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AD-COR Program inovativ de formare in domeniul cardiologiei pediatrice
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MODUL TEORETIC

The ascending aorta dilatation in cono-truncal malformations

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The ascending aorta dilatation in cono-truncal malformations

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IRCCS Policlinico San Donato

Cono-truncal anomalies definition

Cono-truncal heart defects are a group of genetically and embriologically related heart malformation due to a **defective development of the cardiac outflow tract**

Cono-truncal heart malformation
Classification

Tetralogy of Fallot

Transposition of the great a.

Double-outlet ventricle

Truncus arterious

Tipe B interrupted aortic arch

Conal septal defect

Aorto pulmonary window

Anatomical corretted TGA

Cono-truncal heart defects prevalence per 10.000 live births in USA

- Trasposition of the gret arteries 4.73
- Tetralogy of fallot; Pulm. atr. + VSD 3.92
- Truncus arteriosus 0.82
- Double outlet RV 0.3-1.1

ARD cono-truncal anomalies

The majority of the medical litterature focus on the re-operative management of the right-sited outflow tract. Is common, however, for the ascending aorta and aortic root be dilatated at birth and at initial repair

ARD in cono-truncal malformation

While the aortic root is common, surgical interventions was performed in few patients.

Post switch aortic root dilatation (ARD) incidence

Probability of freedom from ARD 98%, 97%, 96% and 93%and from surgery 100%, 100%, 99% and 95% at 1-2-5 and 10 years and 1.2% for severe ARD

ARD and AR continue to develop after ASO but ARD does not tend to be progressive during late follow up

Schwarz et al Circulation 2004

Pathophysiology

a) Increased aortic blood flow

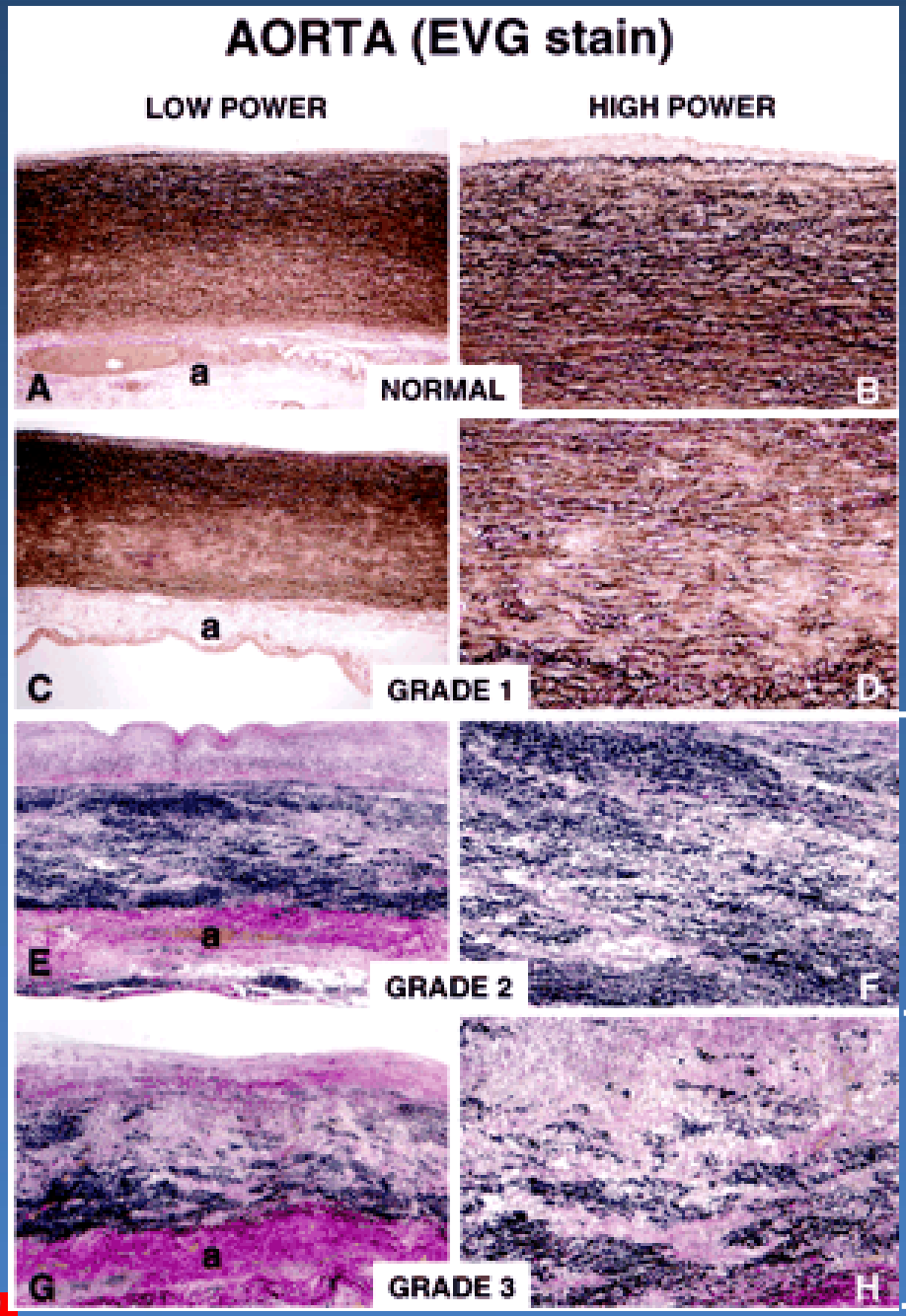
b) Intrinsic vascular structural abnormalities

c) Combination of a and b

Pathophisiology

Given the potential effects of volume overlood on the aortic root it would seen logical that once the cono-truncal defect is repaires, thus eliminalizating the right-to-left ventricular level shunt, aortic sided would stabilize

