

# *Cardiopatie congenite complesse*

*Piccoli cuori crescono : impariamo a conoscerli e a gestirli*



**CARDIOLOGIA E CARDIOCHIRURGIA  
PEDIATRICA E DELL'ETA EVOLUTIVA  
CENTRO PER IL TRATTAMENTO DEL  
CONGENITO ADULTO  
UNIVERSITA DI BOLOGNA  
POLICLINICO S ORSOLA MALPIGHI  
M.Bonvicini**

**BAD NEWS: EVERYONE GETS OLDER.....**



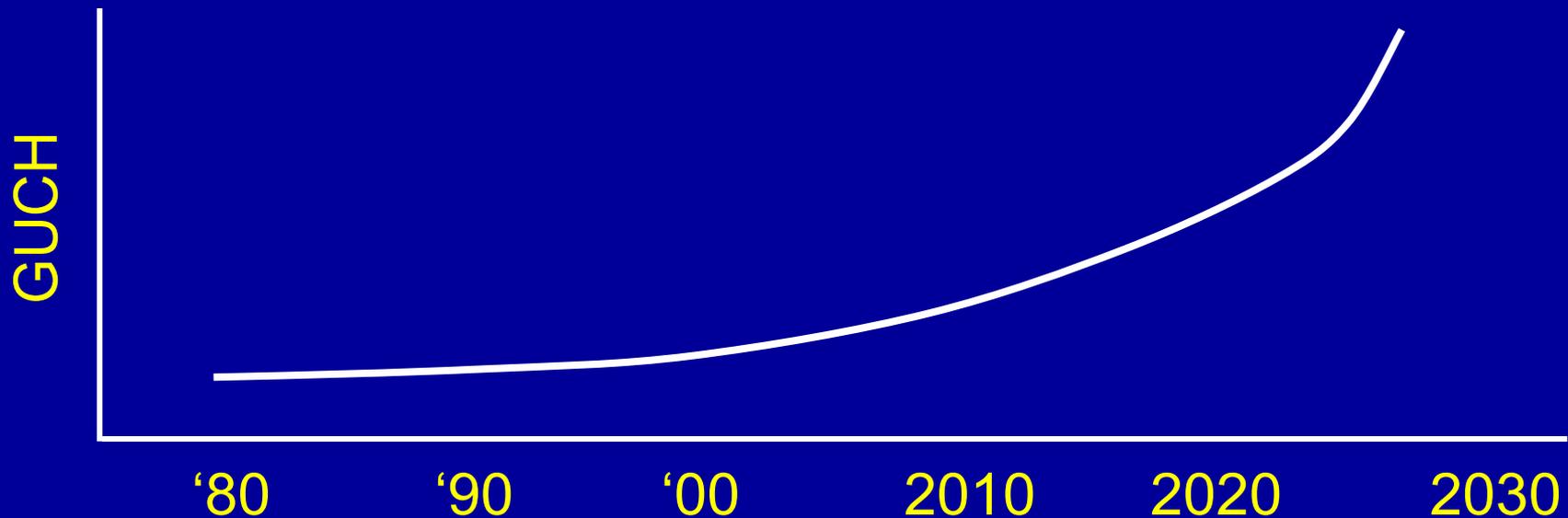
# Il follow-up del cardiopatico congenito adulto

## Dimensione del problema

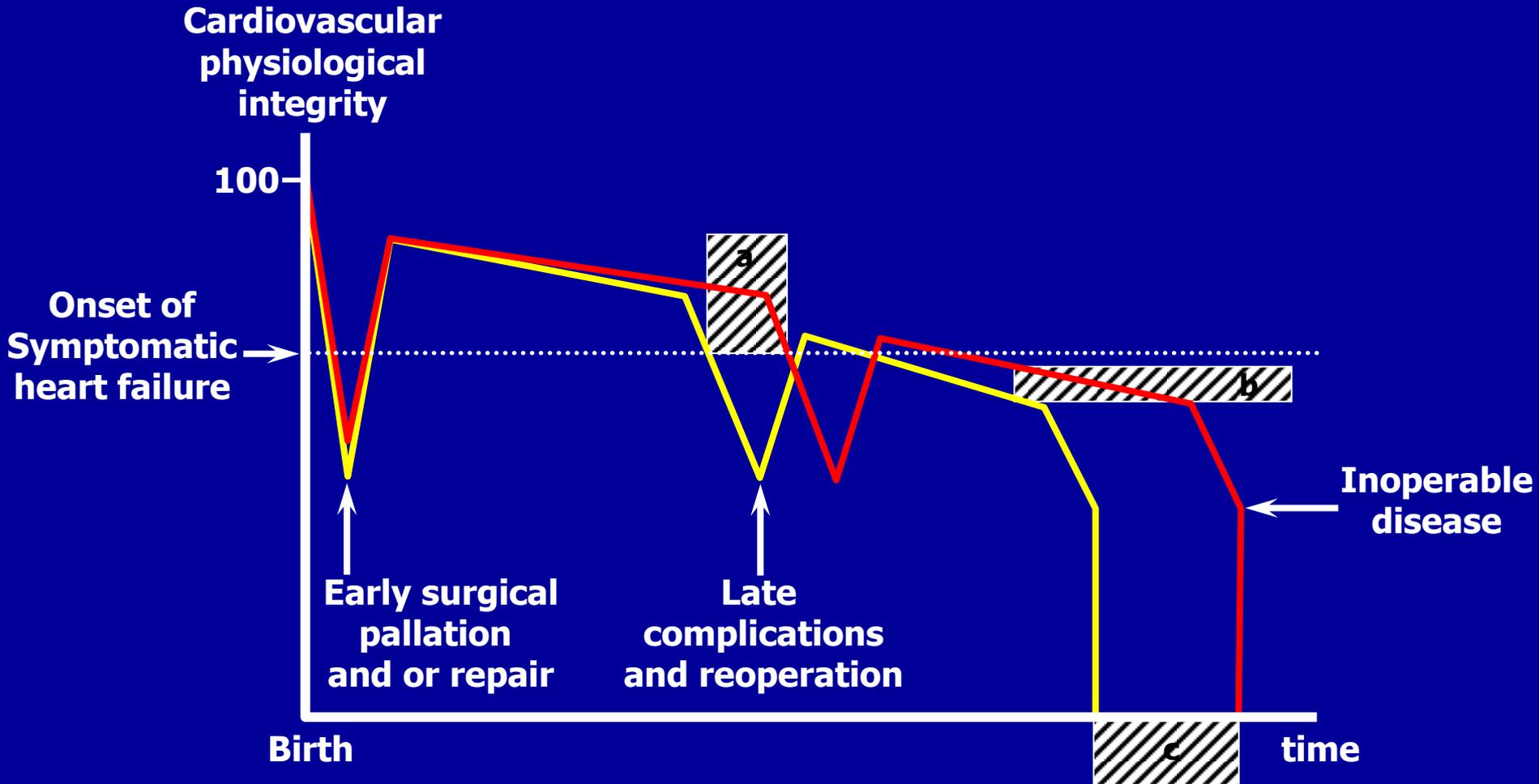
Incidenza delle CC alla nascita stabile nei paesi occidentali

Miglioramento della gestione medica, chirurgica ed anestesiologicala → aumento sopravvivenza

