

Current Issues in Congenital Heart Disease

**Can we Change the Tails of
Congenital Heart Disease?**

**Seoul
2005**

**Glen Van Arsdell
Head, Cardiovascular Surgery
Hospital for Sick Children, Toronto**

Definition of Tails and Tales

- Tails



- Tales/Stories



Walton Heath Golf Club

“Tales/Stories”



Gentleman Bob



Tails and Tales

Tetralogy

Transposition with VSD/Pulmonary stenosis

Corrected Transposition

Complete Repair of Tetralogy of Fallot

August 31, 1954

1st Complete repair

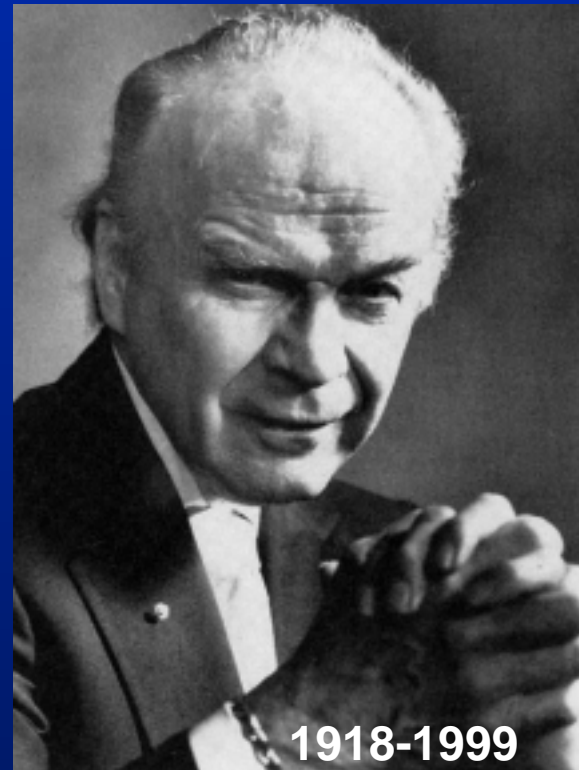
Crosscirculation

Survival

6/10

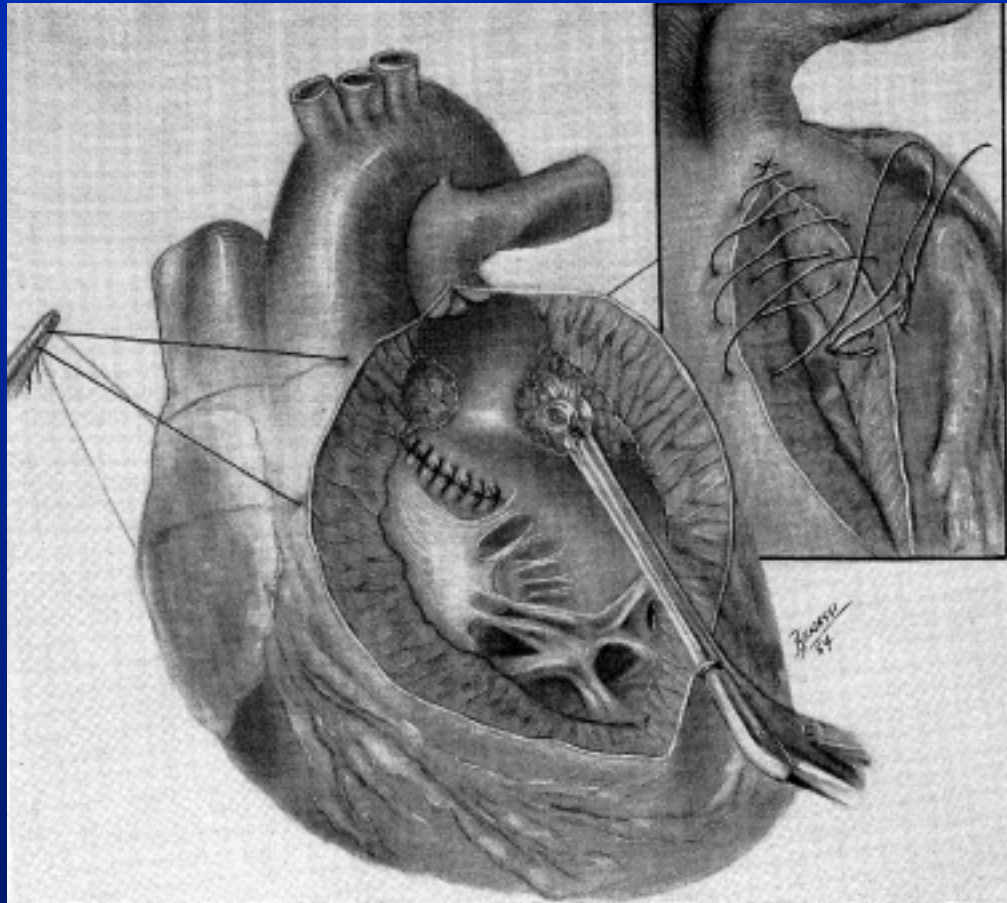
Lillehei Ann Surg 1955; 142:418-445

C. Walton Lillehei



1918-1999

Transventricular Repair of Tetralogy of Fallot



Lillehei Ann Surg 1955; 142:418-445

36 Year Outcomes of ToF – The “Tails”

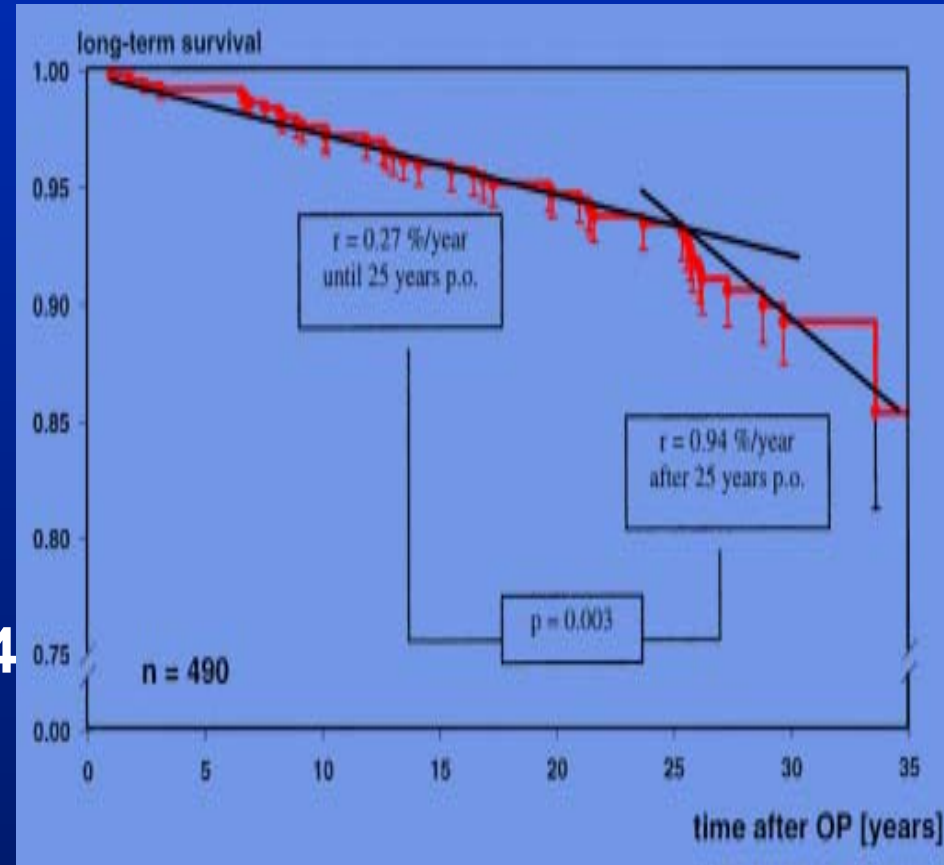
Munich

1958 -1977

490 over 1 year survivors

Late Sudden death 3.1%

Higher RV/LV >0.70 had 94.4% survival vs 83.7% term survival (p=NS)



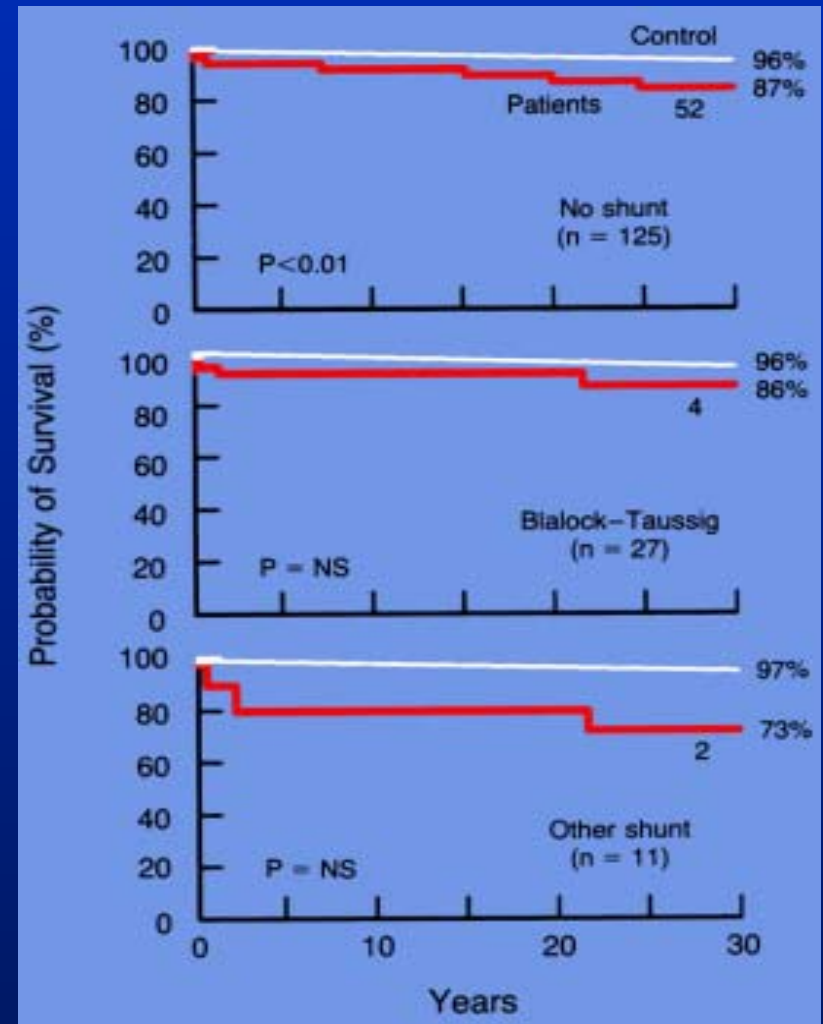
32 Year Outcomes Transventricular Repair ToF

Mayo Clinic – 1955-1960
163 Survivors

Late Sudden death 6%

Higher RV/LV systolic pressure ratio predicted long term survival ($p=0.008$)

