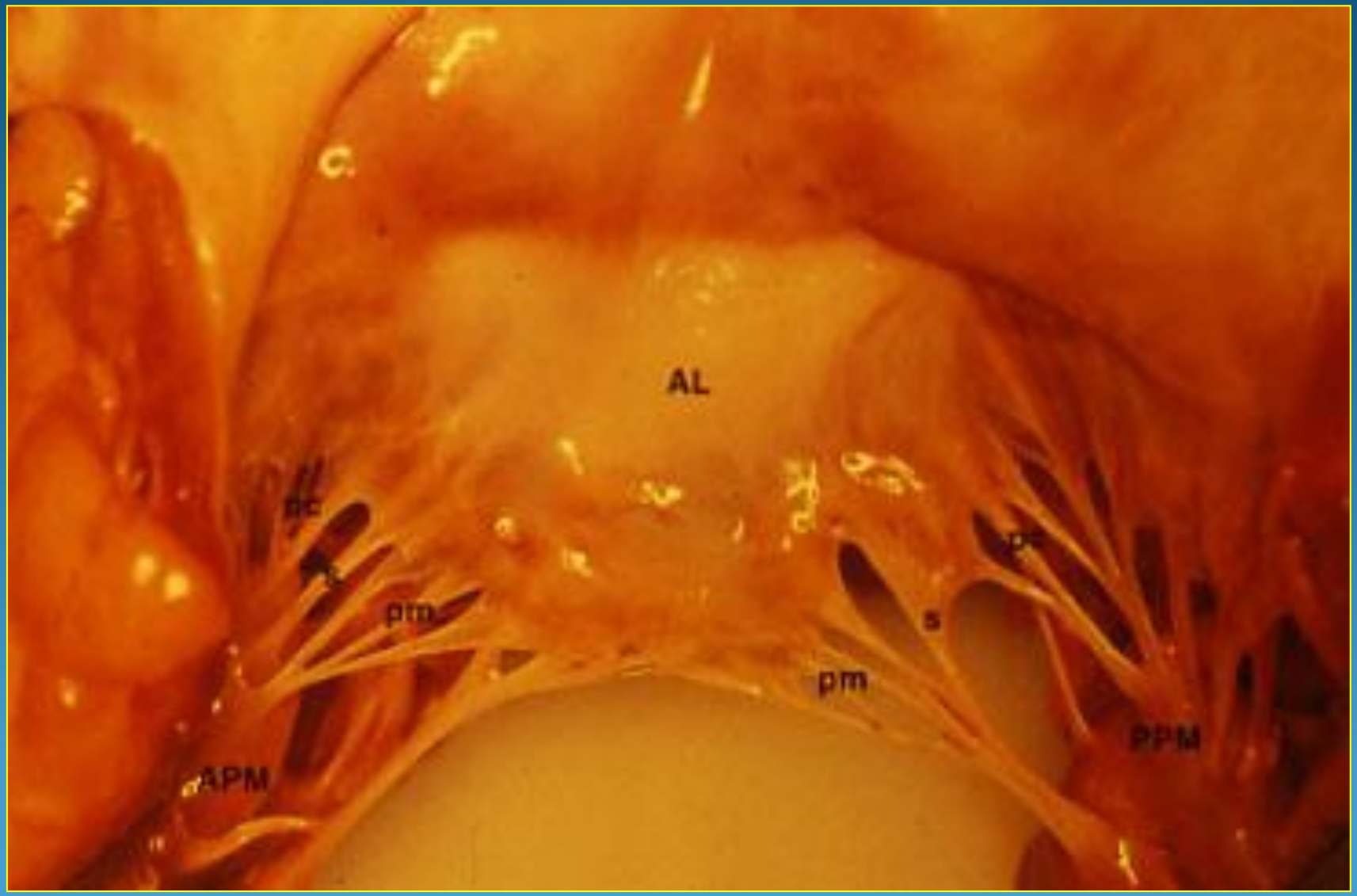
Freedom from all adverse events at 10years follow-up: 17%

"In children every effort should be made to preserve the native valve"

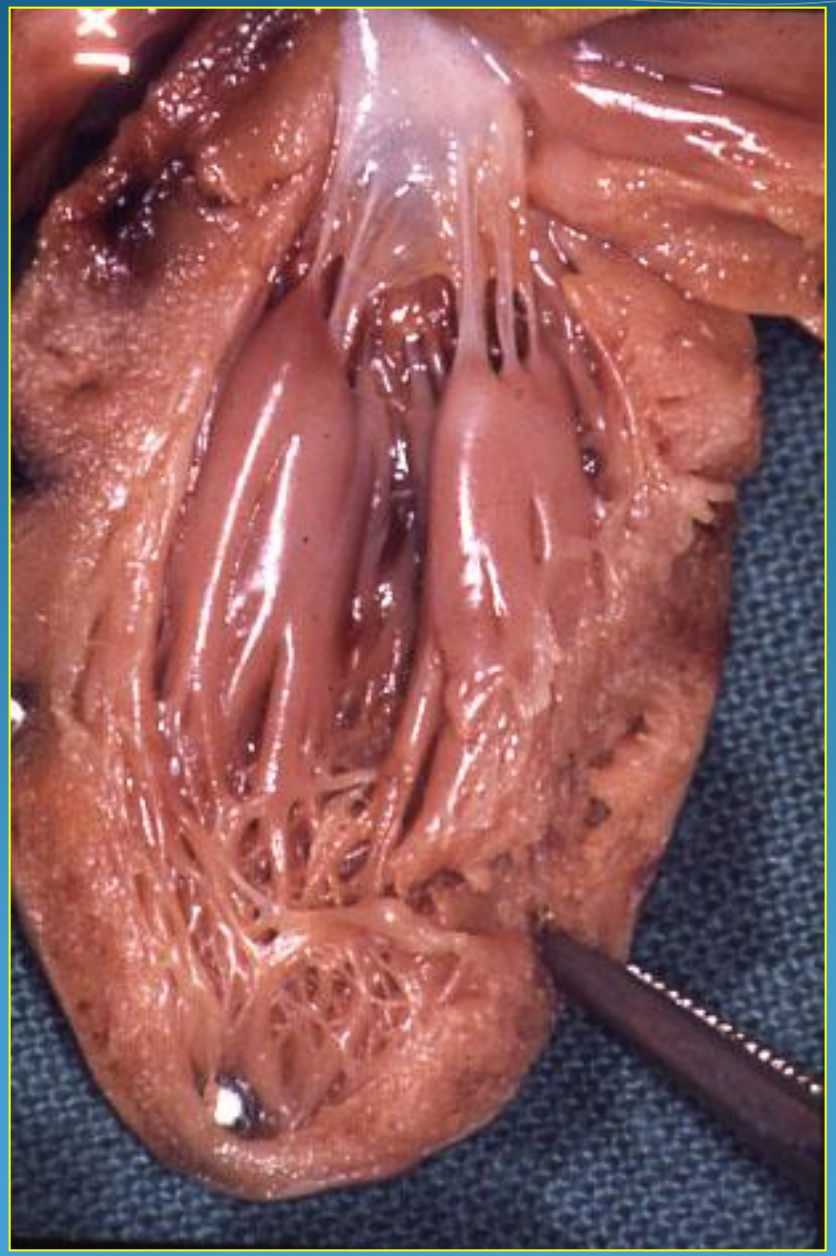
Mortality of Aortic valve surgery in children

Author	Year	No.of patients	Mortality
James L. Monro	2000	56	5.3%
Domenico Mazzitelli	1998	46	17%
JW Brown	1988	257	Age > 6 months 4% Age < 6 months 60%
Mark W. Turrentine	2001	99	8%
Raoul Arnold	2008	30	0%
RUZMETOV Mark	2006	147	7.5%
Nahidh Hasaniya	2004	21	0%
John W. Brown	2003	508	8%
Andreas R. Tiete	2006	27	3.7%

Normal Mitral Valve

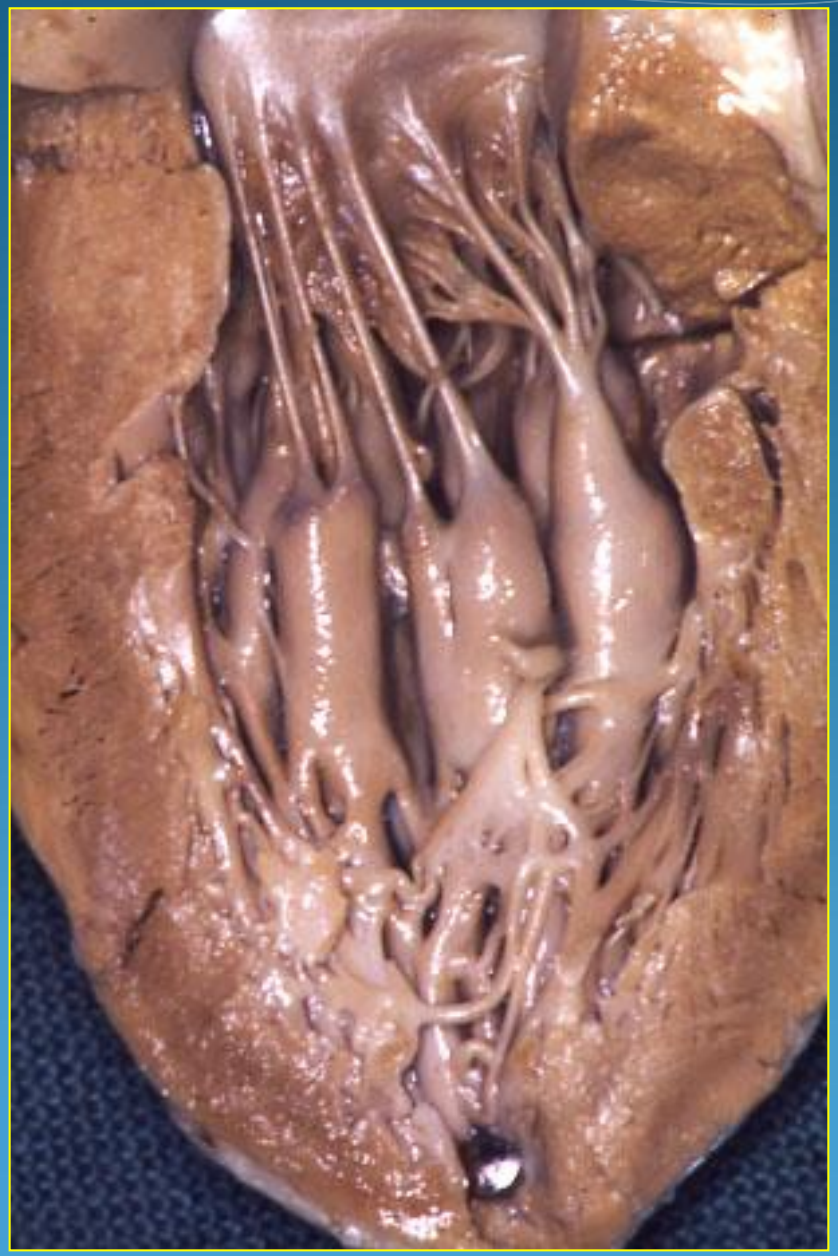


Normal Mitral Valve



Short chordae

Multiple papillary Muscles



Absent Papillary Muscles

Papillary Muscles Fusion

Mitral Valve Disease Anatomical Classification

TYPE 1 ÷ **Supravalar**

TYPE 2 ÷ **Valvar**

- TYPE 2a Annular Defects
- TYPE 2b Leaflet Defects

TYPE 3 ÷ **Subvalvar**

- TYPE 3a Chordal Defects
- TYPE 3b Papillary Defects

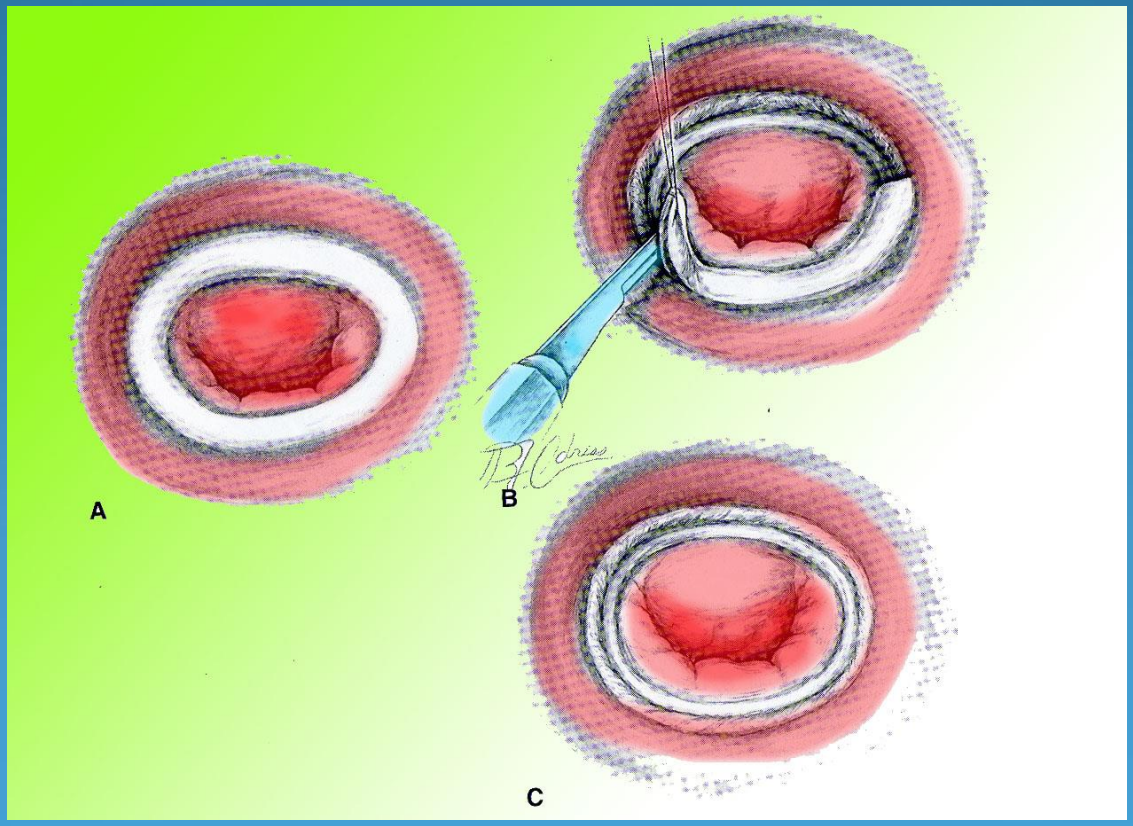
TYPE 4 ÷ **Mixed Lesions**

S. Mitruka – J. Lamberti (Ann. Thor. Surg. 2000)

Mitral Valve Disease Anatomical Classification and Type of disease

TYPE 1 ÷ SUPRAVALVAR

SUPRAVALVAR MITRAL RING



Mitral Valve Disease Anatomical Classification and Type of disease

TYPE 2 ÷ VALVAR

ANNULUS

A) - **Midvalvar ring**
Fibrous mitral ring within the substance of the valve leaflets causing valvar stenosis. Often Associated fused papillary muscles