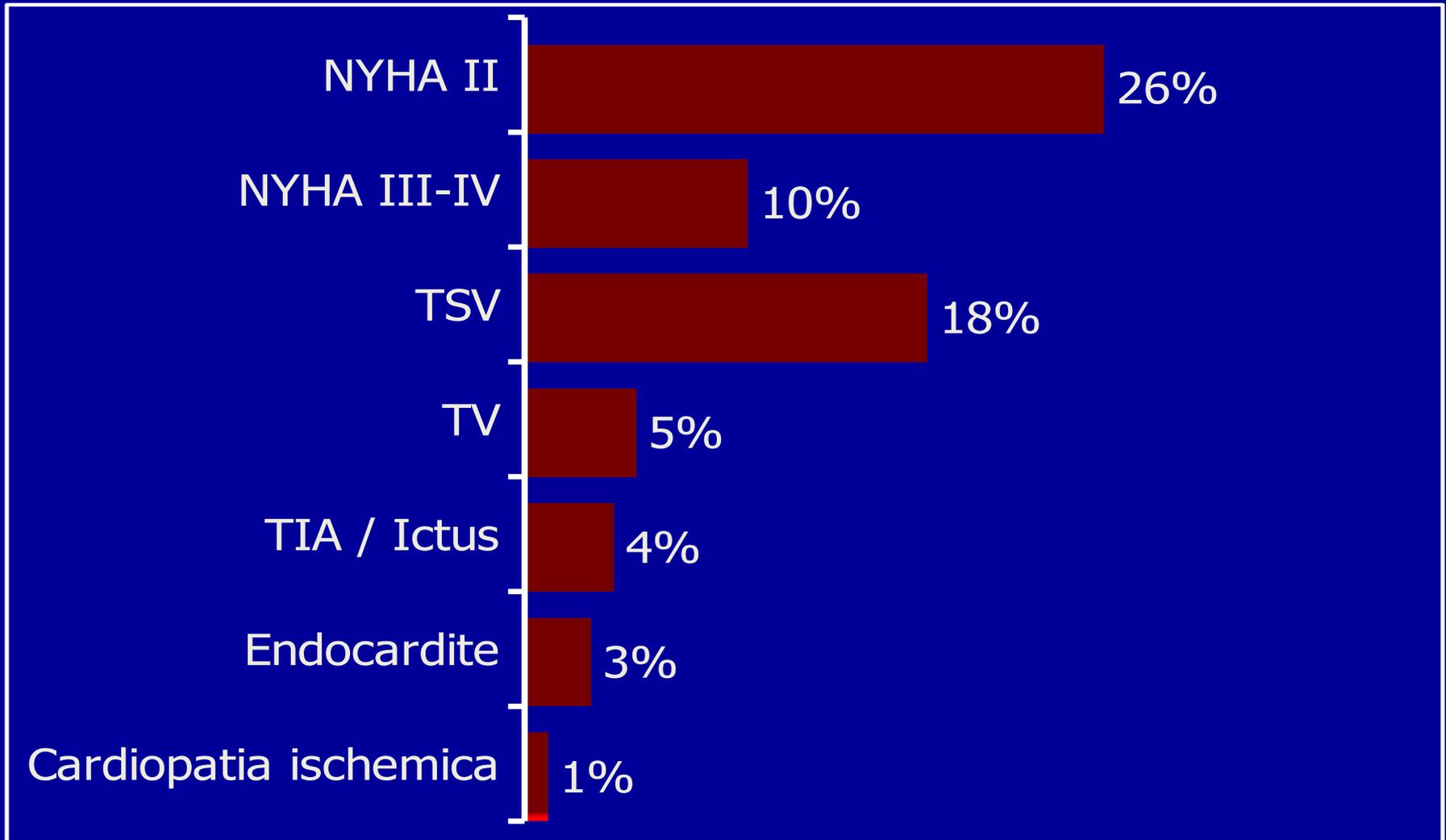
Immigrazione



# Congenital heart disease

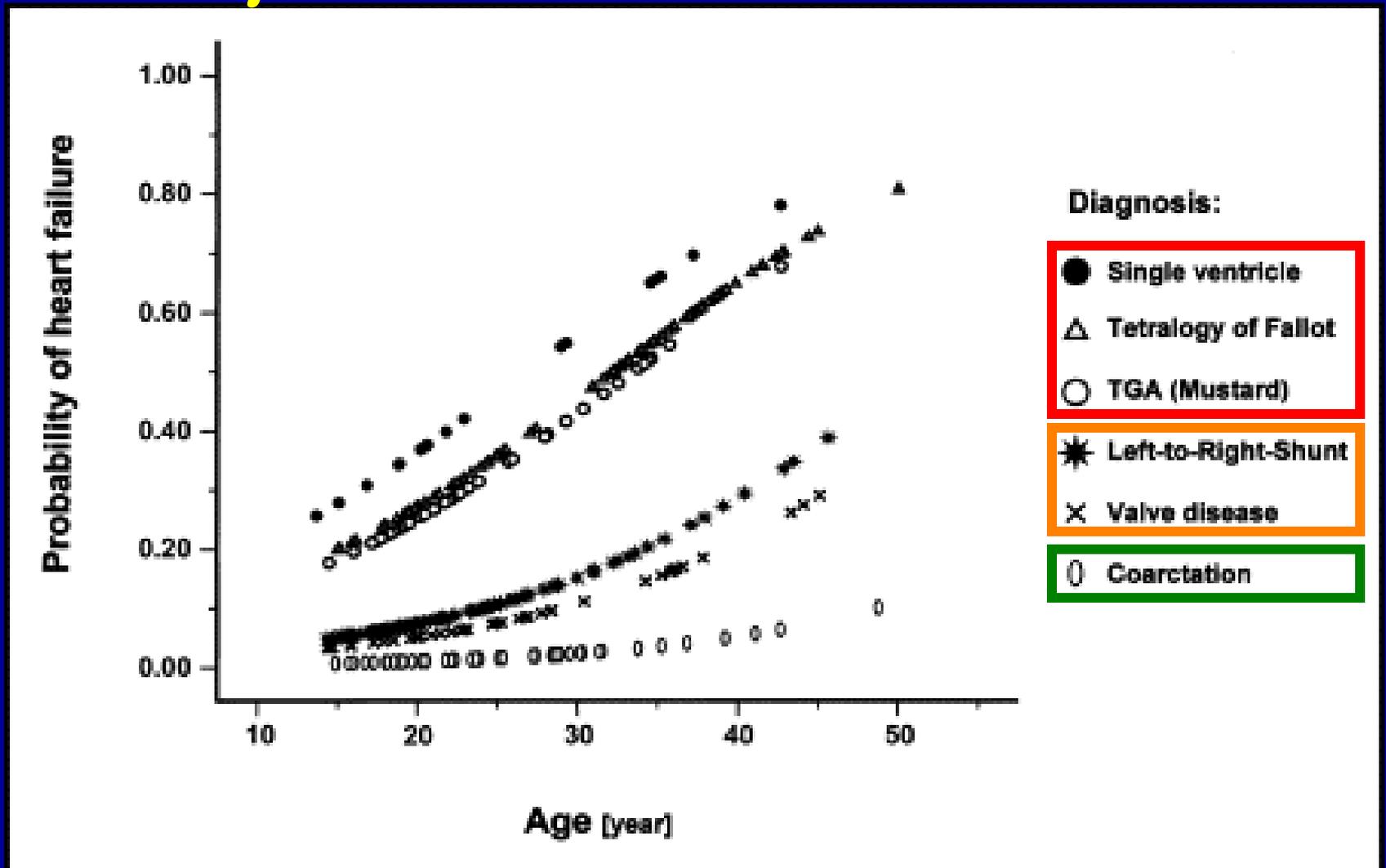


# MORBILITA'



The Euro Heart Survey on adult congenital heart disease, Eur Heart J 2005

# *Scompenso cardiaco nel GUCH*



# *Lo scompenso cardiaco*

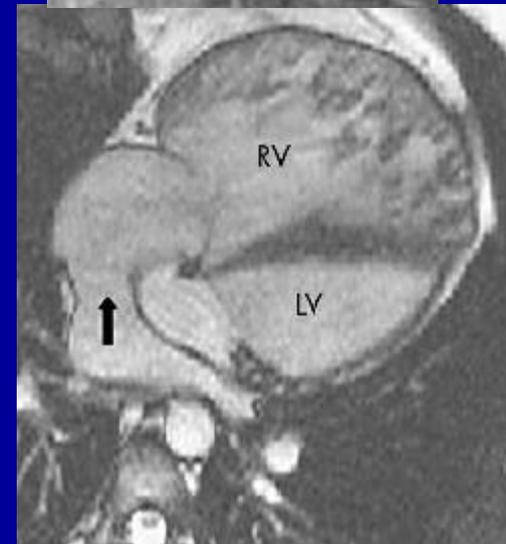
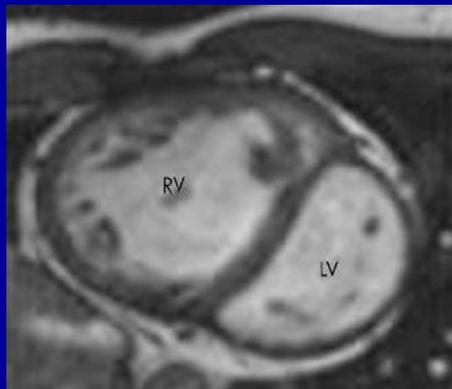
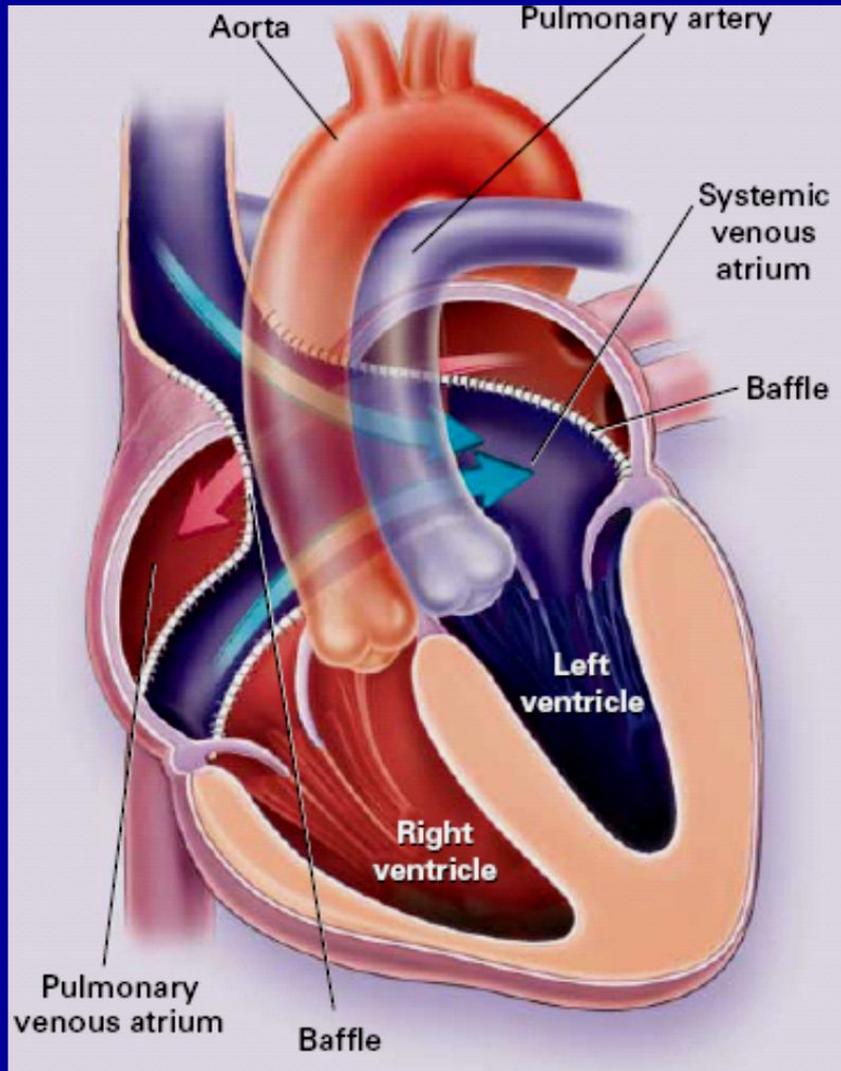
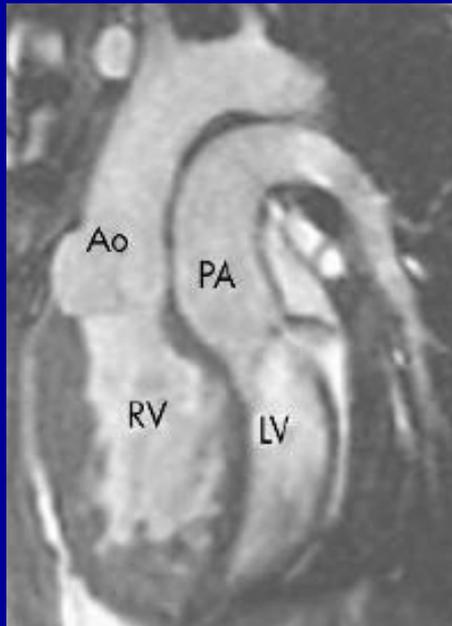
## *Cardiopatie acquisite*

- ❖ *Disfunzione VSn*
- ❖ *Vsn dilatato-ipocinetico*

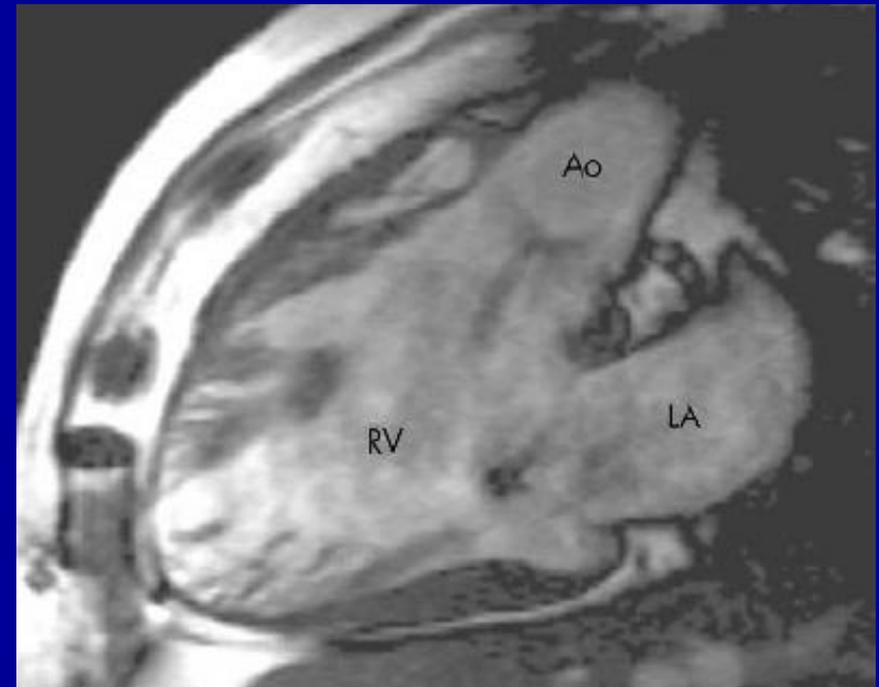
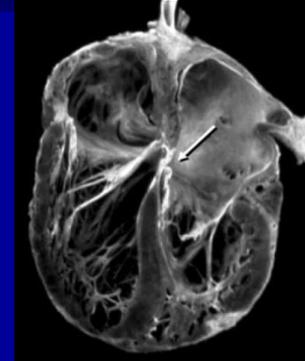
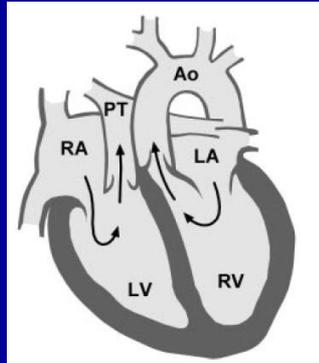
## *GUCH*

- ❖ *VDX iperteso*
- ❖ *VDx dilatato*
- ❖ *Ventricolo unico*

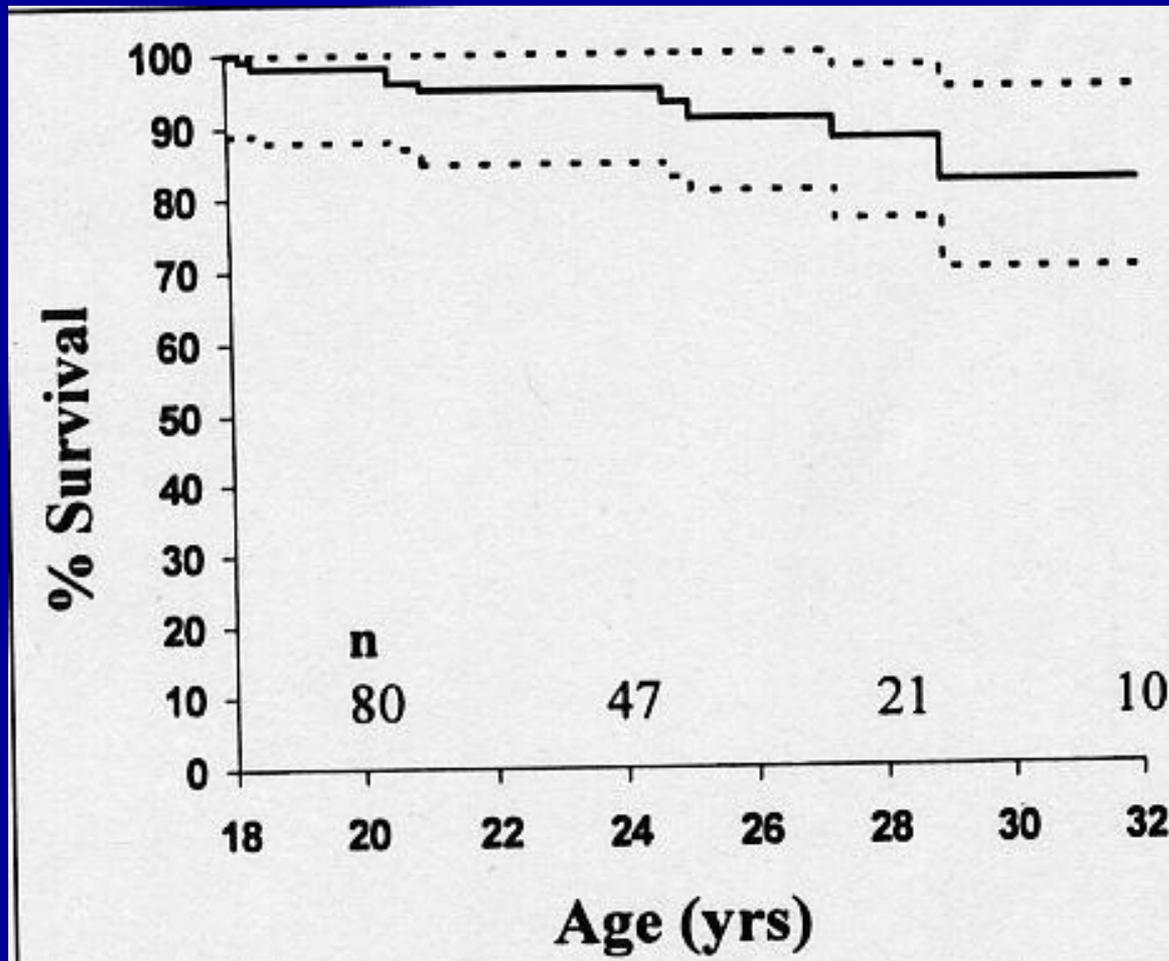
# Ventricolo destro sistemico: TGA S/P Mustard-Senning