Murphy J et al NEJM 1993

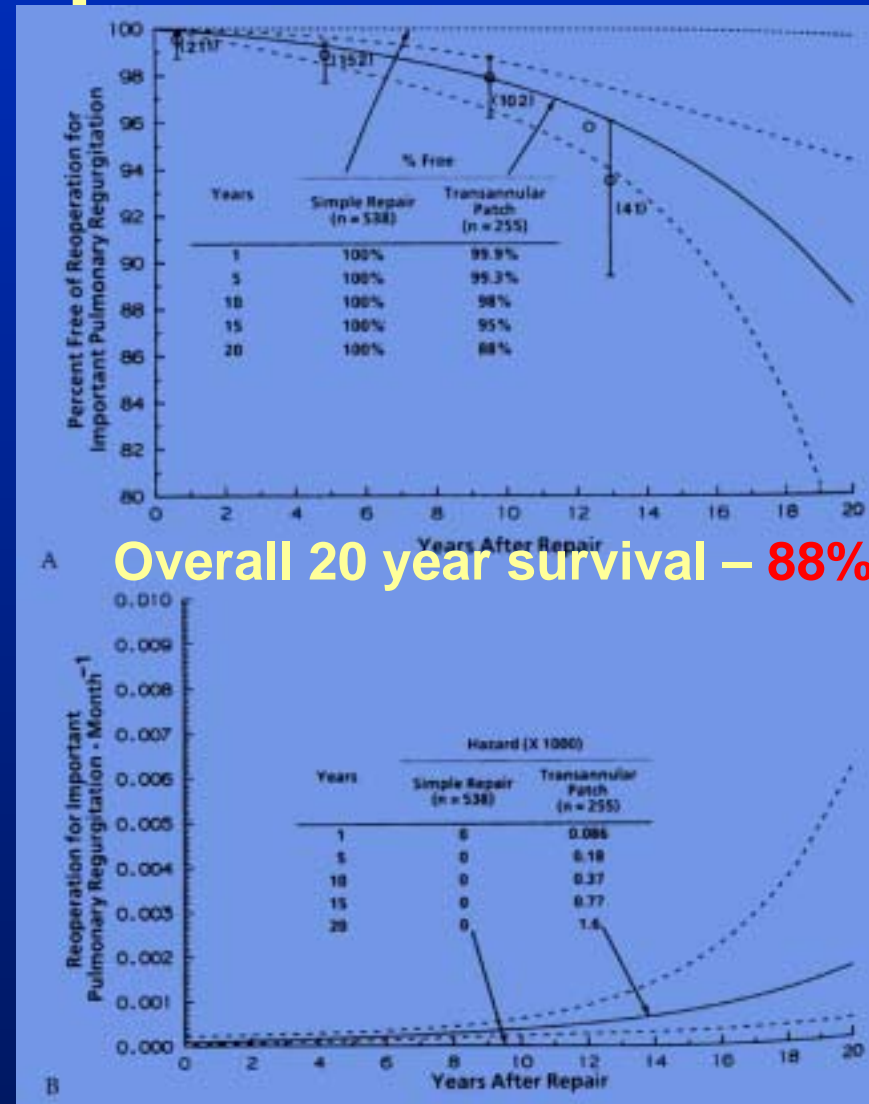


Effect of Transannular Patch on Outcome after repair of ToF

1967 – 1986
814 patients

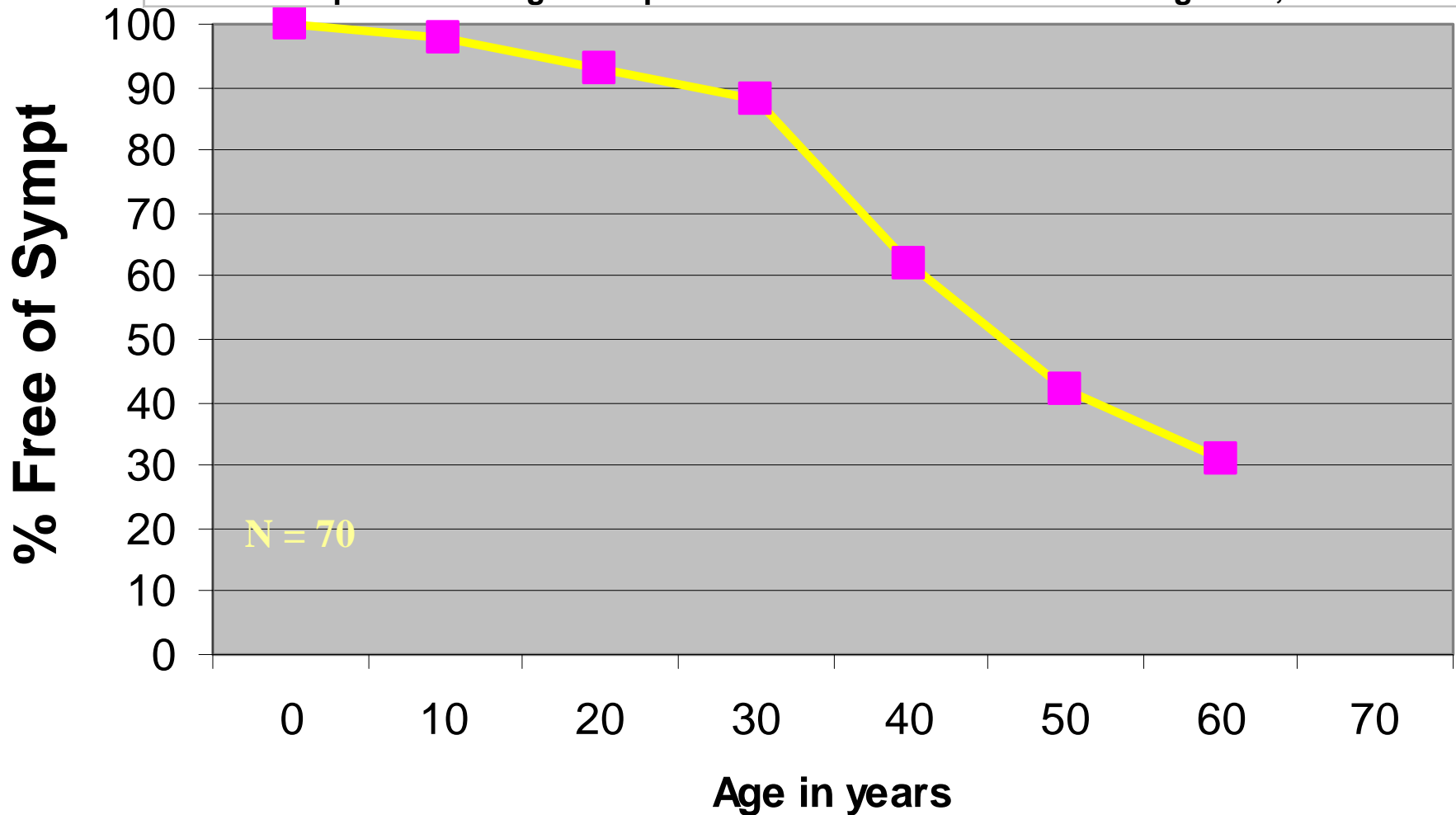
Op mortality
4% vs 1.4%
TAP vs No TAP

Kirklin J et al ATS 1989;48:783-791



Natural History of Isolated Pulmonary Valve Insufficiency

Shimazaki Y, Blackstone EH, Kirklin JW. The natural history of isolated pulmonary valve incompetence: surgical implications. *Thorac Cardiovasc Surg* 1984;41:478-82



Late Outcomes - Tetralogy

Mechanoelectrical interactions

Andrew Redington

Restrictive physiology – QRS 129 ms
CTR 0.51

No restriction - QRS 157ms
- CTR 0.54

Ventricular Tachycardia - QRS 199 ms
- CTR 0.67

QRS > 180 ms - 100% sensitive for VT/SD



Gatzoulis MA, Redington et al. Circ. 1995;92:231

Late Outcomes - Tetralogy

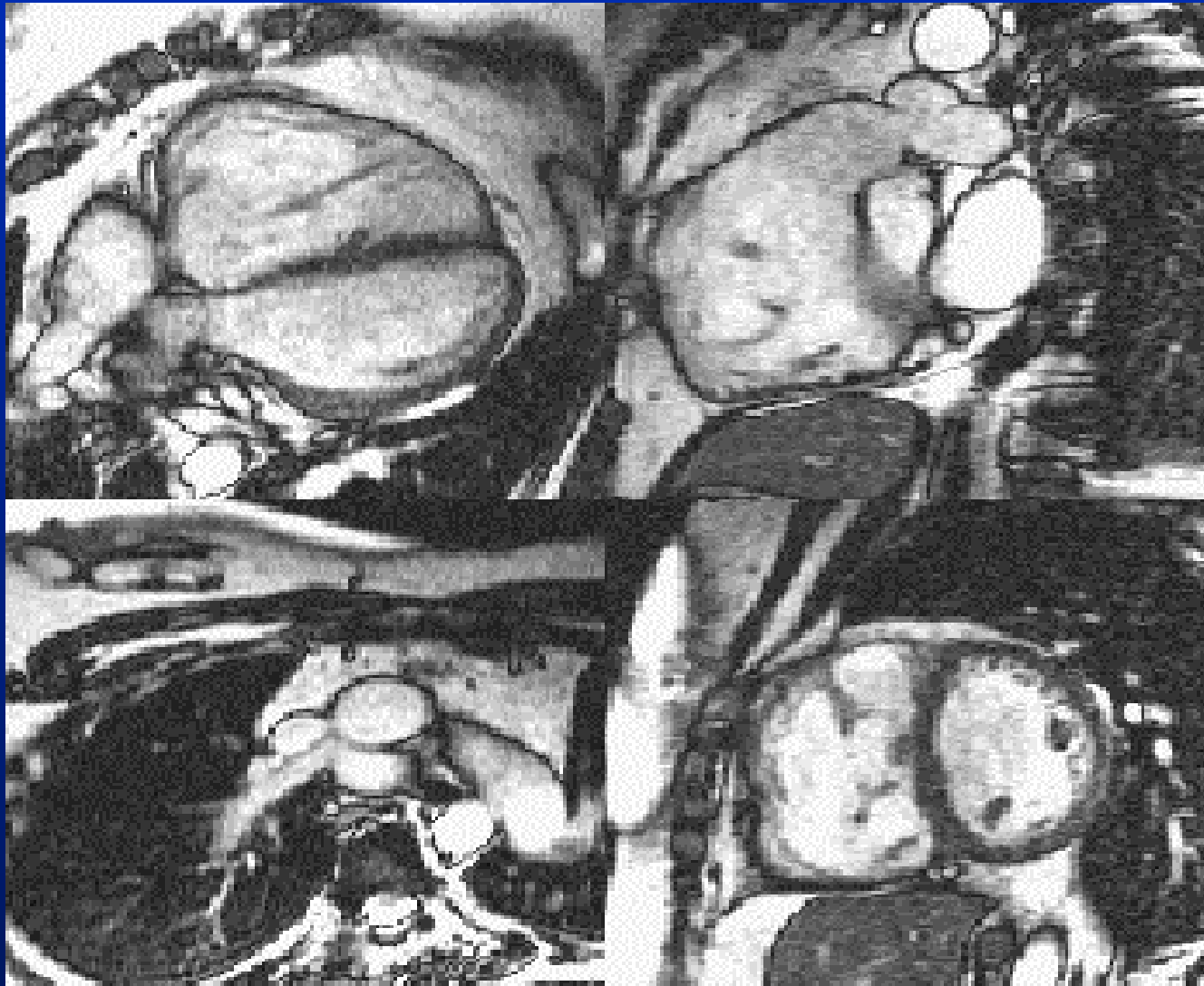
793 patients – multi-institutional

- Older age of repair – risk for sudden death and atrial tachyarrhythmia
- PI – risk for VT and sudden death
- Tricuspid insufficiency – risk for afib/flutter
- Transannular patch was associated with SD and ventricular tachycardia

Transatrial/Transpulmonary Repair

- **10 yr f/u - minimal muscle release – Bove**
 - **Smaller RV size RV/LV – 0.66 vs 0.81**
 - **Small CTR - 0.53 vs 0.58**
 - **Shorter QRS 126 vs 143**
 - **Better RV function**
 - **Less ventricular ectopy**

Ventricular Function Pulmonary Insufficiency



Ventricular Function Pulmonary Stenosis



Spectrum of Ventricles

Supreme Notes



Just Right Dr. Kim



Governor of California



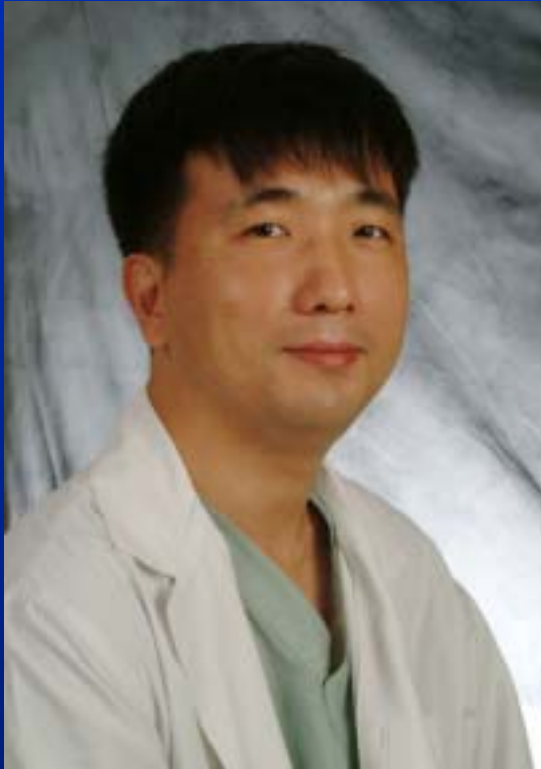
Impact of a Strategy for Preservation of the Pulmonary Annulus – July 1997

Bias that all pulmonary annuli can be
preserved (exception - 10 %)