B) - **Hypolplasia**
Associated with hypolastic left ventricles

C) - **Dilatation**

Mitral Valve Disease Anatomical Classification and Type of disease

TYPE 2 ÷ VALVAR

LEAFLET

A) - Cleft

B) - Excessive tissue
(Barlow Prolaps – mixomatous)

C) - Double orifice

Mitral Valve Disease Anatomical Classification and Type of disease

TYPE 3 ÷ SUBVALVAR

CHORDAE

A) - Chordae tendineae agenesis

B) - Shortened – tunnel valve (splitting)

C) - Chordae elongated

Mitral Valve Disease Anatomical Classification and Type of disease

TYPE 3 ÷ SUBVALVAR

PAPILL. MUSCL.

A) - Hypoplasia/agenesis

B) - Papillar muscl. Shortened
The chordae are short and papillary muscles hypertrophied

C) - Papillar muscl. Elongated
Papillary muscles are thin and elongated

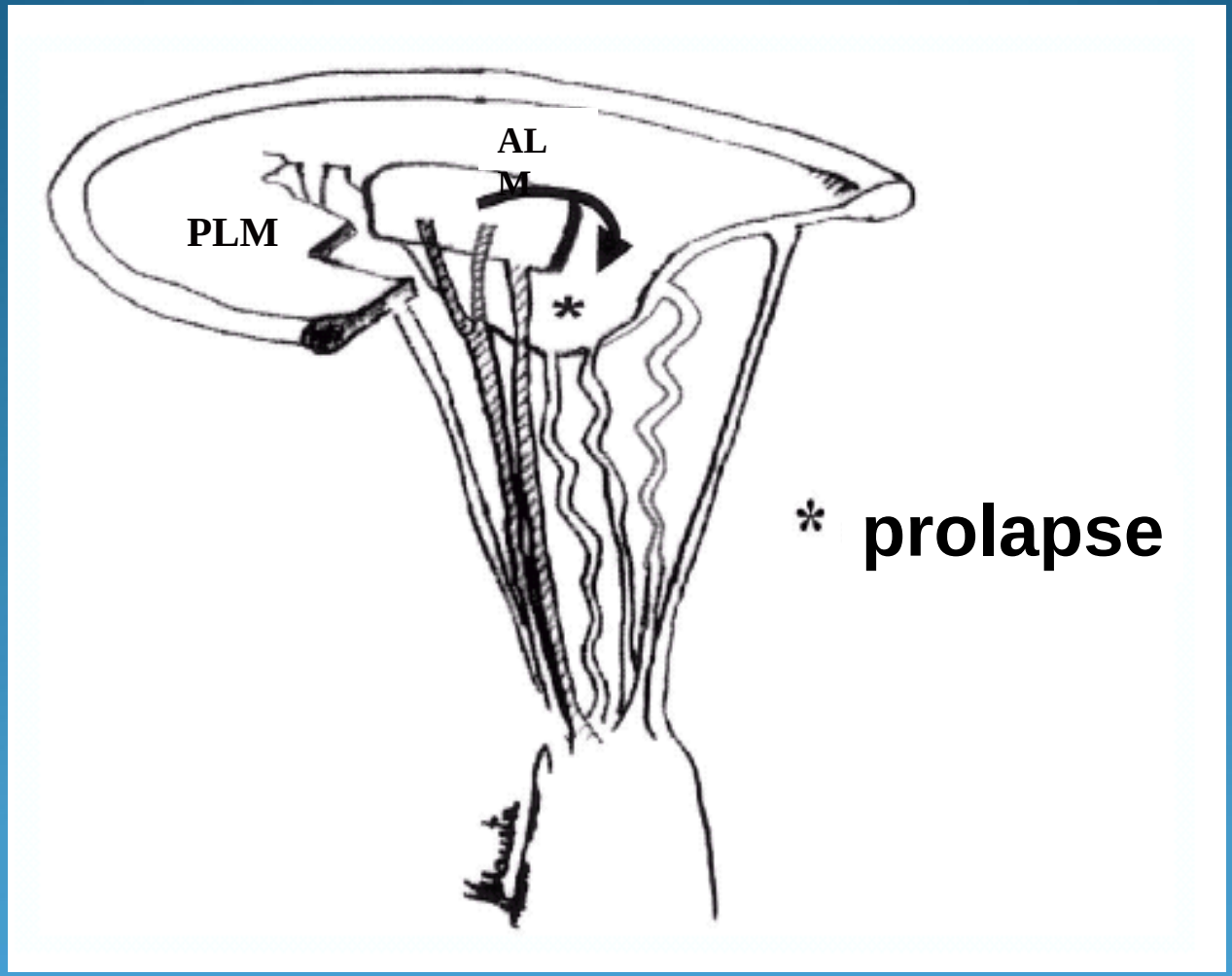
D) - Single: parachute valve

E) - Hammock valve

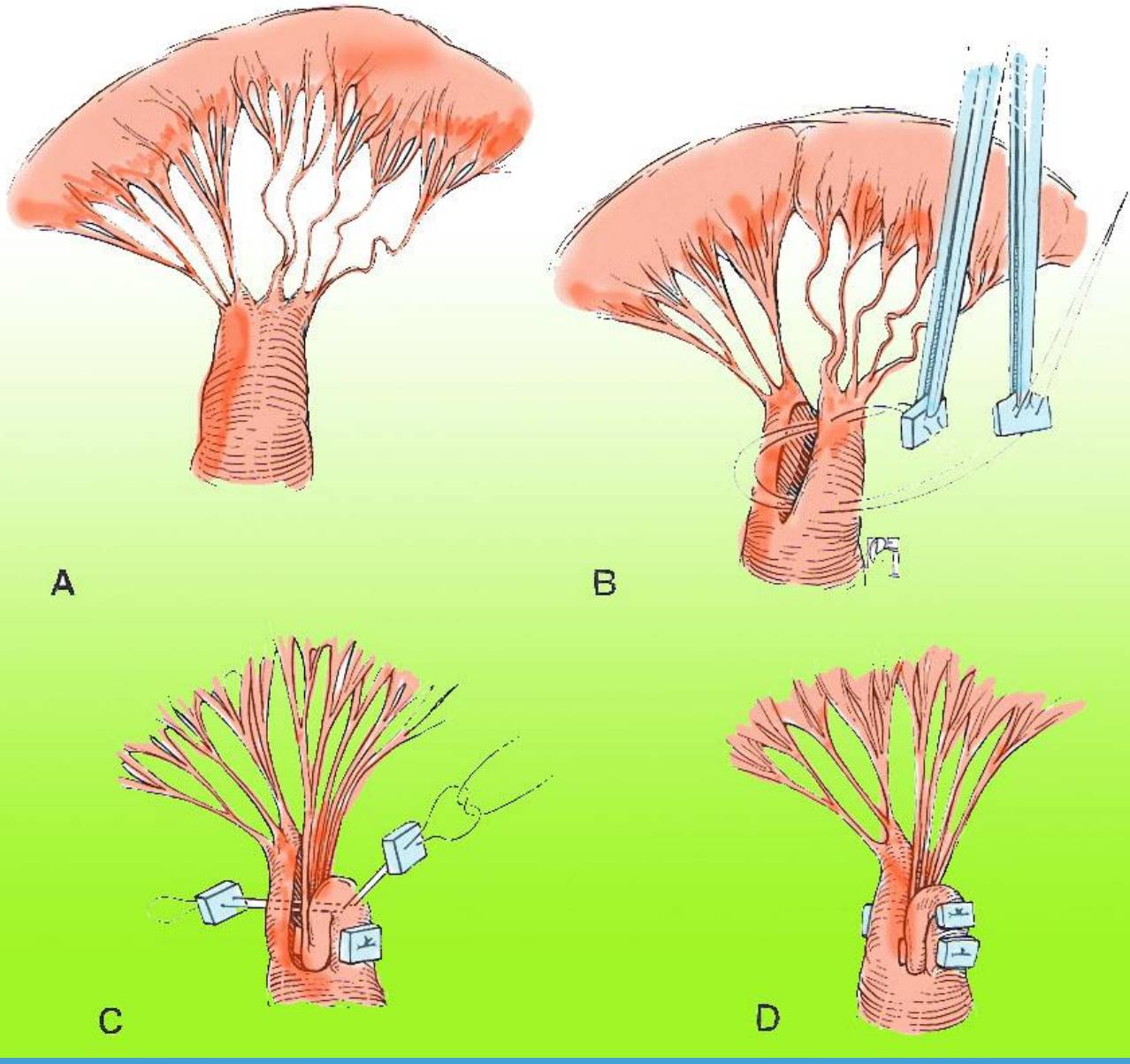
Mitral valve repair techniques

- Commisurotomy
- Papillary muscle splitting
- Quadrangular resection
- Chordal shortening
- Edge to Edge technique
- Annuloplasty
- Cleft closure
- Artificial chordae
- Chordal transpositionn
- Fenestration