Acending aortic structure in CHD



Grade 2
wide spread elastic fiber fragmentation

Grade 3
complete loss of elastic fibers

Niwa K. et al 2001, Circulation

Clinical features

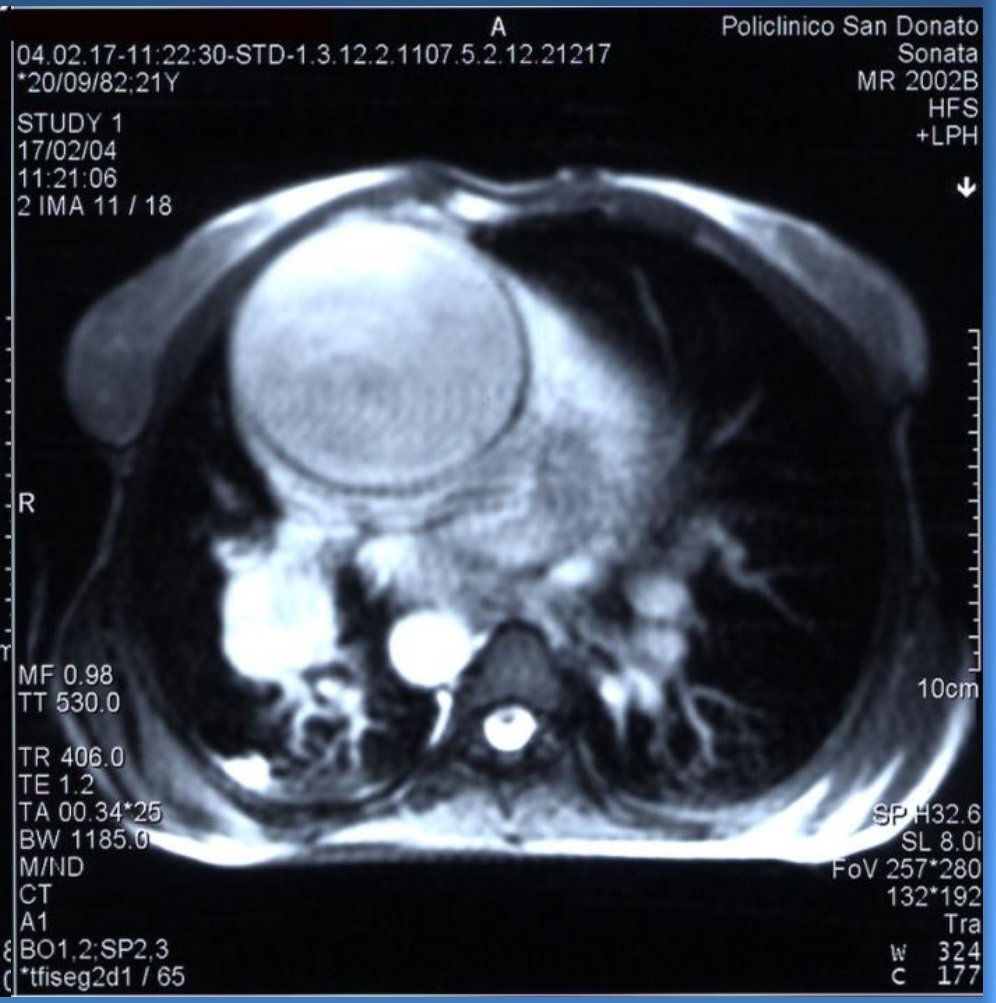
- The majority of pts come to medical attention because of right-sided abnormalities
- Symptoms only in the presence of aortic regurgitation
- Chest or back pain indicate acute dissection are very rare

Diagnostic studies

- Chest x-ray
- Echocardiography
- When the ARD is present : CT/CTA, MRI
- Coronary angiography when we need to know preoperatively the coronary anatomy

Pulmonary atresia + VSD and aortic dilatation

Pulmonary atresia + VSD and aortic dilatation



ARD
Surgical Indication (I)

- Moderately dilated aorta: **observation alone**
- Ascending aorta diameter ≥ 55 mm : **surgery**
- Ascending aorta diameter < 55 mm when there is moderate or greater aortic regurgitation : **surgery**

ARD
Surgical Indication (II)

- Ascending aorta diameter < 50 mm: 10% increase in size during 6 to 12 months period: **surgery**

- Ascending aorta diameter < 50 mm : rapid growth (> 1mm in a year), a family history of aortic dissection and the presence of moderate or worse aortic regurgitation : **surgery**

Intraoperative complications

- Cardiac structures injury during re-sternotomy
- Bleeding during structures dissection
- Presence of arterial collaterals