Parietal bundles wedge excision

Preservation of septal band

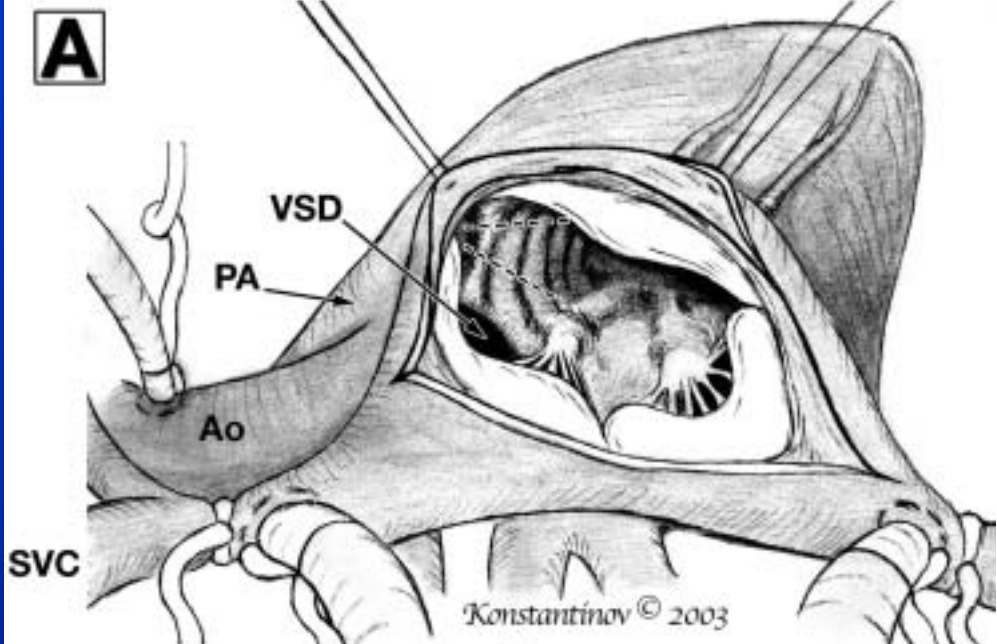
Tae-Jin Yun



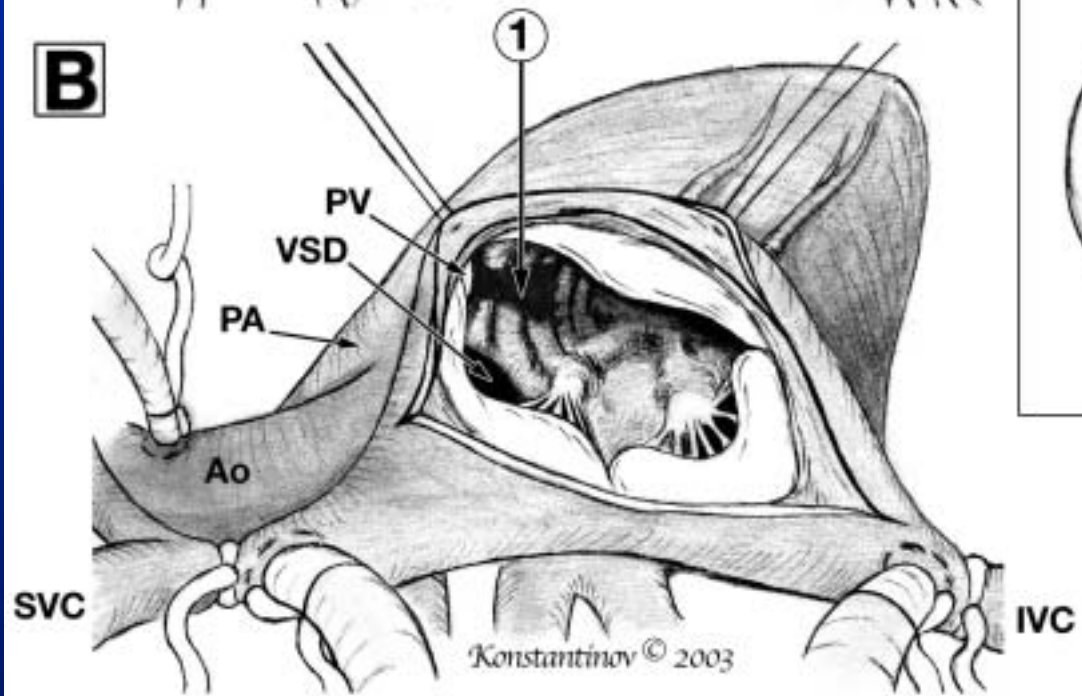
Michael Cheung



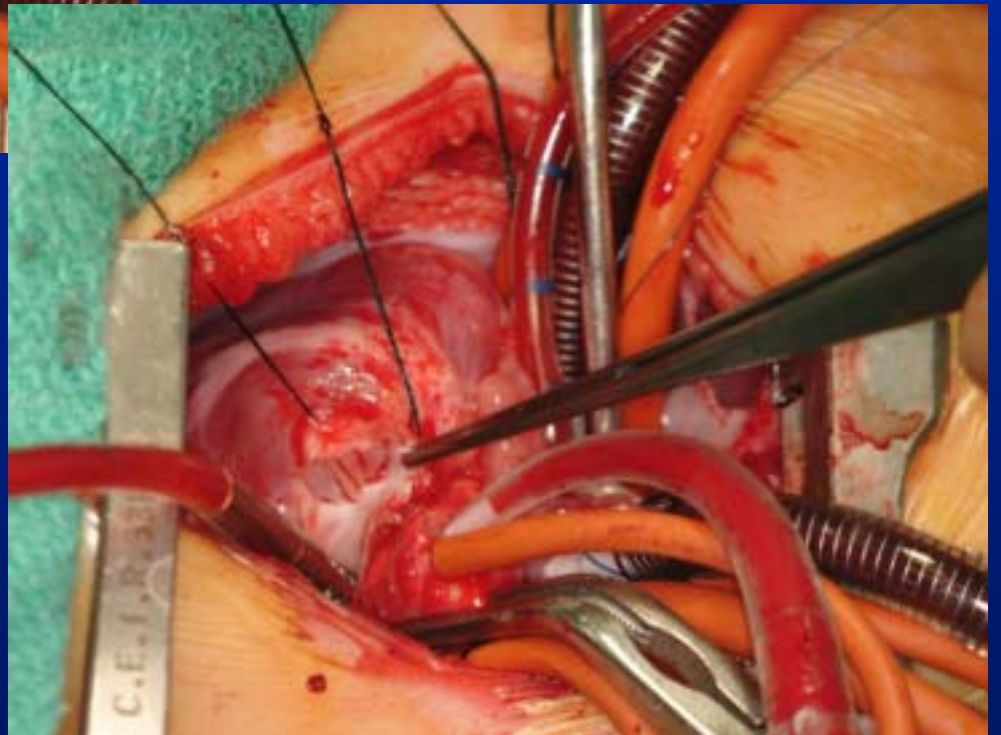
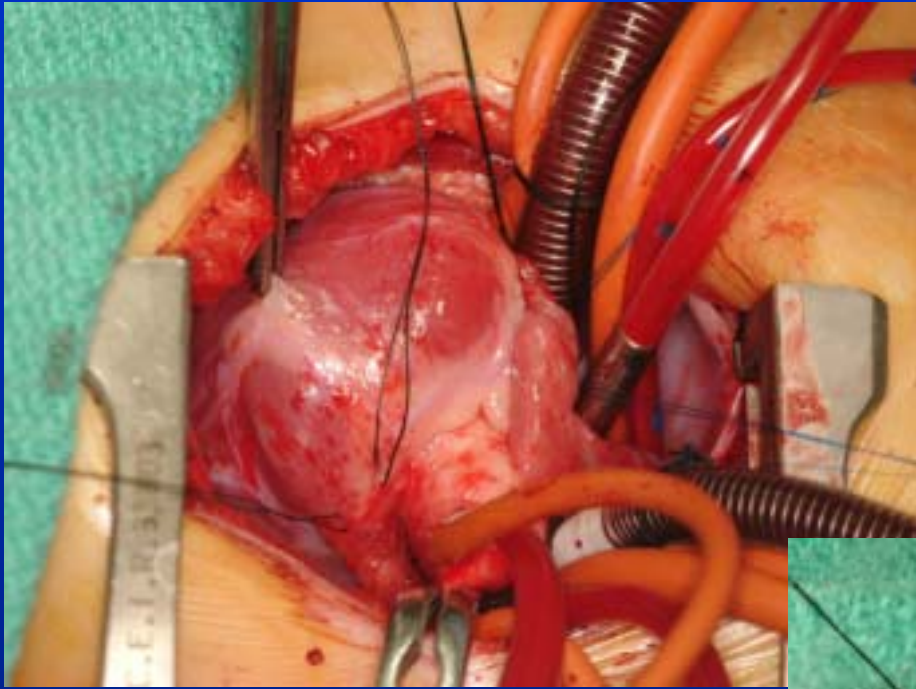
David Ashburn

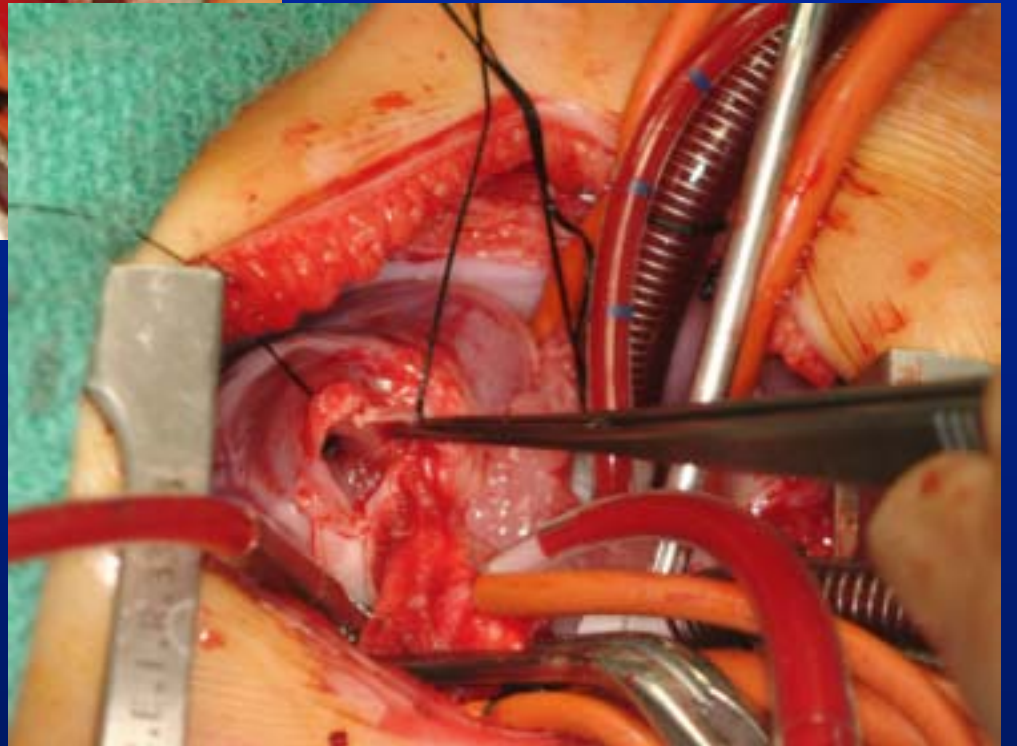
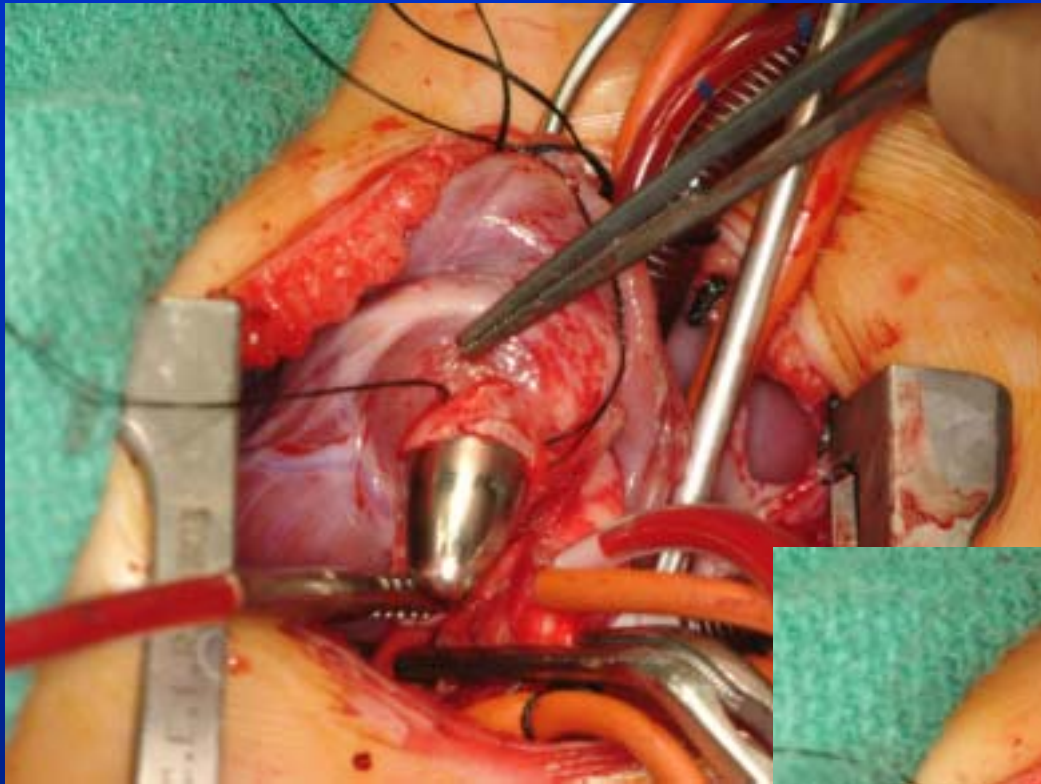
A