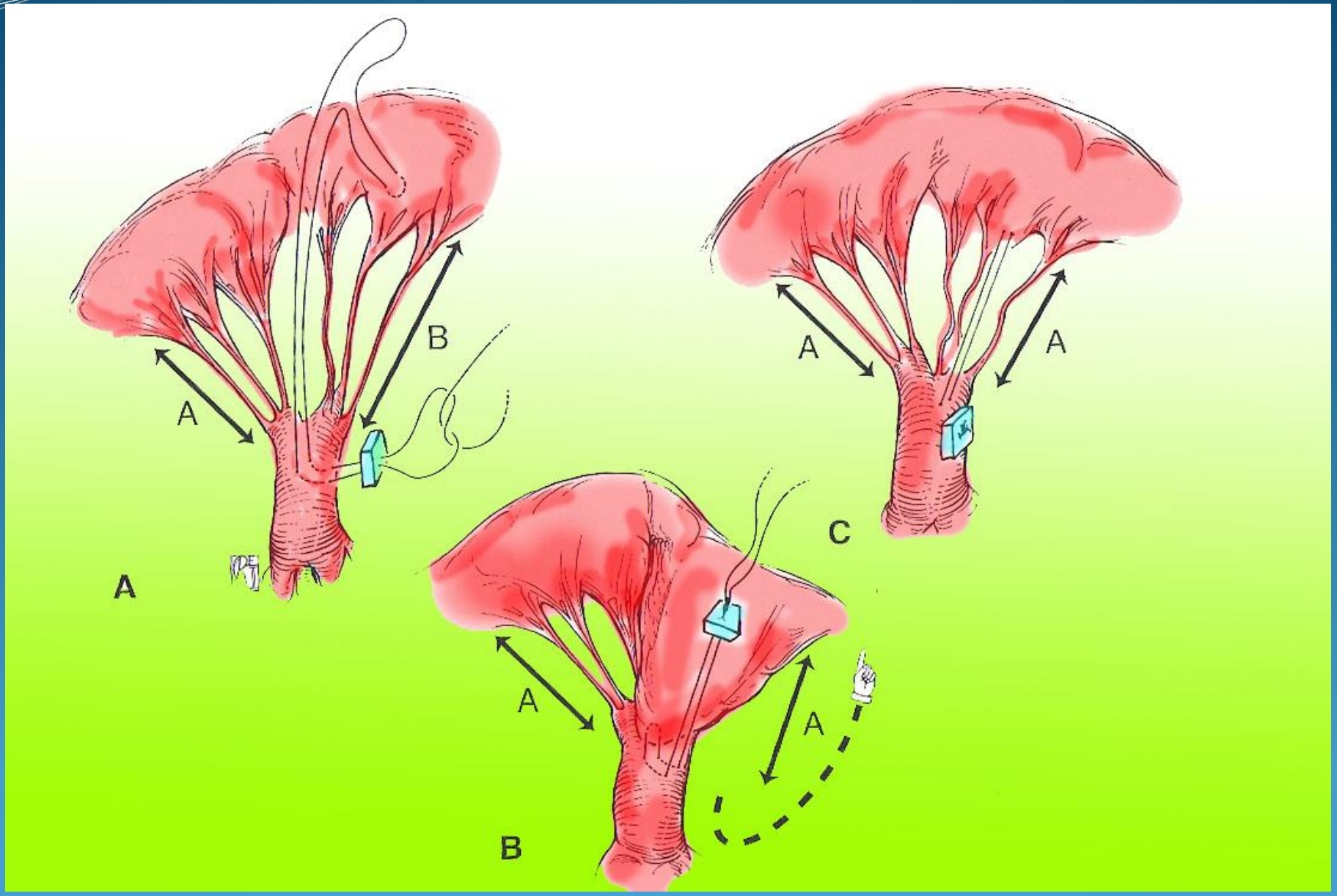
Chordal Transposition



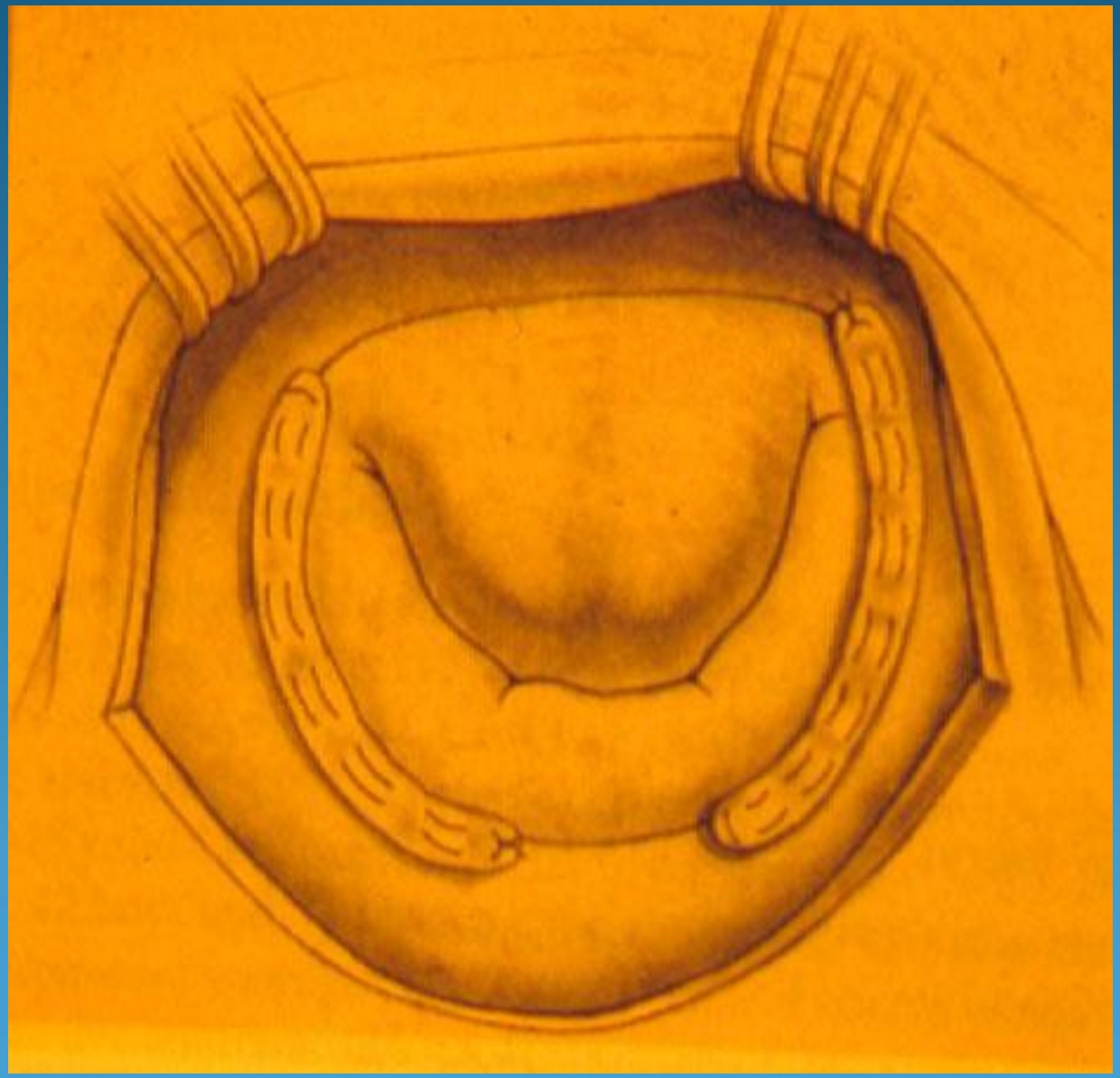
Chordal Shortening



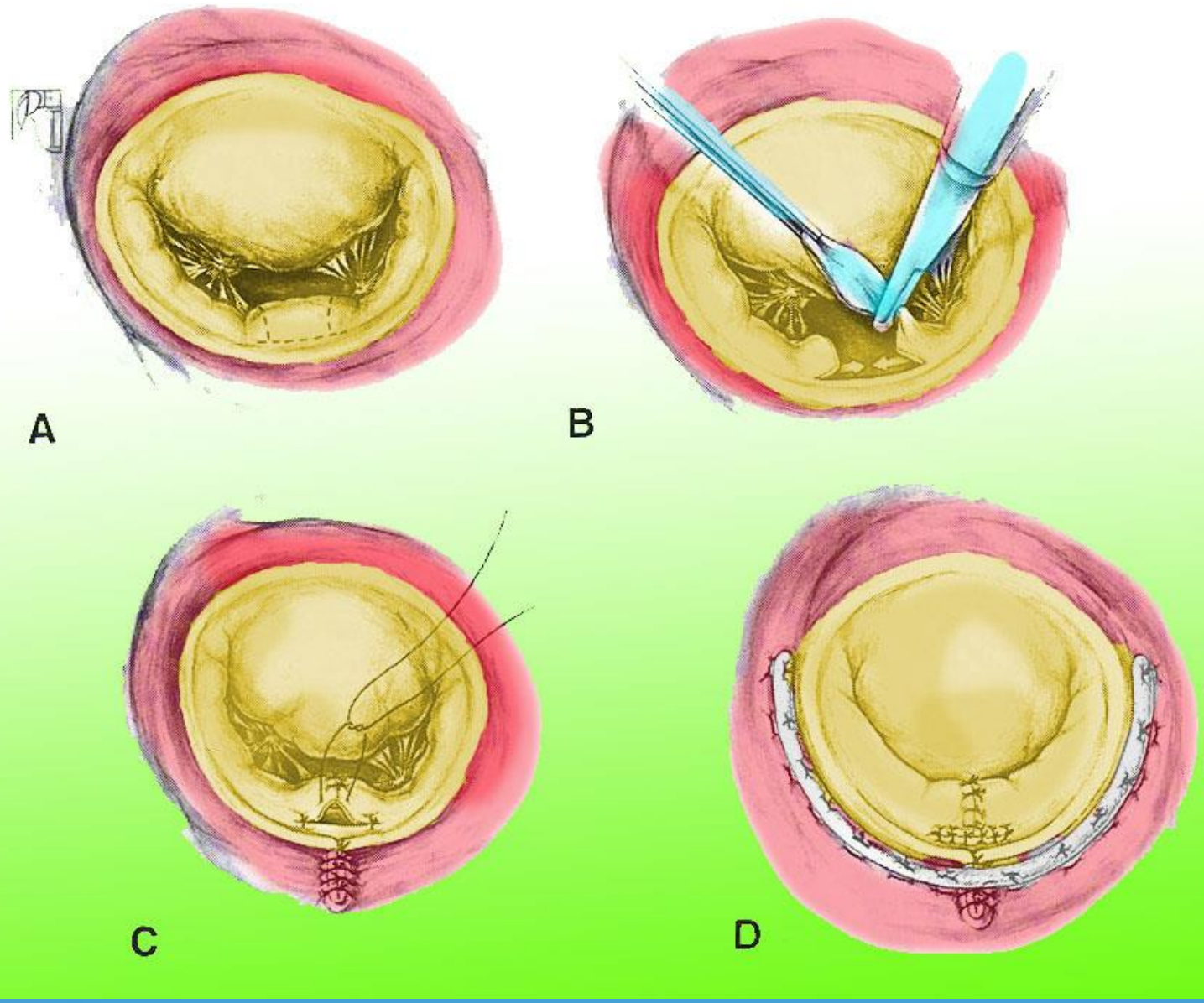
Chordal Shortening with Artificial Chordae



