

# Indications and Outcomes of the Double Switch in ccTGA

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**No Disclosures**

# ccTGA: The Problem

ccTGA does not fit into neat, clinically discrete sub-groups

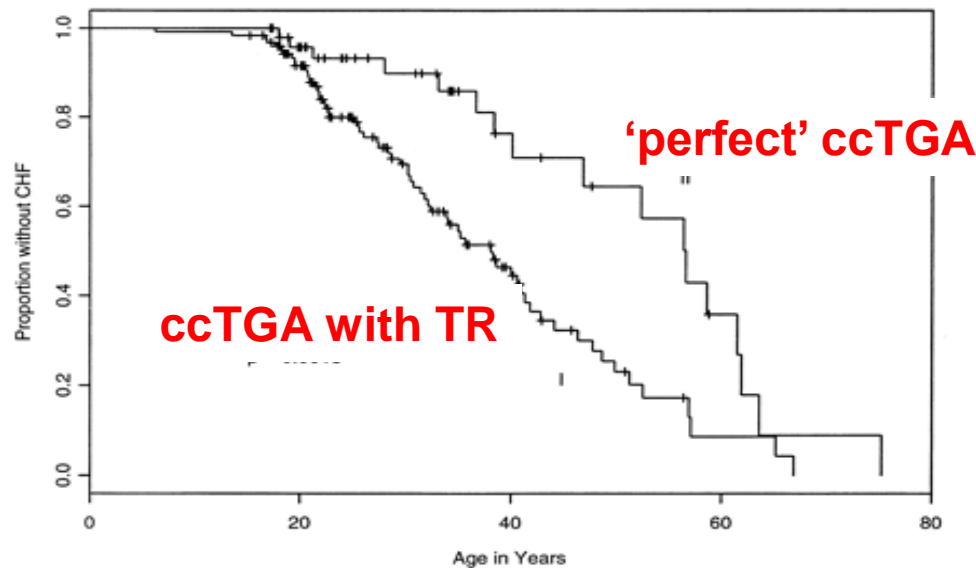
		Frequency	
	Atrial Situs		
Wide range	Normal	80- 85%	
	Inversus	10-15%	
Wide range	Isomeric	2-4%	in age and severity
	Position		
Wide range	Laevocardia	70-75%	Conventional Repair
	Mesocardia	10-15%	Chemical Repair
	Dextrocardia	15-20%	and
	VSD	70-80%	nothing!
	LVOTO	40-80%	
	Pulmonary Atresia	3-8%	
	Arch Hypoplasia/CoA	3-15%	
	Ebsteinoid Tricuspid Valve	10-20%	
	Heart Block	10-15%	
	DORV	3-10%	

# Do Nothing vs Do Something

Natural History of ccTGA in Symptomfree adults

Freedom from CCF

n=168

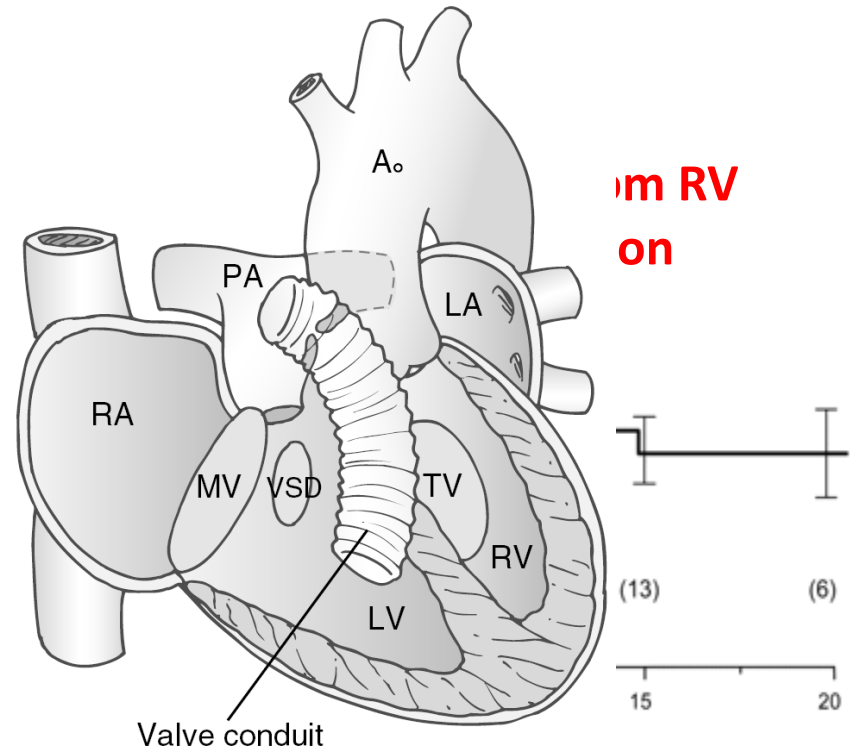
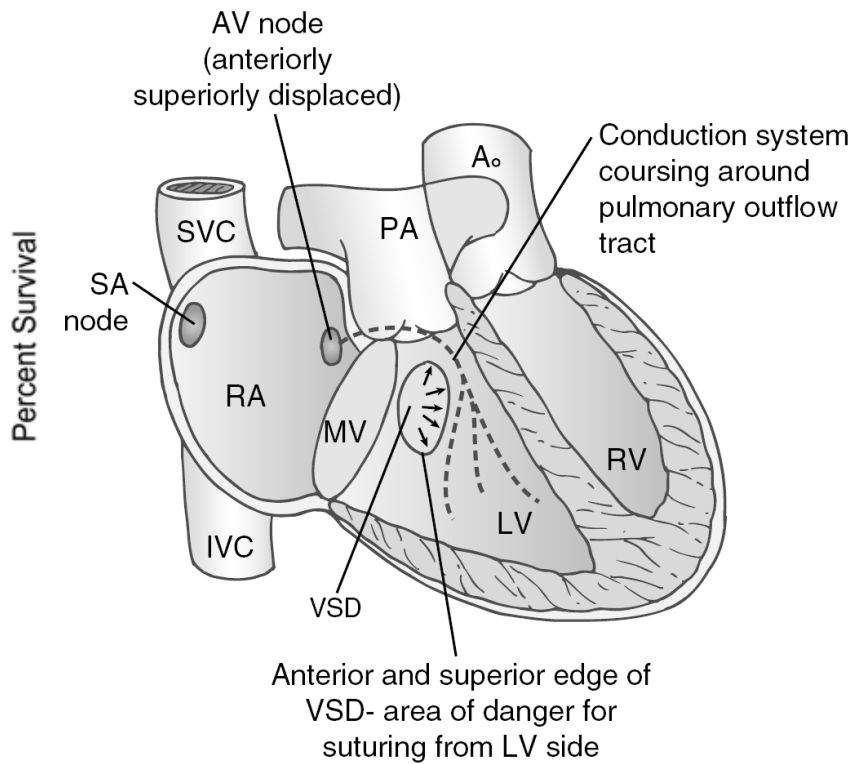