# *Ventricolo destro sistemico: TCCGA*

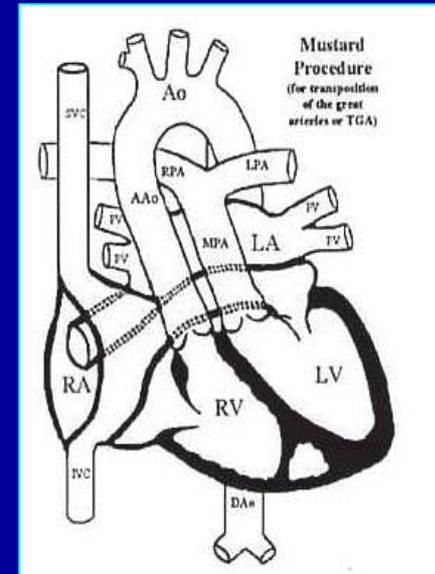
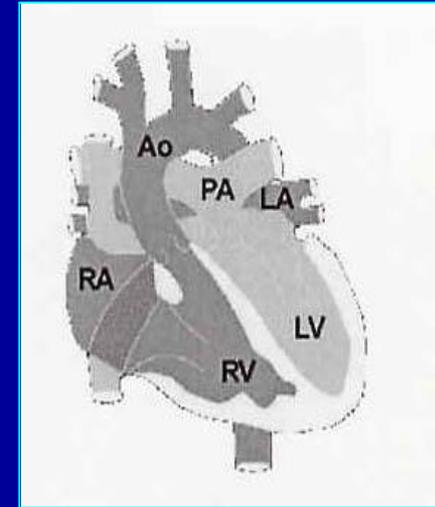


# Switch atriale secondo Mustard/Senning per TGA



# Switch atriale: complicazioni

- Ostruzione o difetti del baffle
- Disfunzione del RV  $\pm$  IT
- Aritmie da dilatazione atriale
- Sviluppo di bradiaritmie
- Ipertensione polmonare



# TGA + atrial switch (Mustard/Senning) Diagnostic Work-up

- **Echocardiography**

First line diagnostic technique providing systemic and sub-pulmonary ventricular size and function, sub-pulmonary outflow tract obstruction, TR, leakage or obstruction of the atrial baffles and assessment of pulmonary venous return. SVC stenosis is, however, mostly difficult to assess and may require TEE. *Contrast echo* is helpful for baffle leakage or stenosis.

- **CMR (CT)**

Indicated for assessment of systemic RV function and patency of the atrial baffles.

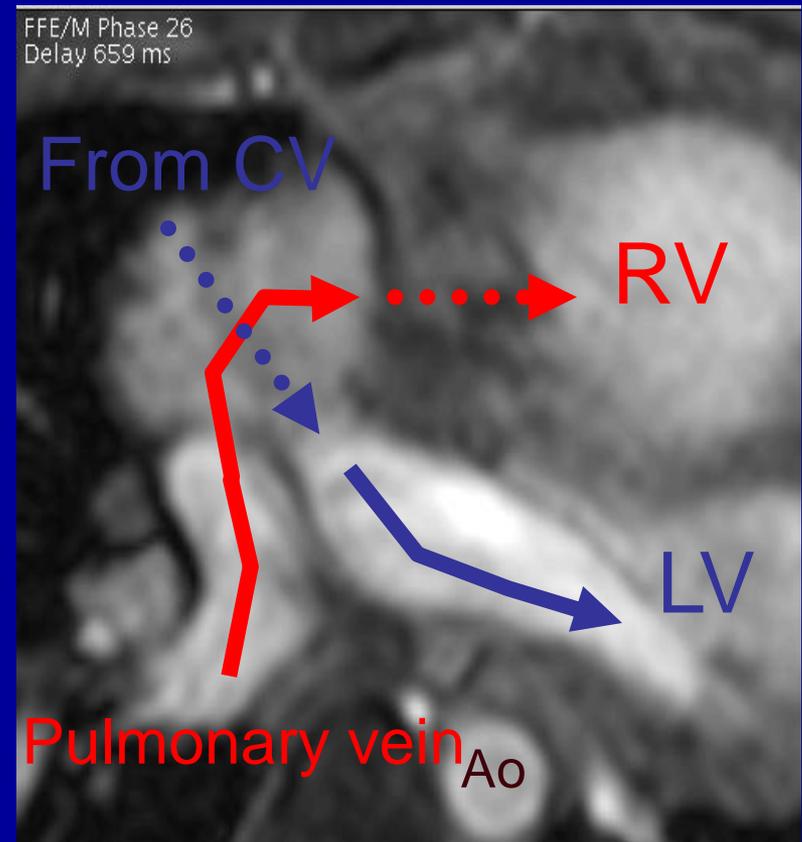
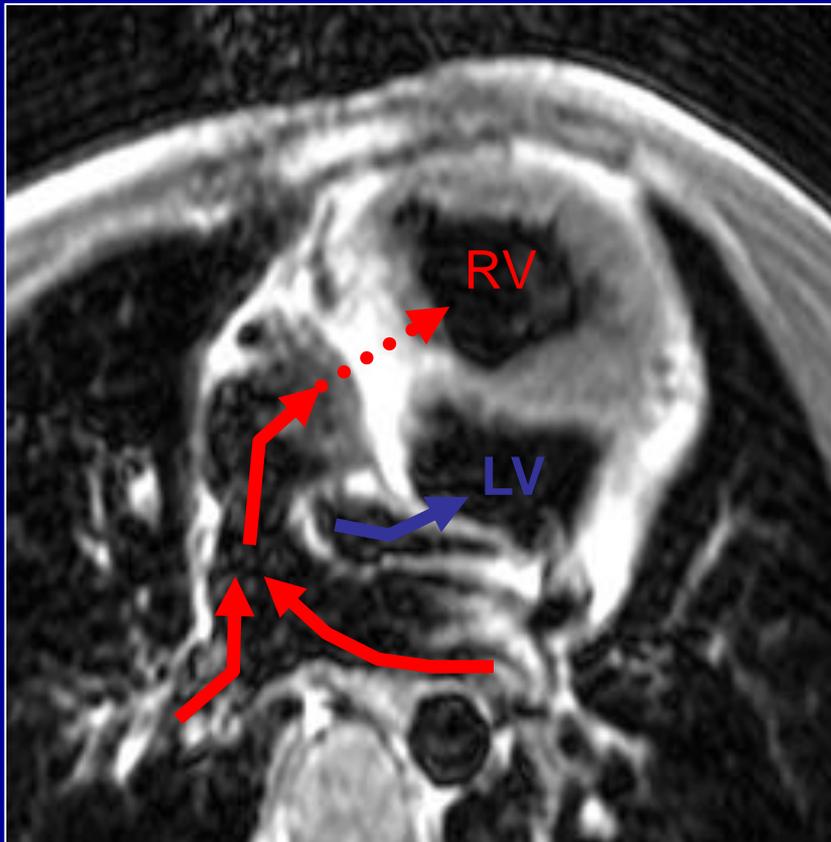
- **Holter monitoring, event recorder**

Required for selected patients (high-risk, investigated for suspected or clinical arrhythmia).

- **Cardiac catheterization**

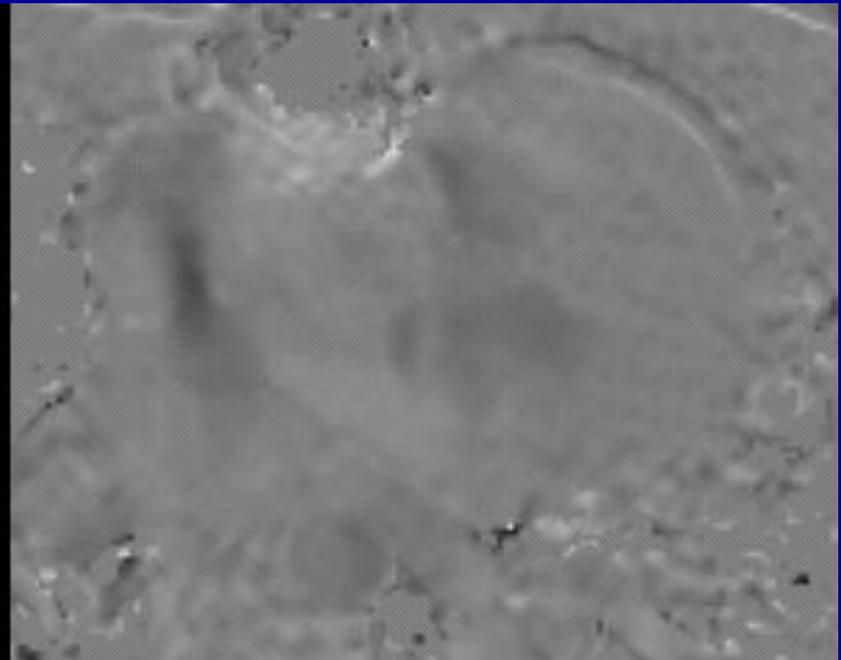
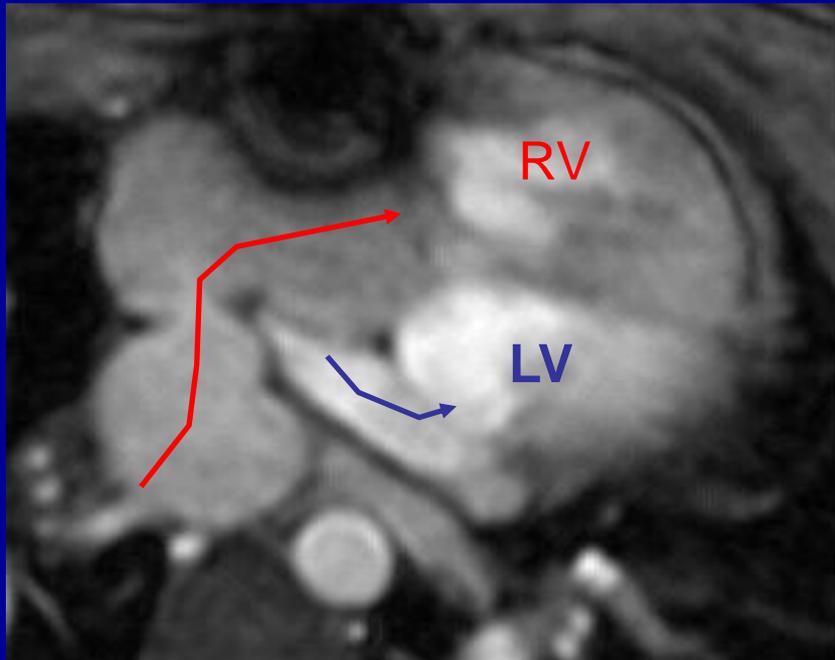
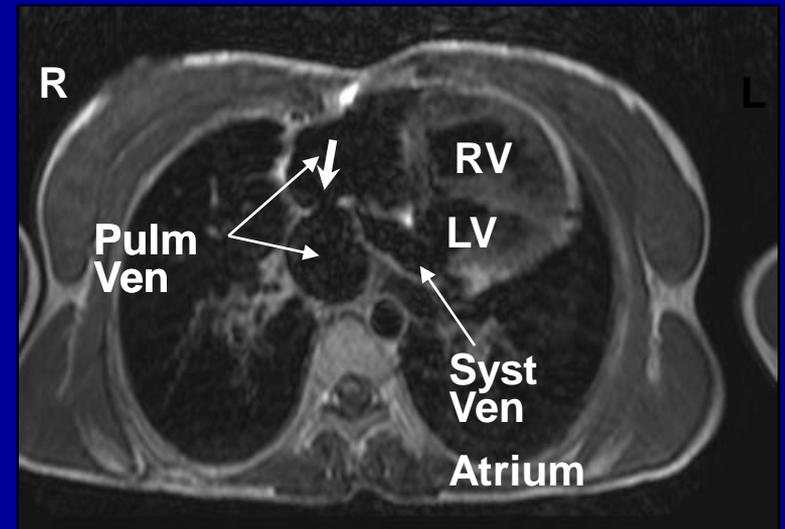
Indicated when non-invasive assessment is inconclusive or PAH requires evaluation.