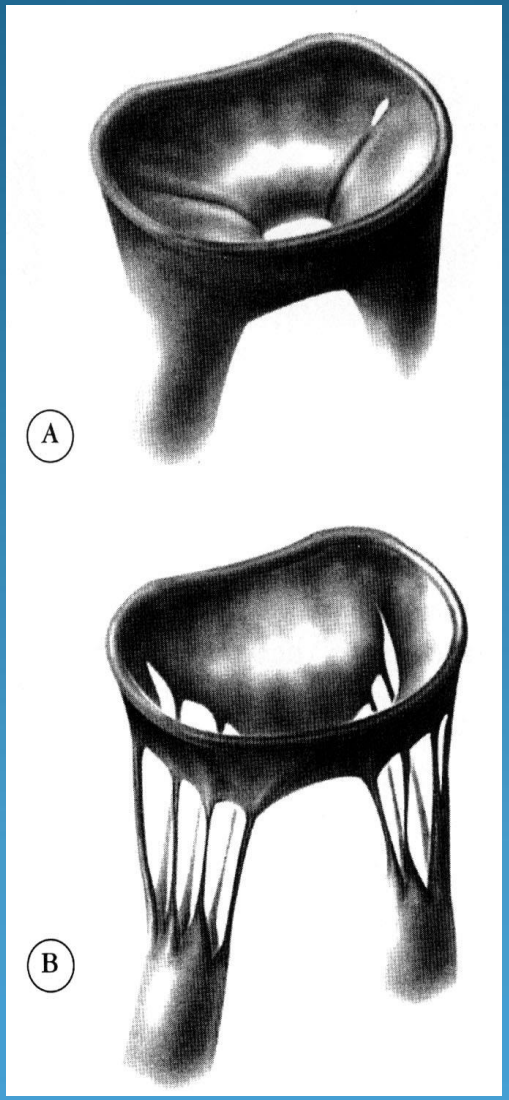
Annuloplasty



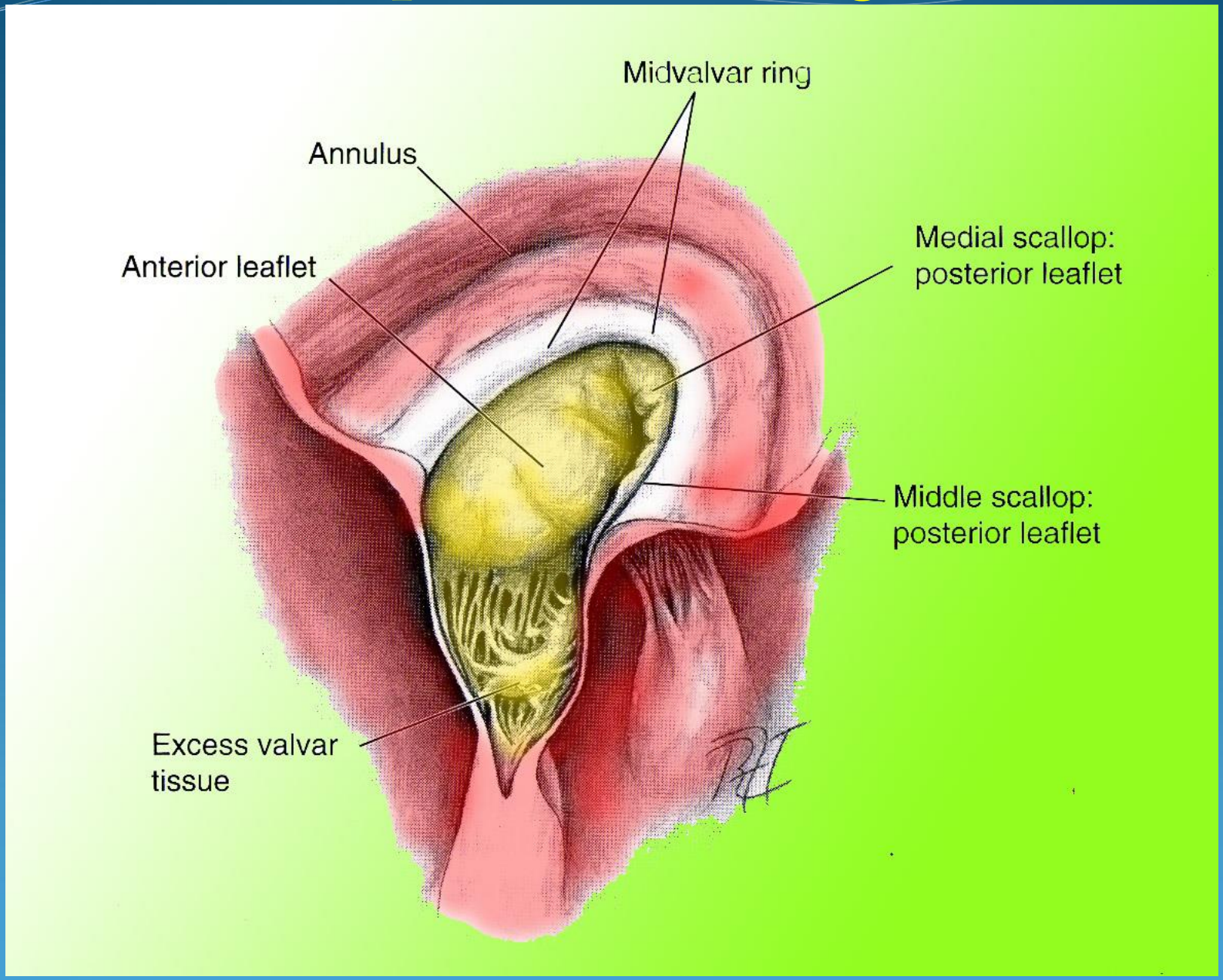
Quadrangular Resection



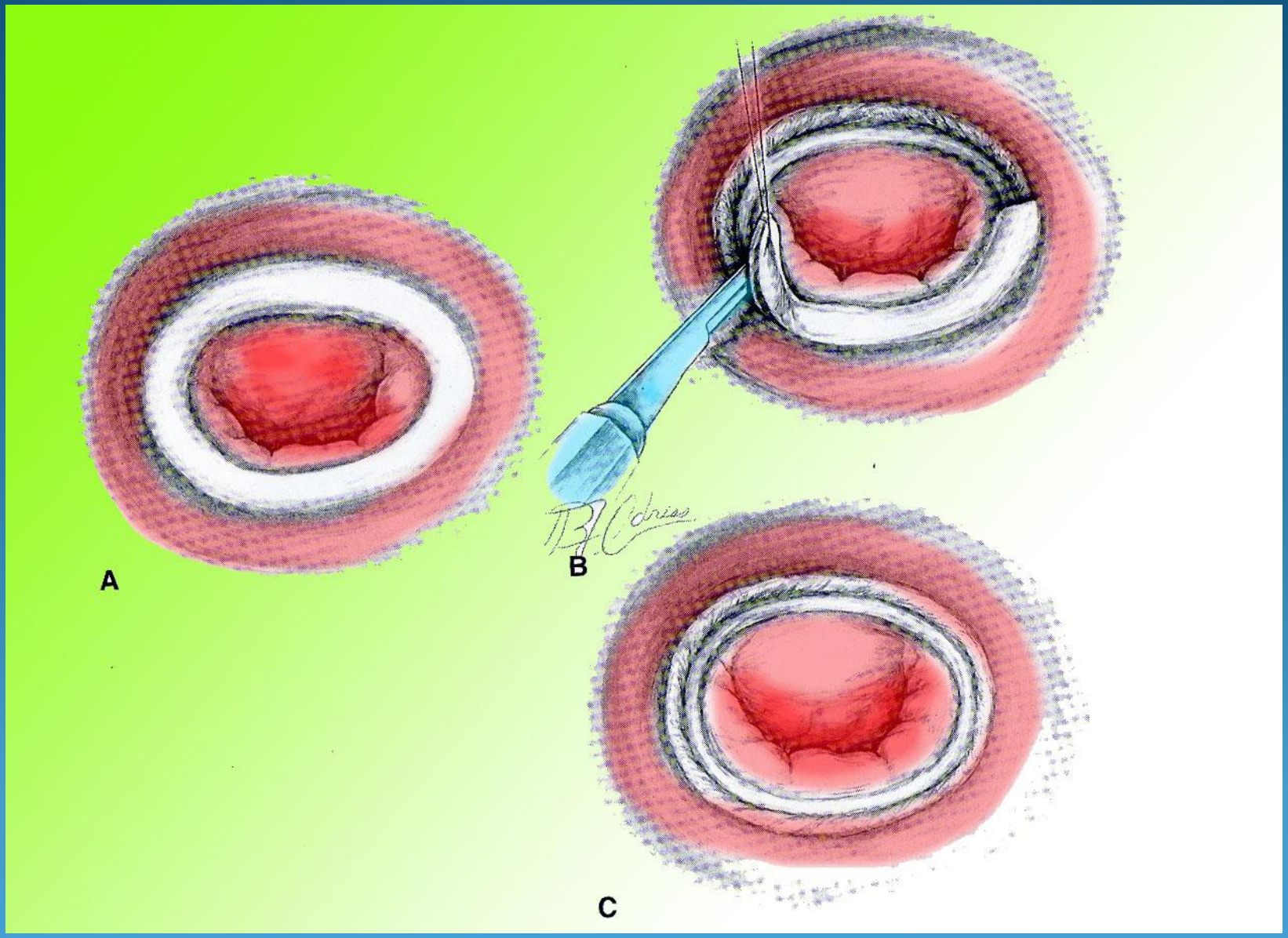
Mitral Stenosis: commisurotomy and fenestration