**Even the best patients:  
50% in heart failure within 20y**

JACC 36: 255, 2000

JTCVS 117: 1190, 1999

# Conventional repair vs Anatomical repair



**Boston n=123**

**JTCVS 129: 182, 2005**

**Paris:**

**Conventional repair : All attempts at TV repair failed.**

**80% of all patients needed TVR within 5 years**

**Anatomical repair: All attempts at TV repair succeeded.**

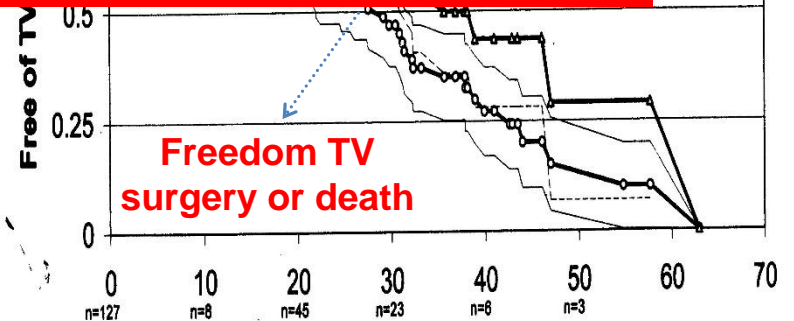
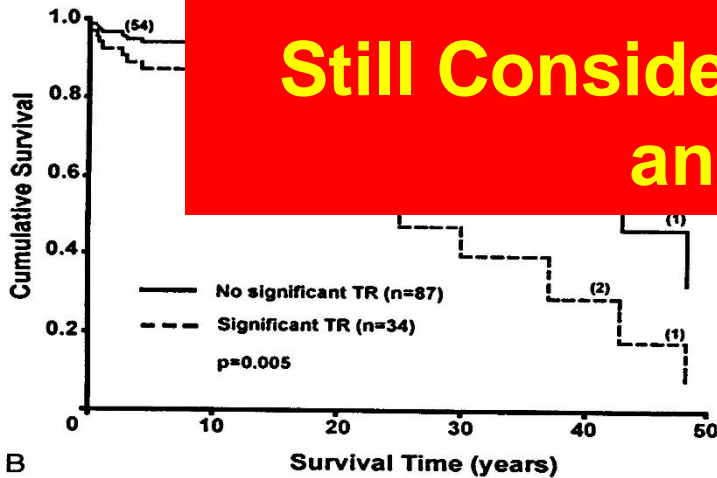
**Heart 80: 479, 1998**

# Conventional ('Physiological') Repair

Texas

**Generally disappointing outcomes  
but  
Still Consider if RV function good  
and  $\leq$  mild TR**

n=121



Mean age 9 m at entry point

TR  $\geq$  mod at time of surgery STRONGEST risk facto

20 year mortality 50%

# Choice of Anatomical Repair

**Normally developed LVOT  
+/- VSD**

Acyanotic

**Small/Stenotic LVOT  
With VSD**

Cyanosis (variable in degree)

**DOUBLE-SWITCH  
DS**

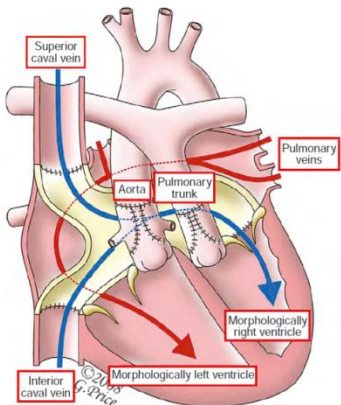
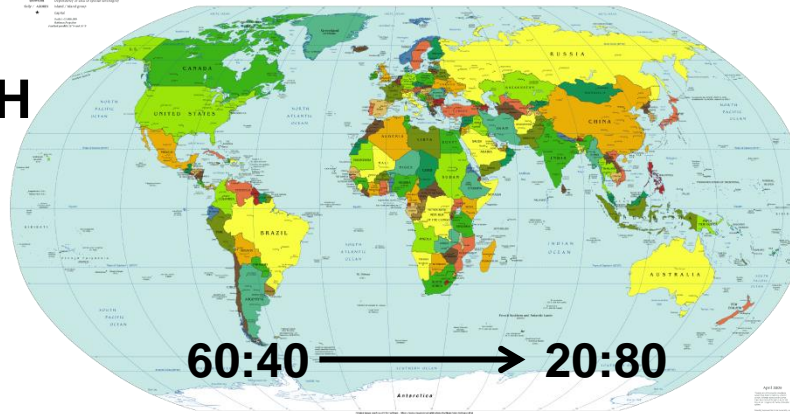


Figure 39-28 The cartoon shows the steps involved in the so-called double-switch procedure.

Political Map of the World, April 2006



**RASTELLI-SENNING  
RS**

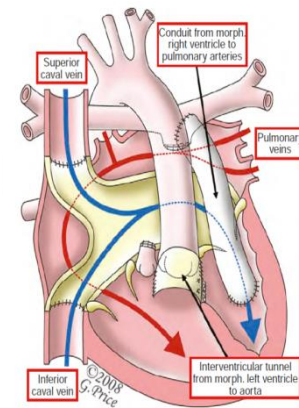
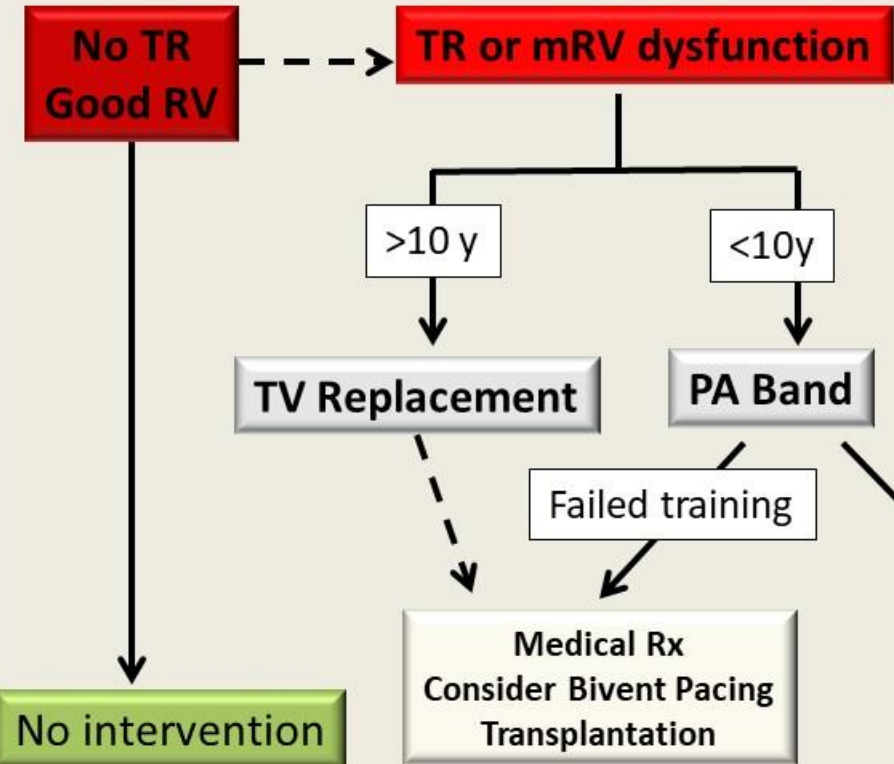
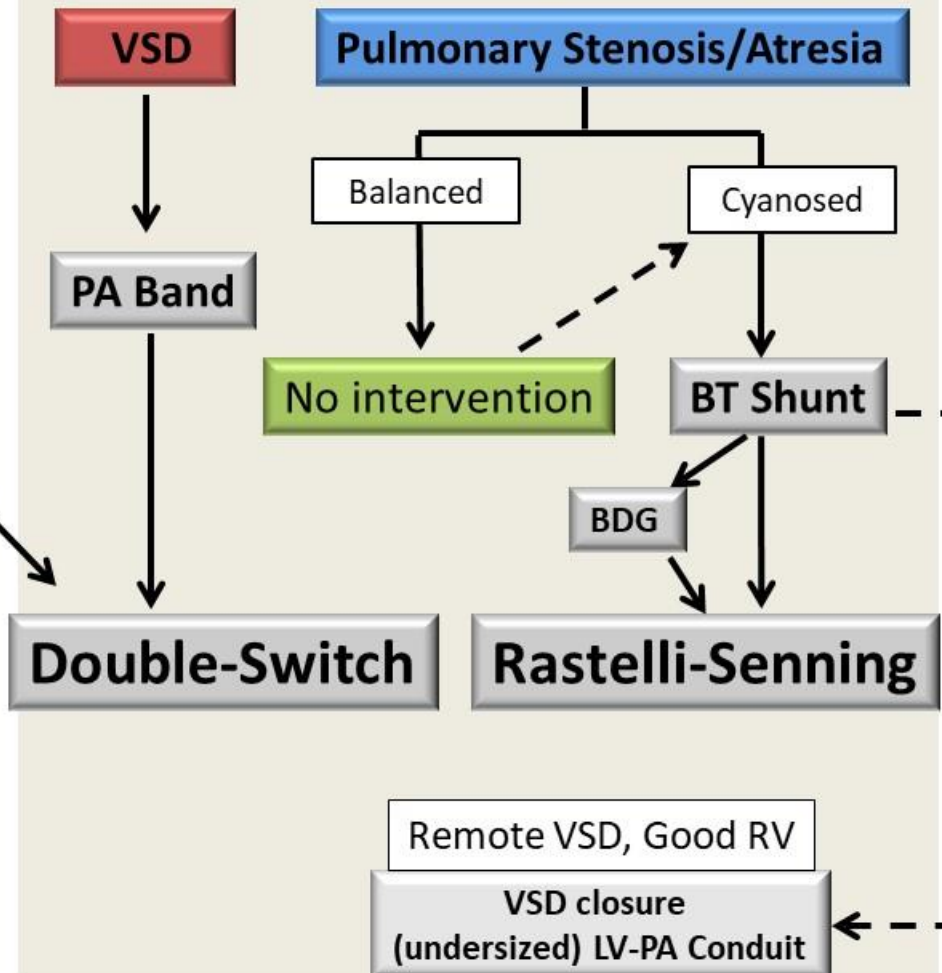