# Tecnica di Mustard/Senning per TGA

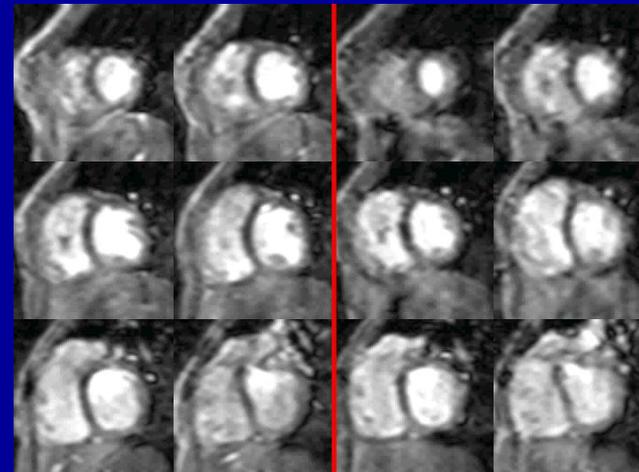
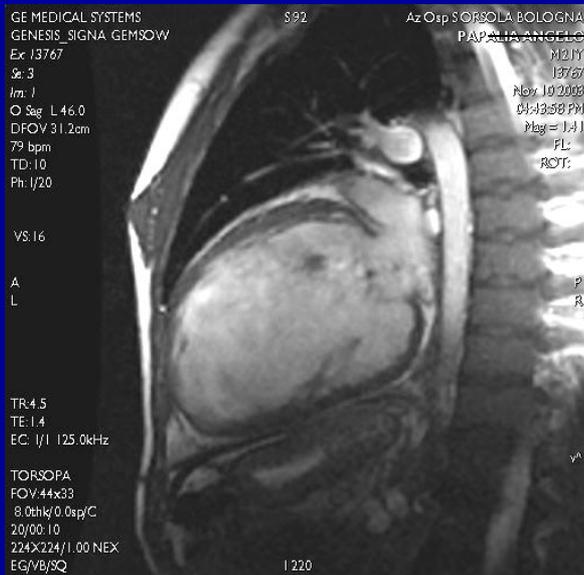
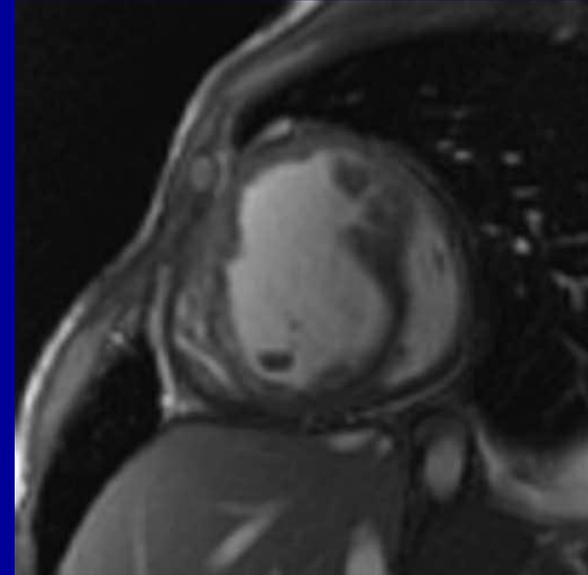
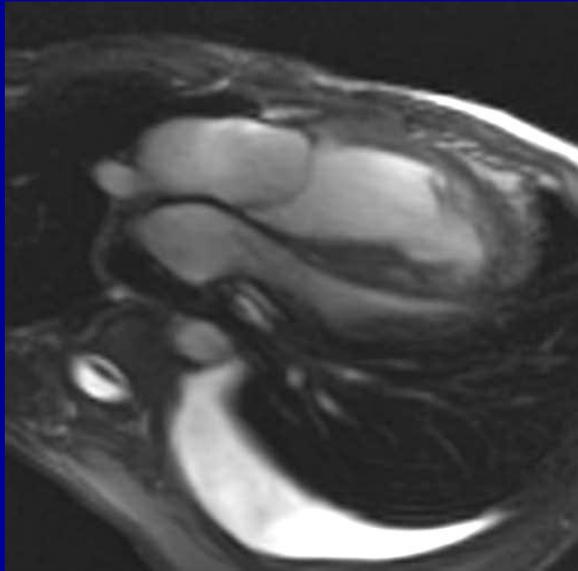


# Tecnica di Mustard per TGA

atrio venoso polmonare  
stenotico



# Switch atriale: Studio funzionale del VDx



ORIGINAL ARTICLE

Volume 316:1429-1435 June 4, 1987 Number 23

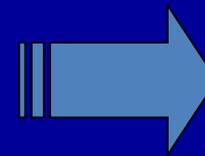
[Next ▶](#)

**Effects of enalapril on mortality in severe congestive heart failure. Results of the Cooperative North Scandinavian Enalapril Survival Study (CONSENSUS). The CONSENSUS Trial Study Group**

**Effect of enalapril on survival in patients with reduced left ventricular ejection fractions and congestive heart failure. The SOLVD Investigators**

**Effect of captopril on mortality and morbidity in patients with left ventricular dysfunction after myocardial infarction. Results of the survival and ventricular enlargement trial. The SAVE Investigators**

*MA Pfeffer, E Braunwald, LA Moye, L Basta, EJ Brown, TE Cuddy, BR Davis, EM Geltman, S Goldman, GC Flaker, and et al.*



**GUCH**

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Number 21

**THE EFFECT OF CARVEDILOL ON MORBIDITY AND MORTALITY IN PATIENTS WITH CHRONIC HEART FAILURE**

MILTON PACKER, M.D., MICHAEL R. BRISTOW, M.D., PH.D., JAY N. COHN, M.D., WILSON S. COLUCCI, M.D., MICHAEL B. FOWLER, M.B., B.S., EDWARD M. GILBERT, M.D., AND NEIL H. SHUSTERMAN, M.D.,  
FOR THE U.S. CARVEDILOL HEART FAILURE STUDY GROUP\*

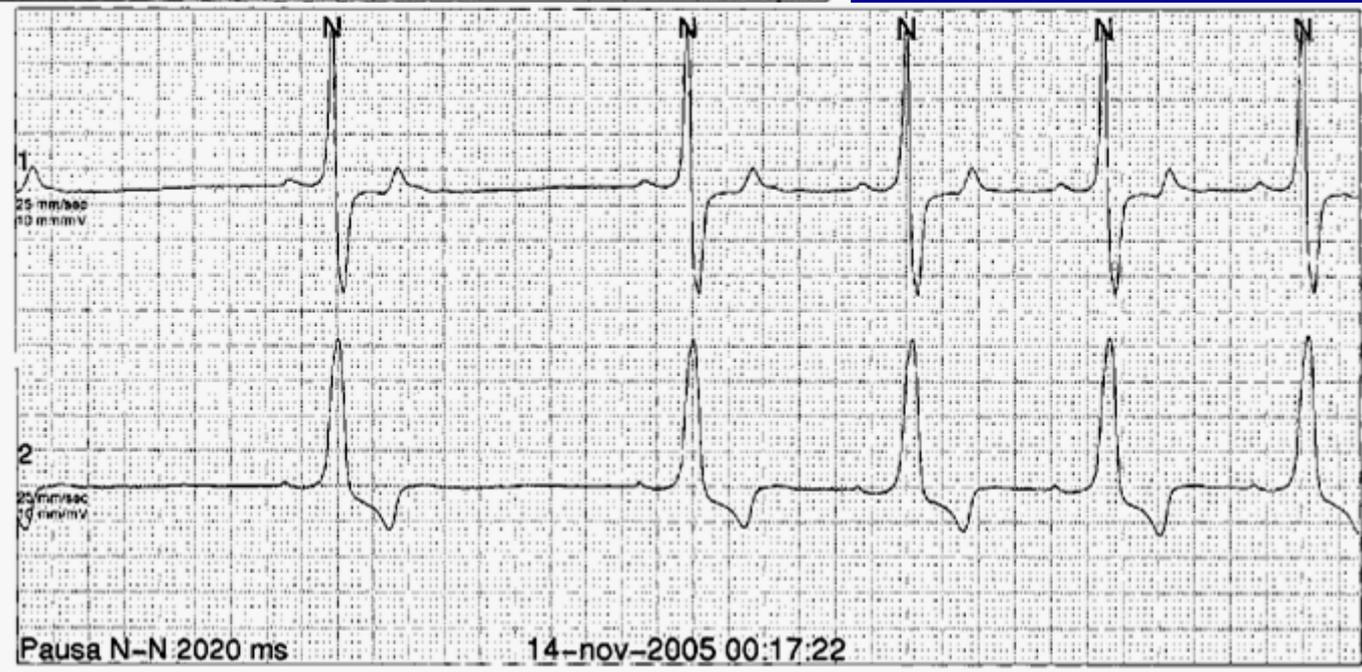
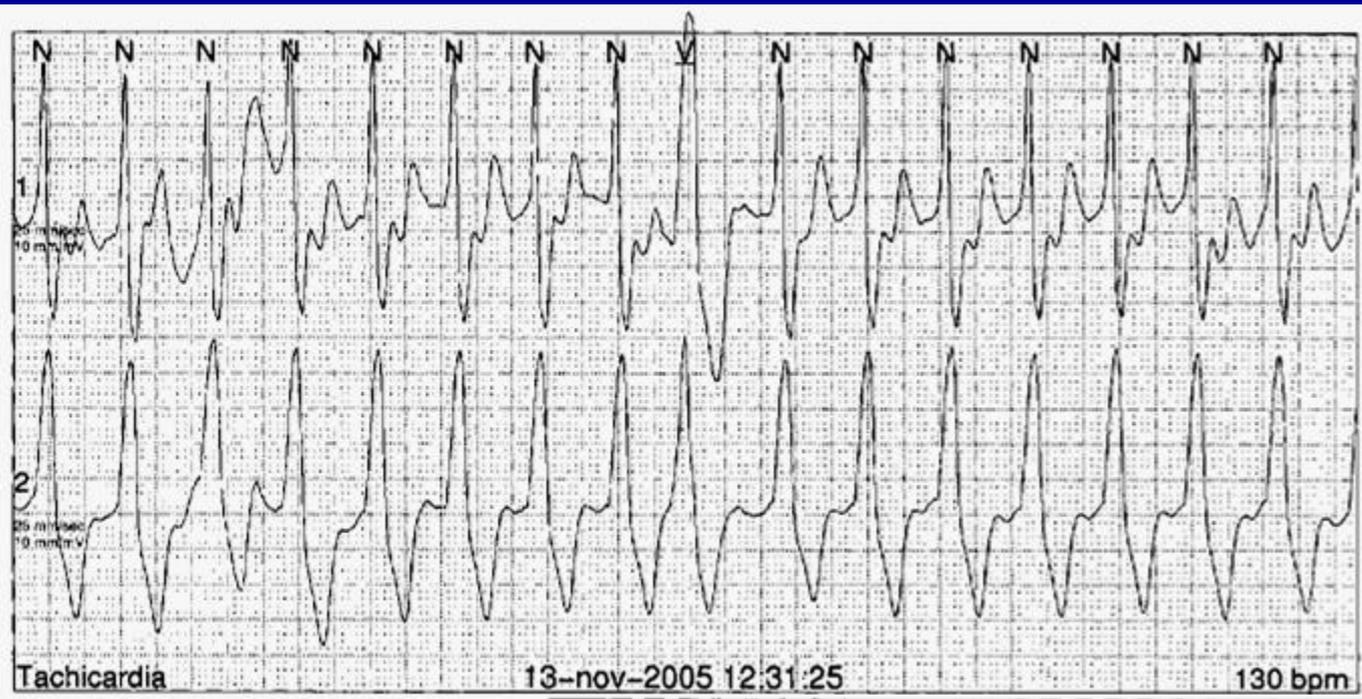
# Ventricolo dx sistemico

	Agent	TGA/ccTGA	No	Follow-up (months)	MRI*	VO <sub>2</sub> max	NYHA	Pro/retro	
<b>β-blocker</b>									
	Lindenfeld <i>et al</i> <sup>17</sup>	Carvedilol	ccTGA	1	7	↑	ND	ND	pro
	Giardini <i>et al</i> <sup>18</sup>	Carvedilol	both	8	12	↑	=	↑	pro
	Josephson <i>et al</i> <sup>16</sup>	Various	TGA	8	36	ND	ND†	↑	retro
	Doughan <i>et al</i> <sup>15</sup>	Various	TGA	31	4	ND	ND	↑	retro
<b>ACE inhibitor</b>									
	Hechter <i>et al</i> <sup>21</sup>	Various	TGA	14	24	=	=	ND	retro
	Robinson <i>et al</i> <sup>22</sup>	Enalapril	TGA	9	12	ND	=	ND	pro
	Therrien <i>et al</i> <sup>23</sup>	Ramipril	TGA	17	12	=	=	ND	pro
<b>ATII antagonist</b>									
	Dore <i>et al</i> <sup>26</sup>	Losartan	both	29	3.5	ND	=	ND	pro
	Lester <i>et al</i> <sup>24</sup>	Losartan	TGA	7	2	↑‡	ND	ND	pro

\*Right ventricular ejection fraction as determined by MRI; †determined in minority of patients; ‡right ventricular ejection fraction as determined by echocardiography. ACE, angiotensin-converting enzyme; ATII, angiotensin II; ccTGA, congenitally corrected transposition of the great arteries; MRI, magnetic resonance imaging; ND, not determined; NYHA, New York Heart Association class; pro, prospective study design; retro, retrospective study designs; ↑, significant improvement; =, no significant change.