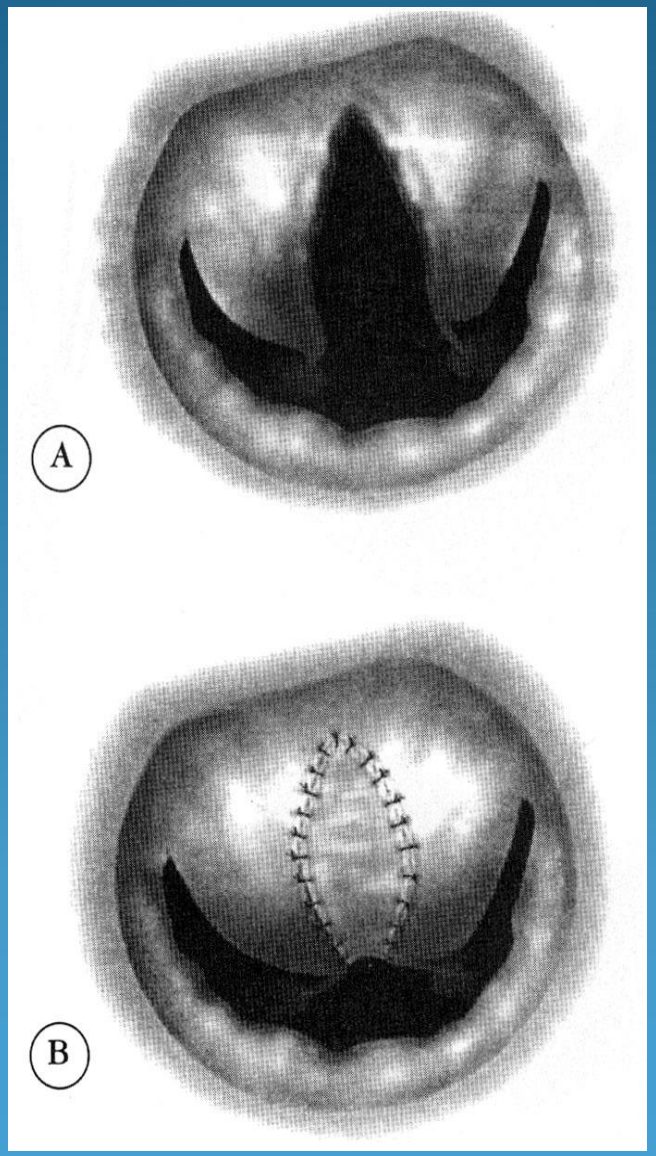
Supra - Mitral ring



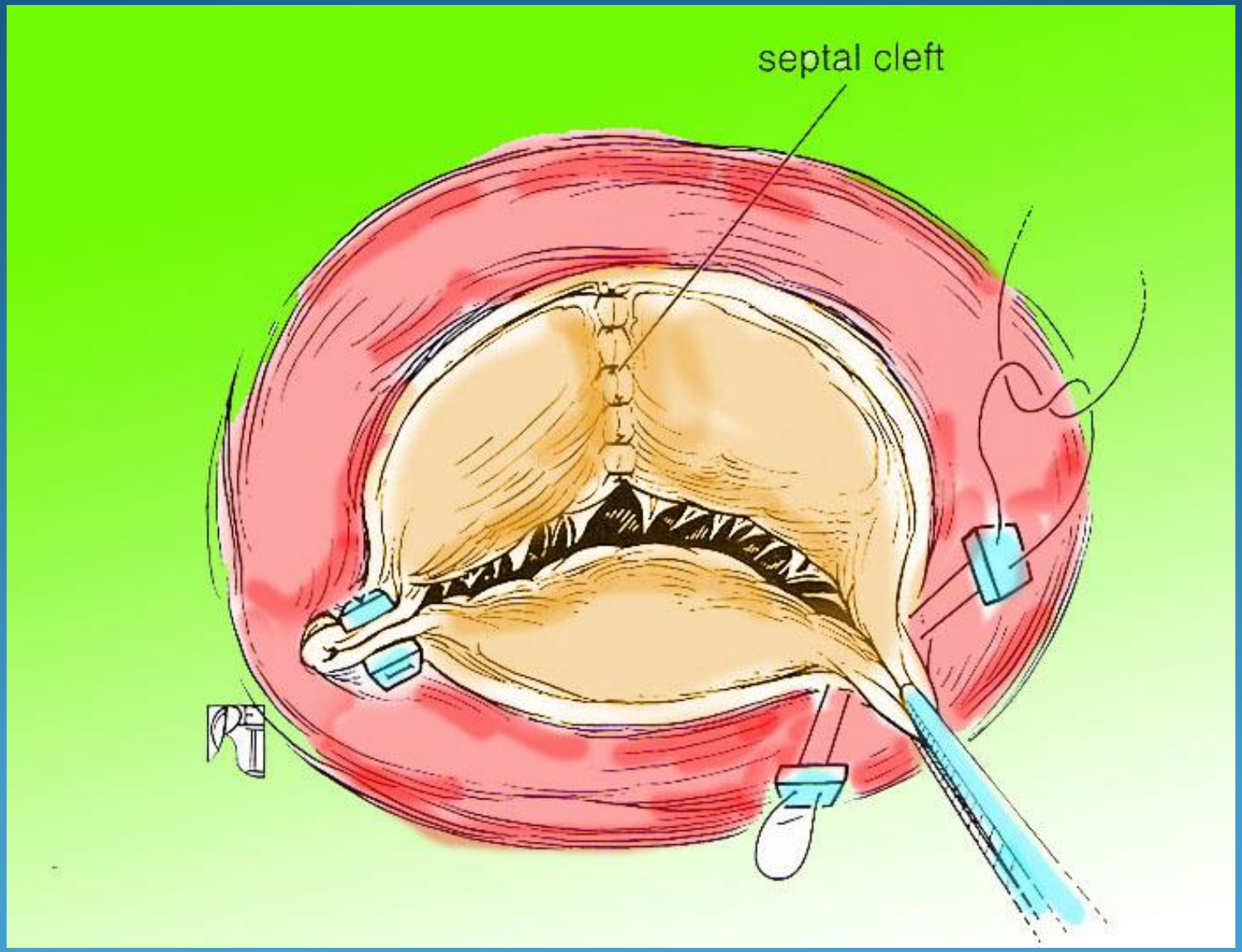
Supravalvar Mitral Ring



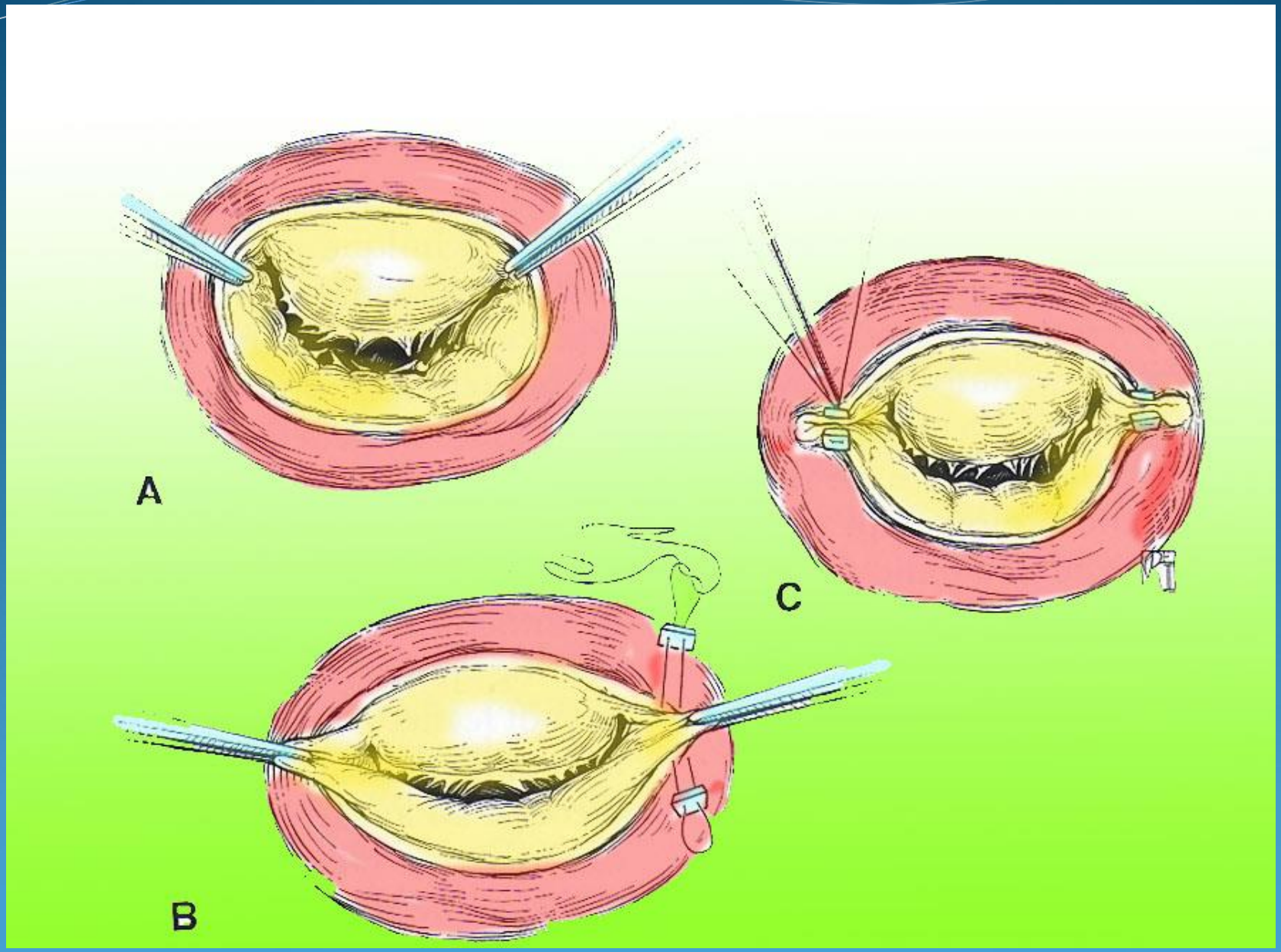
Anterior Leaflet Cleft



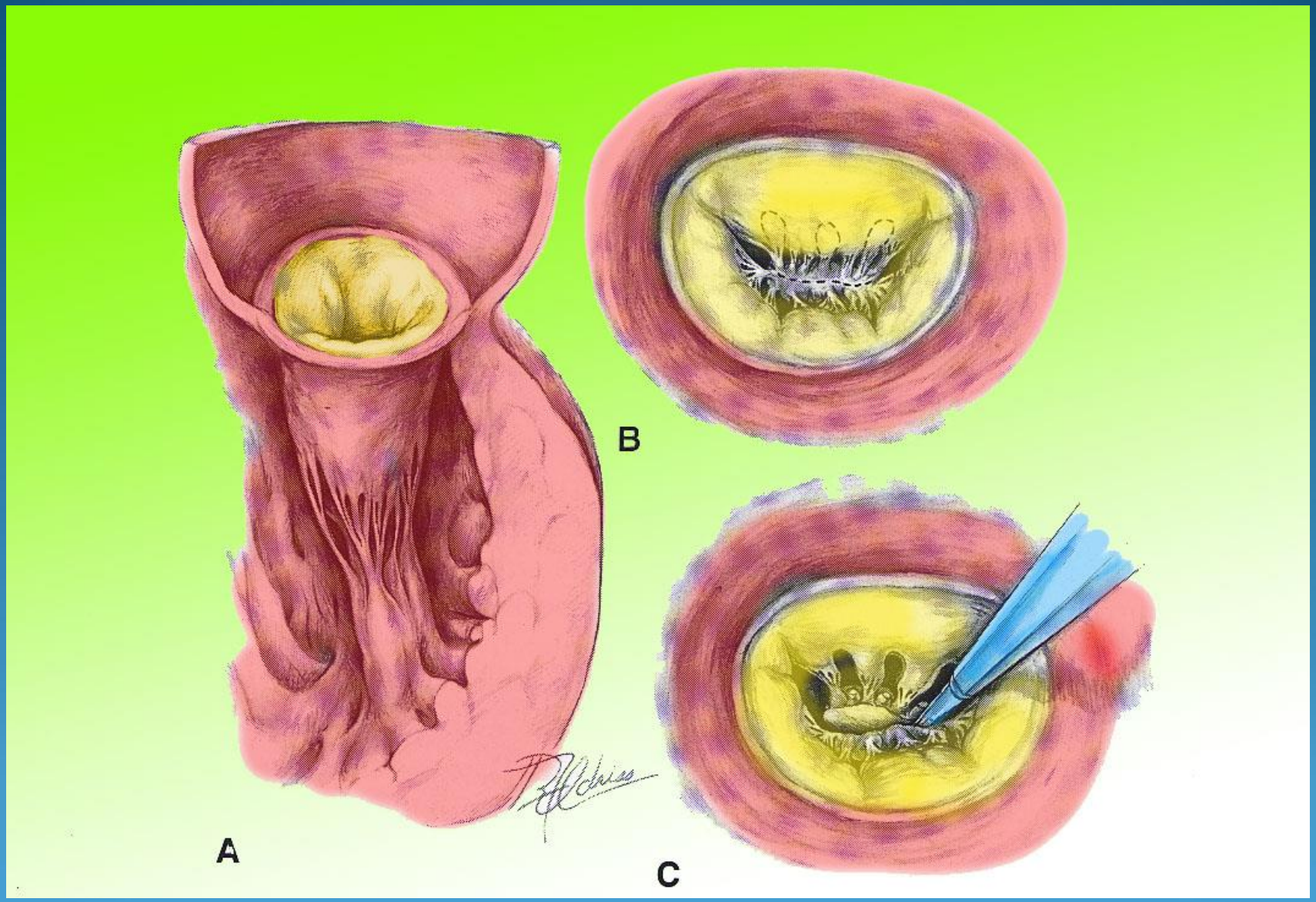
Cleft Closure and Commissuroplasty



Commisuroplasty: Wooler Technique

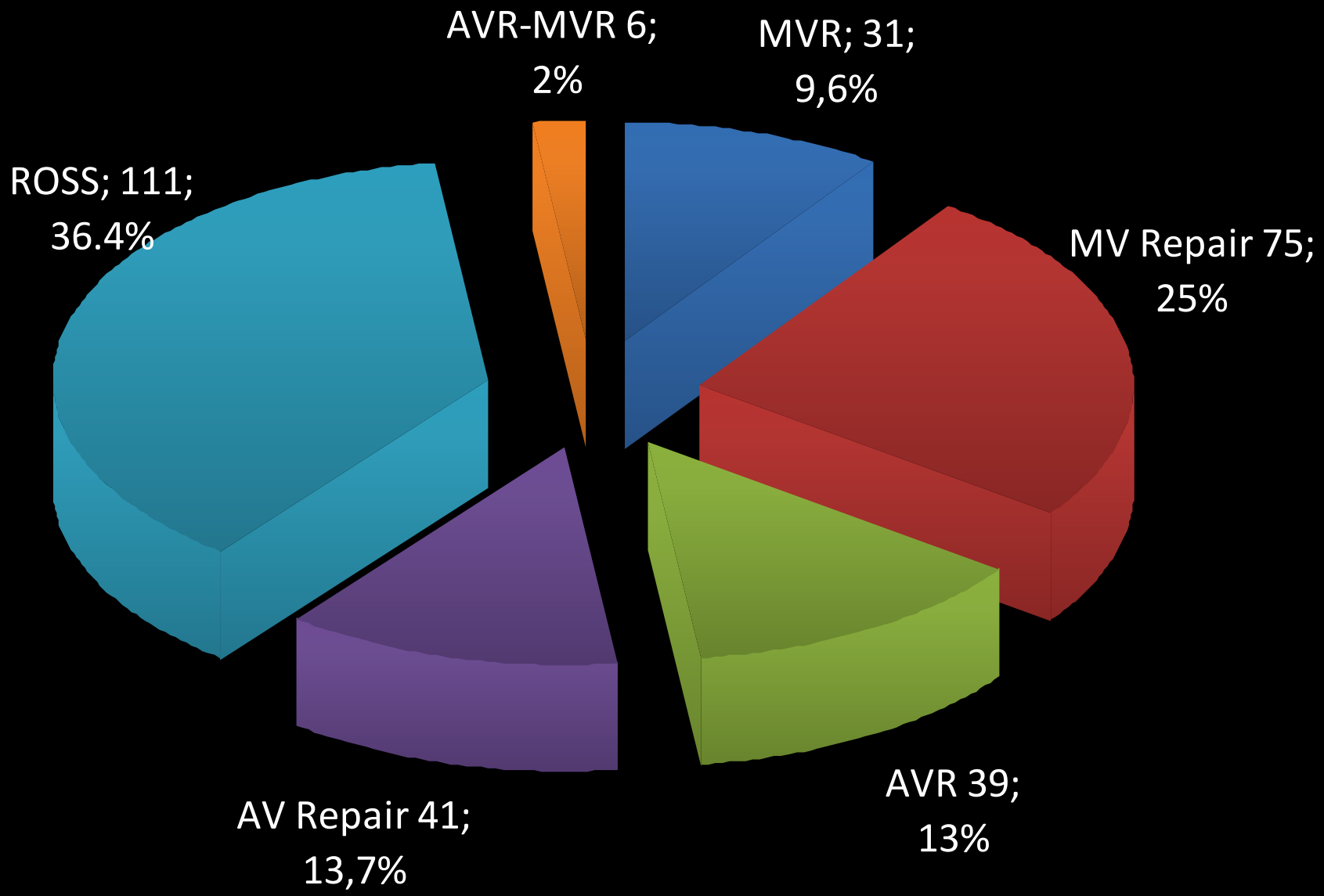


Parachute Mitral Valve: Leaflet fenestration



OUR EXPERIENCE 1988-2008

VALVULAR PEDIATRIC OPERATIONS (303 pts)



Paediatric valve disease

Surgical Experience: San Donato

Mitral Valve Disease

1988-2008 n° cases: 106

	N° cases	Mortality
Mitral Valve Replacement	29	5 (17%)
≤ 1 year	10 pts	4 death (40%)
Mitral Valve Repair	75	3 (4%)
≤ 1 year	12 pts	2 death (16%)
Ross- Kabbani	2	0
	106	7 (6,8%)

EDGE-TO-EDGE AND ARTIFICIAL-CHORDAE TECHNIQUE IN CHILDREN

- May 1997-june 2008
- 15 children
- AGE 1-17 years (6±5)

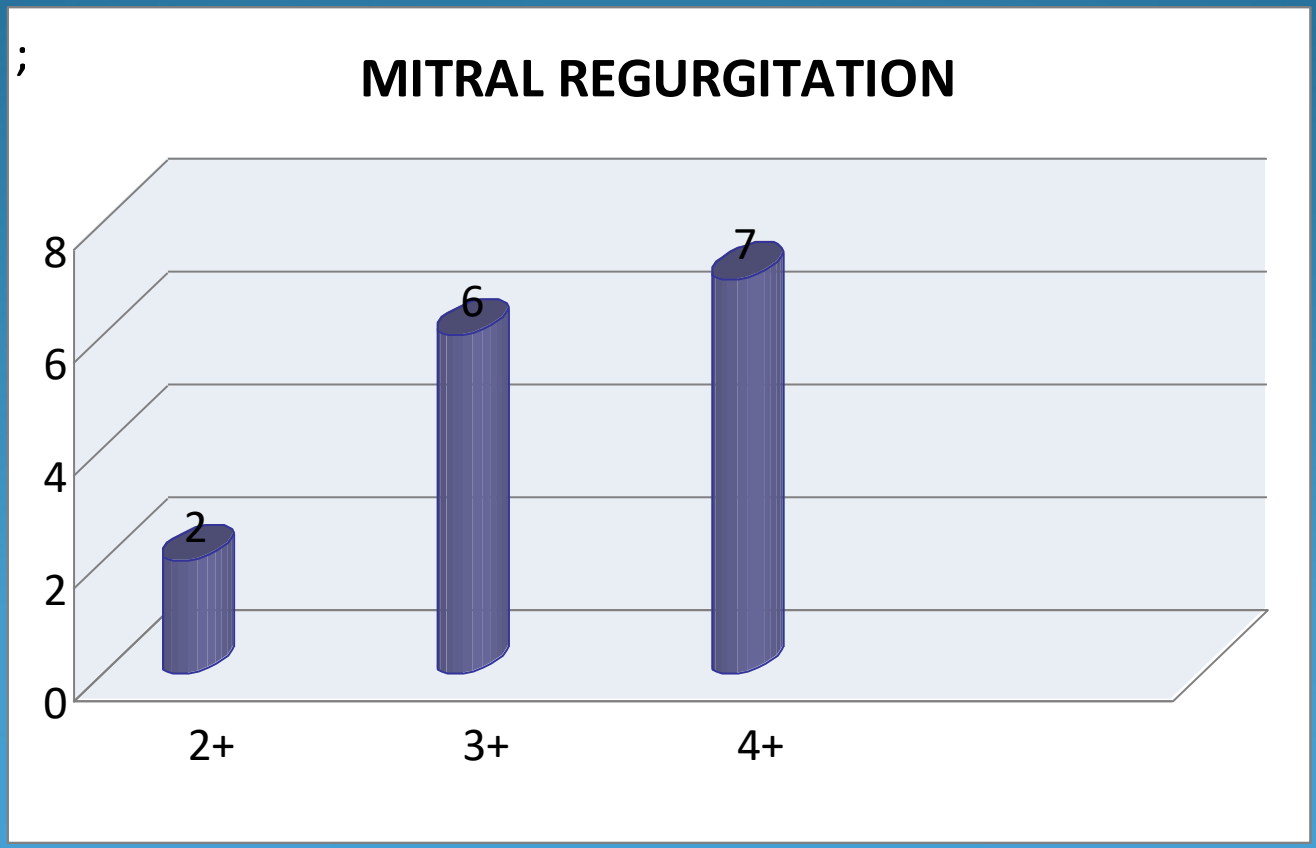
EDGE-TO-EDGE AND NEO-CHORDAE TECHNIQUE IN CHILDREN