Figure 39-29 The cartoon shows the end-result after an atrial redirection procedure combined with intraventricular rerouting of the ventricular septal defect to the aorta, and placement of a conduit from the morphologically (morph.) right ventricle to the pulmonary arteries.

No Associated Abnormalities

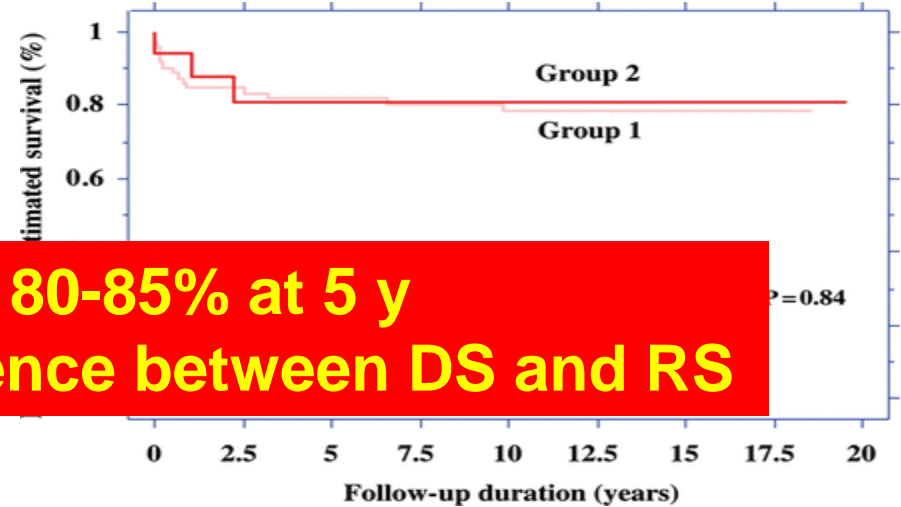
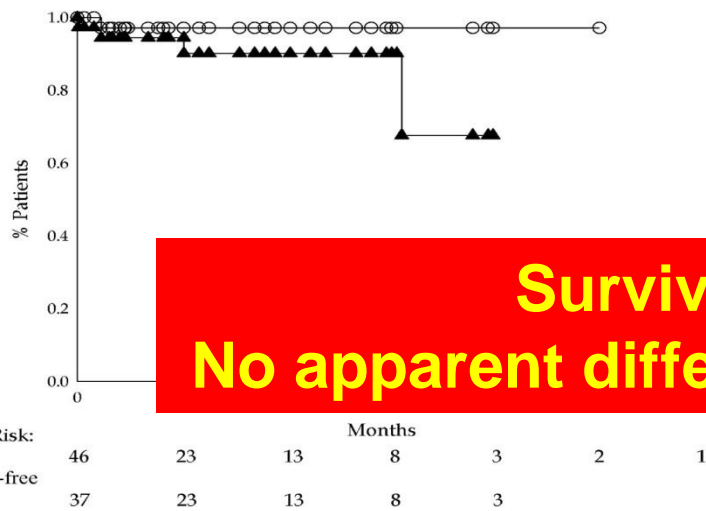