Pulmonary Valve

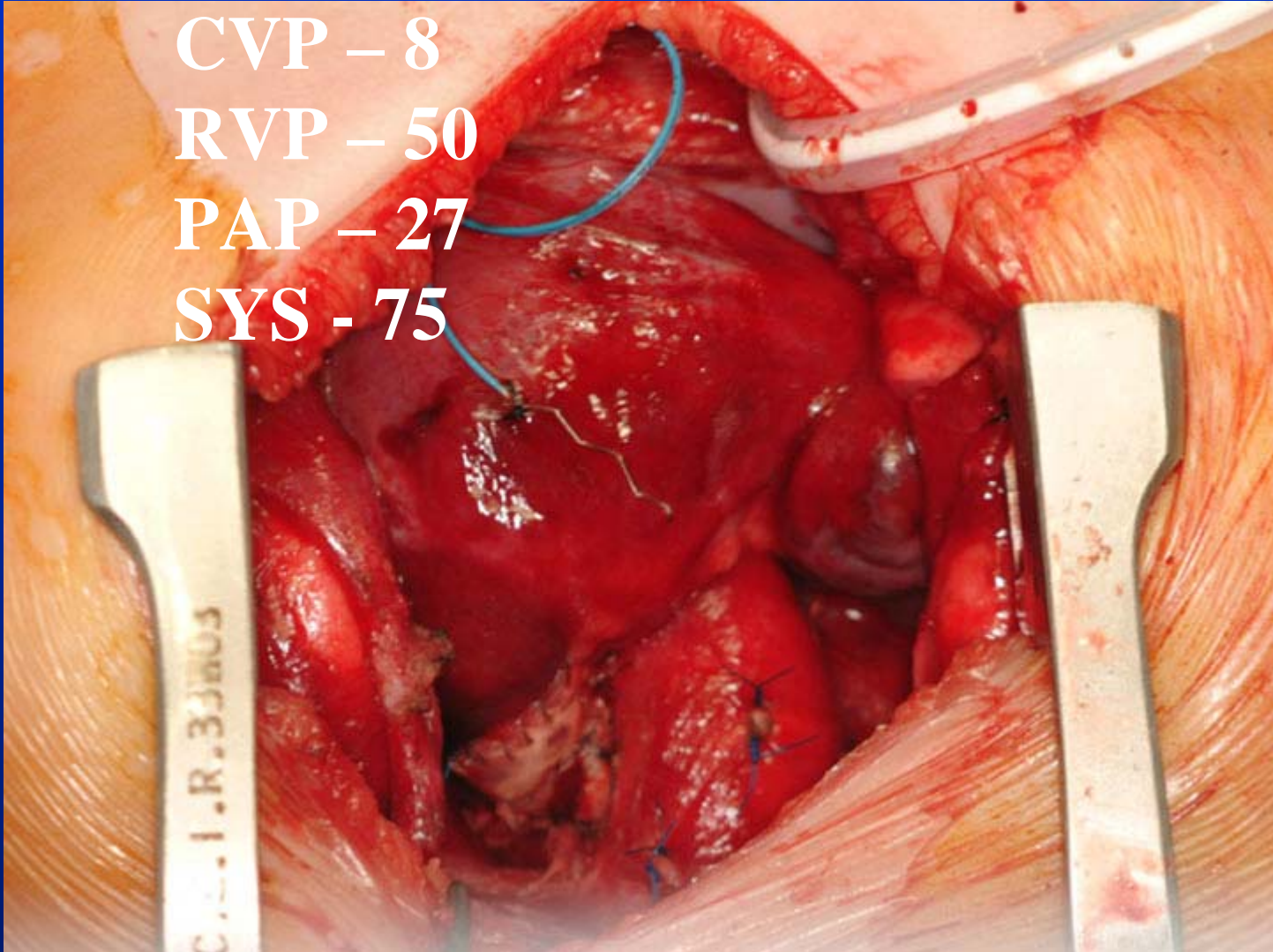
B**A****B**

Konstantinov © 2003

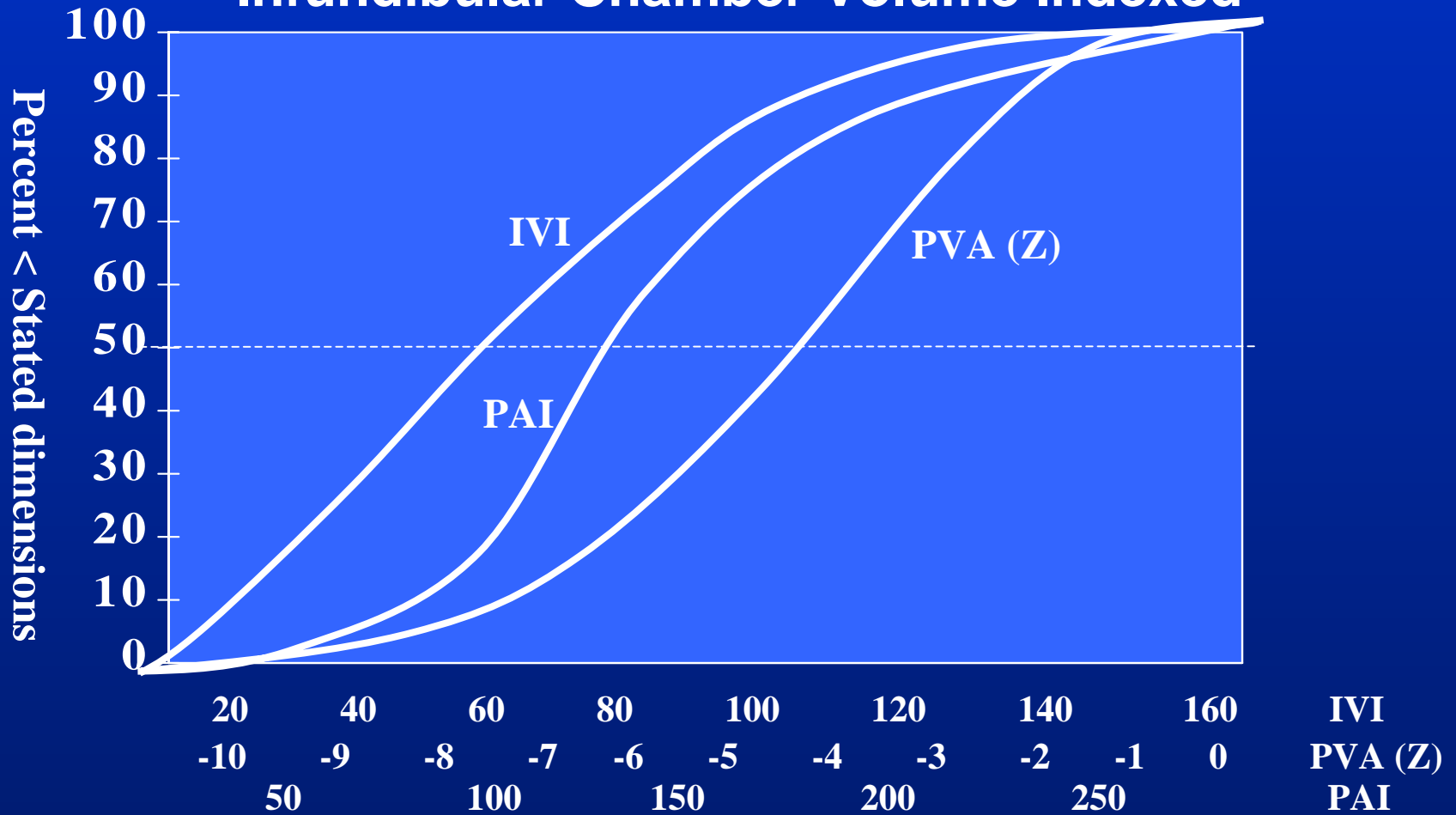




CVP - 8
RVP - 50
PAP - 27
SYS - 75



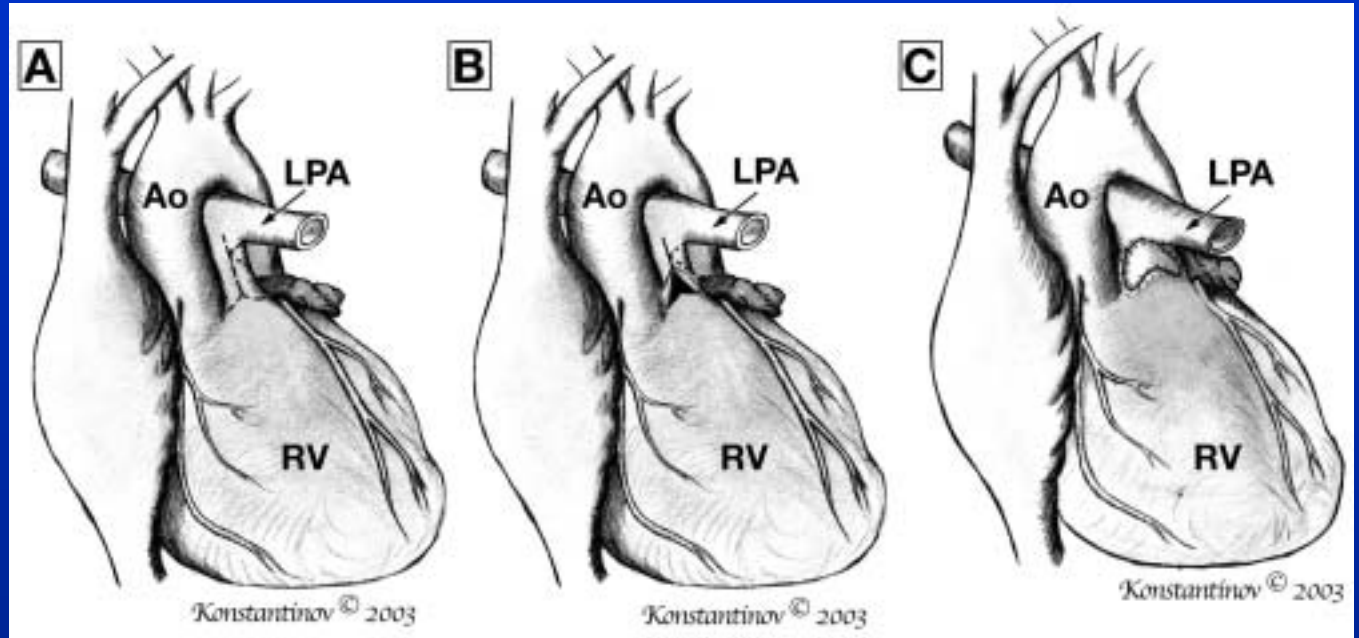
Pulmonary Valve z Value Pulmonary Artery Index Infundibular Chamber Volume Indexed



Frequency

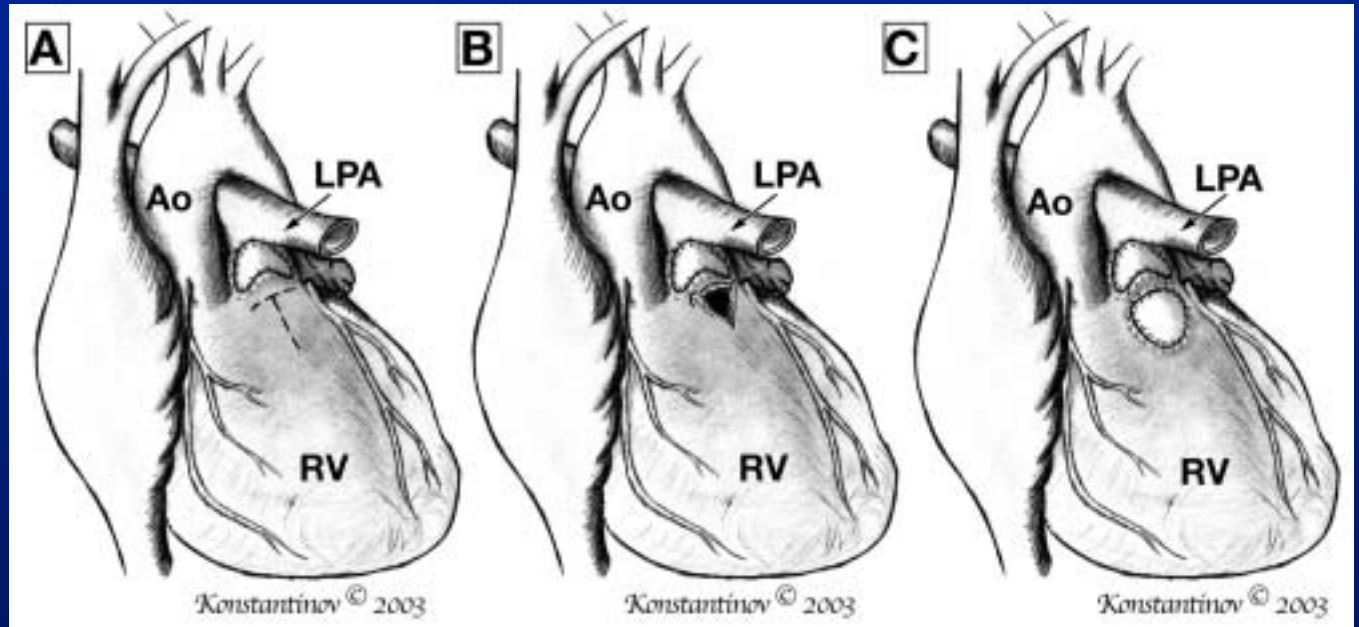
N=118

53%



28%

TAP 19%
PV n=1

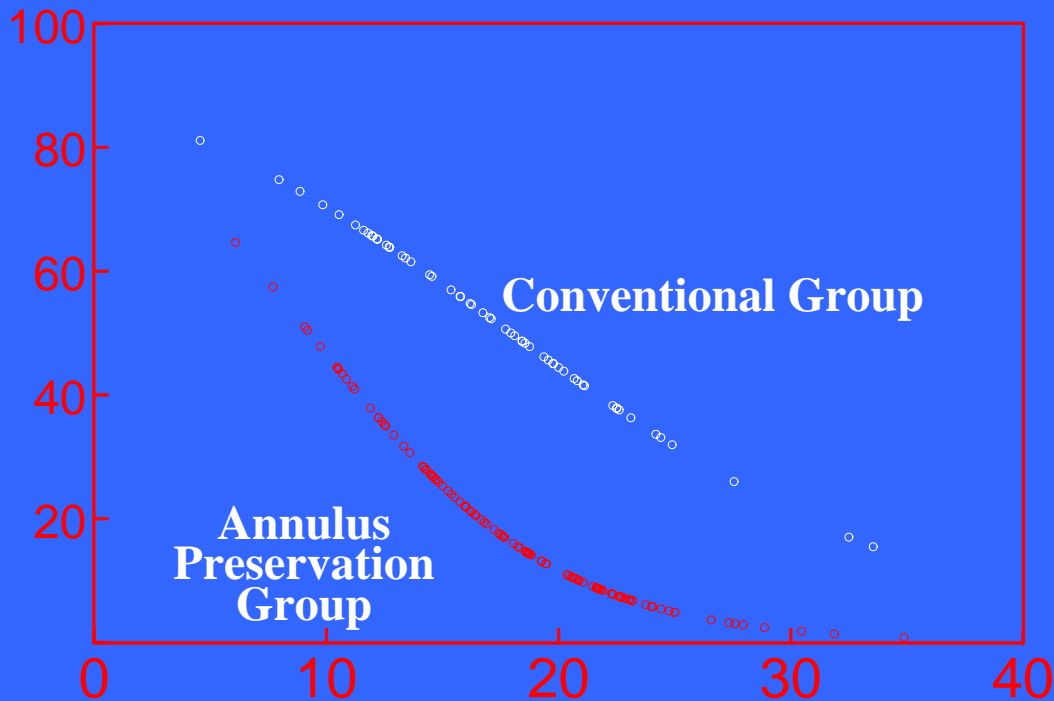


Preservation of the Pulmonary Annulus and Septal Band

- Sep 1996 – Sep 2002 n=185
- AP strategy : 118 (64% of ToF patients during same period)
- Age at repair: 1.4m – 185m (median: 6.6m)
- No operative mortality AP strategy group
- Reoperation for RVOTO: 4/118 (3.5%)
Reoperation for RVOTO in conventional group: 0/67 (0%)

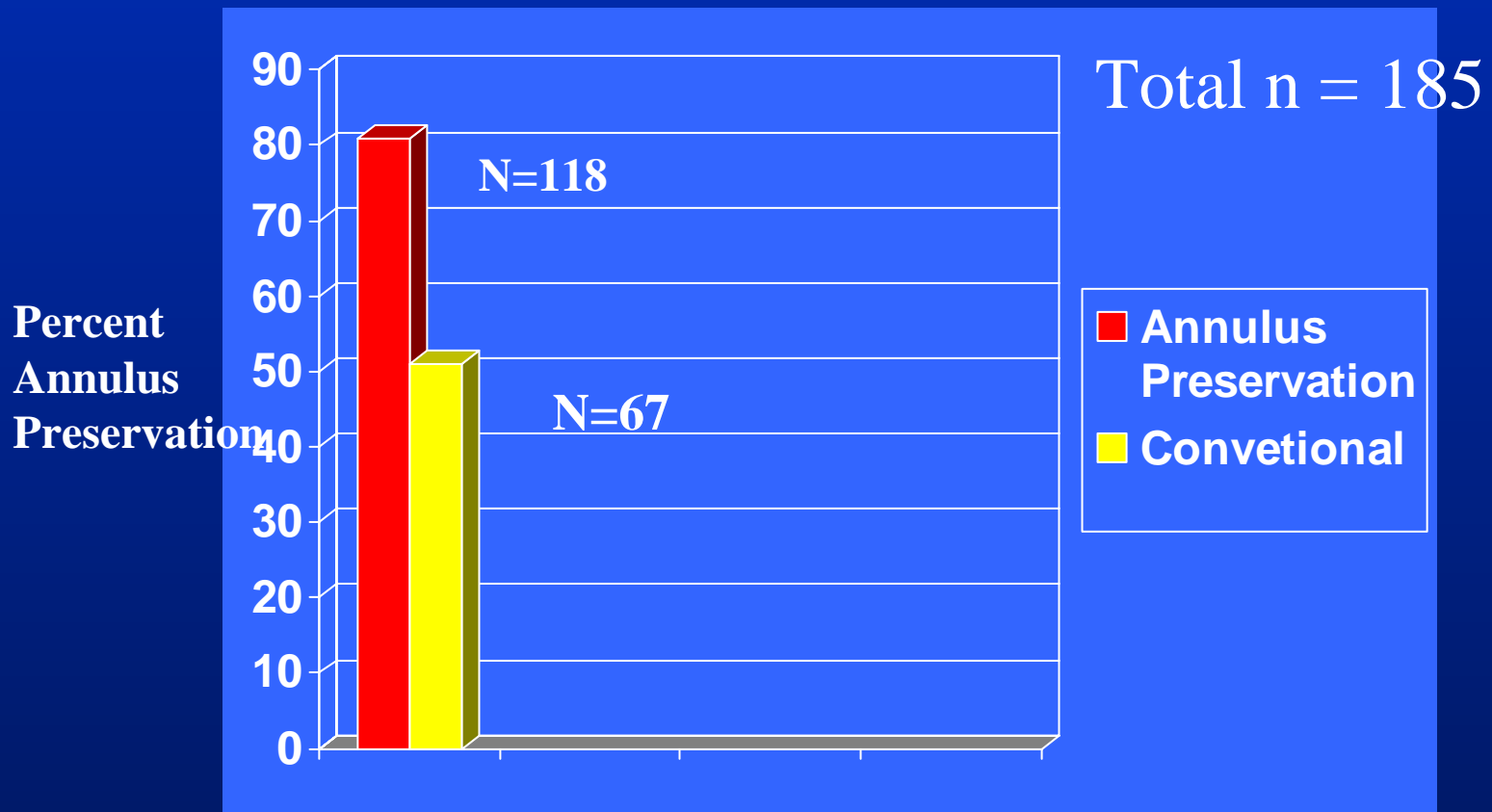
Surgical Strategy Impacts Incidence of TAP for Equivalent Anatomy

Probability of TAP (%)