Diagnosis

- Degenerative disease 12 pts
- Rheumatic disease 3 pts

EDGE-TO-EDGE AND NEO-CHORDAE TECHNIQUE IN CHILDREN

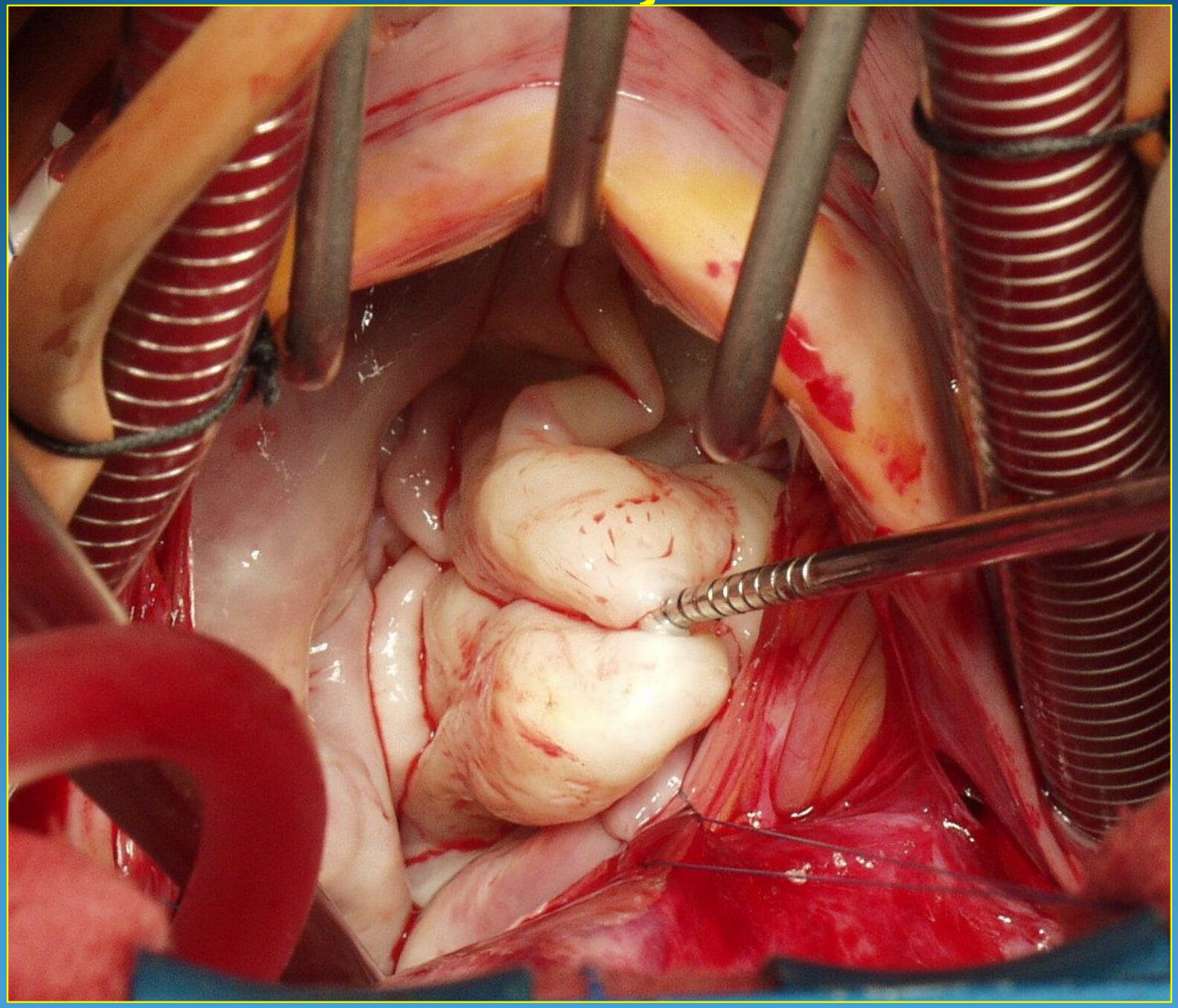
Pre-operative Data



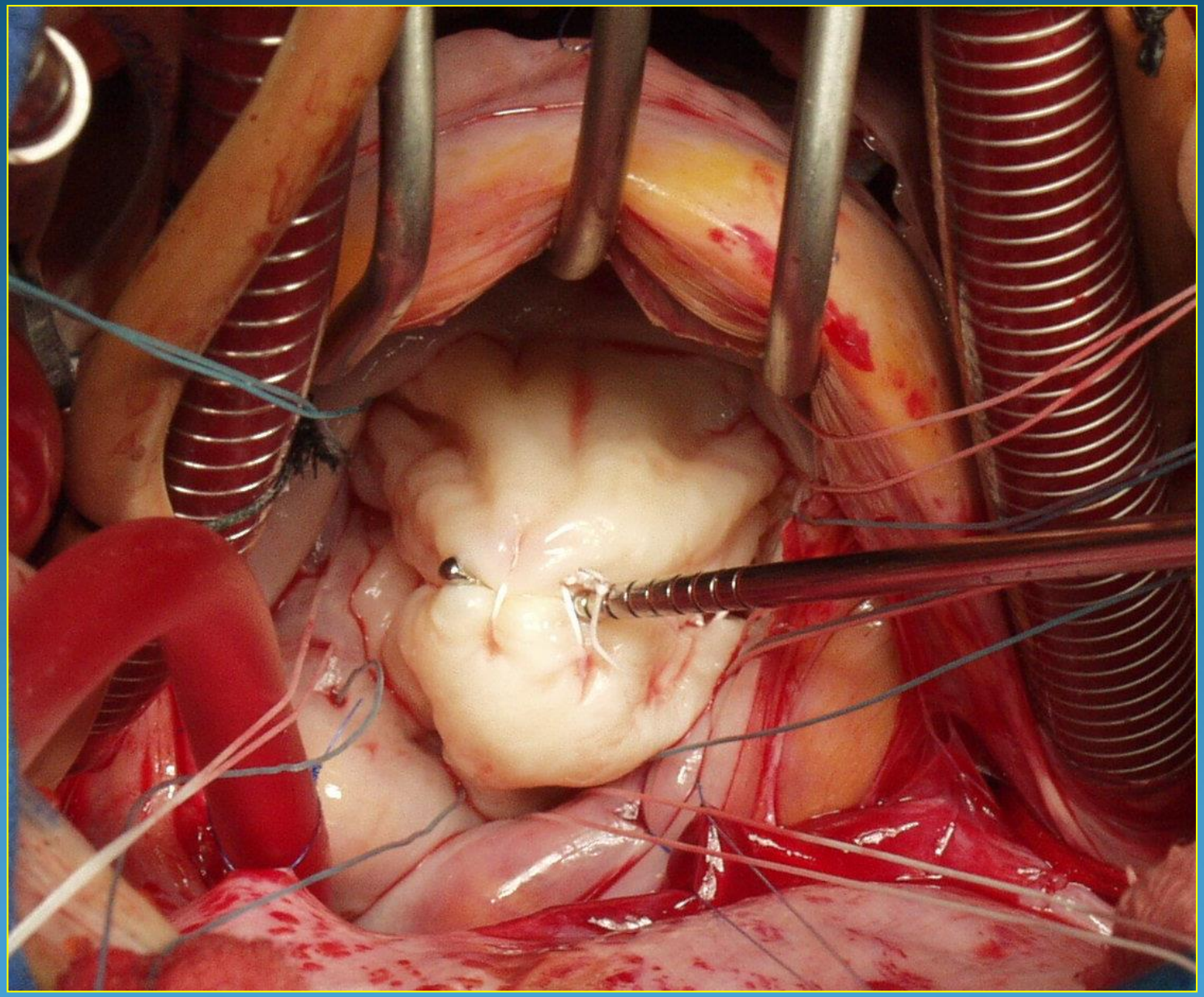
EDGE-TO-EDGE AND NEO-CHORDAE TECHNIQUE IN CHILDREN