Associated Lesions





# Early & Mid-Term Outcome



**Survival 80-85% at 5 y**  
**No apparent difference between DS and RS**

**Cleveland Clinic n=46 60% DS**

**Tokyo n= 90 20% DS**

**EJCTS 24:11-20, 2003**

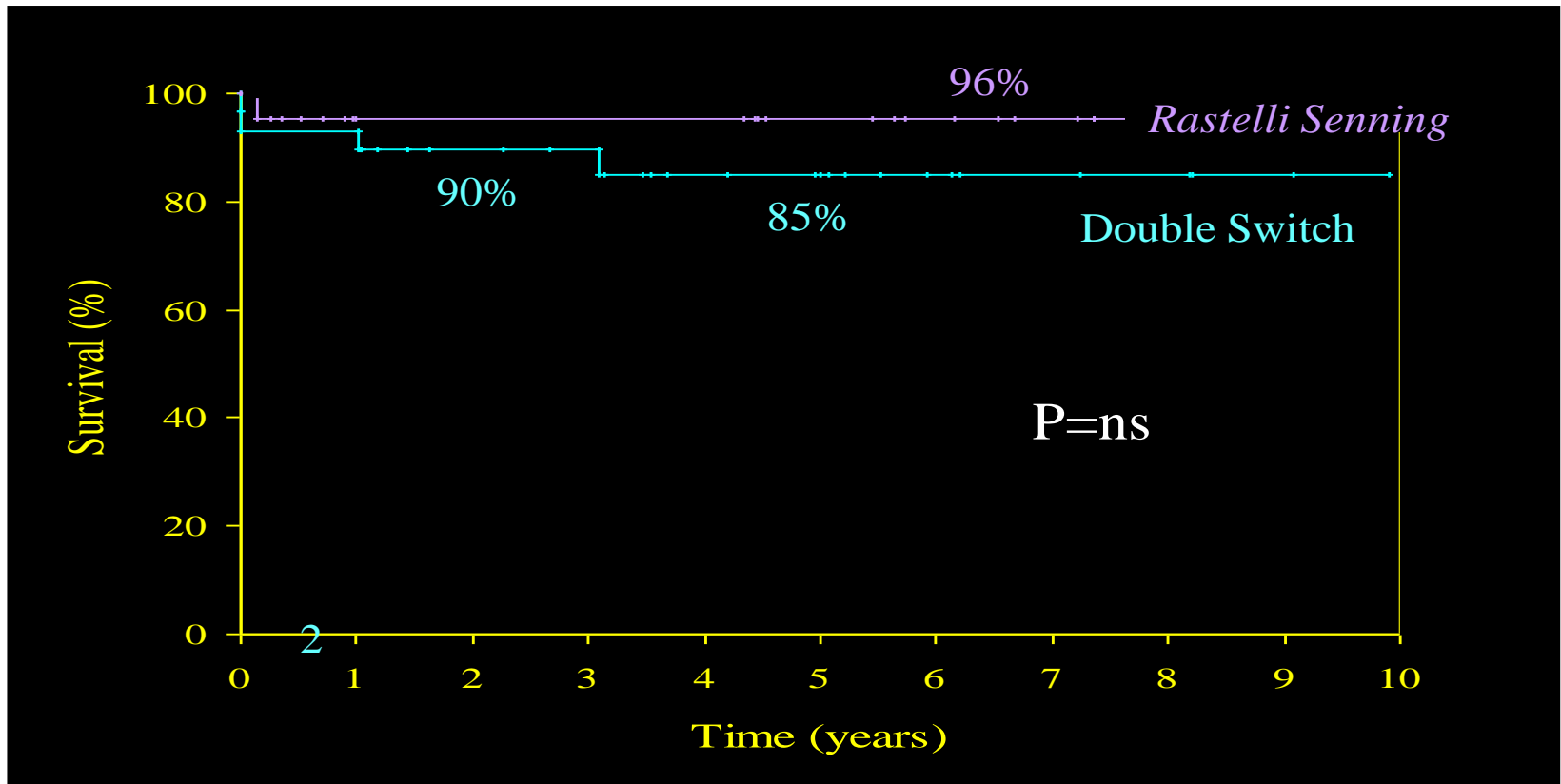
**EJCTS 42:1004, 2012**

# Early & Mid-Term Outcome

0-10% Early Mortality across all series

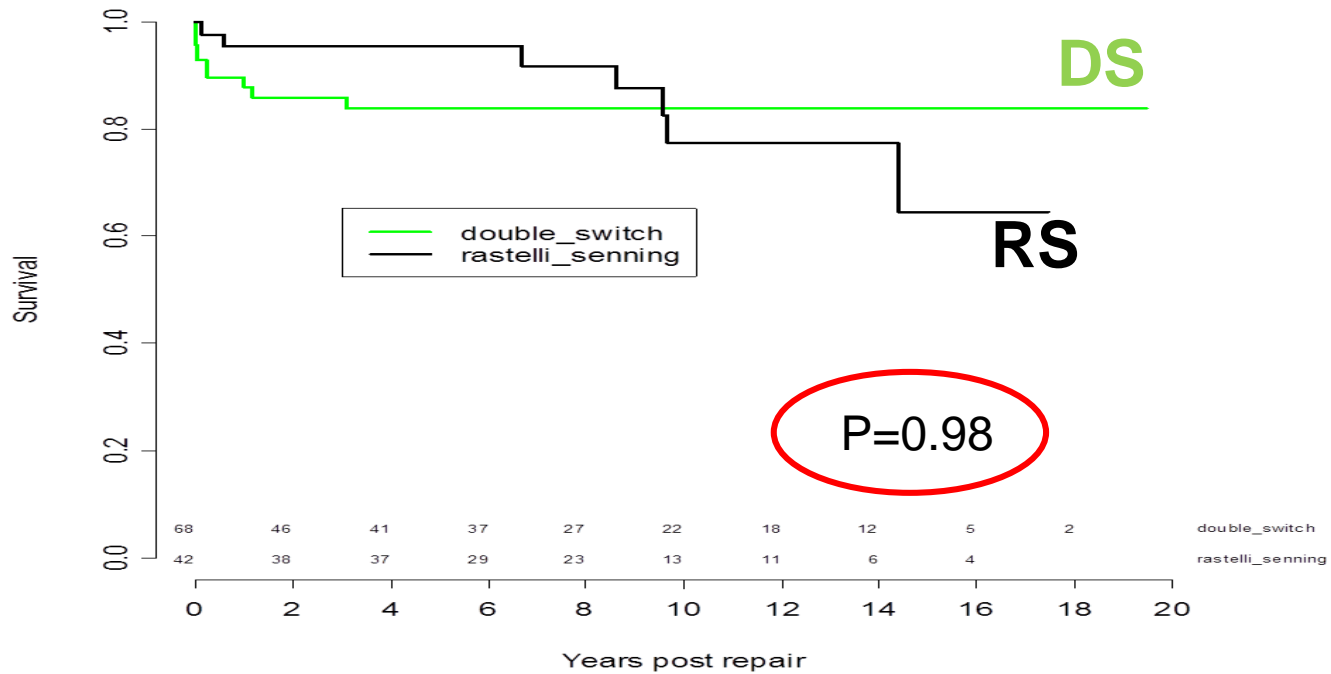
No difference worldwide in DS vs RS early outcomes