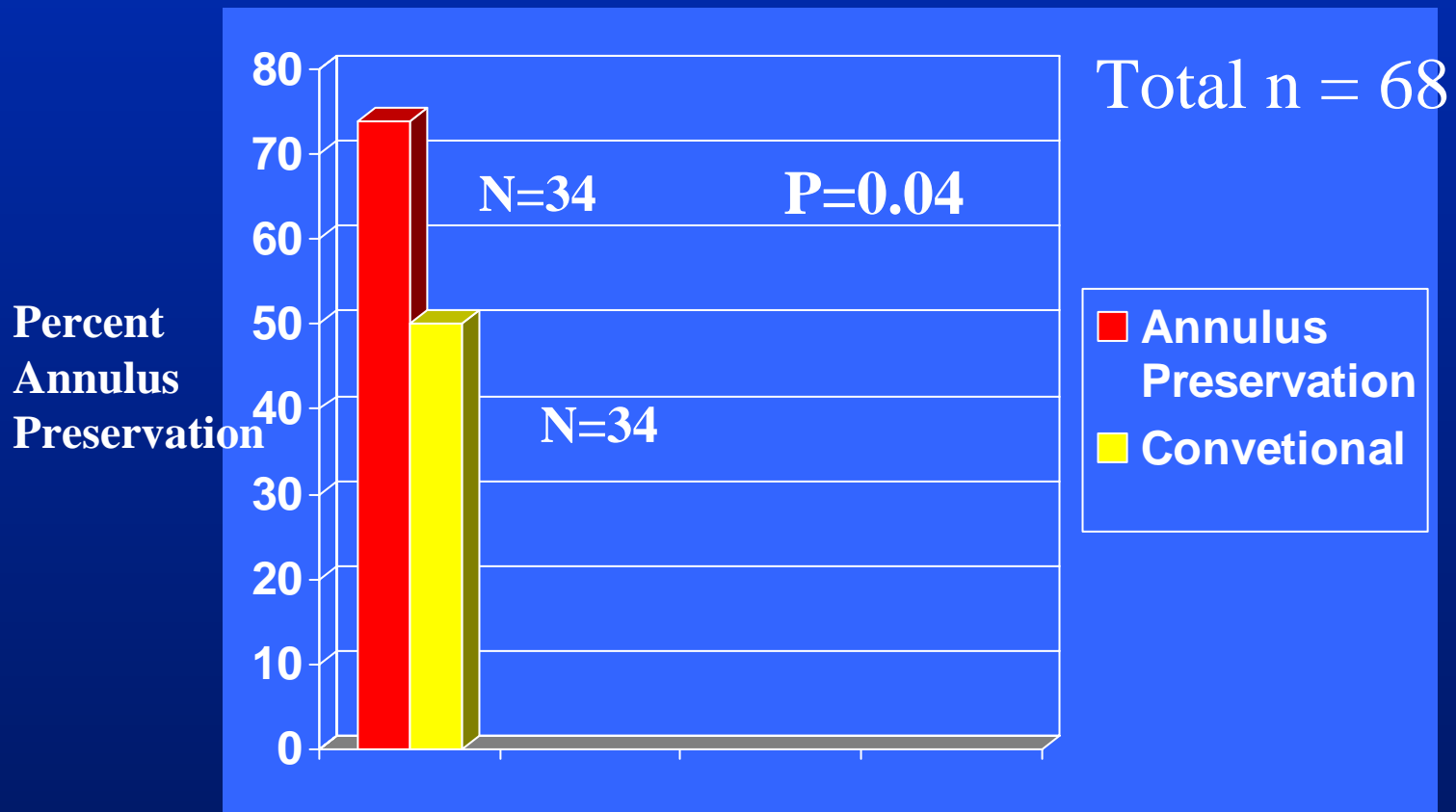


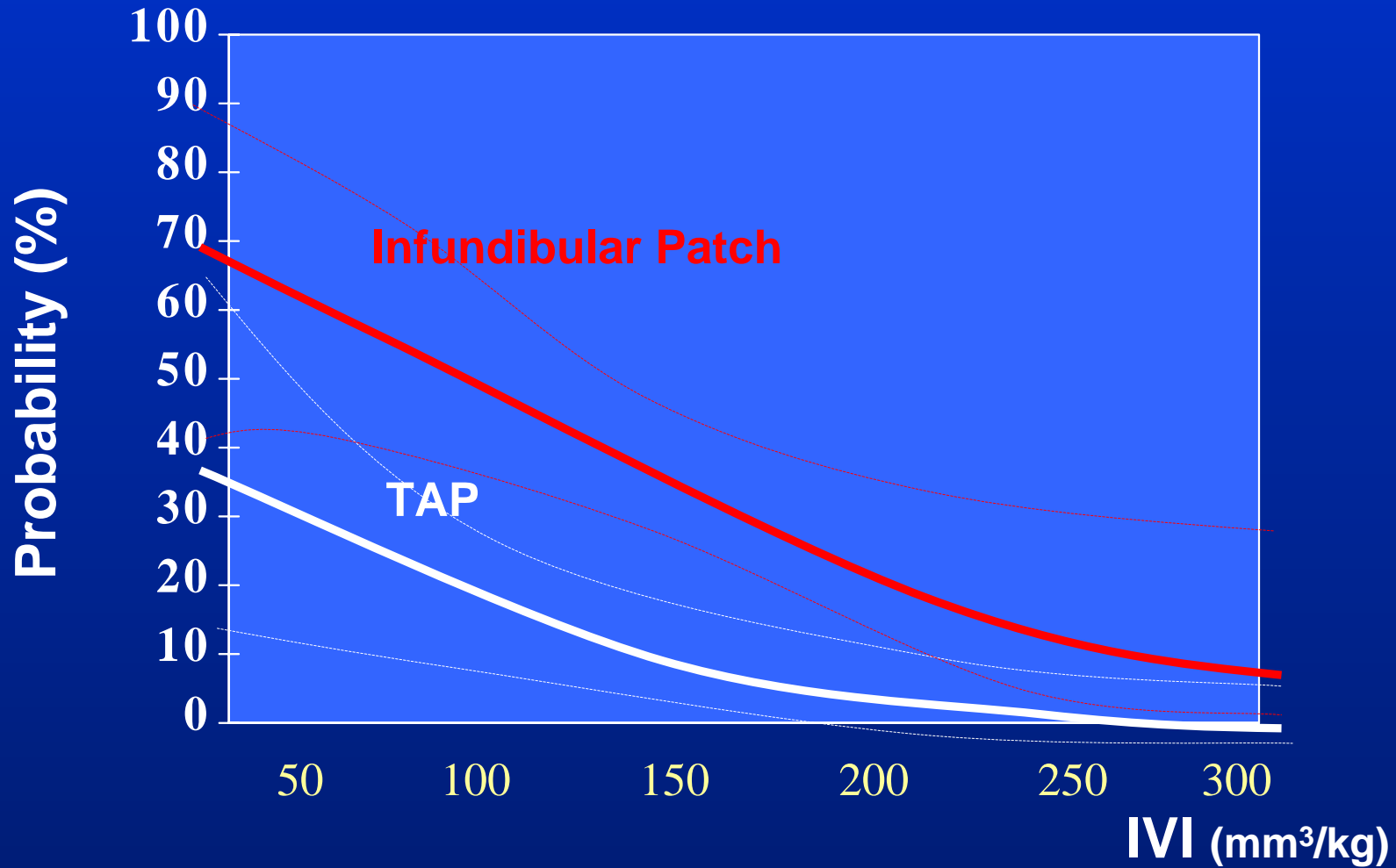
PV annulus/BSA (mm/m²)

Conventional and Annulus Preservation 9/96 – 9/02



Propensity Matched Comparison of Conventional and Annulus Preservation Strategies

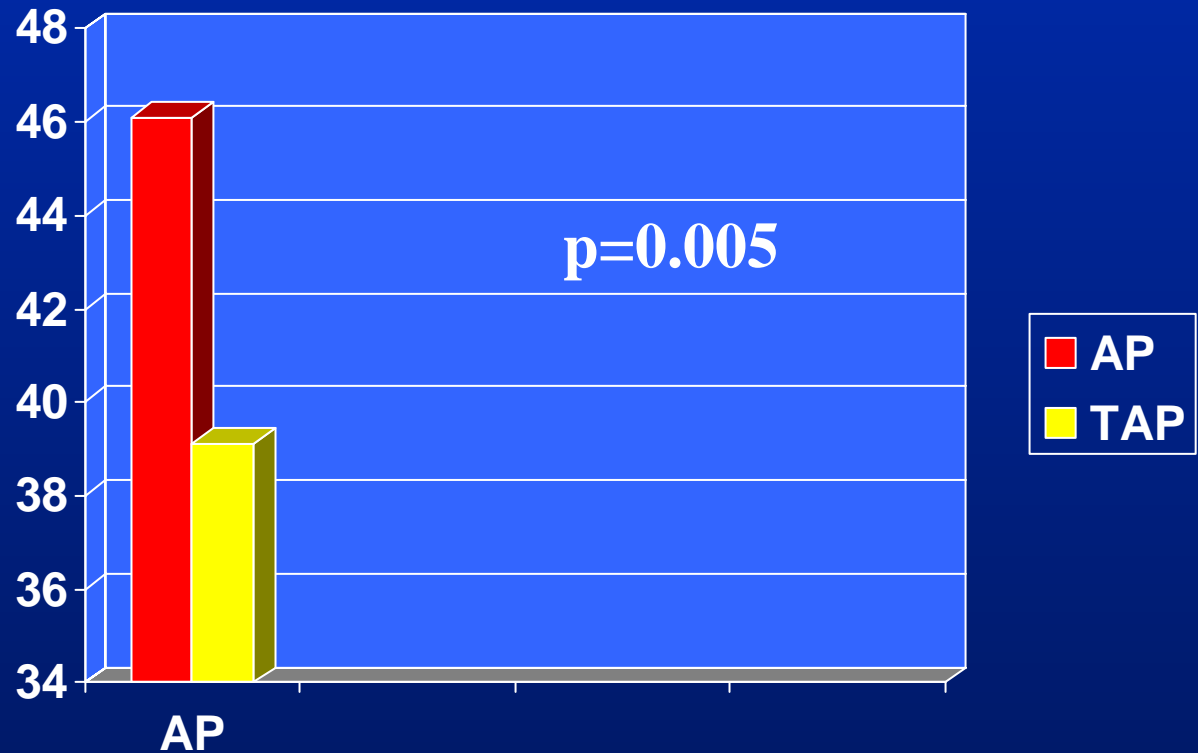




Influence of Infundibular Volume Indexed on Infundibular and Transannular Patch

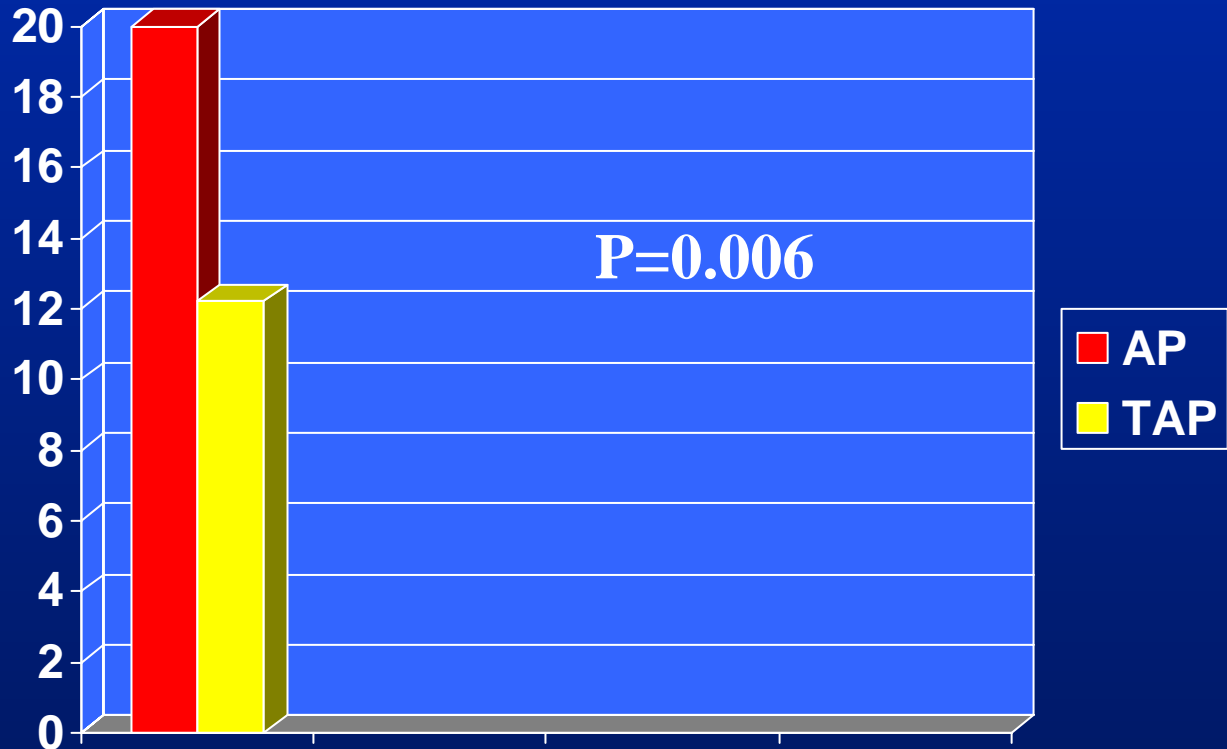
Comparison of AP and TAP in All “Marginal” Patients

RV
Pressure
Intraop



Comparison of AP and TAP in “Marginal” Patients

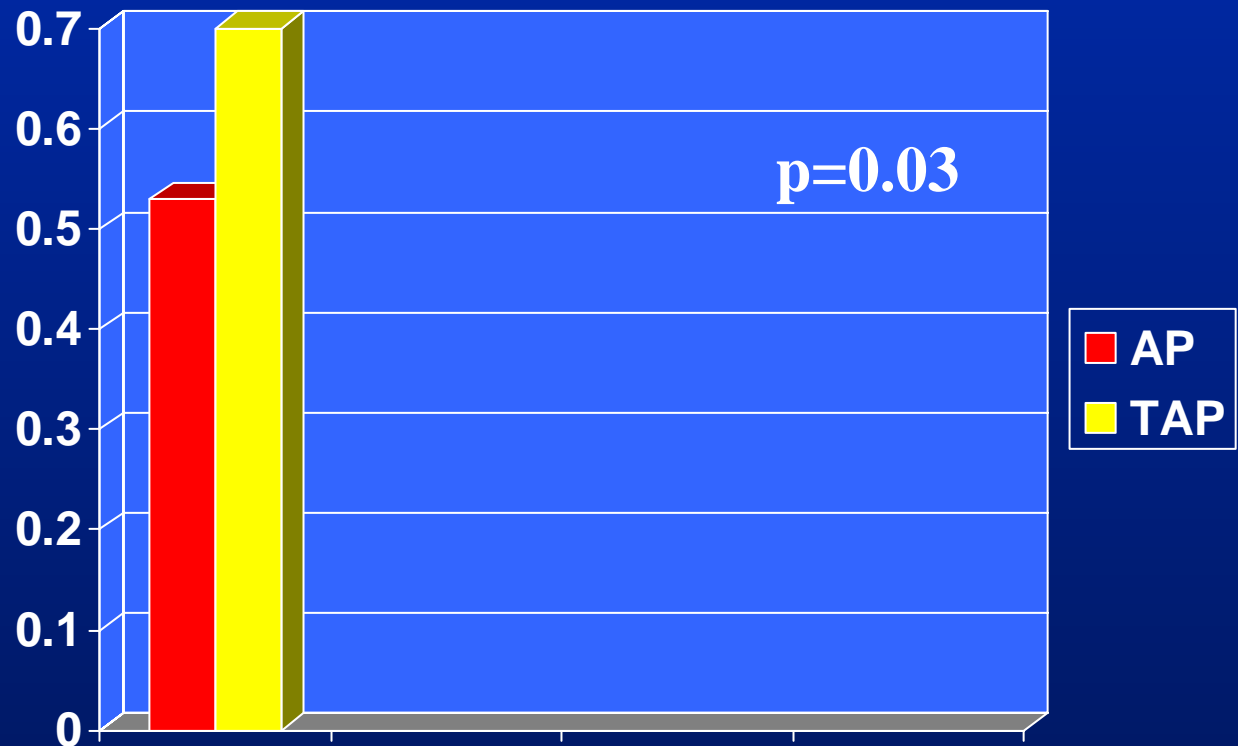
RVOT
Gradient
Intraop



Comparison of AP and TAP in “Marginal” Patients

Median f/u 16mo. (8-72 mo)