Technique	
Edge-to-edge	8
Artificial chordae	7

BARLOW'S Syndrome



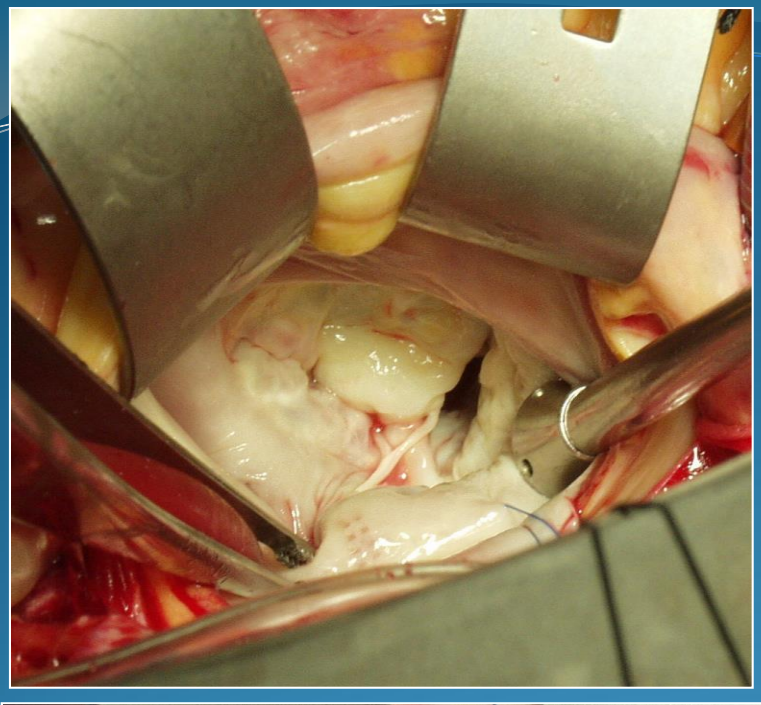
EDGE-TO-EDGE

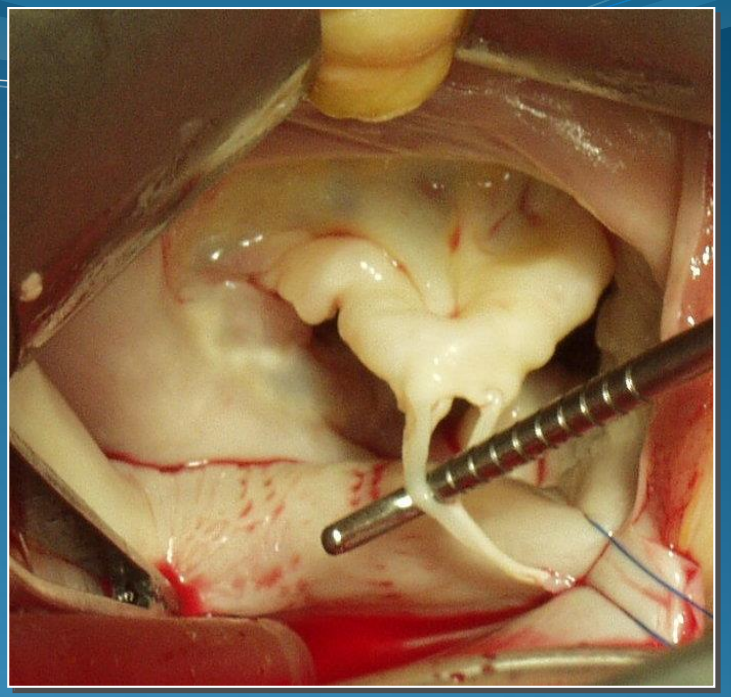


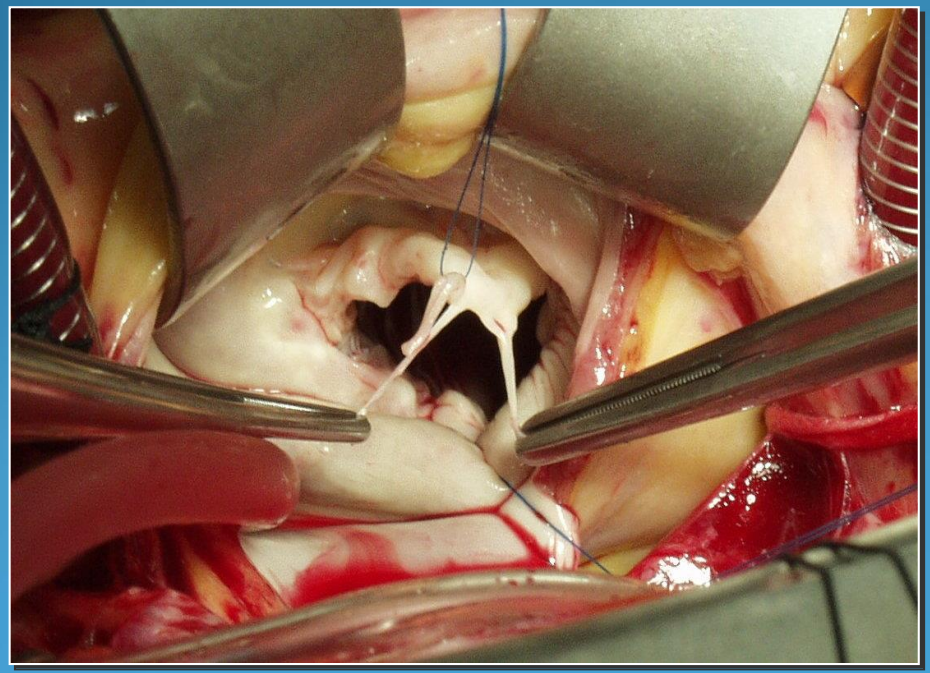
EDGE-TO-EDGE

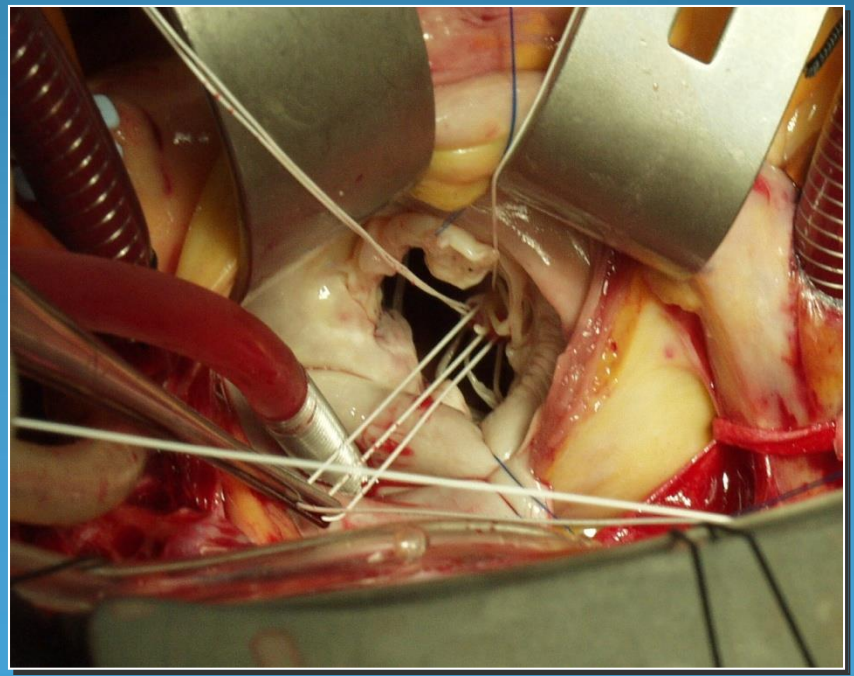

/3.7-T
PAZ T: 37.0°C
TEE T: 38.0°C
3.7MHZ
ELAB.2/0/E/E/A
HEWLETT-PACKARD
CARDIACO ADULT
ID:BONESHI
PADLO AN TOR
COL DIS.
50MM/S
TRAS:A
12CM
9HZ

Chordal Rupture

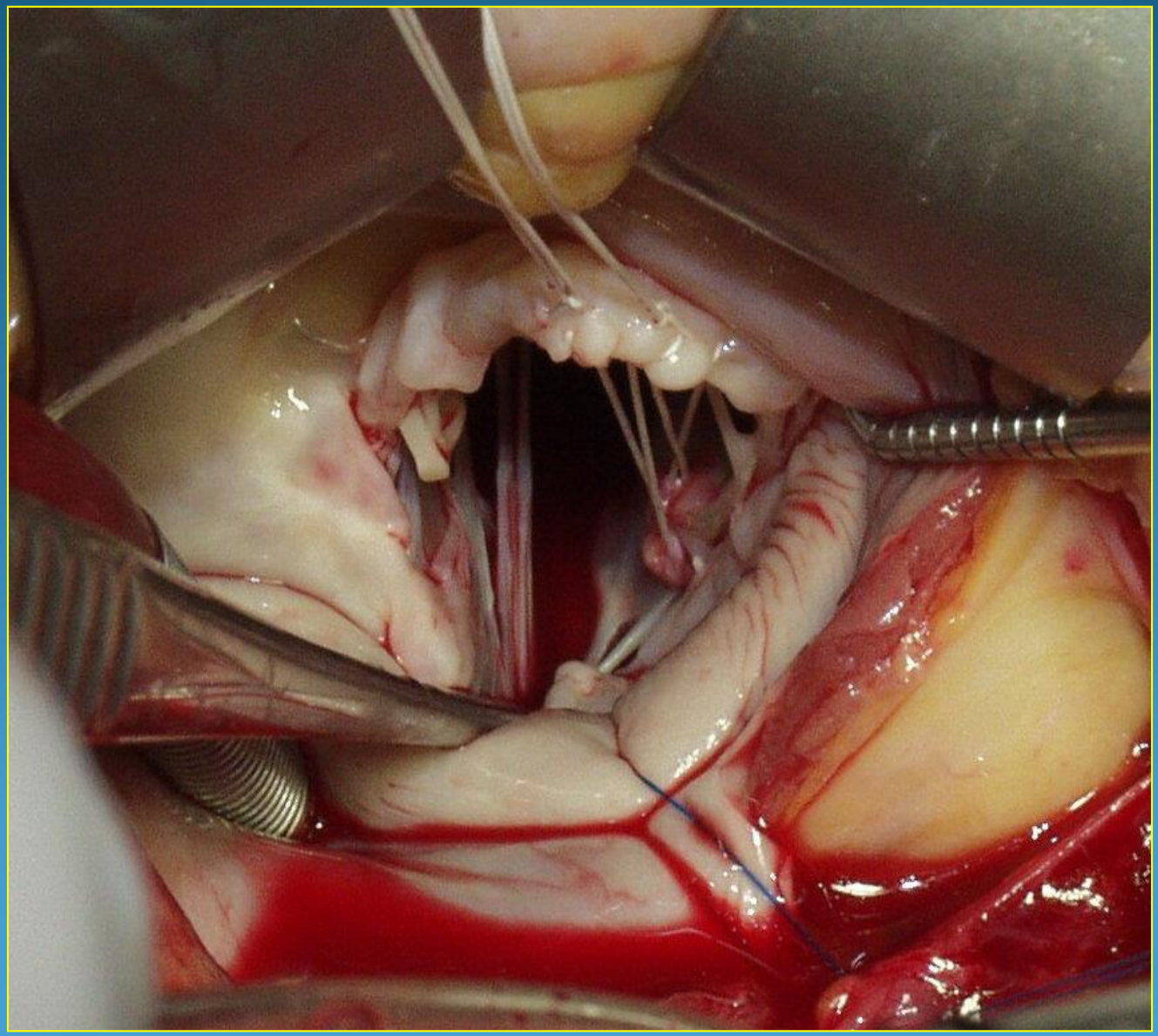






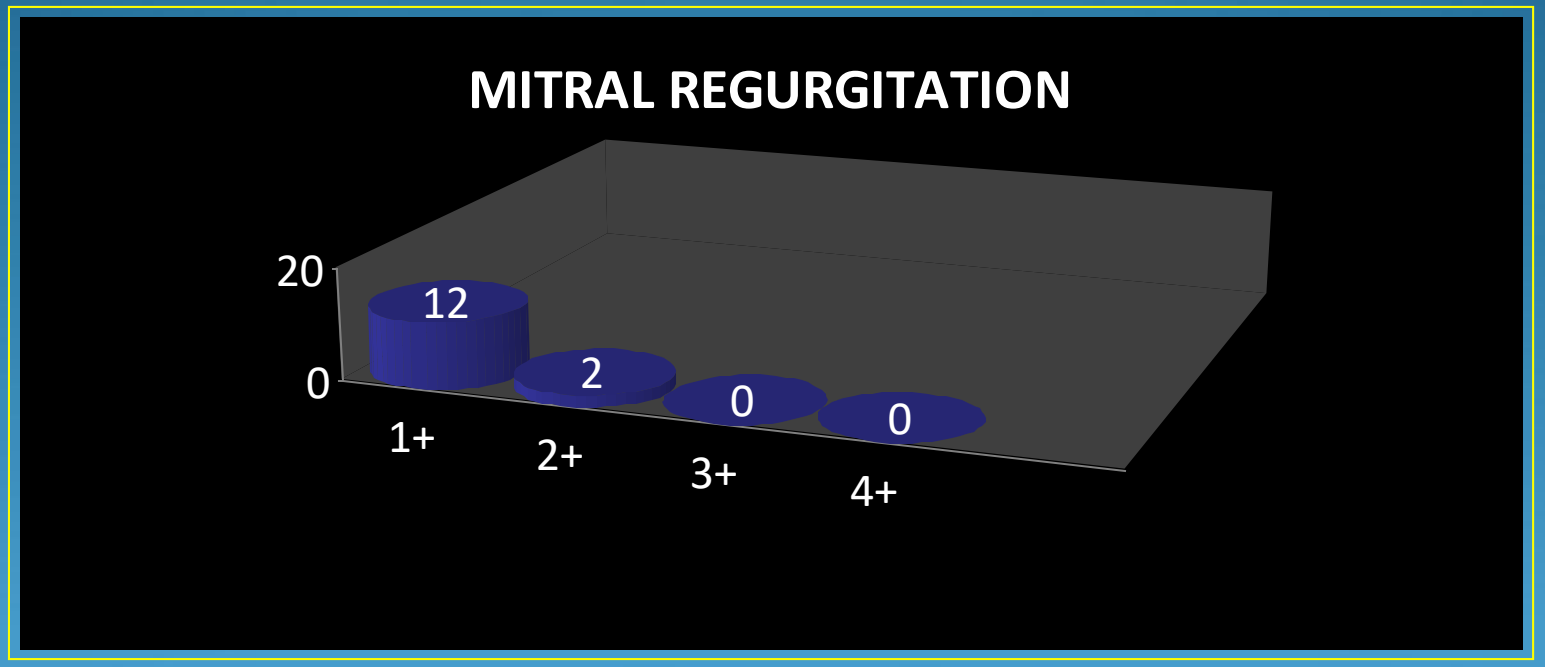


ARTIFICIAL-CHORDAE



EDGE-TO-EDGE AND ARTIFICIAL-CHORDAE TECHNIQUES IN CHILDREN

Hospital discharge



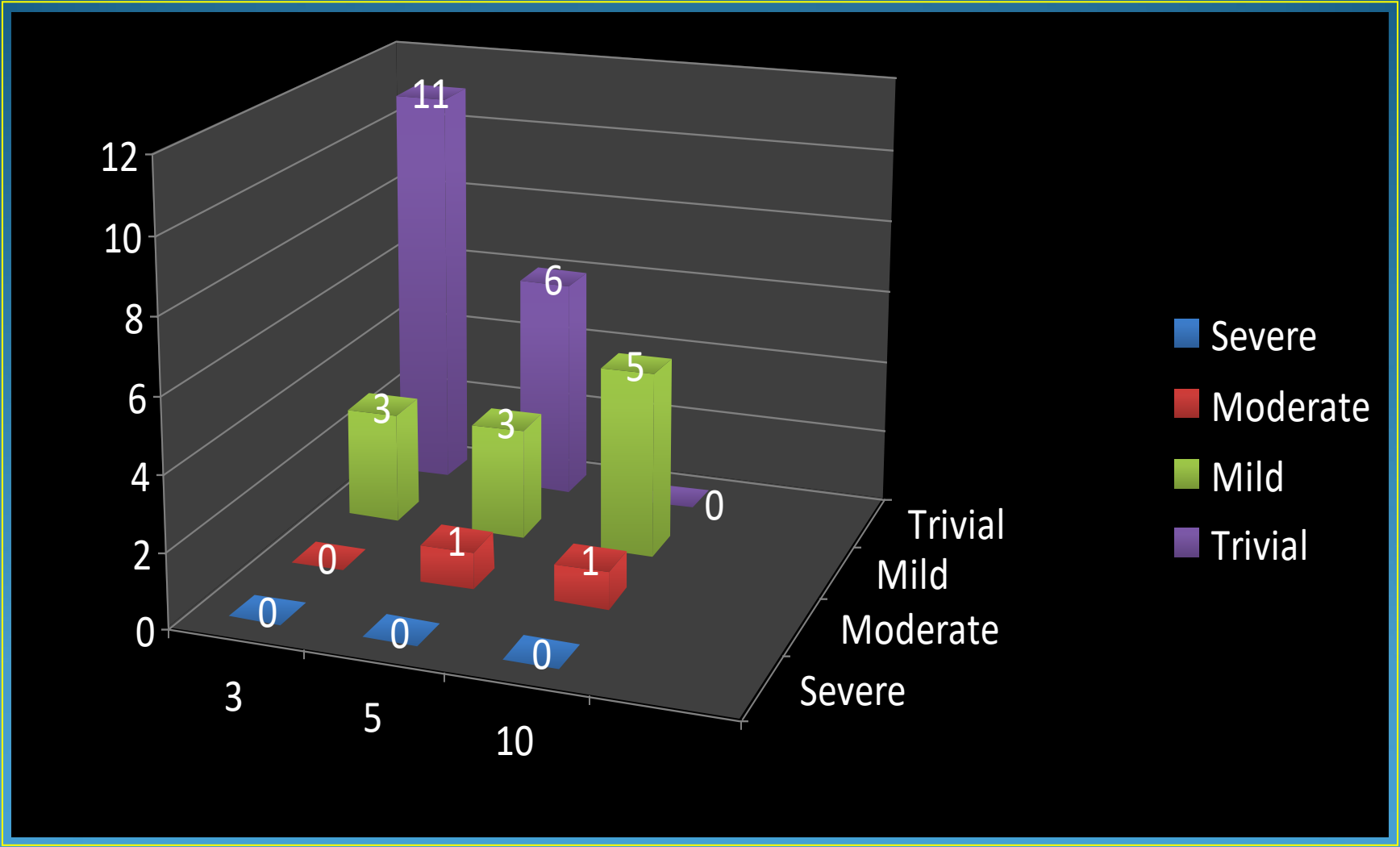
Hospital death : 1
Unrelated to type of technique