Birmingham n=113



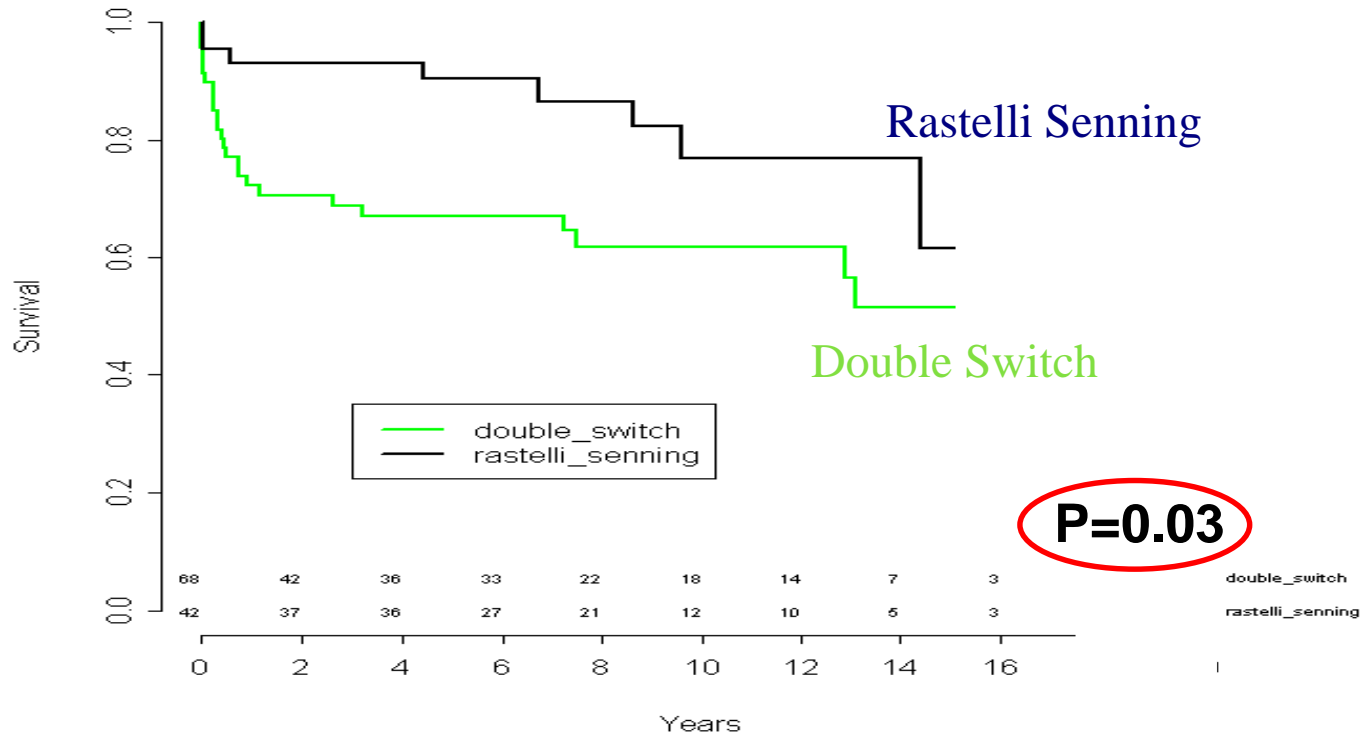
# Late Survival

Birmingham n=113



JTVCS 142: 1348, 2011

# Freedom from Death/Transplantation/Poor mLV Function

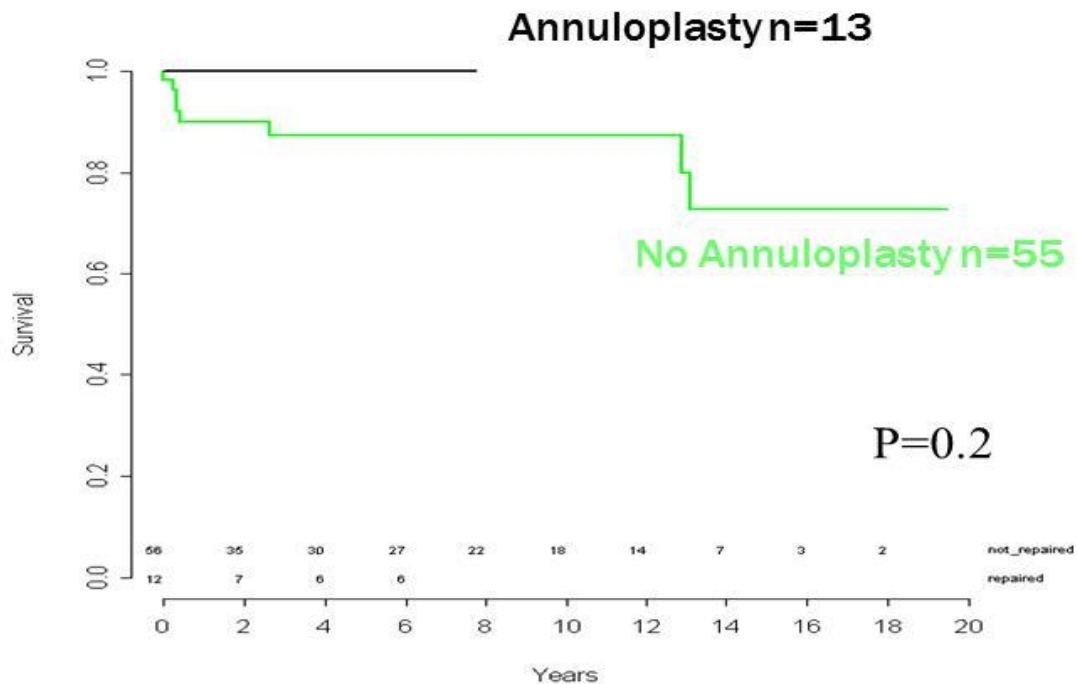


# Aortic Incompetence at 20 years

	DS	RS
≥ Mild AI	40/58 (70%)	8/38 (21%)
≥ Mod AI	6/58 (10%)	0/38 (0%)
AV Replacement	6/ 58 (10%)	1/38 (3%)

# Impact of Aortic Root Annuloplasty

## Freedom from $\geq$ mild AI or AVR

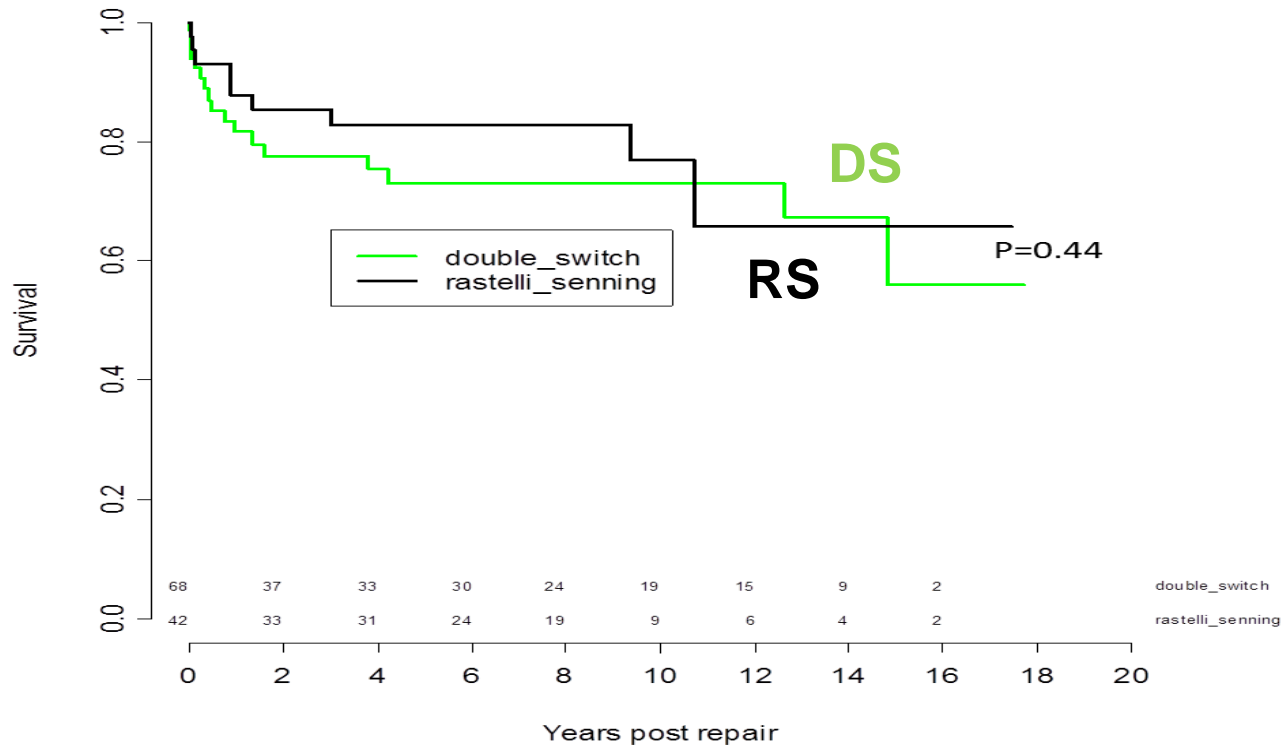


# Reinterventions

Revision- Pacing procedures excluded

	DS	RS
AVR	6	1
MV repair	1	0
TV repair	3	0
RF ablation for Aflutter	4	0
Multi-site pacing	3	0
Residual VSD	3	0
LVOTO resection	0	4
Senning Pathways	7 (3 balloon)	5 (4 balloon; 1 stent))
Pulmonary Arteries	11 (3 balloon/stent)	8(5 balloon)
RVOT enlargement	2	0
RV-PA conduit	n/a	14 (2 balloon)

# Freedom from Reintervention





# Poor mLV Function at Follow-Up

15% developed late mLV dysfunction during follow-up

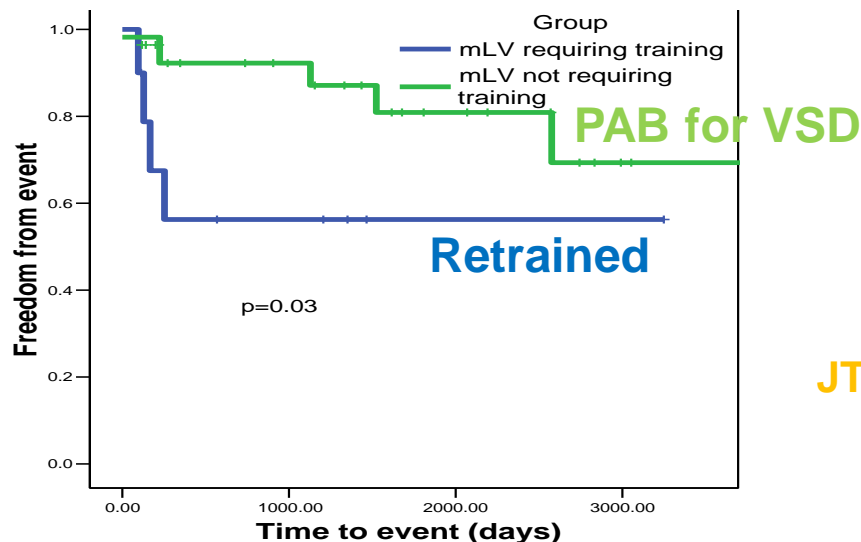
**ALL were in the DS group**

Not associated with aortic regurgitation

Not associated with 'High Risk' Group

## Impact of PA Banding to *Retrain* the mLV

Survival with good LV function



Median age at banding 6 y

JTCVS 135: 1137, 2008

# Late Outcomes - Boston

## Boston Data

n=25 're-trained' mLVs

**20% LATE dysfunction: *all* in cases banded at >2 y  
linked to longer duration of 'training' >6/12**

(all had good function early post-op)