Re-sternotomy: risk of injury

First reoperation	7,8%
Second reoperation	14,7%
Third or more reoperation	17,1%

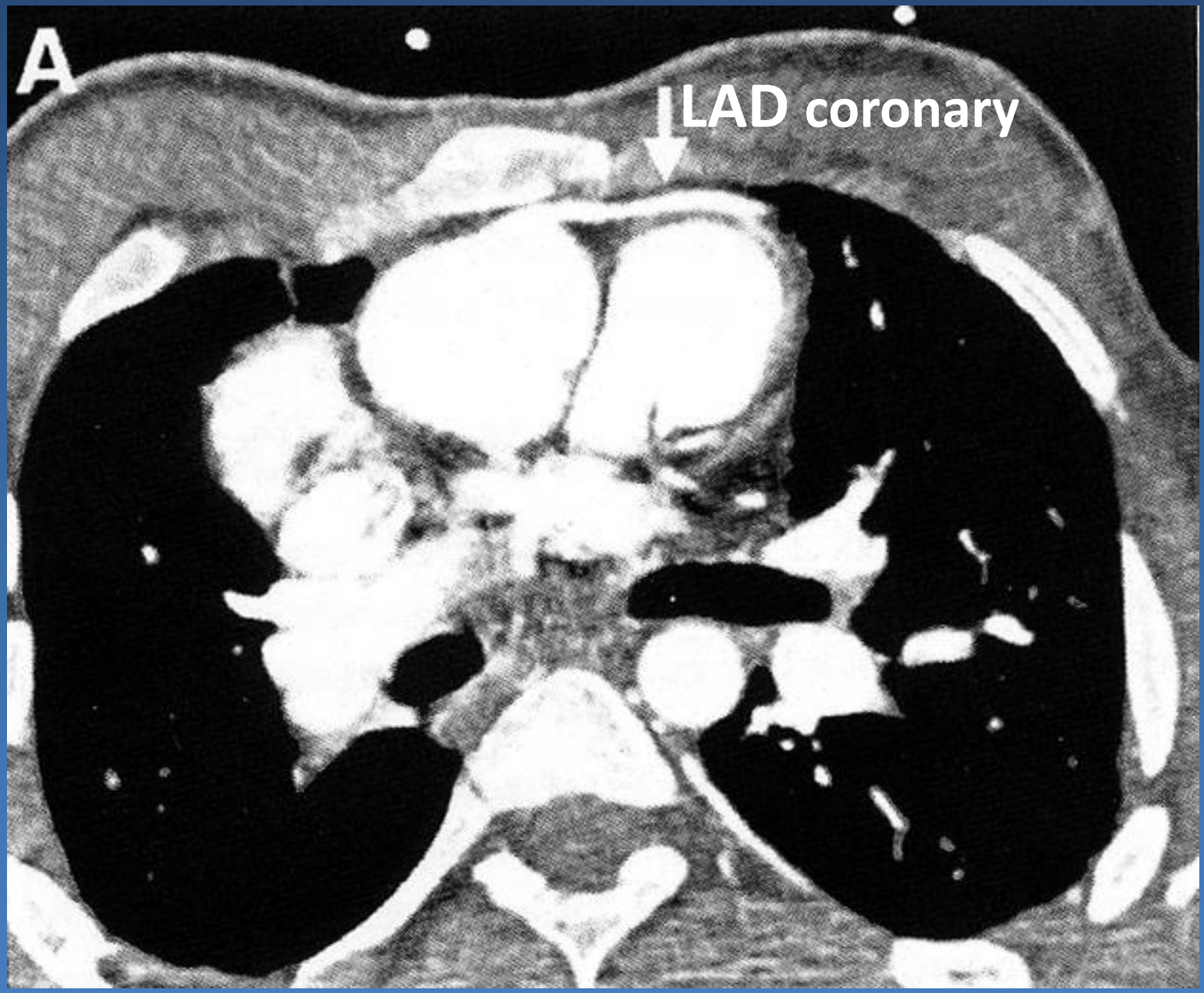
Chan B. Park – JTCS 2010

Strategies for re-sternotomy

- Angiography for periferal vessels
- Clinical evaluation of periferal vessels
- Femoral vessels exposure
- Periferal cannulation + deep hypother.
- Left anterior thoracotomy for LV venting
- Mantain a positive CVP (>5 mm Hg) if intracardiac shunt is present

Preoperative computed tomography for repeat median sternotomy : is it useful?