## *Aritmie nel GUCH*

- ❖ Motivo principale di accesso in PS
- ❖ Frequente associazione di bradi-tachiaritmie (disfunzione nodo SA o AV)
- ❖ Causa di disfunzione ventricolare, trombosi, morte improvvisa
- ❖ Morbilità: farmaci, CVE, device, chirurgia



**C.C.♂ 22 anni**  
**S/P Mustard**

# TGA + atrial switch (Mustard/Senning) EP Testing, Ablation and ICD

- These procedures are complicated by the fact that the atria are not normally accessible for catheters and “normal” EP procedures because of the course of the baffles and should only be done in specialized centers with specific expertise.
- Patients are at increased risk of SCD.

Atrial tachyarrhythmia, impaired systemic RV function and QRS duration  $\geq 140$  msec have been reported to be risk factors.

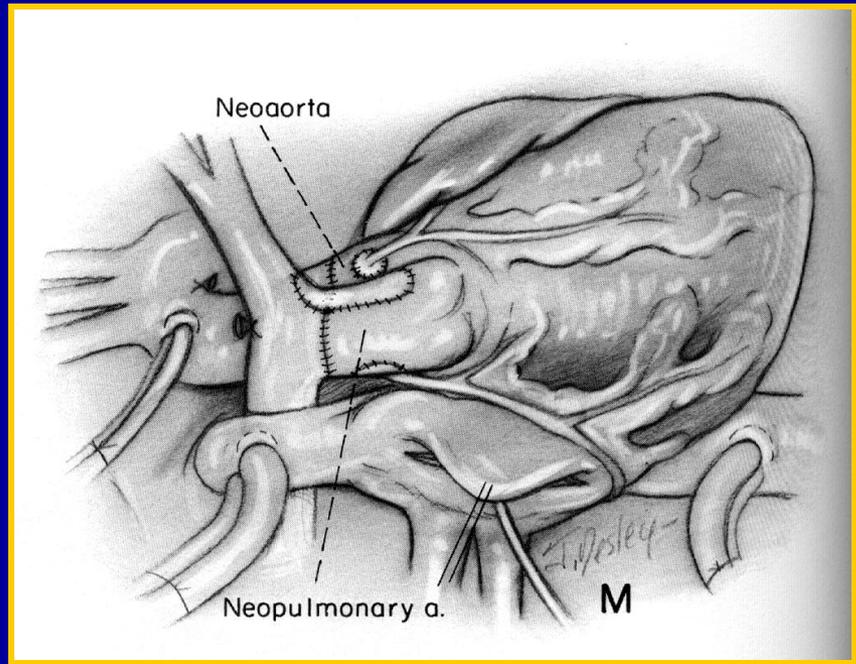
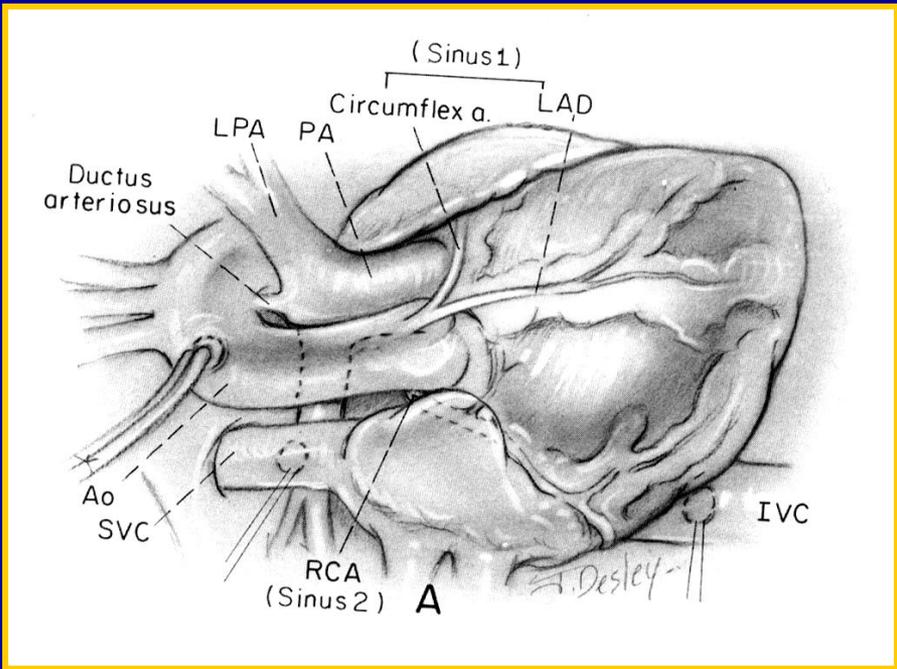
*See general recommendations for ICD implantation.*

# TGA + atrial switch (Mustard/Senning) Follow-up

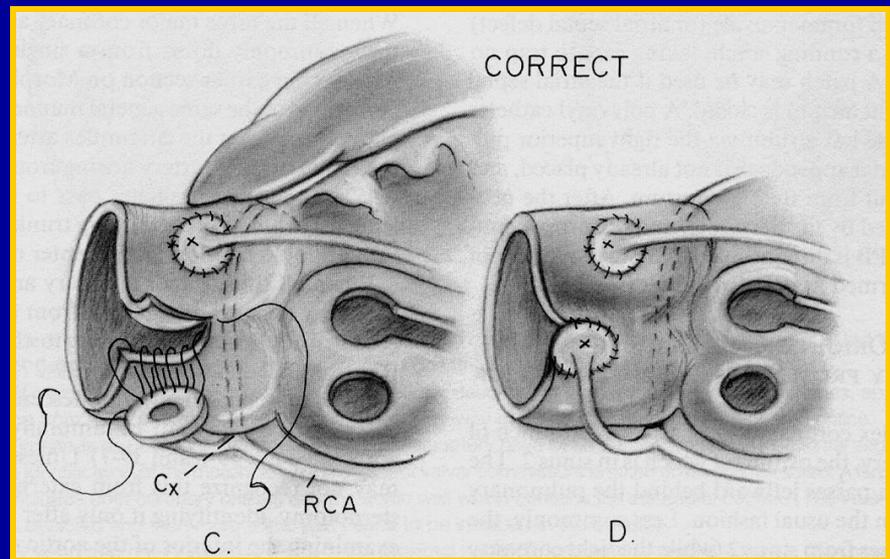
**All patients should be seen at least annually  
in a specialized center**

## **Frequent complications to look for:**

- Dysfunction of the systemic RV.
- TR: often develops as sign of RV dilation and progresses.
- Tachy-arrhythmias: atrial flutter is most typical, but atrial fibrillation and all other types of SVT can occur. VT and VF are reported, associated with SCD. Brady-arrhythmias: ongoing loss of sinus node function frequently necessitates pacemaker therapy.
- Baffle (intra-atrial tunnel) leaks: may cause left-to-right or right-to-left shunt.
- Obstruction of systemic venous and/or pulmonary venous drainage.
- Sub-pulmonary outflow tract obstruction: can occur due to leftward bulging of the interventricular septum (systemic ventricle may benefit).



# Arterial Switch operation for TGA



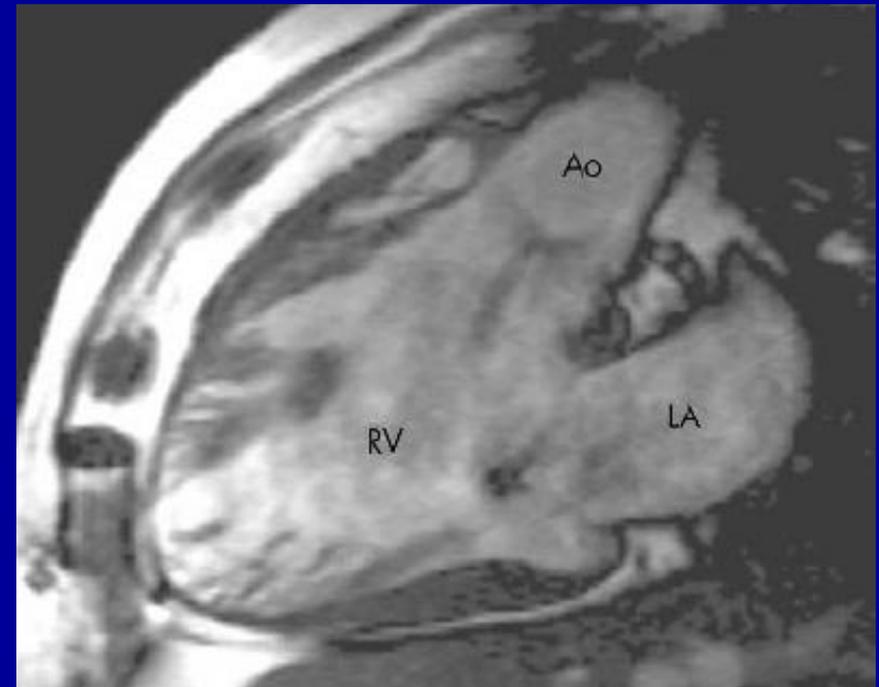
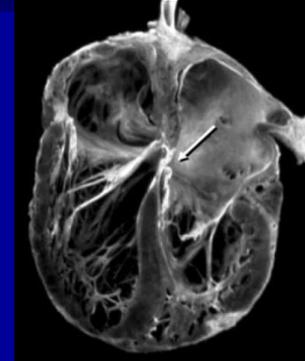
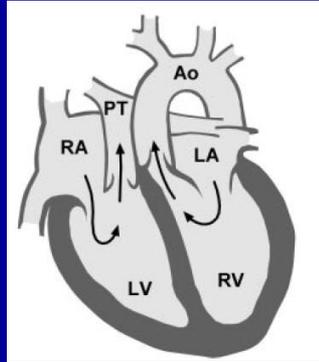
# TGA + arterial switch Follow-up

**All patients should be seen at least annually in a specialized GUCH center**

## **Frequent complications to look for:**

- LV dysfunction and arrhythmias: both may be related to coronary artery problems (re-implanted ostia).
- Dilatation of the proximal part of the ascending aorta resulting in AR.
- Supravalvular PS, pulmonary branch stenosis (unilaterally or bilaterally).