In the entire cohort, n=106

Late dysfunction and AI associated with older age (>10 y at repair)

Significant benefit of RESYNCHRONISATION (biventricular pacing)

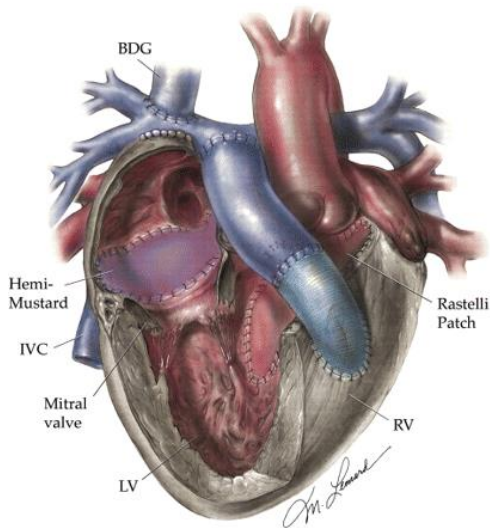
## Risk of late mLV dysfunction

**Banding > 2y  
Double Switch >3 y**

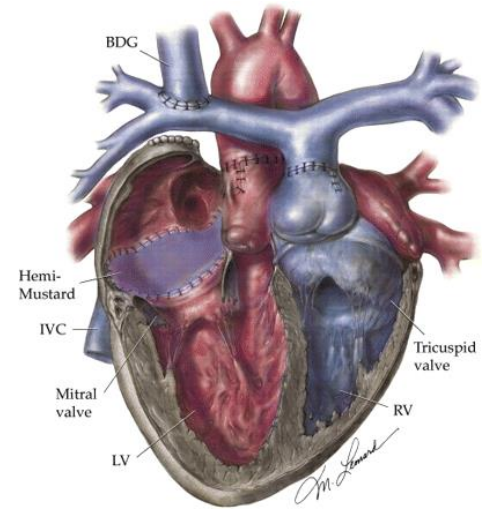
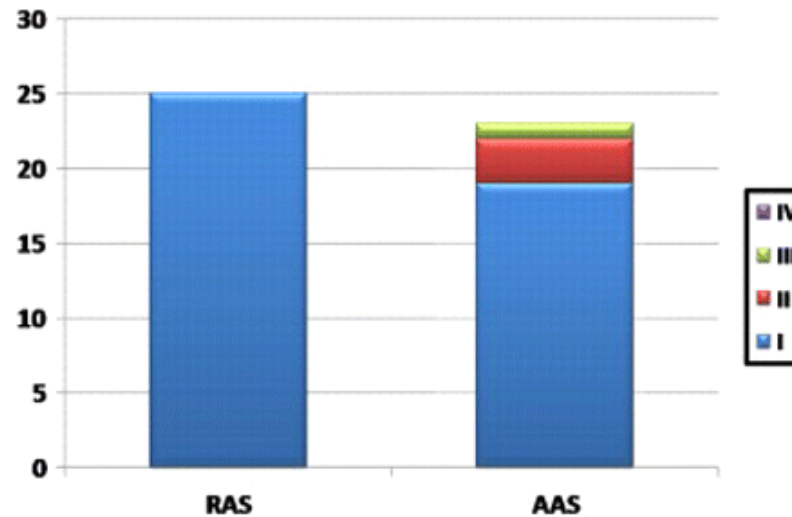
JTCVS 147: 537, 2011

ATS 96: 603, 2013

# One-and-a-Half Repair - Stanford



Prolong Conduit life  
Technically easier



n= 48. One early death and no late deaths.

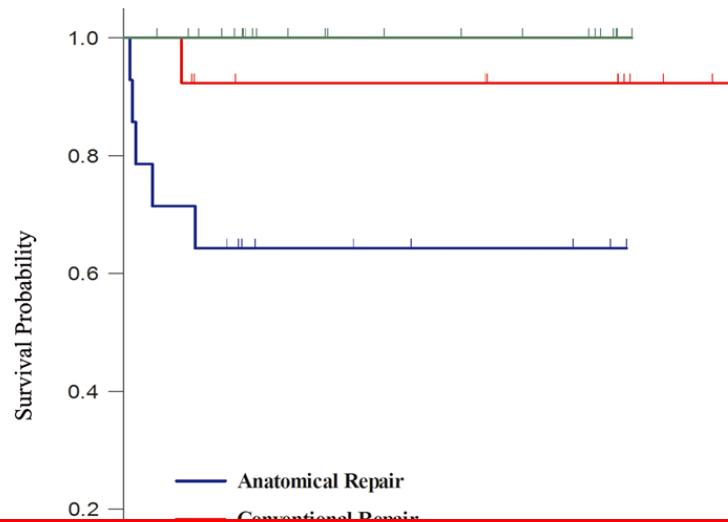
**13% have sig impaired LV function - all were in the DS group**

Reop rate no different between RS and DS

JTCVS 141:162, 2010

# The Counter-Argument

Taiwan 1995-2012: n=56



**Fontan can give good outcomes for some patients (eg remote VSD)  
But not good if impaired RV or >modTR**

No. at risk	Survival Times (years)			
	0	1	2	3
Conventional Repair	13	9	7	2
Anatomical Repair	14	5	3	0
Single Ventricular Palliation	23	12	8	0

EJCTS 49: 522, 2016

# Early prophylactic pulmonary artery banding in isolated congenitally corrected transposition of the great arteries<sup>☆</sup>

Olivier Metton<sup>a</sup>, Régis Gaudin<sup>a</sup>, Phalla Ou<sup>b</sup>, Sébastien Gerelli<sup>a</sup>, Shafi Mussa<sup>a</sup>, Daniel Sidi<sup>c</sup>, Pascal Vouhé<sup>a</sup>, Olivier Raisky<sup>a,\*</sup>

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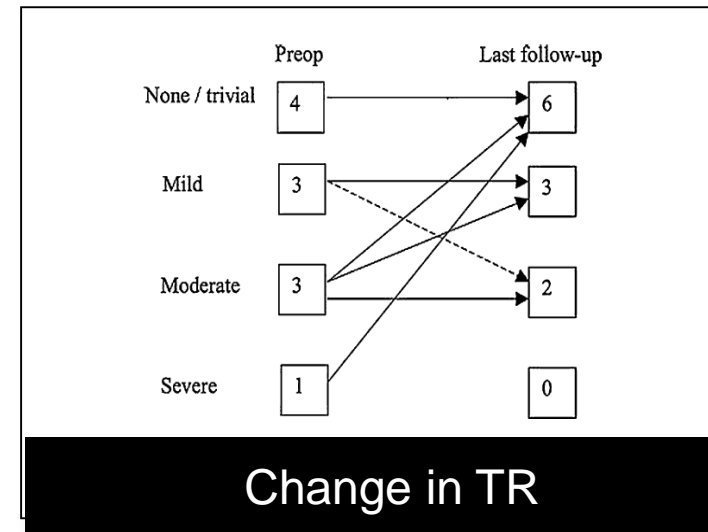
<sup>c</sup>Department of Pediatric Cardiology – University Paris Descartes and Necker Sick Children Hospital, Paris, France