Incidence of injury	8,1% without CT
Incidence of injury	15,2% with CT



J. Deaerani et al, S.T.C.S.P.C.S.A. 2009

Aortic root surgical procedures

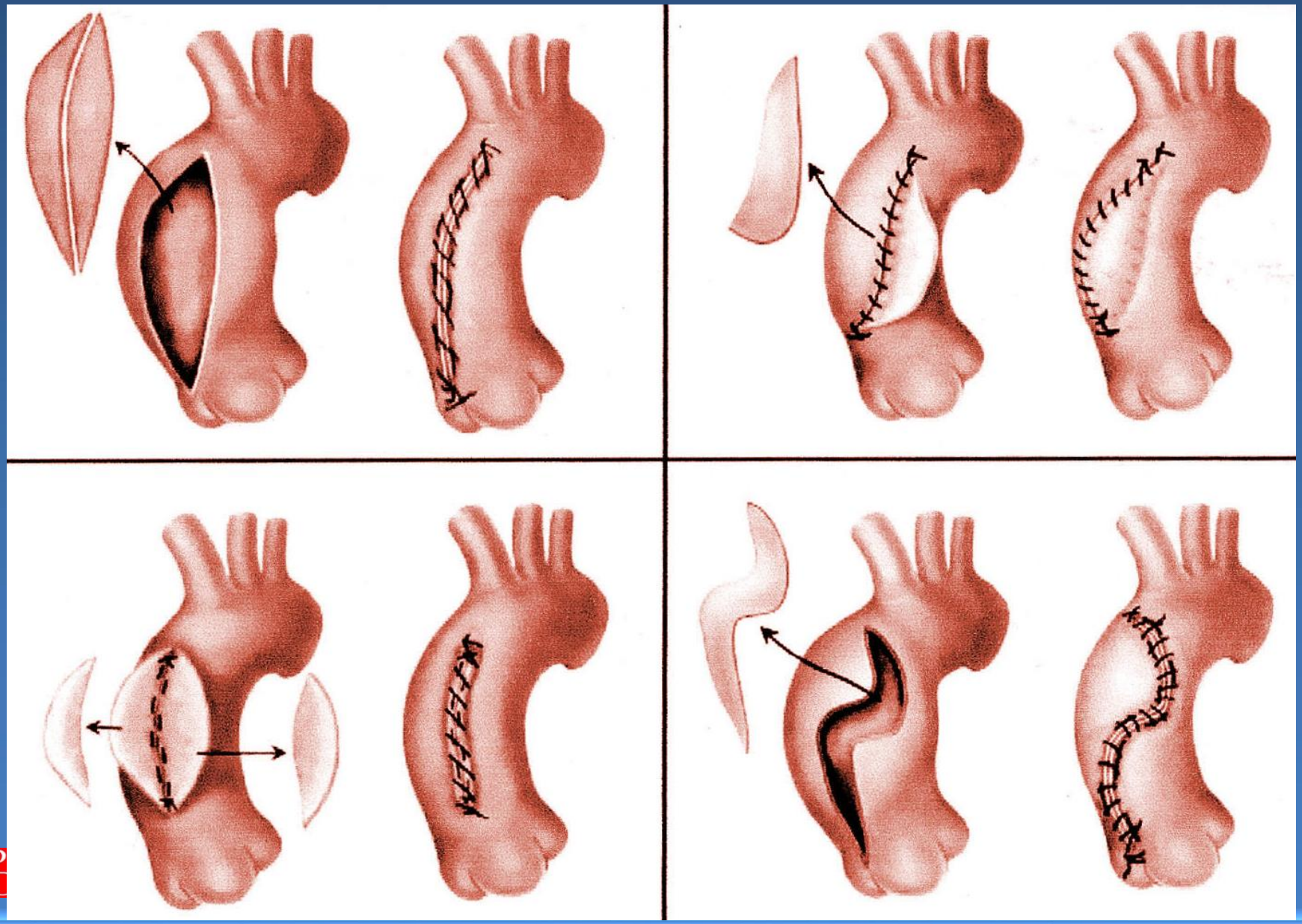
Aortic valve replacement/repair

Reduction ascending aortoplasty

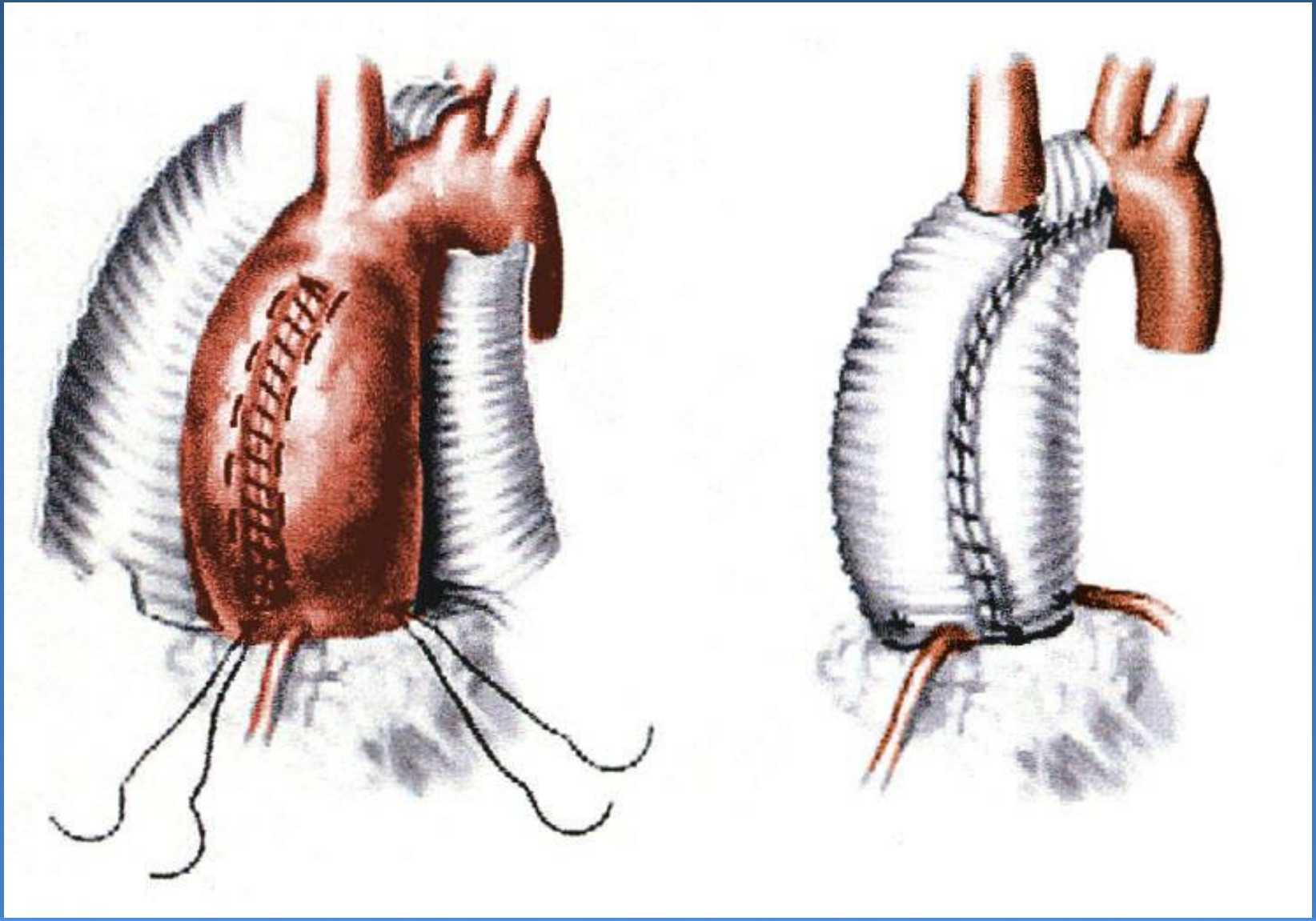
Aortic root replacement: Bentall, T. David