Ratio
RVEDD/LVEDD
F/U



Annulus and Septal Band Preservation Strategy

Strategy impacts the incidence of TAP for **equivalent** patients

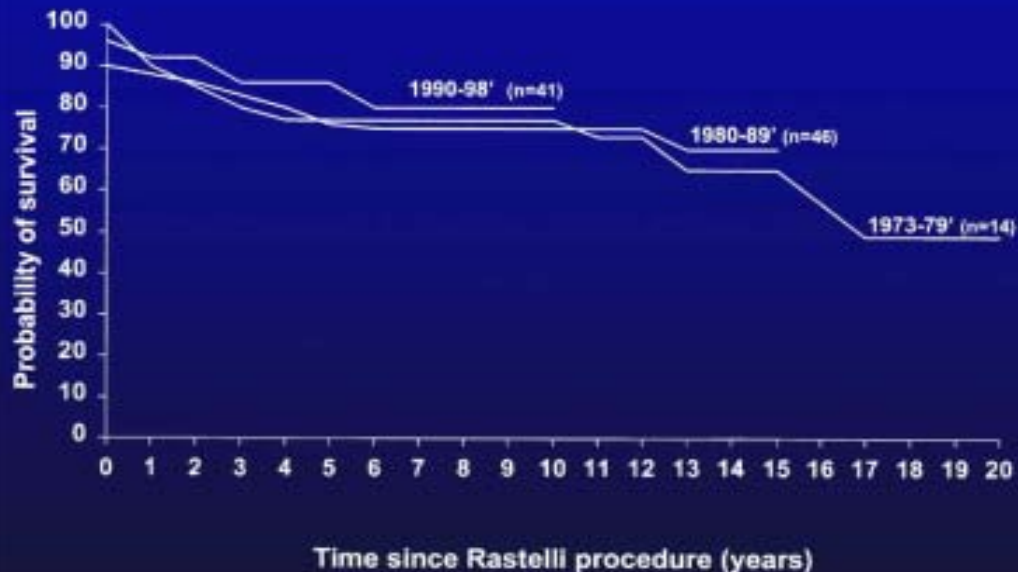
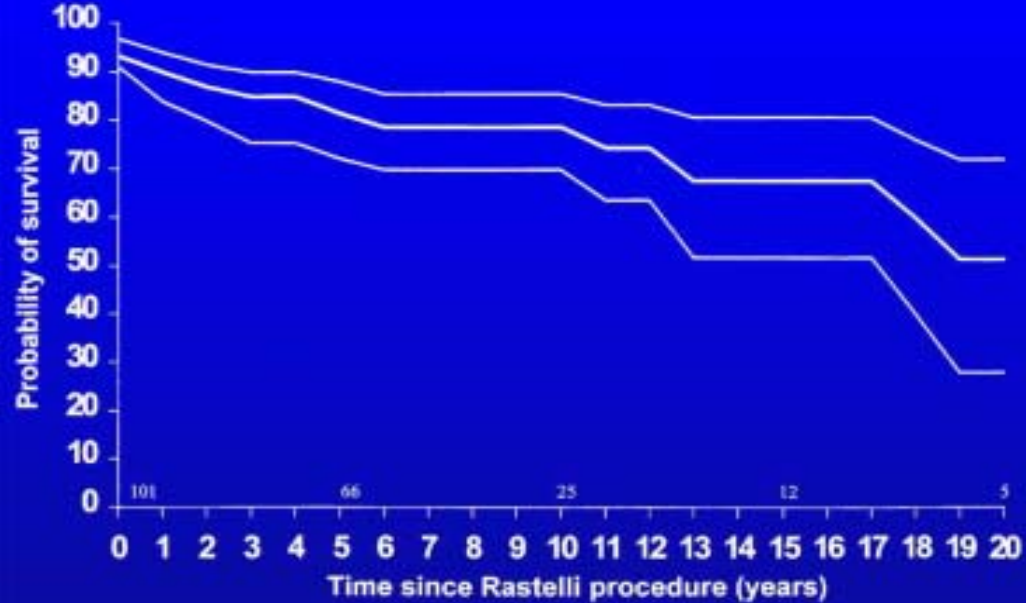
No apparent detrimental early physiologic outcomes to AP strategy

Annulus Preservation provides **improved right ventricular health** – better RVEDD/LVEDD at 16 months follow up

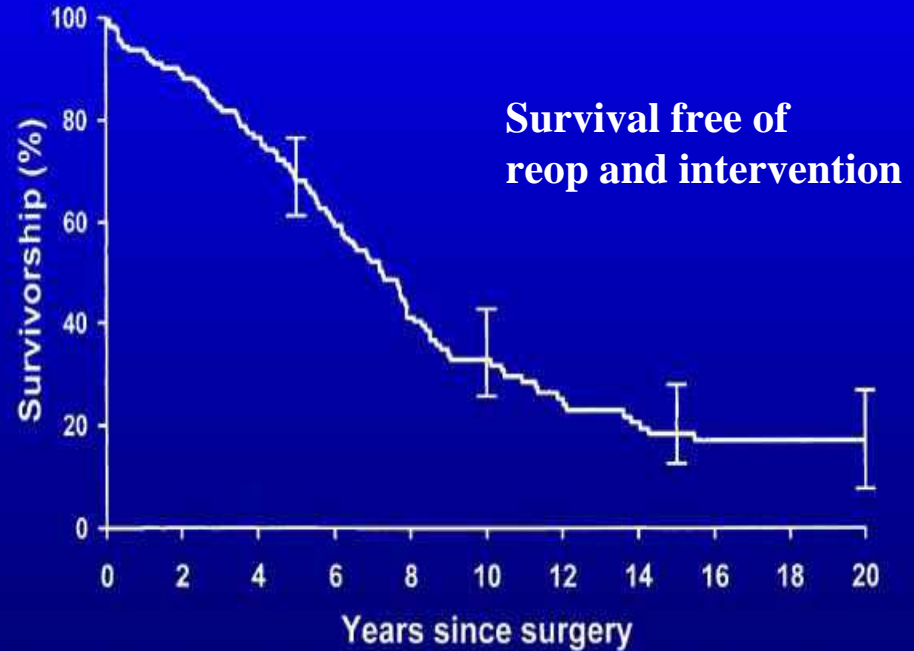
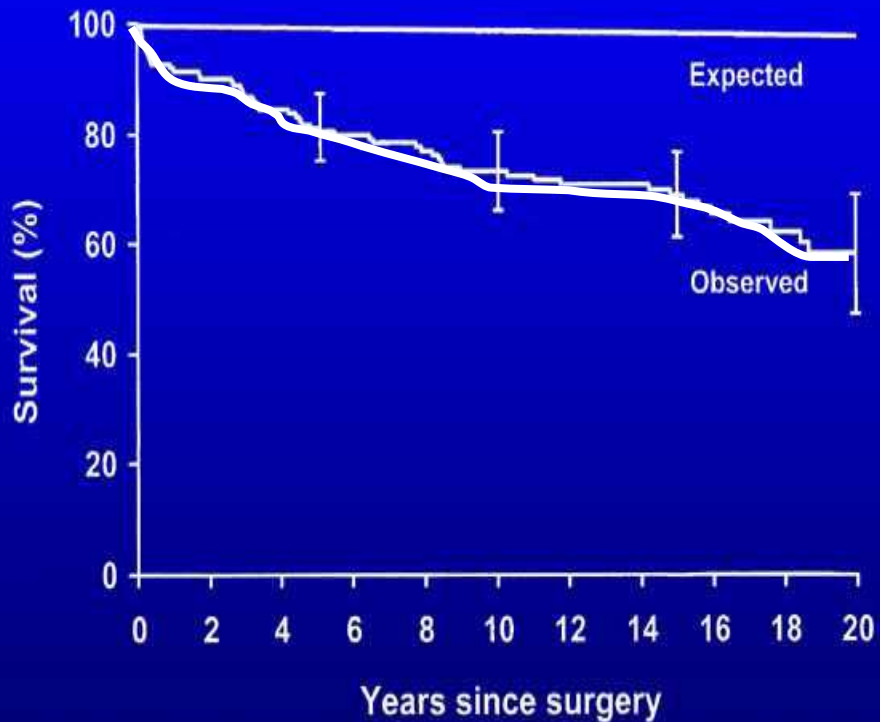
Changing the Tails of Congenital Heart Disease

- Converting tetralogy of Fallot to Pulmonary Stenosis
- Converting d-TGA/VSD/PS to tetralogy of Fallot

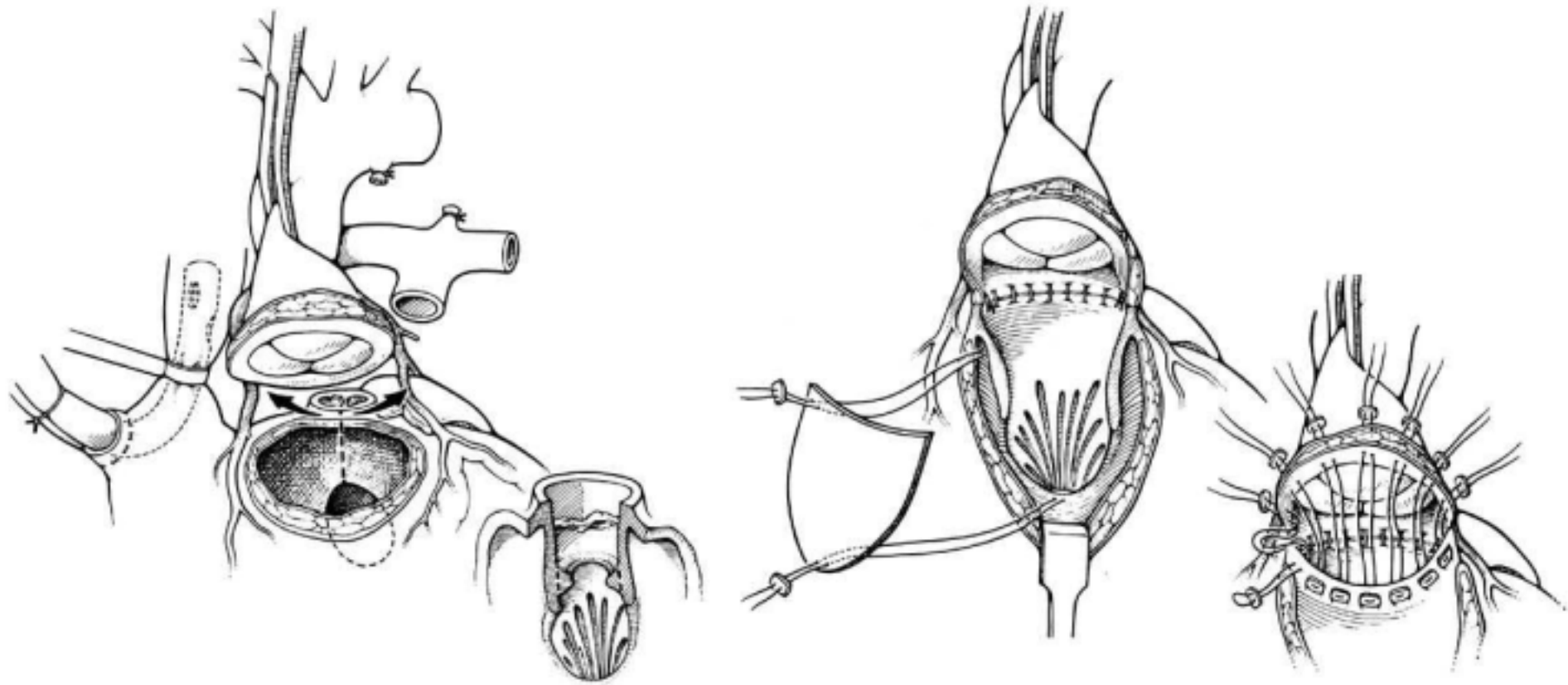
Rastelli Survival



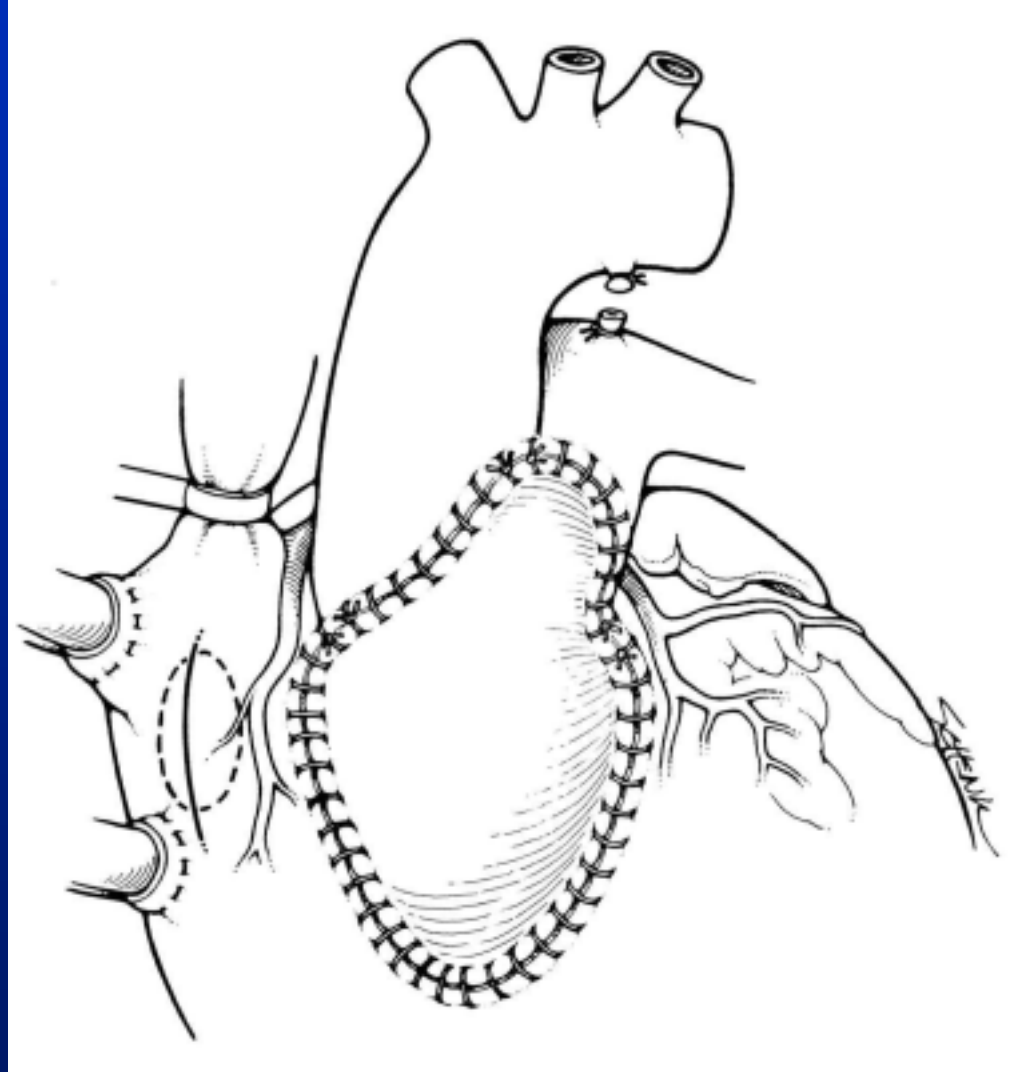
Rastelli Survival Excluding Early Mortality