Received 7 October 2009; accepted 25 March 2010

n=11 Asymptomatic infants

## ? Banding as Destination Therapy

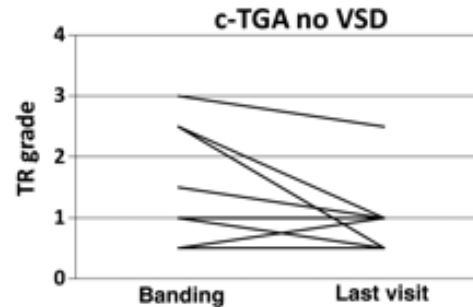
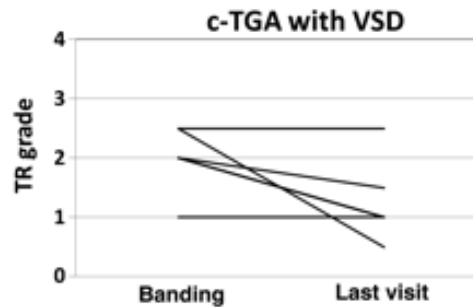
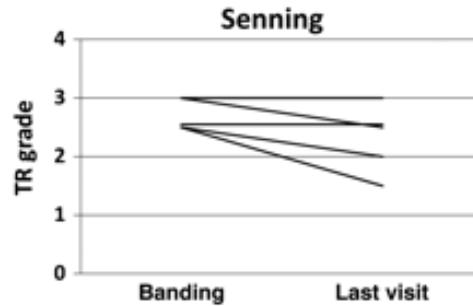
EJCTS 38: 728, 2010



“The aim of this early prophylactic PA banding is not to offer a double-switch procedure for all patients but to preserve an optimal cardiac status”

# PA Band: Open-Ended Palliation?

Lueven



n=20

Median F/U 7 years

Sustained improvement in TR

EJCTS 41: 913 2012

# Conclusions

- . Anatomical Repair offers the best long term outcomes.....  
BUT we are still learning who will benefit most
- . Significant early and late reinterventions.
- . Late Follow-up:
  - . Late mLV dysfunction is the concern.  
? Related to mLV re-training
  - . Aortic regurgitation is important in the DS group
  - . High risk groups have very rewarding outcomes
- . Overall survival is still >85% for all-comers at 15-20 y

# Changing Indications

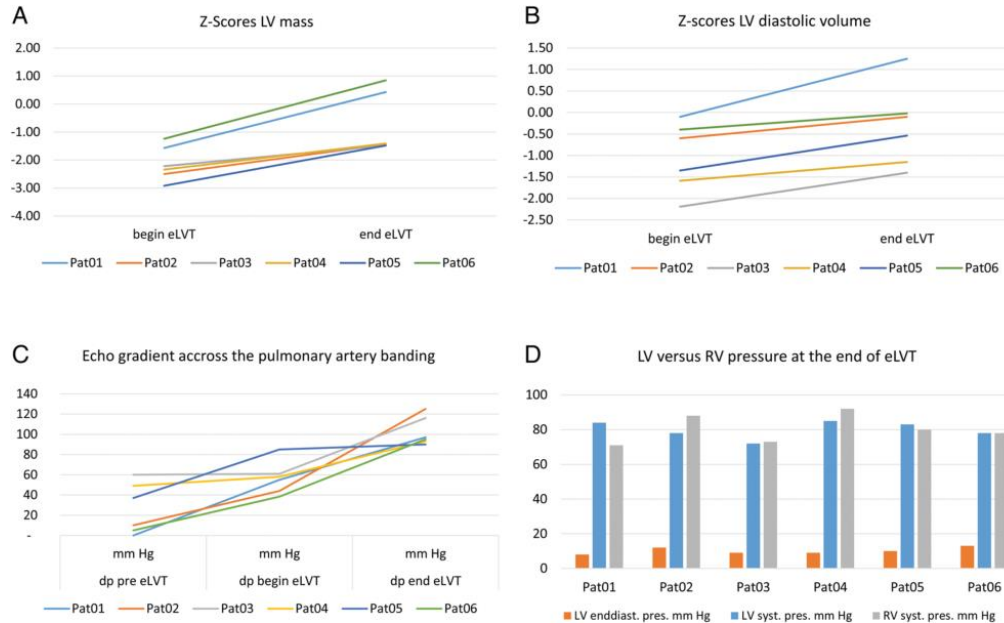
- . Should we be banding Earlier?
- . Have we underestimated the risk of re-training  
( ? Maximum age 3 y ?)
- . Role of Enhanced training (create ASD and better assessment)
- . Role of Banding as definitive treatment?
- . Role of the 1 ½ repair?
- . Need to be more selective to identify who will benefit most
- . Some patients (eg remote VSD, borderline size RV)  
may do better with Fontan



# Enhanced Retraining

PA Banding AND creation of ASD to volume load the mLV

n=6



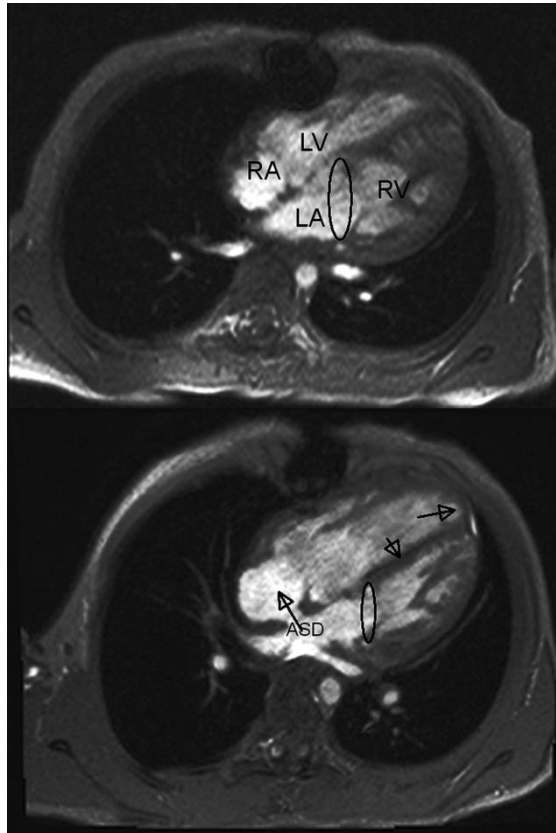
Double-Switch after median 1.2 y training

Median Age at DS 3.8 (1.4-6.8) y

EJCTS 49: 1571, 2016

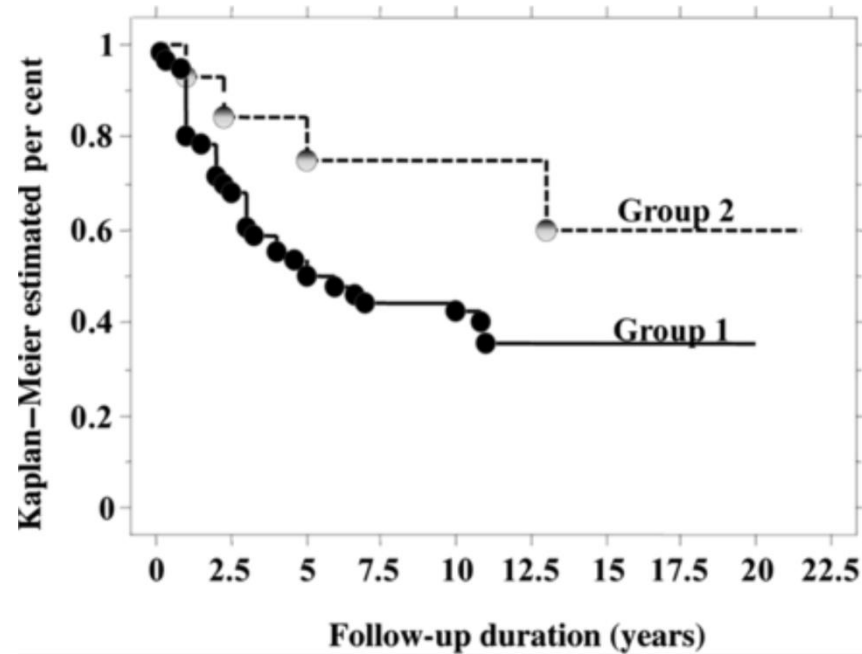


# Enhanced Retraining





Tokyo paper again – freedom from all cardiovascular events



From: **Long-term prognosis of double-switch operation for congenitally corrected transposition of the great arteries†**