Ascending aortic replacement

Aortoplasty



Aortoplasty and Wrapping



Bentall

Tirone David

Ascending Aorta Replacement

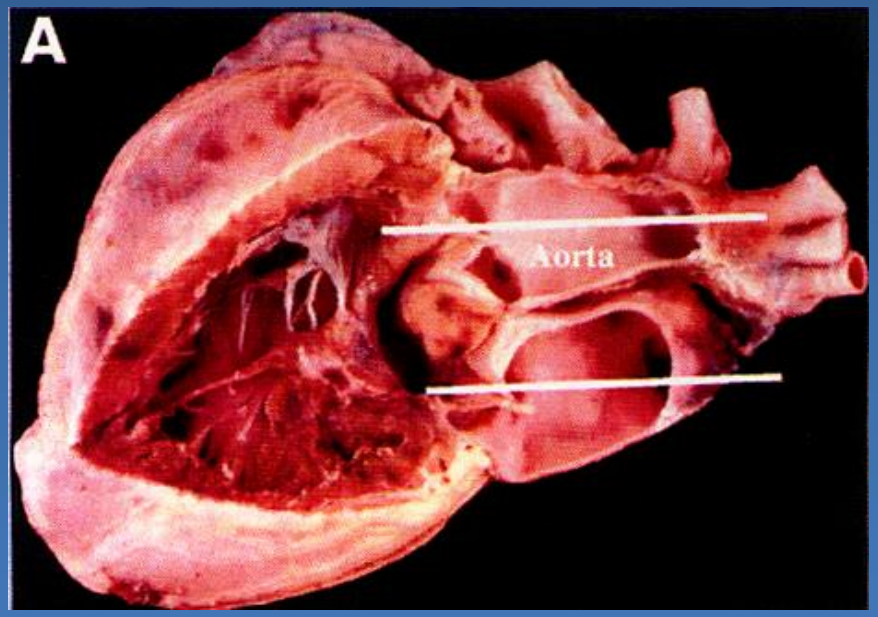
Prosthetic valve selction

- Mechanical aortic valve for AVR
- Mechanical valved conduit for aortic root replacement
- Homograft as root replacement is used in cases of active endocardity
- Valve sparing root replacement (VSRR)

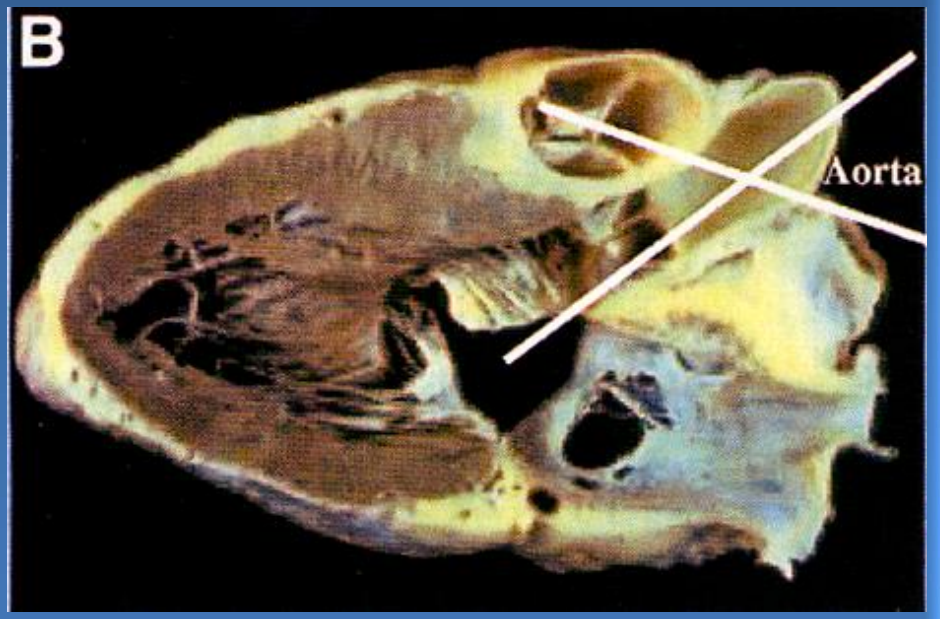
Technical issues for VSRR

1) There is more horizontal position of the aortic root in a cono-truncal anomaly
2) Difficult mobilization of coronary arteries and skeletonization of the aortic root in the presents of right-sided extracardiac conduit
3) Should be mild or less AR if VSRR is going to be considered
4) In the presents of others intracardiac diseases that will lengthen the cross-clamp time
5) Possibility of future re-operation wich is an important risk factor

DORV



Normal heart



Concomitant surg. procedures

- Ablation for A.F.
- Tricuspid annuloplasty
- Residual VSD closure
- Coronary diseases

INCIDENCE OF SURGERY FOR ACHD

IRCCS Policlinico San Donato

January 2000 – December 2011

N° CHD operated 4523

ACHD 995 (22%)

Reoperations 339 (34%)

Reoperation for ACHD

Surgical approach

Re-sternotomy	**270/276 (98%)**
CPB	262 (95%)
Ao cross-clamp	182 (66%)
FV isolation	36 (13%)
FV cannulation	16 (6%)

Reoperation for ACHD

Type of reoperations

RVOT Disease	**102 (37 %)**
PVI	73
Conduit replacement	29
LVOT procedures	**46 (17%)**
AoV Repl/repair	26
Aortic root surgery	10
Sub Ao S	10
Fontan conversion	**29 (10%)**
Residual ASD/VSD	**28 (10%)**
Other	**56 (21%)**

LVOT surgical procedures in pts wih cono-truncal malform.