# *Ventricolo destro sistemico: TCCGA*



# Congenitally corrected TGA (cc TGA) Follow-up

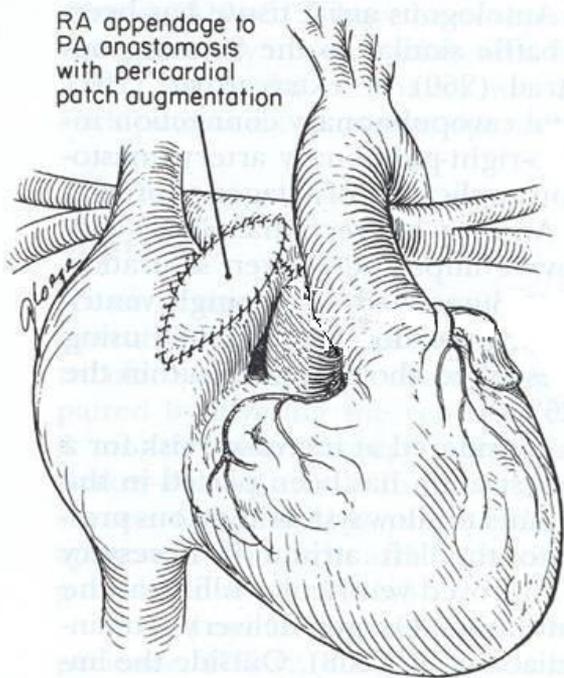
- Patients with ccTGA need life-long follow-up in a specialized GUCH center with annual intervals particularly because of conduction disturbances (AV block), systemic ventricular and systemic AV valve dysfunction.

# Single Ventricle Physiology and Fontan Circulation

- Tricuspid atresia
- Hypoplastic right heart syndrome variants, e.g. pulmonary atresia with intact ventricular septum variants
- Hypoplastic left heart syndrome variants, including mitral atresia
- Double-inlet LV
- Double-inlet RV
- Extreme forms of unbalanced complete AV septal defects
- Single ventricle with undefined morphology.

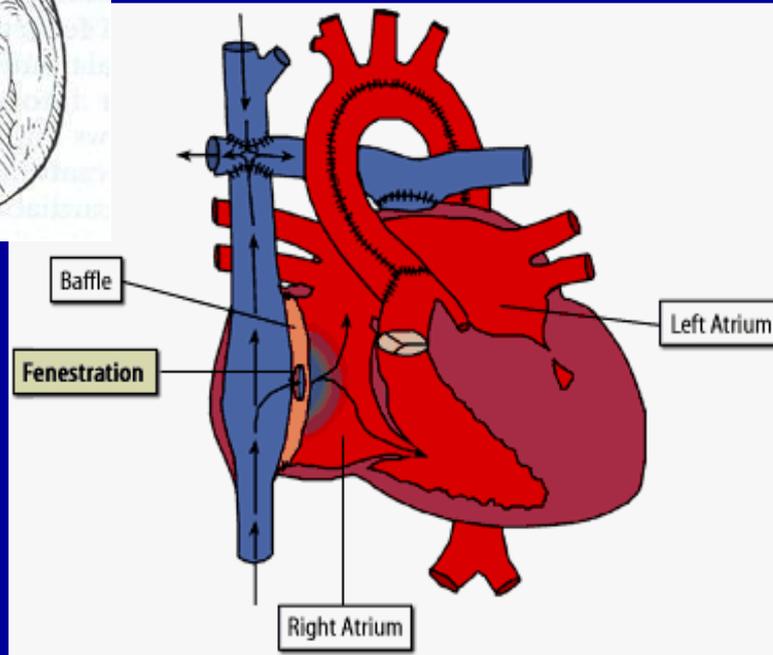
**Biventricular repair can not be done**

# L'evoluzione della Fontan

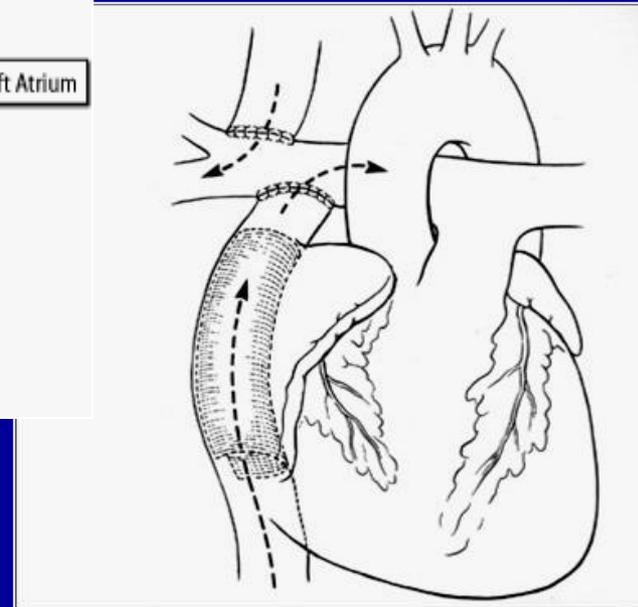


LATERAL TUNNEL  
TCPC  
1980

ATRIO-PULMONARY  
CONNECTION  
1971

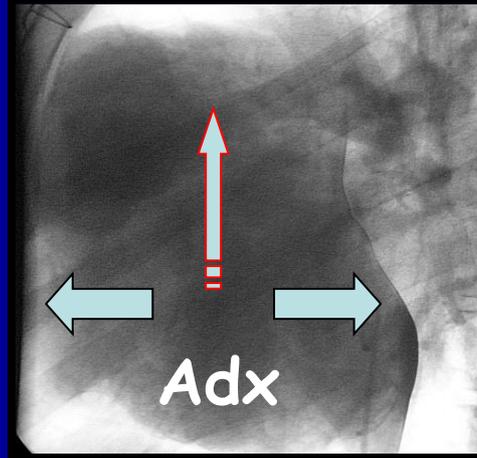
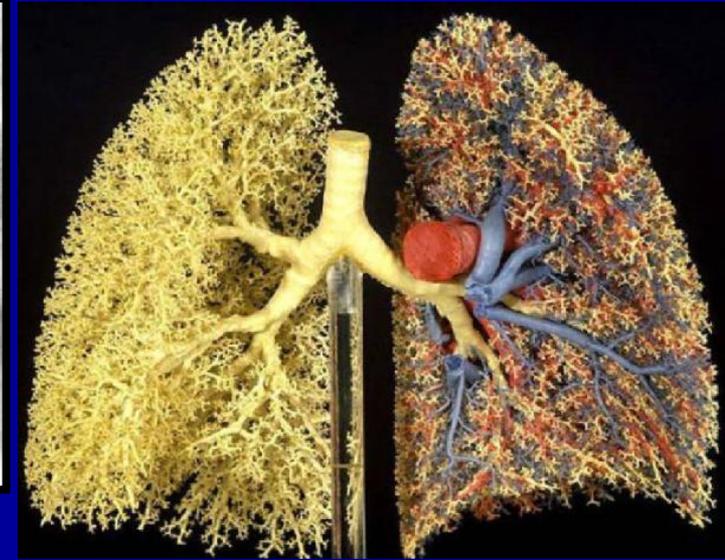
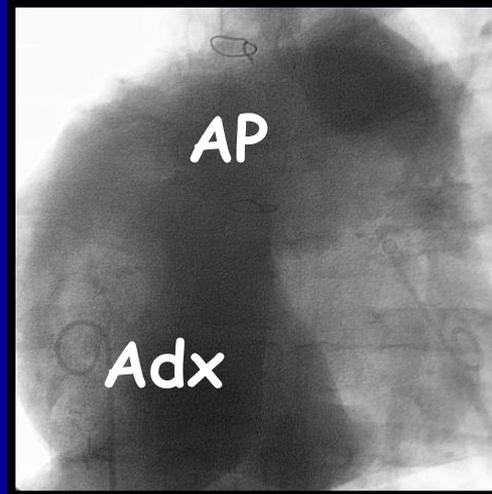
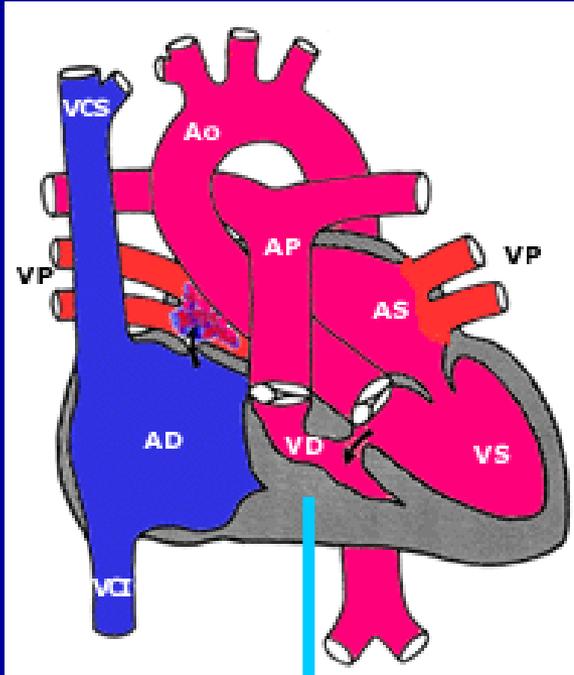


EXTRACARDIAC  
CONDUIT  
1990 - 95



# La circolazione tipo Fontan

## Il ventricolo "poco riempito"



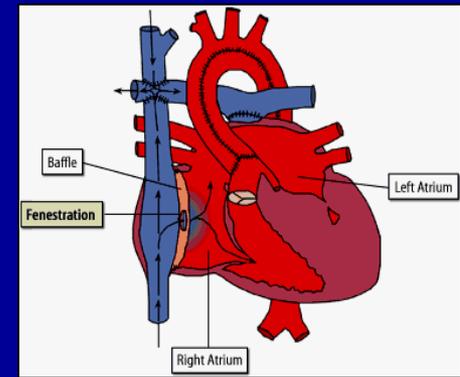
↑ PCO<sub>2</sub>    Perdita flusso pulsatile  
                   ( ↓ NO)

Assenza del ventricolo sottopolmonare

Perdita energia cinetica anterograda

↑ RVP

# *Il ventricolo unico - La Fontan*



*Stenosi circuito*

*Stenosi A. polmonari*

*Compressione VPo*



*RAP*

*Disfunzione ventricolare*

*Insuff. valvole AV*

*Aritmie*

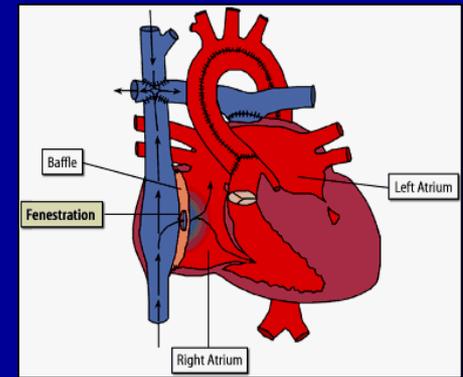
*Enteropatia*

*Proteino-disperdente*

**Scompenso  
cardiaco**

# *Ventricolo unico - Fontan*

## *Terapia dello scompenso cardiaco*



*Angioplastica  
± stent*

*Embolizzazione  
Collaterali sist-Po*

*Fenestrazione circuito*

*Conversione a  
Cavopolmonare totale*

*Trapianto cardiaco*

*Terapia medica  
convenzionale ??*

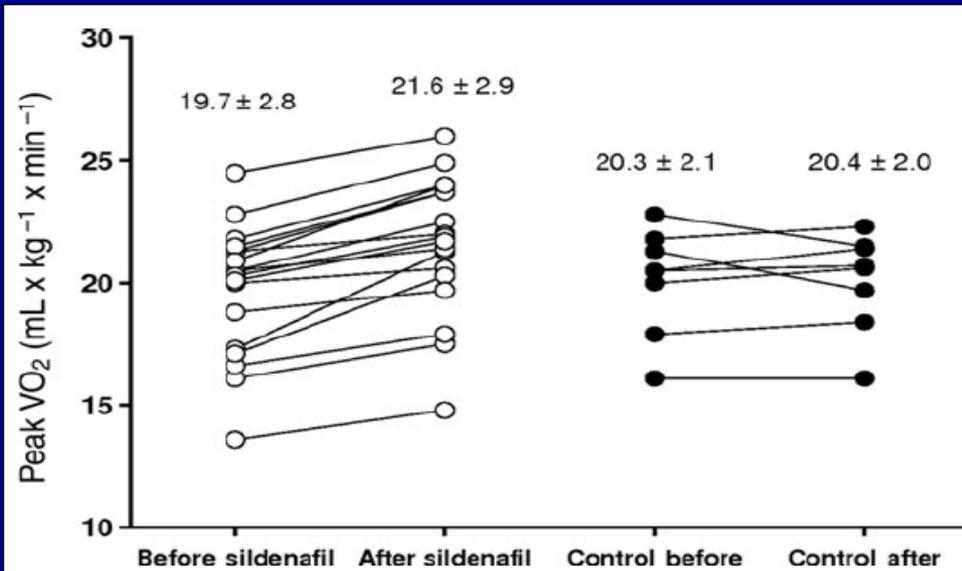
## *Ventricolo unico - Fontan*

- ENALAPRIL per 10 settimane
- 18 pazienti
- Prospettico, randomizzato, cross-over
- Non modifica la capacità funzionale
- Non modifica la funzione diastolica
- Non modifica RVS nè IC a riposo