Nikaidoh Operation for d-TGA/VSD/PS



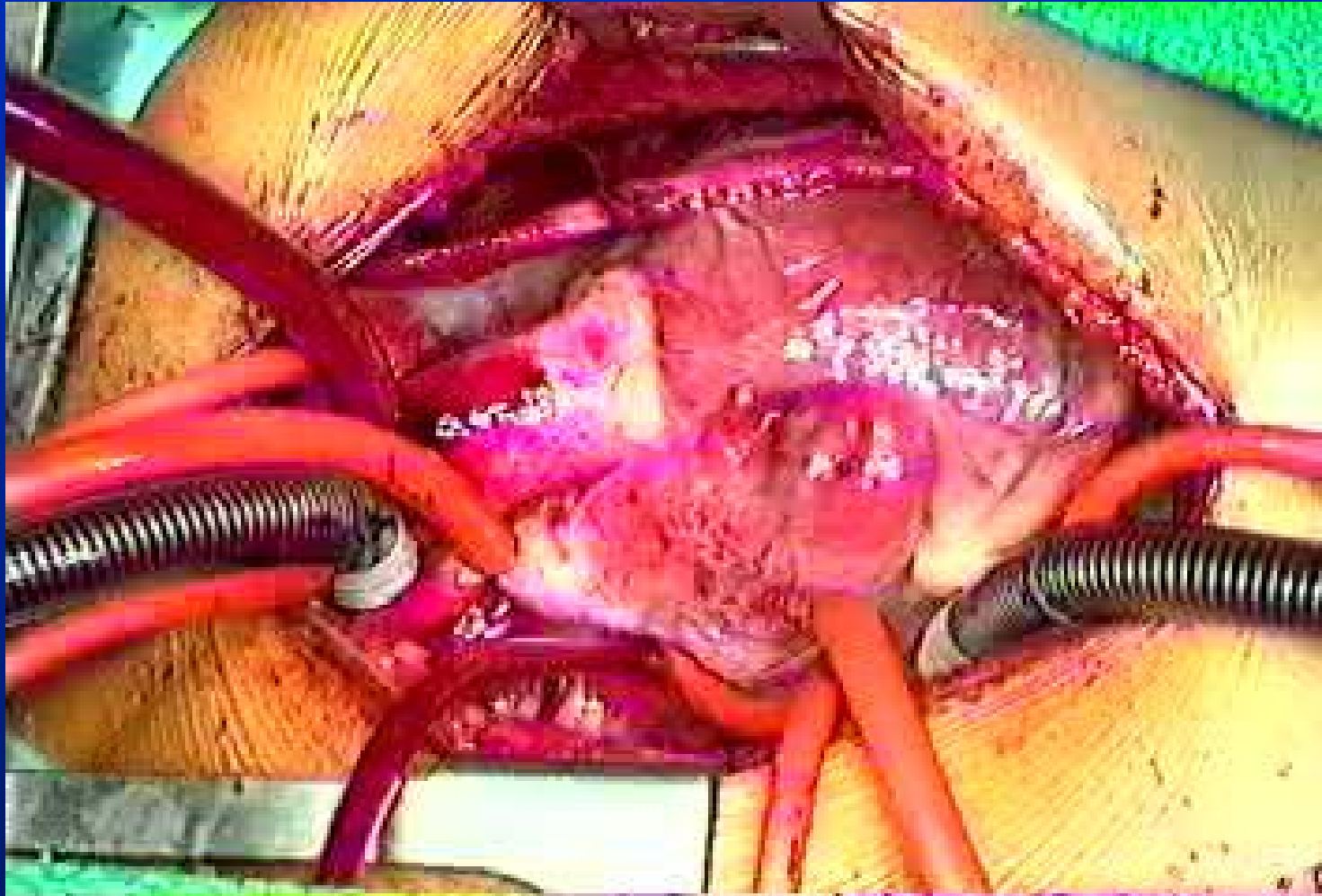
Nikaido Repair d-TGA/VSD/PS



D-TGA/VSD/PS



Nikaido Procedure



Nikaido Postoperative Echo

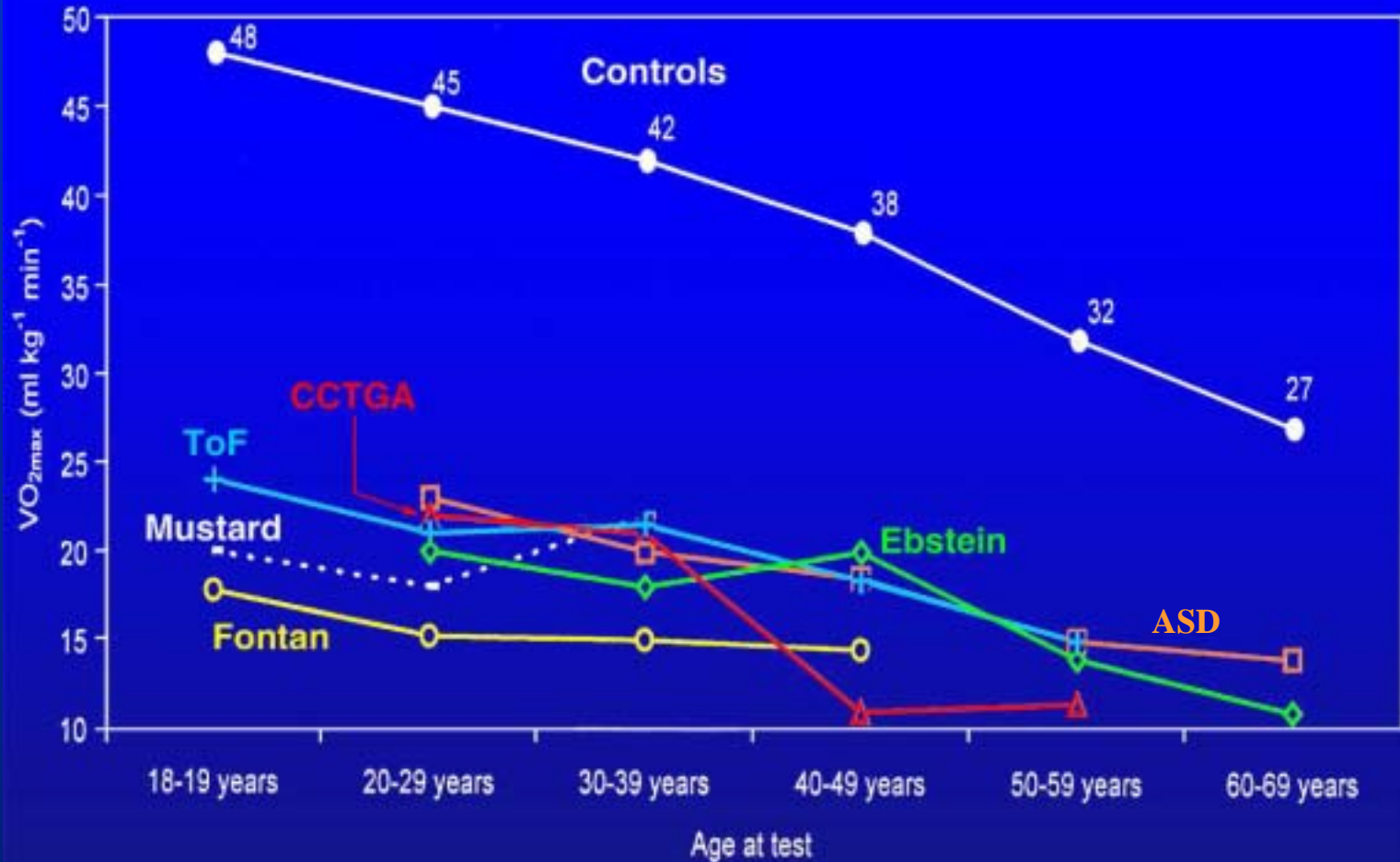


Corrected Transposition

Corrected TGA – Physiologic Repair Survival by Associated Lesions



Exercise Capacity



ASD	18	27	18	16	14
CCTGA	15	8	4	2	
Ebstein	9	11	9	5	3
Fontan	5	30	12	4	
Mustard	16	57	8		
ToF	7	63	50	30	17

CCTGA Surgical Outcomes

Rastelli/Atrial Switch

Tokyo experience 6-89 to 12-00
61 patients

VSD enlargement - 1/2 of patients

Total experience op mortality 8.1%
4 late deaths (16% total)
Generally favourable TV function

CCTGA

Comparison of anatomic vs physiologic n=26/10

Freedom from rhythm disturbance identical:

<u>1</u>	<u>5</u>	<u>10 yrs</u>
92%	82%	61%
80%	67%	67%

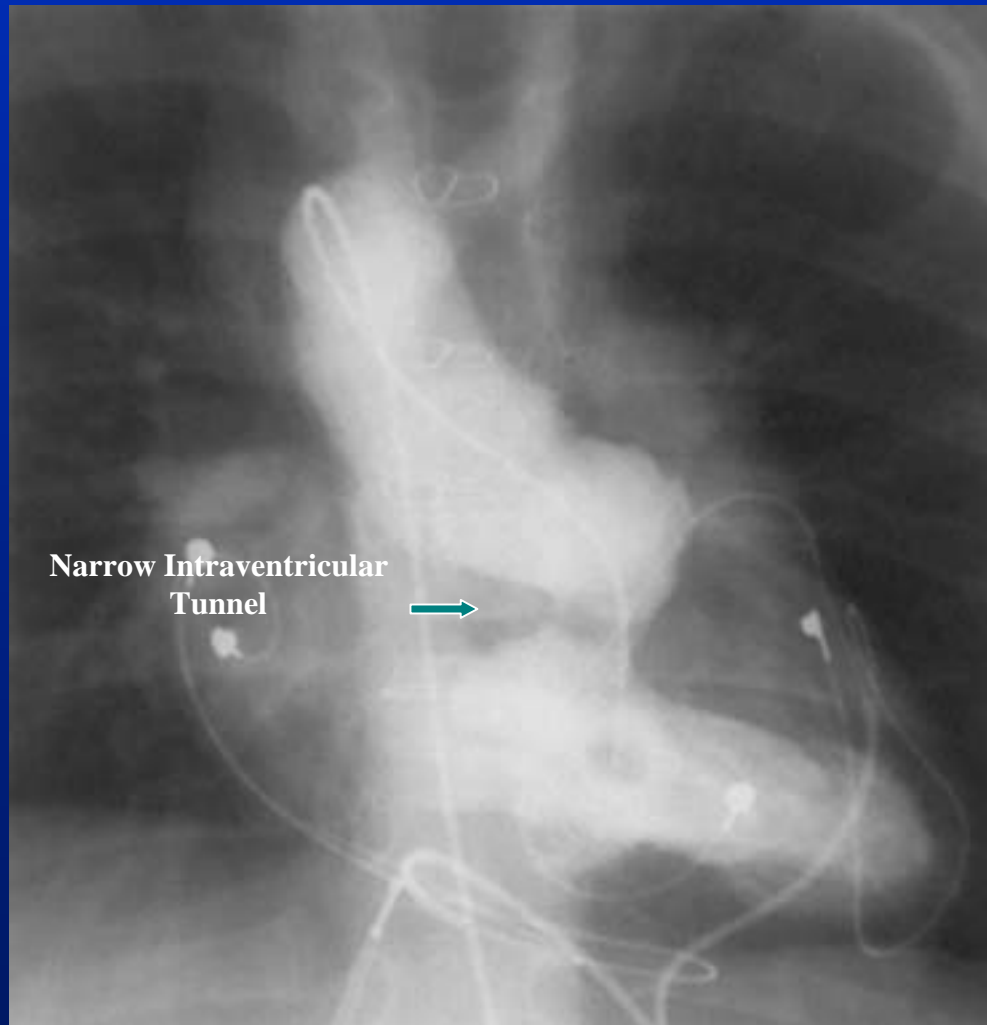
Impaired LV function in about 1/3 of double switches

LV function impairment related to VSD enlargement

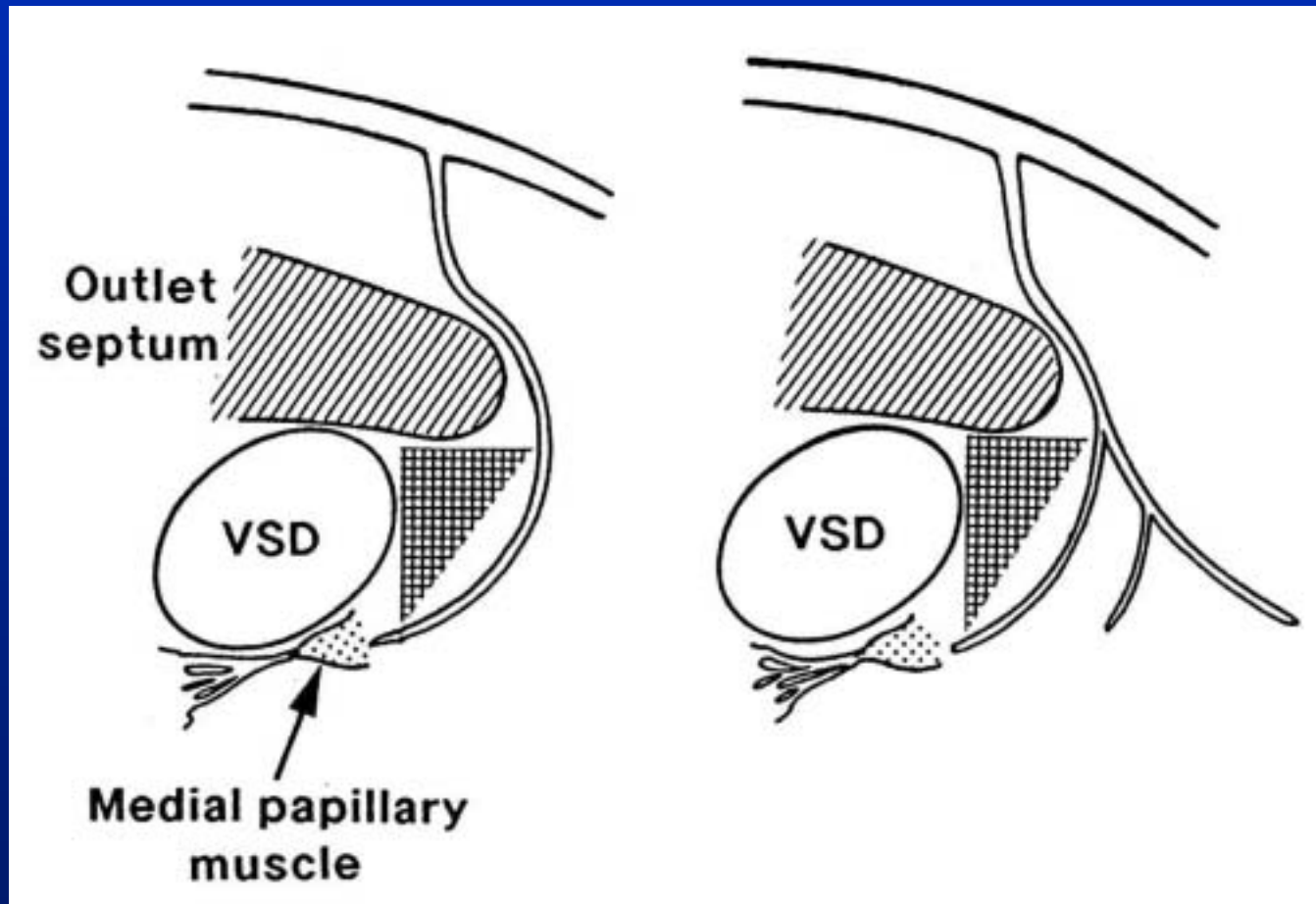
**Is there a
Misconception?**

Why?