Eur J Cardiothorac Surg. 2012;42(6):1004-1008. doi:10.1093/ejcts/ezs118

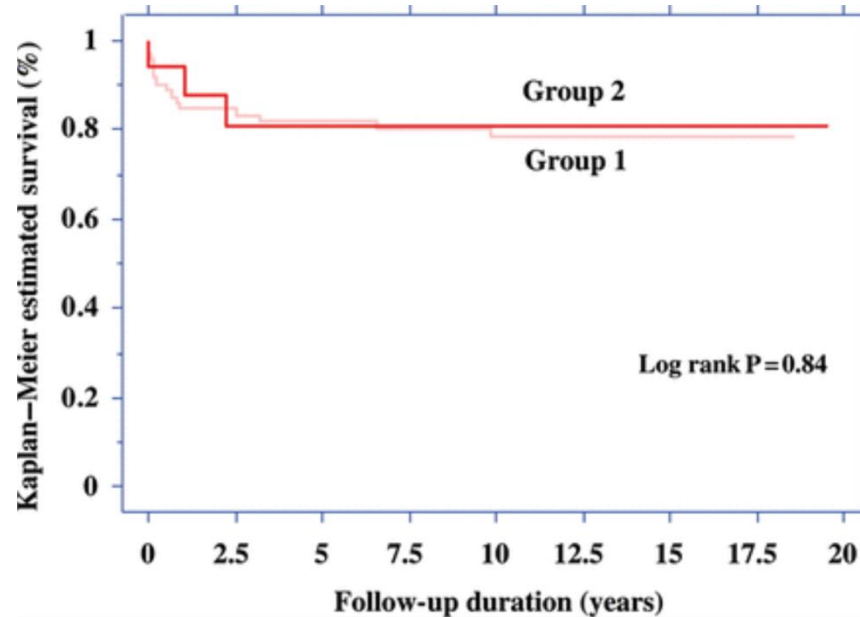


Figure Legend:

Long-term survival curve. The Kaplan-Meier actuarial survival rate including hospital and late mortality at 20 years was similar.



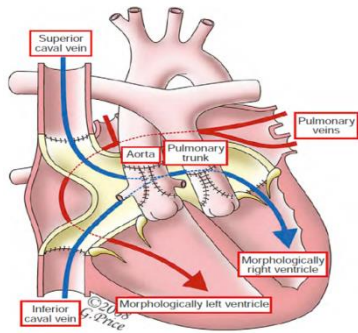


Figure 39-28 The cartoon shows the steps involved in the so-called double-switch procedure.

# Double Switch

VS

# Rastelli Senning

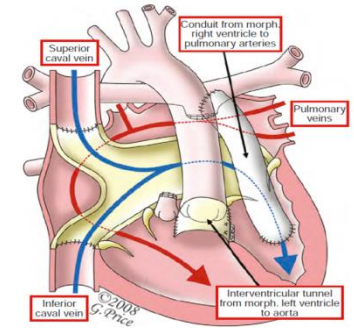


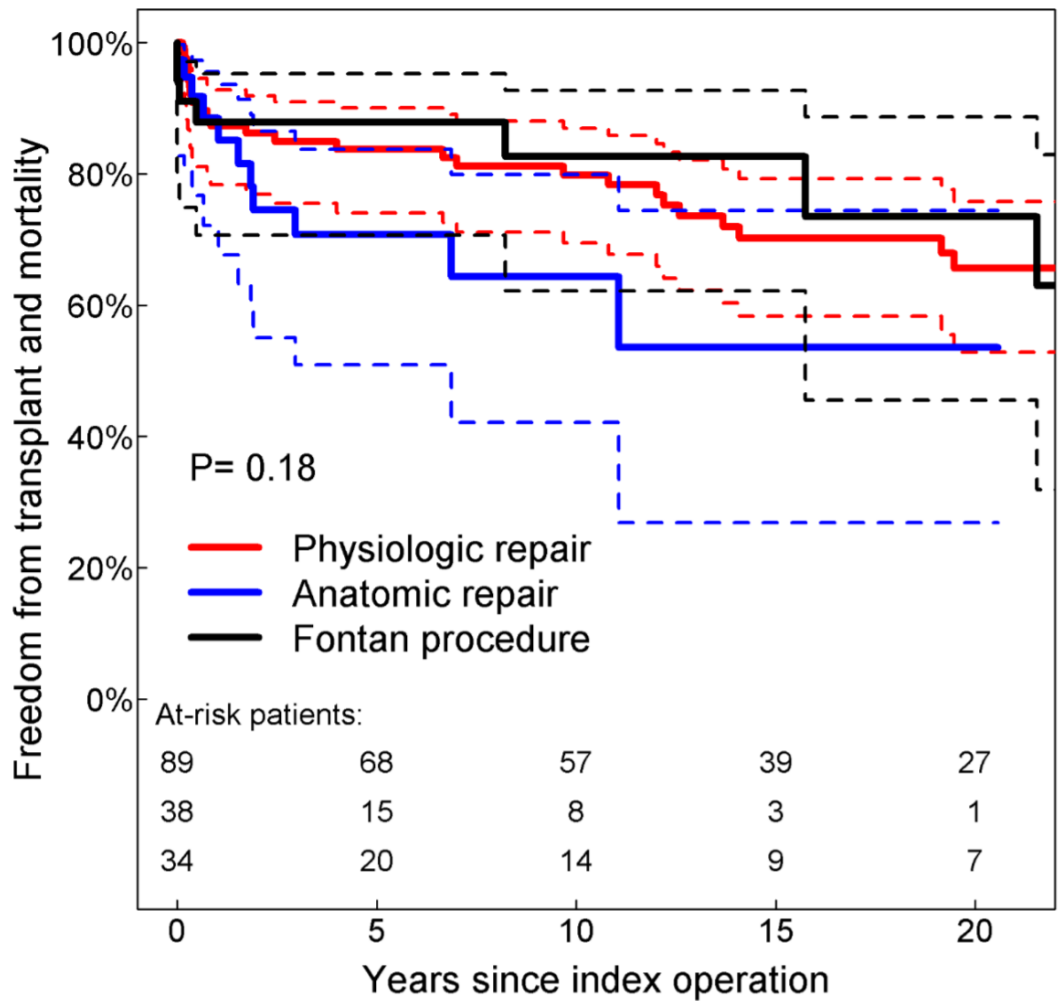
Figure 39-29 The cartoon shows the end-result after an atrial redirection procedure combined with intraventricular routing of the ventricular septal defect to the aorta, and placement of a conduit from the morphologically (morph.) right ventricle to the pulmonary arteries.

Arterial switch  
 Coronary problems  
 Aortic Root distortion/AR

VS

Rastelli  
 LVOT Distortion/Stenosis  
 Conduit problems

....but it's more subtle than that:  
 Pre-operative state  
 Preparation of the mLV  
 Tricuspid valve function  
 Age at operation  
 High-risk presentation



Toronto –JTCVS but ? Still under review. Experience over 30 years, suggests fontan as good as other options

# Long Term Outcome:

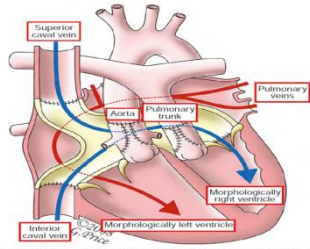


Figure 39-28 The cartoon shows the steps involved in the so-called double-switch procedure.

Double  
Switch

vs

Rastelli  
Senning

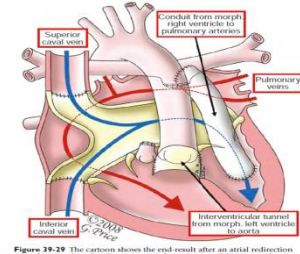


Figure 39-29 The cartoon shows the end result after an aortic redirection procedure, combined with intraventricular remodeling of the ventricular septal defect to the aorta, and placement of a conduit from the morphologically (morph.) right ventricle to the pulmonary arteries.

What do we expect?

- .Re-intervention - More in the RS group
- .Late Aortic Regurgitation – More in DS group
- .Late mLV dysfunction – variable ? More in DS