# Effect of sildenafil on haemodynamic response to exercise and exercise capacity in Fontan patients

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- ❖ 27 pazienti post Fontan
- ❖ Età : 22.8±5 aa
- ❖ 70% classe NYHA II/III

**Table 2** Change in cardiopulmonary and haemodynamic variables observed in exercise test nos 2 vs. 3 in the sildenafil treatment and in the control group at each exercise stage

Variable	Rest				Peak exercise			
	Sildenafil		Control		Sildenafil		Control	
	Before	After	Before	After	Before	After	Before	After
Cardiac index (L/min/m <sup>2</sup> )	2.9 ± 0.8	3.7 ± 1.0	2.9 ± 0.9	2.9 ± 0.9	5.1 ± 0.9	5.6 ± 0.9	5.1 ± 0.9	5.2 ± 0.9
PBF index (L/min/m <sup>2</sup> )	2.2 ± 0.6	2.8 ± 0.5	2.4 ± 0.5	2.4 ± 0.6	4.2 ± 0.5	4.7 ± 0.6	4.4 ± 0.6	4.4 ± 0.6

# Conversion of atriopulmonary Fontan to extracardiac total cavopulmonary connection improves cardiopulmonary function

Alessandro Giardini <sup>a,\*</sup>, Carlo Pace Napoleone <sup>b</sup>, Salvatore Specchia <sup>a</sup>, Andrea Donti <sup>a</sup>, Roberto Formigari <sup>a</sup>, Guido Oppido <sup>b</sup>, Gaetano Gargiulo <sup>b</sup>, Fernando M. Picchio <sup>a</sup>

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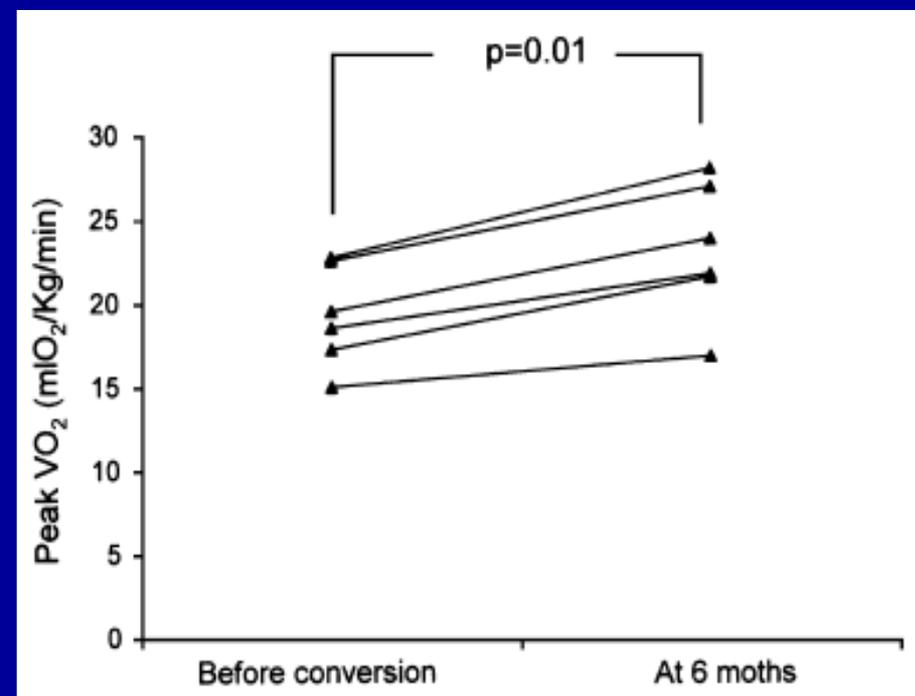


Table 2

Cardiopulmonary test results before vs. 6 months after conversion to extracardiac total cavopulmonary connection

	Before	After conversion	<i>p</i> value
Peak O <sub>2</sub> pulse (ml O <sub>2</sub> /kg/beat)	4.9 (2.5–10.5)	5.7 (3.5–11.5)	0.01
Peak HR (beats/min)	136 (117–160)	134 (121–159)	0.88
Peak VE (l/min)	55.4 (23.9–68.5)	56.5 (32.5–68.2)	0.63
VC (l/min)	3.5 (2.2–4.7)	3.4 (2.3–4.6)	0.66
FEV1 (l/s)	2.8 (1.9–3.9)	2.9 (1.9–3.9)	0.06
Arterial O <sub>2</sub> saturation at peak exercise, %	91 (87–94)	92 (88–93)	0.7

# Patients after Fontan Operation Follow-up

- After Fontan operation patients should be followed in specialized GUCH centers, usually at least annually including:
  - echo,
  - ECG,
  - blood controls and,
  - exercise testing.

Intervals for CMR and hepatic ultrasound (CT) must be decided on an individual basis.

- Comprehensive assessment is mandatory for patients with manifestations of the “failing Fontan” complex, with particular care to exclude even minor obstructions to cavo-pulmonary and pulmonary venous return which may have major haemodynamic impact.

# Patients after Fontan Operation

## Medical Therapy (1)

- **Anticoagulation**

- Right atrial blood stasis and disturbed coagulation may predispose to thrombosis.
- The potential for sub clinical, recurrent pulmonary embolism leading to a rise in PVR has led to a recommendation by some groups for life-long anticoagulation. There is, however no evidence for benefit and practise varies among centers.
- Anticoagulation is definitely indicated in the presence of atrial/Fontan pathway/PA thrombus, atrial arrhythmias or thrombo-embolic events.

# Patients after Fontan Operation

## Medical Therapy (2)

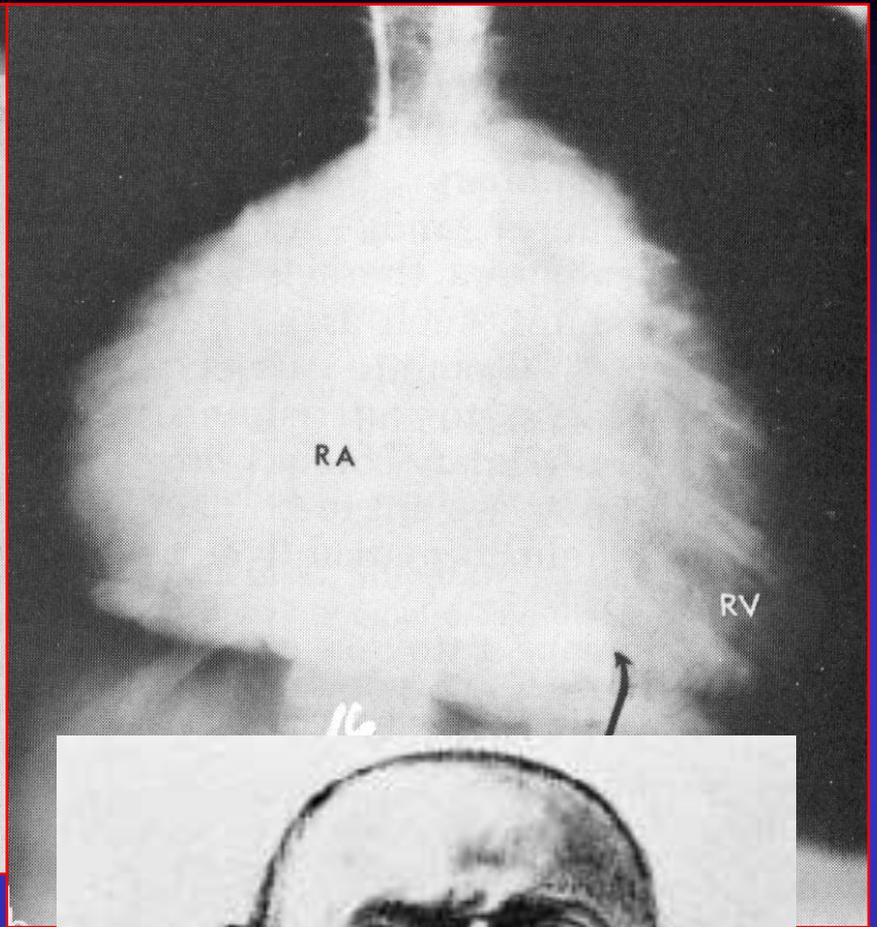
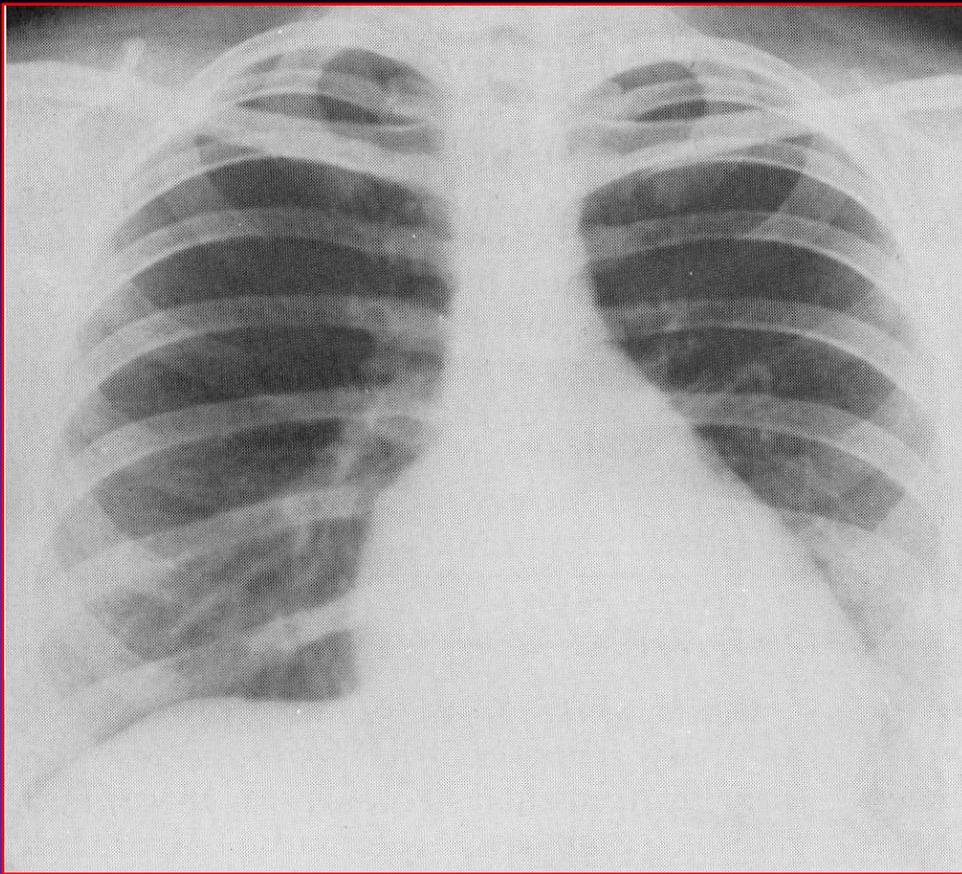
- **Antiarrhythmic therapy:**

- loss of sinus rhythm may precipitate rapid haemodynamic decline and sustained arrhythmia should be considered a medical emergency,
- electrical cardioversion is the mainstay of treatment as drug therapy is often ineffective,
- Amiodarone may be effective in preventing recurrence but it has many long-term side effects. Sotalol can be an alternative,
- there should be a low threshold for radiofrequency ablation although these arrhythmias are difficult to treat in the catheterization laboratory,
- anti-tachycardia atrial pacemakers may assist,
- if AV pacing is required, this will need an epicardial approach,
- occurrence of arrhythmias should prompt haemodynamic evaluation.