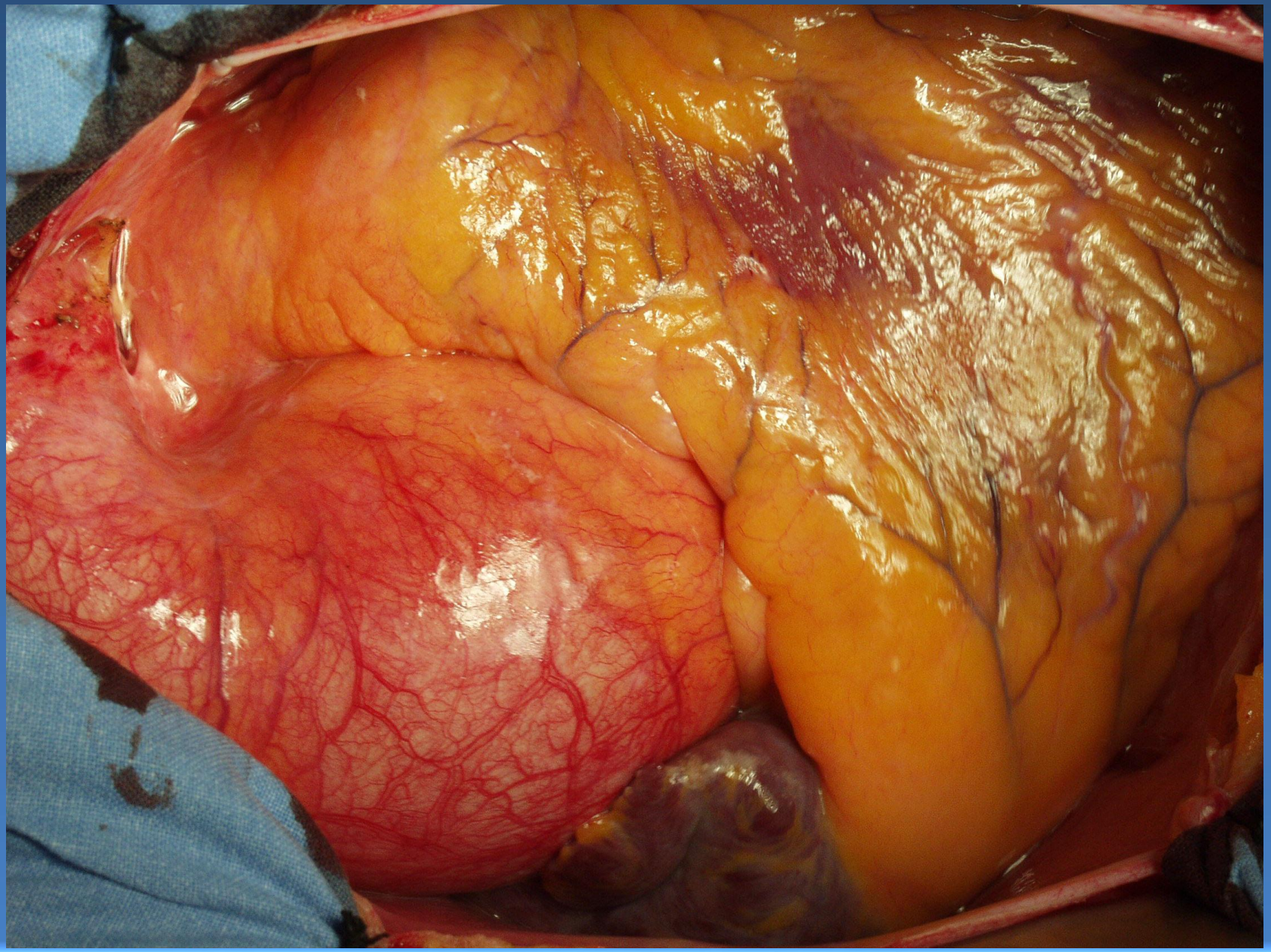
Ao V. Replacement/Repair	26
Aortic root replacement (Bentall)	7
Reduction ascending aorta + AR	2
Ascending aortic replacement	1

(5 Fallot - 2 Pulm. Atresia + VSD - 1TGA - 1 C.TGA - 1 Truncus)

Conclusion

In our experience, moderate ascending aortic enlargement is common among patients with conotruncal anomalies coming to aortic valve surgery. Because aortic dissection and the subsequent need for re-operation apper to be low in this setting, we belive that the moderately dilated aorta can be observed.

ARD



Reoperation for ACHD

High incidence of associated surgical procedures

N° procedures 193 on 124 patients (45%)

Arrhythmia surgery	54
RV remodelling	47
TV repair	32
Res VSD closure	13
PAs enlargement	11
Res ASD closure	11
PMK	10
Other	15

Type of reoperation for ACHD

Comparative data

Dore et al	Berdat et al	IRCCS PSD
LVOT/Ao 36%	LVOT/Ao 46%	**PVI 25%**
Cond repl 17%	PS 12%	LVOT/Ao 22%
Palliations 8%	PVI 11%	Cond repl 10%
Reco Ao 6%	Reco Ao 8%	Res ASD 8%
PVI 3%	Cond Repl 6%	**Conv Fontan 7%**
MVR/Repair 3%	MVR/Repair 6%	MVR/Repair 6%
Fontan 3%		TVR/Repair 4%
Res ASD 3%		Fontan 4%
		Palliation 4%
		Reco Ao 1,5%