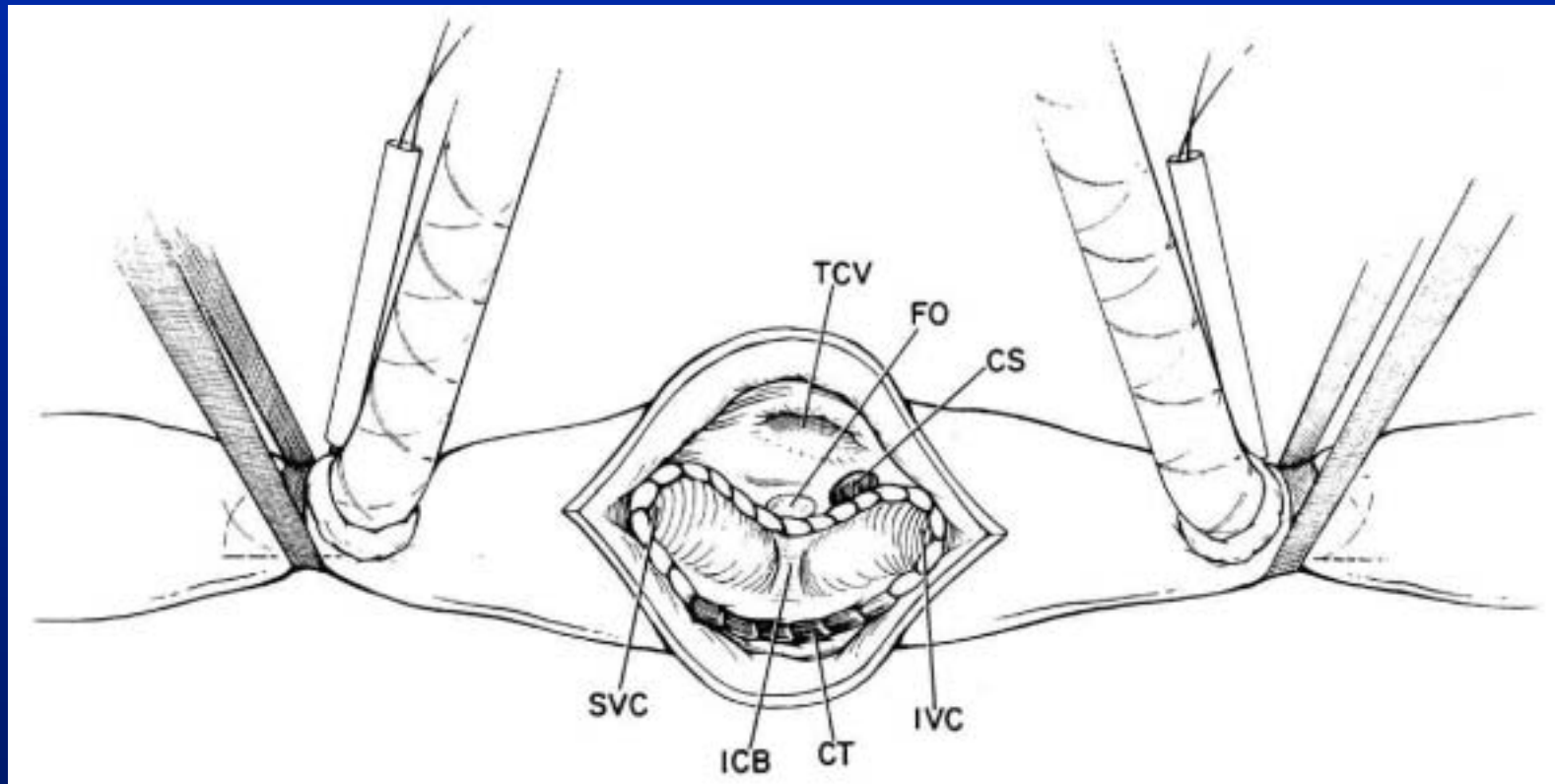
Corrected TGA - VSD



First Septal Artery Course



Spontaneous Atrial Flutter in a Canine Model



Gandhi S et al JACC 1997;30:1095

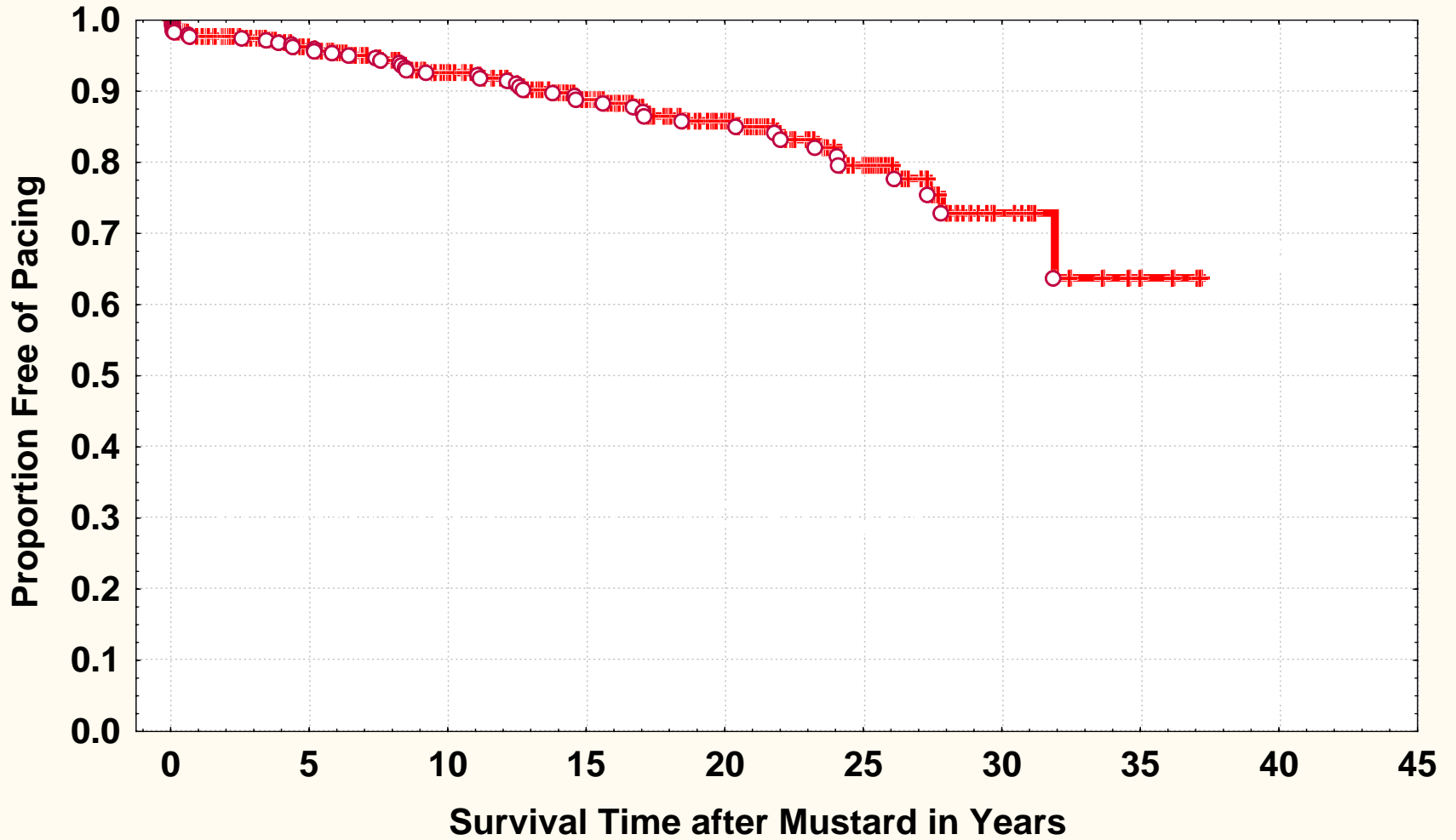
Are the ideas as good as they Seem?

**Best case scenario – ideal
Rastelli
(Conduit changes, LVOTO)**

**Complications with atrial switch
– Rhythm disturbances
Baffle obstruction**

Freedom from Pacing after Mustard Operation

HSC 1963 - 2001 N = 438

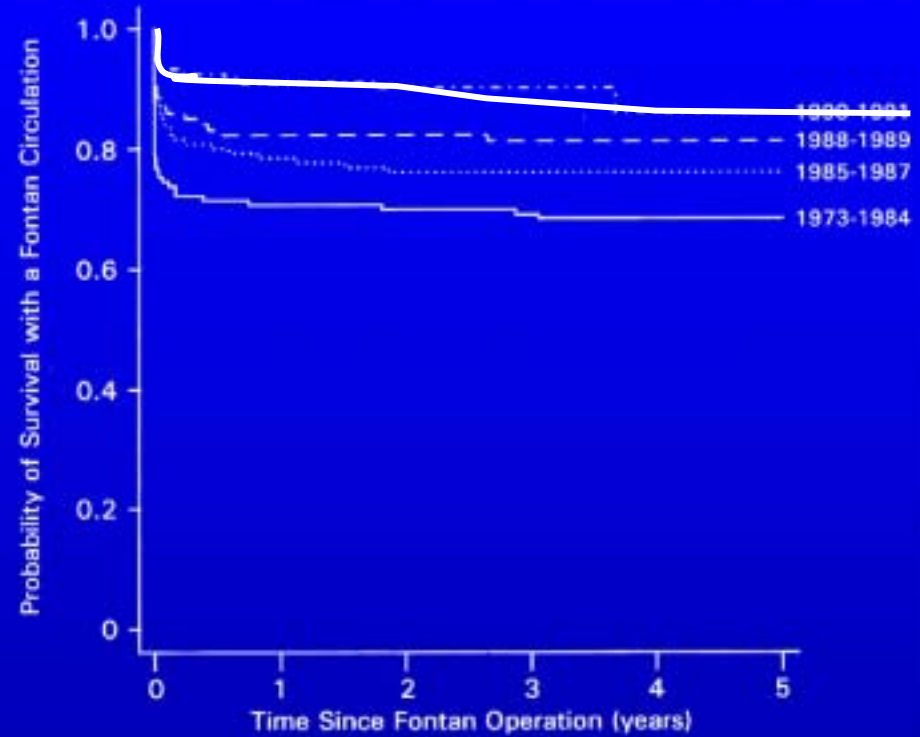
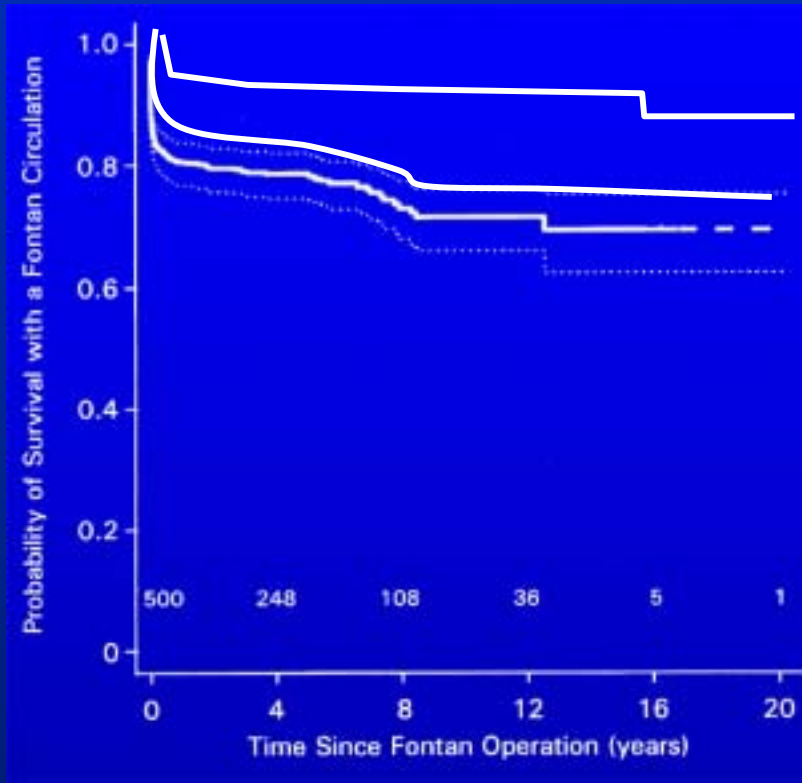


**Houston – We have
a PROBLEM!**

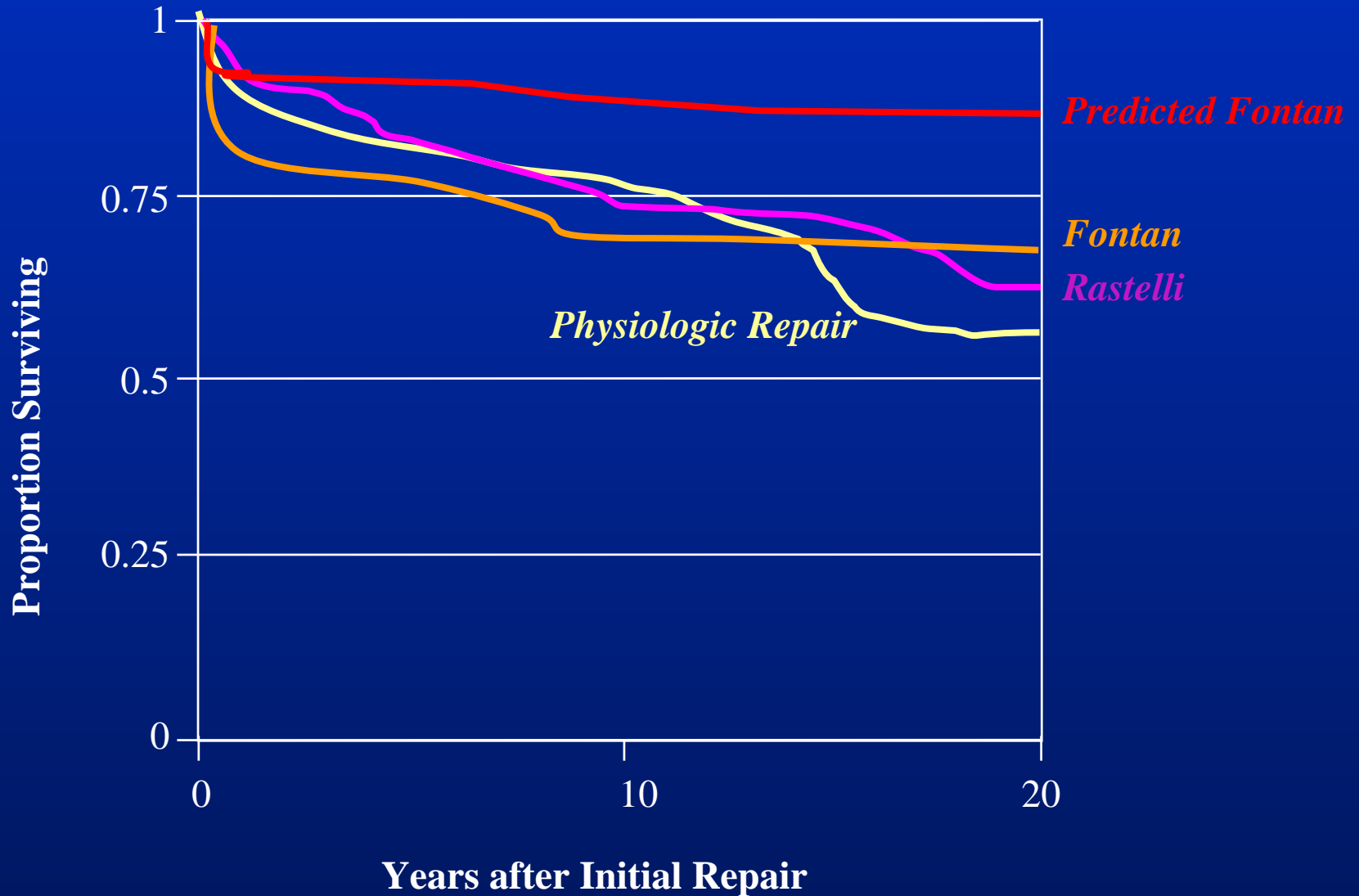
Conception

**Biventricular repair is
always better than a Fontan**

Fontan Outcomes



Tails of Congenital Heart Disease



CCTGA Summary

- **Data Still limited Data**
- **Rastelli comparison most appropriate**
- **Need for multicenter observational study**

Summary

Repaired Congenital Heart Disease
Chronic Disease

The Tails are Long

Overall Summary

Some RVOT obstruction is beneficial in TOF

Aortic Translocation may improve long term outcomes of TGA/VSD/PS

Further analysis of CCTGA is needed

Can We Change the “Tails” of Congenital Heart Disease?

Yes

Change the Tales of repairing the
disease