15 PTE PEDIATRIC

53%

8 PTE ADULT

EDGE-TO-EDGE AND NEO-CHORDAE TECHNIQUES IN CHILDREN



Ross-Kabbani in children

Our experience

- N° pts 2
- Age 2-6 mos
- BW 3.3-4.3 Kgs

- Diagnosis : dysplastic MV with abnormal chordae apparatus and severe regurgitation and moderate stenosis

Ross-Kabbani in children

Literature search (MEDLINE)

Author	N° pts	age	follow-up
• Ishihara (89)	1 pt	11 mos/exitus	
• Al Halees (99)	1 pt	12 yrs	6 yrs
• Serraf (01)	1 pt	4 mos	2 mos
• Mitchell (01)	1 pt	36 mos	8 mos
• Yamagishi (03)	1 pt	11 mos	5 mos
• Kabbani (04)	no spec	12 yrs	no spec
• Frigiola (05)	2 pts	2.5-6 mos	15 mos /5 yrs
• Brown (06)	2 pts	12-14 yrs	25 mos

Ross-Kabbani in children

Results

- Survival 2/2
- Reoperation 2/2

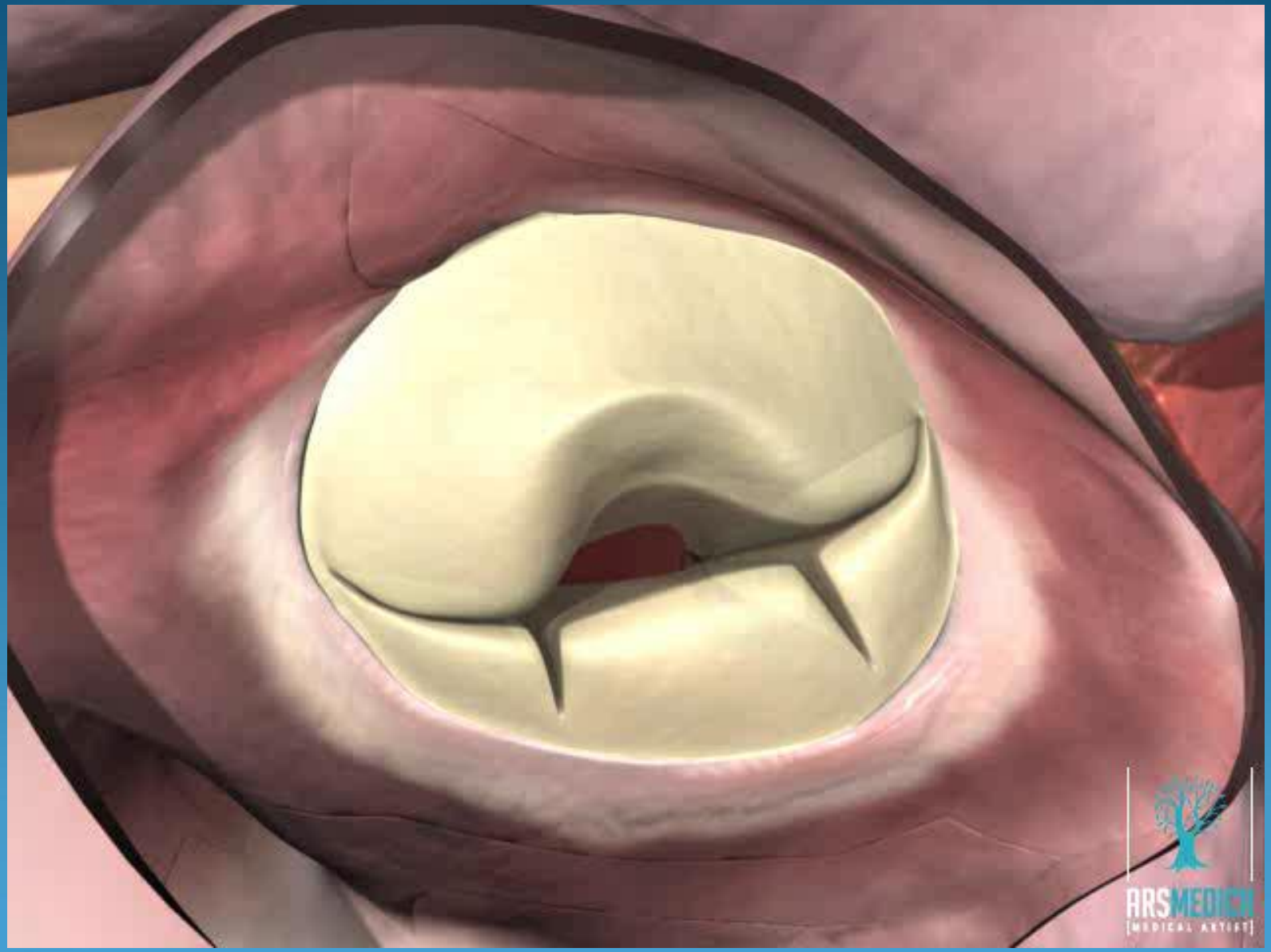
1st: 27 mos later for RV-PA conduit replacement
58 mos follow-up autograft with moderate stenosis

2nd: 15 mos later for MVR with Carbomedics 16 mm
death on 12# POD for LCOS

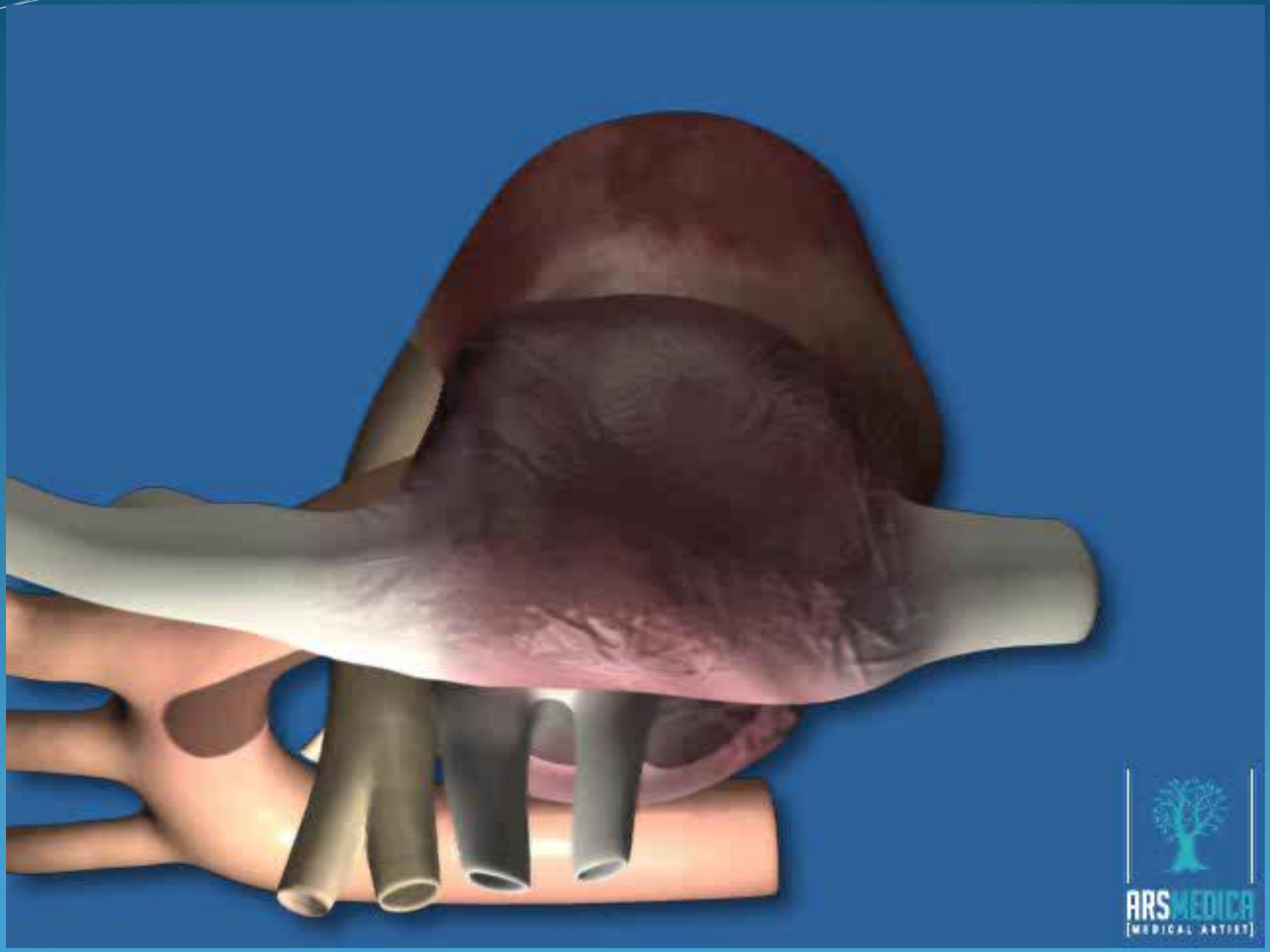
Conclusions

- In children (<10 years old) reparative surgery should be considered the best surgical option
- Is better to have a repaired valve with some degree of regurgitation then a mechanical valve
- The Ross Kabbani operation should have to be take in consideration for small children with not reperable valve

Mitral Stenosys

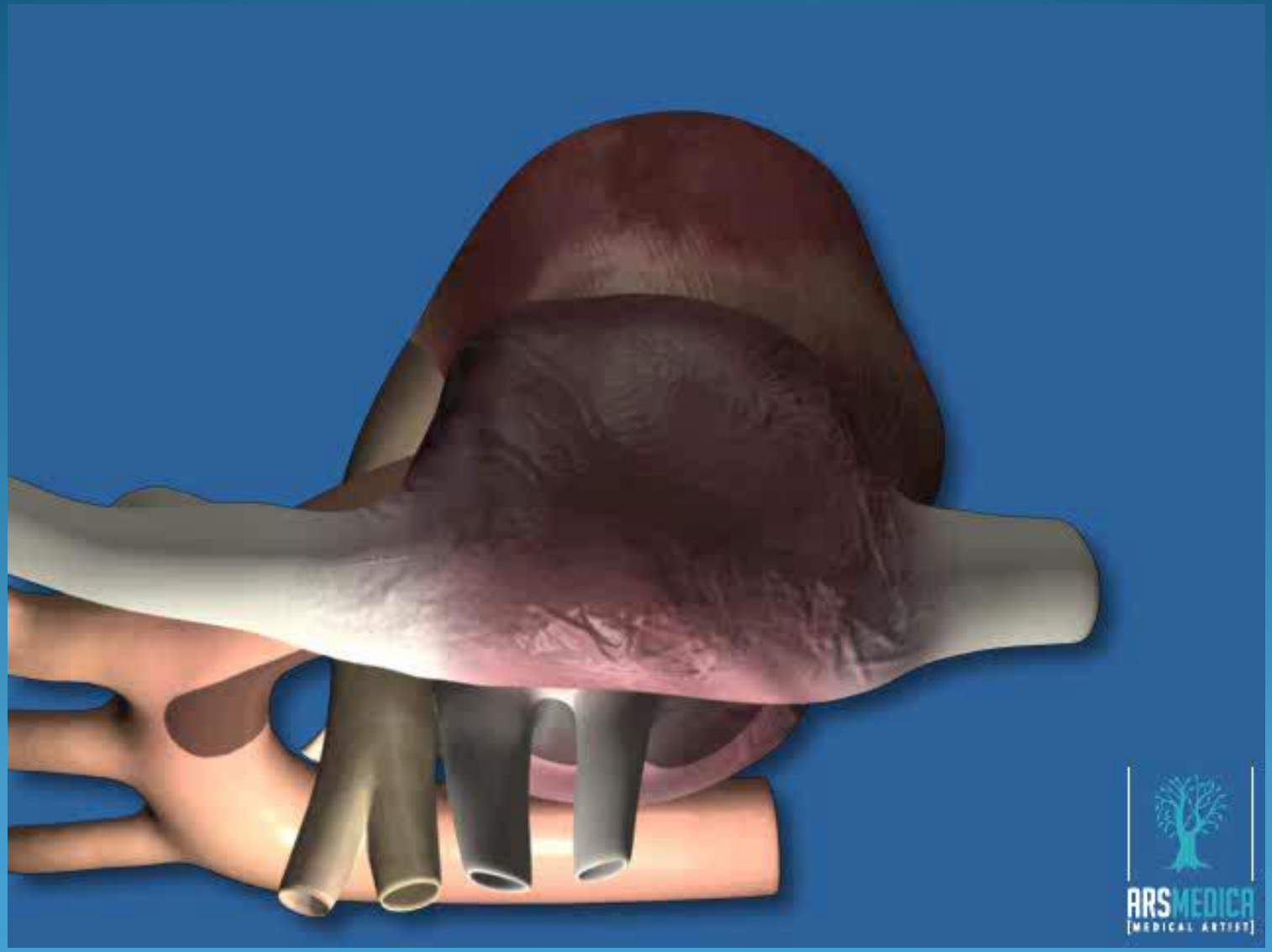


Quadrangular Resection



Shorthening of chordae

Trasposition of chordae



Trasposition of chordae