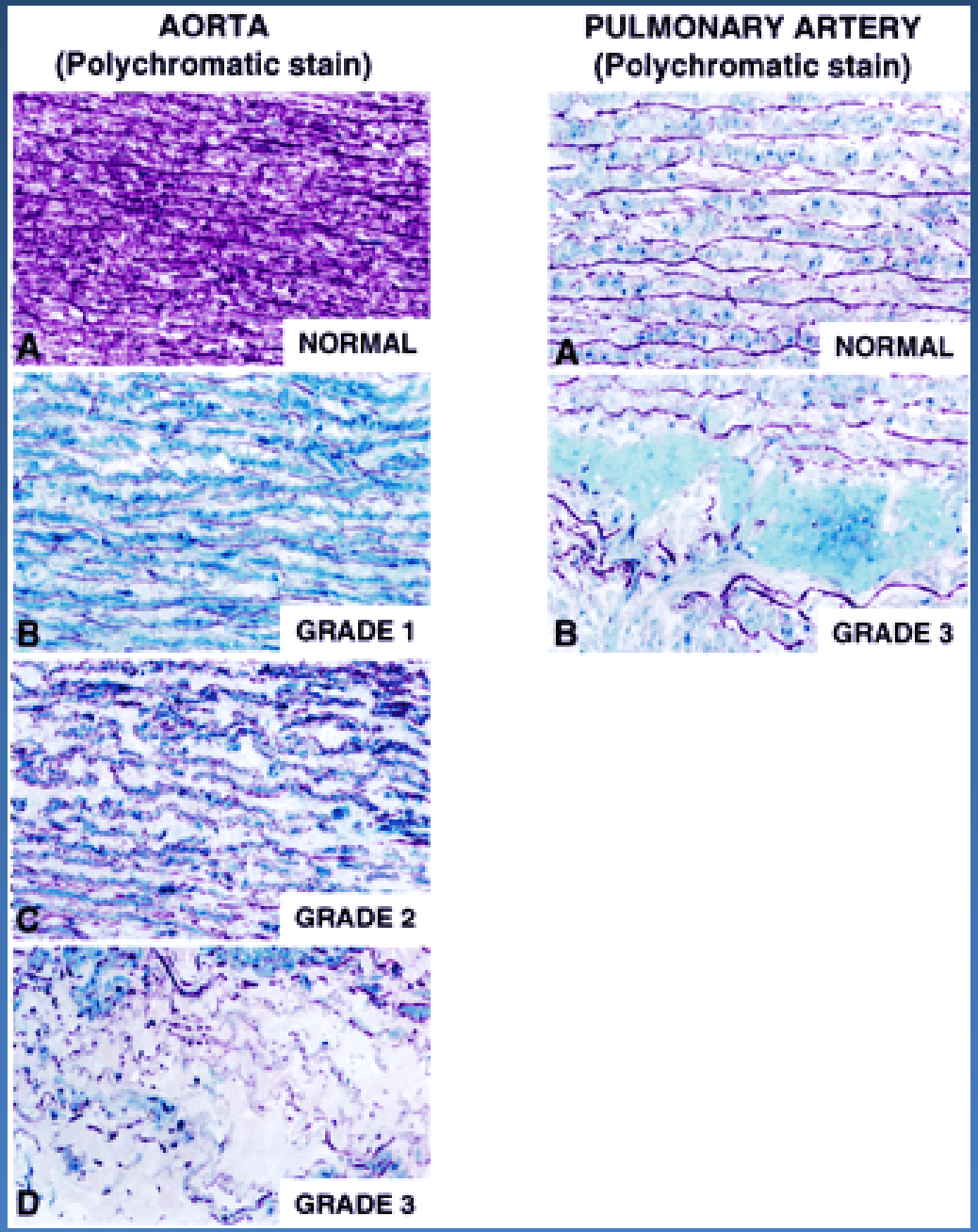
Incidence of reoperations in ACHD

Comparative data

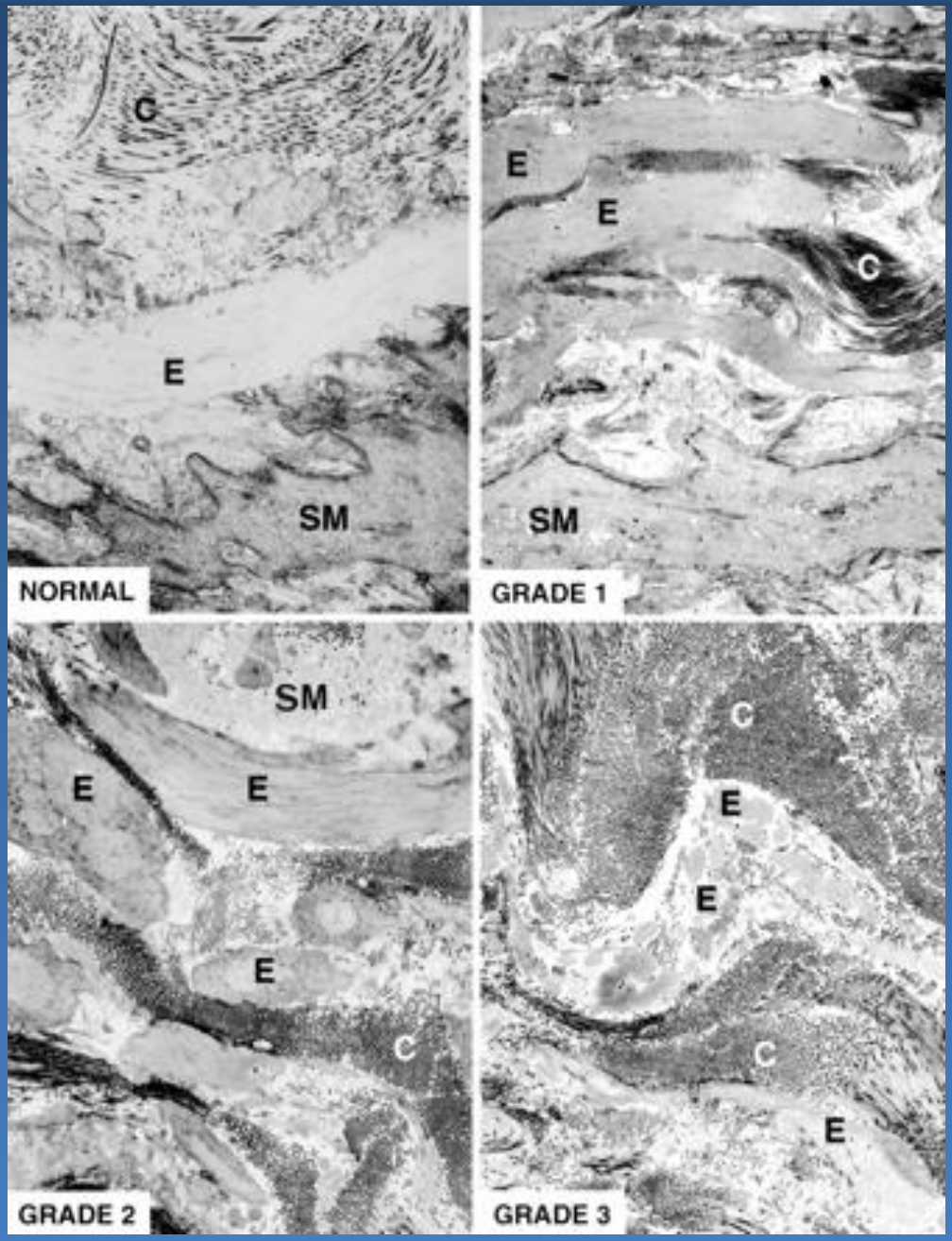
	PERIOD	N. PTS	RE-OP
Dore et al	1991-1994	295	179 (58%)
Berdat et al	1987-2000	277	95 (34%)
IRCCS PSD	2000-2007	664	201 (30%)

Surgical strategies S.H.D.

- Mechanical valve conduit for ARD
- Aortic valve replacement: mechanical
- Aortic homograft for active endocarditis



Niwa K. et al 2001, Circulation



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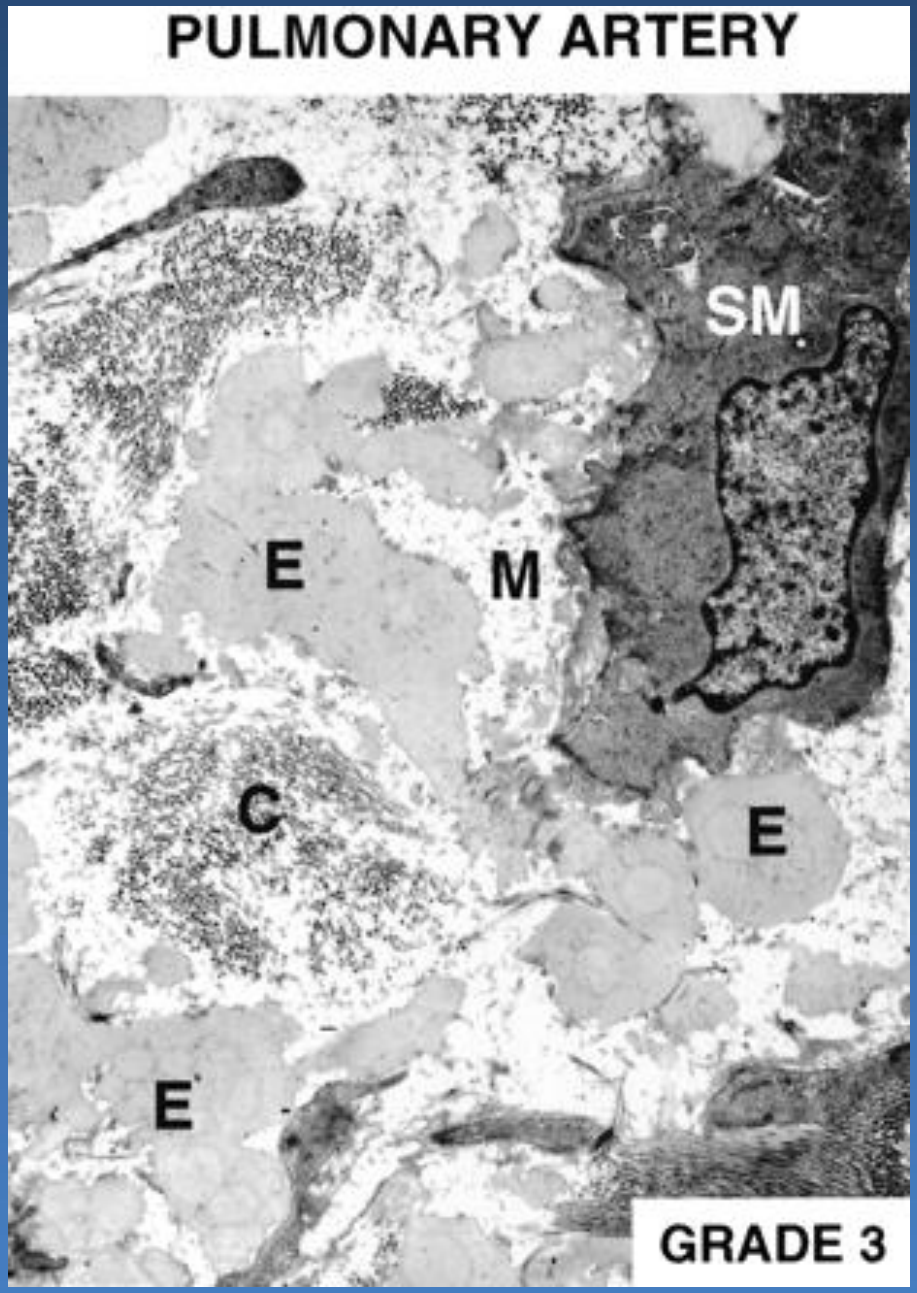

Niwa K. et al 2001, Circulation

ARD