# Patients after Fontan Operation

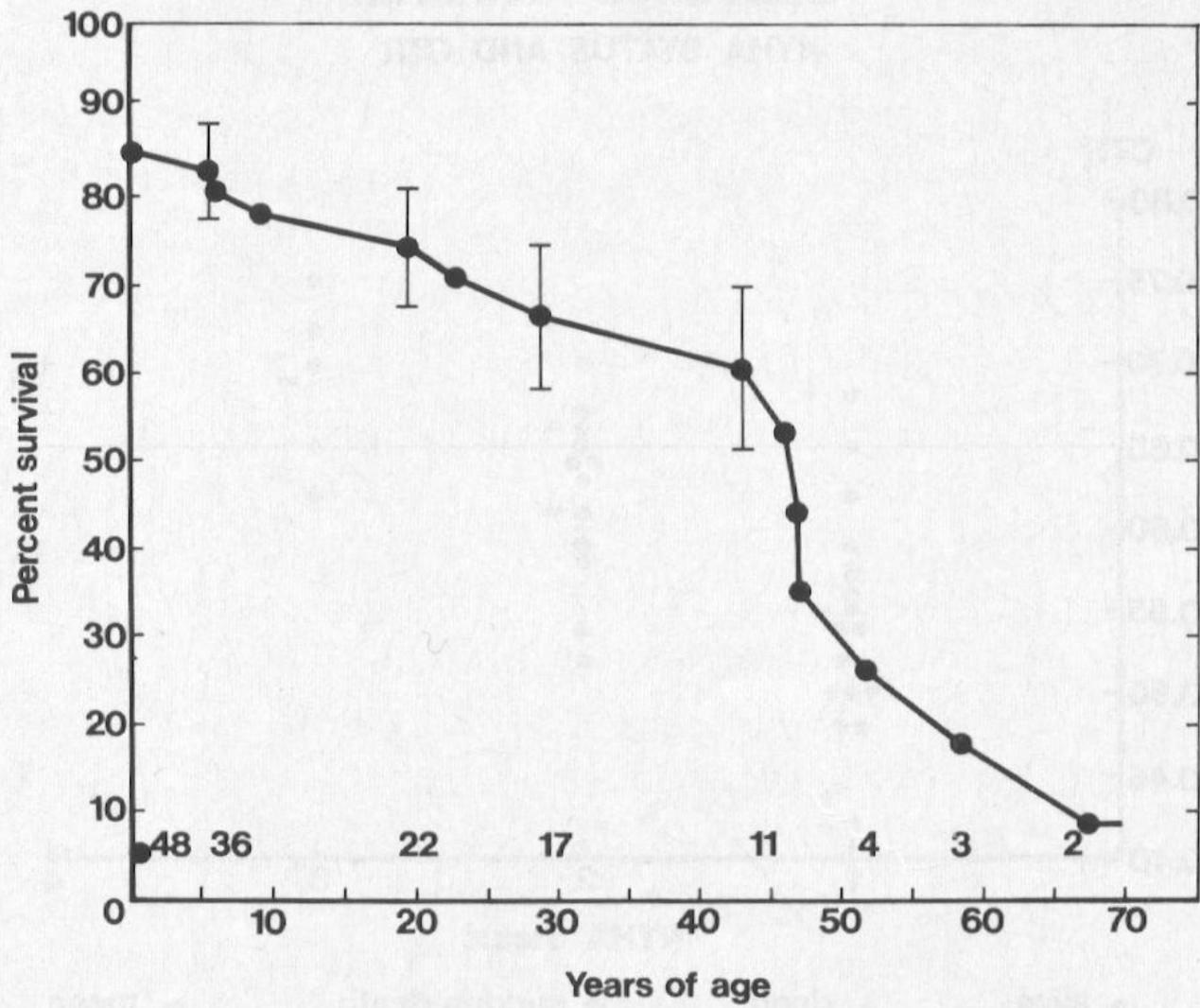
## Medical Therapy (3)

- **Medical therapy of protein losing enteropathy (PLE):**
  - remains challenging and various treatments have been proposed after exclusion of haemodynamic problems including:
    - ▶ salt restriction,
    - ▶ high protein diet,
    - ▶ diuretics,
    - ▶ ACE-inhibitors (may be poorly tolerated),
    - ▶ steroids,
    - ▶ albumin infusion,
    - ▶ chronic subcutaneous heparin,
    - ▶ creation of a fenestration (by interventional catheter),
    - ▶ eventually consideration of transplantation.



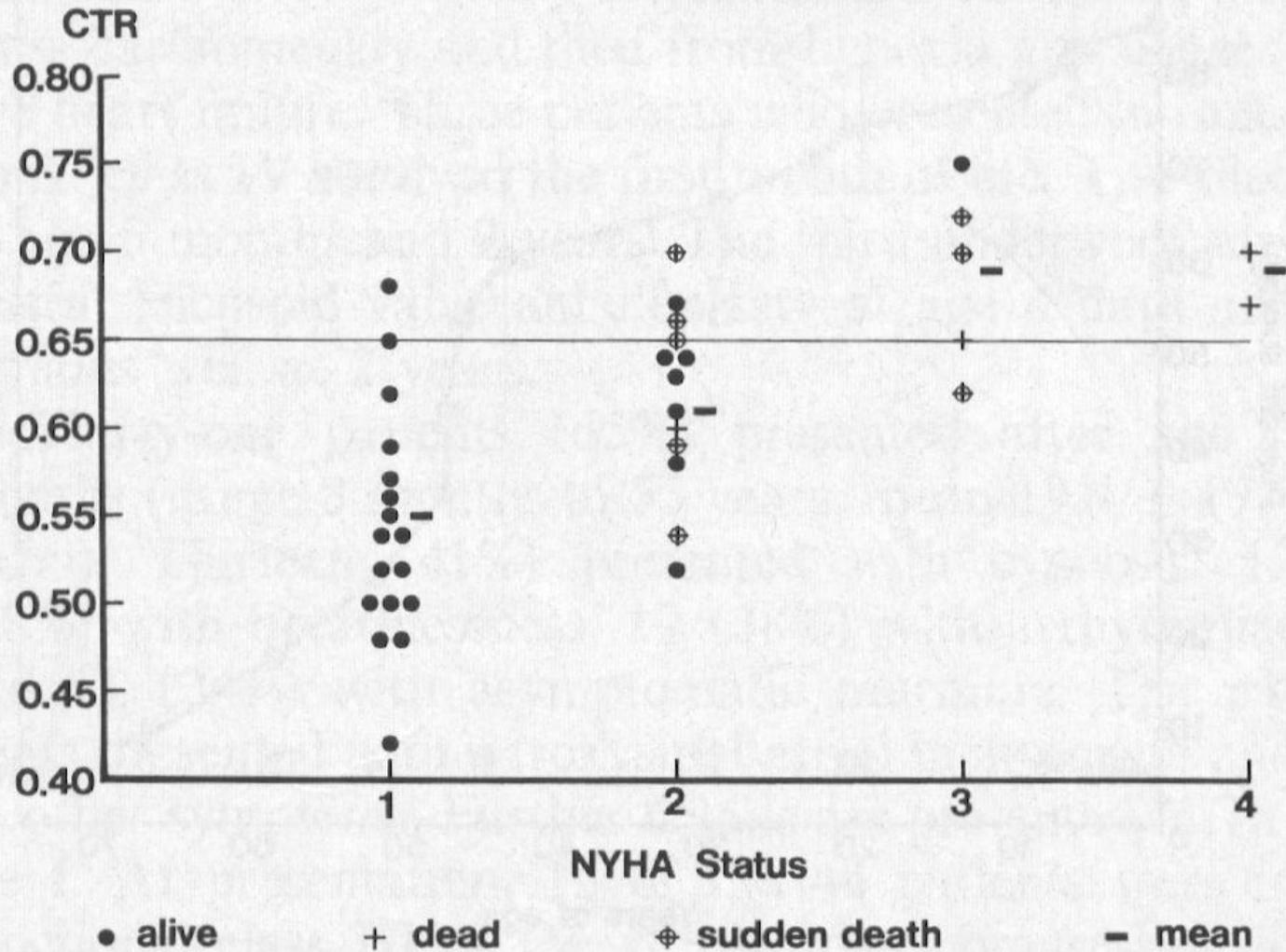
ANOMALIA  
DI  
EBSTEIN





# EBSTEIN'S ANOMALY

## NYHA STATUS AND CTR



# Ebstein's Anomaly

## Diagnostic Work-up

- **Echocardiography**

Key diagnostic technique providing anatomy and function of the tricuspid valve, apical distal displacement of the septal or posterior leaflet (in adults  $\geq 0,8 \text{ cm/m}^2$  body surface area), size of the anterior leaflet, “tethering” of the septal or posterior tricuspid valve leaflet on the septum or ventricular wall, size and function of the different cardiac sections (right atrium, atrialised ventricle, remaining functional RV, LV), RVOTO and associated lesions.

- **CMR**

Has value with regard to evaluation for surgery as it offers unrestricted views for assessment of the dilated right heart, RV function and the tricuspid valve.

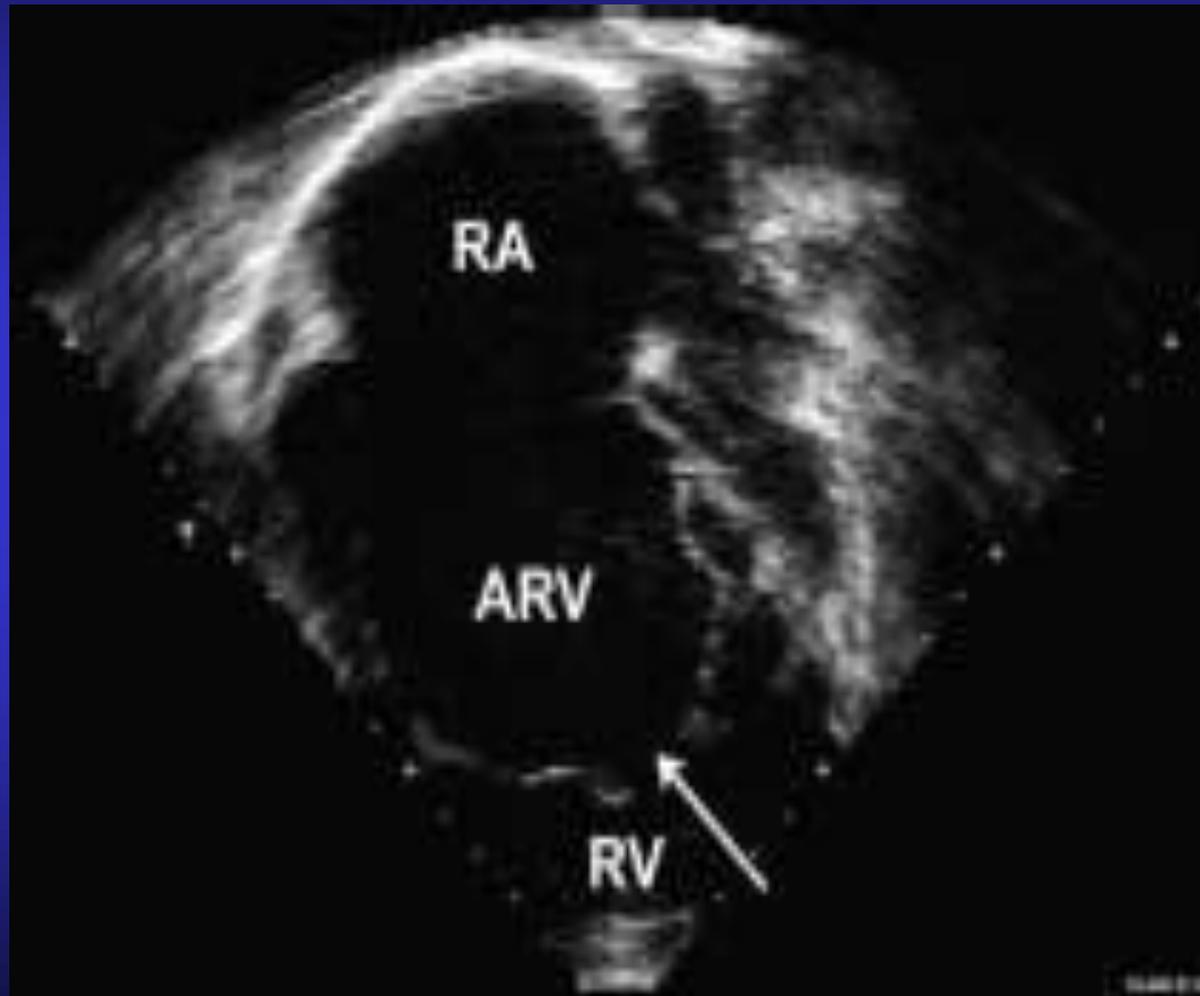
# Anomalia di Ebstein – Score

## ▪ Gose Score:

**Ratio = (RADx + ATRIALISED RV ) / Functional RVdx**

- Grade 1:  $< 0,5$
- Grade 2:  $0,5 - 0,99$
- Grade 3:  $1 - 1,49$
- Grade 4:  $> 1,5$

# Anomalia di Ebstein: ECO



# Anomalia di Ebstein – Prognosi

- Se **Ratio**  $> 1,5$  (grado 4), mortalità del 100%
- Se **Ratio** tra 1 e 1,49 (grado 3), 10% di mortalità precoce e 45% di mortalità nella prima infanzia
- Se **Ratio**  $< 1$  (gradi 1 e 2), la sopravvivenza è del 92%

## **Anomalia di Ebstein**

Fattori predittivi di una cattiva prognosi sono:

- 1) Scompenso cardiaco*
- 2) Sintomi*
- 3) Cianosi*
- 4) Difetti cardiaci associati*

*Anomalie del ritmo anche se frequenti non  
rappresentano una causa di morte  
in questi pazienti*

# Anomalia di Ebstein

## Trattamento

### Medico

- Neonati: PGE

Trattamento dello scompenso

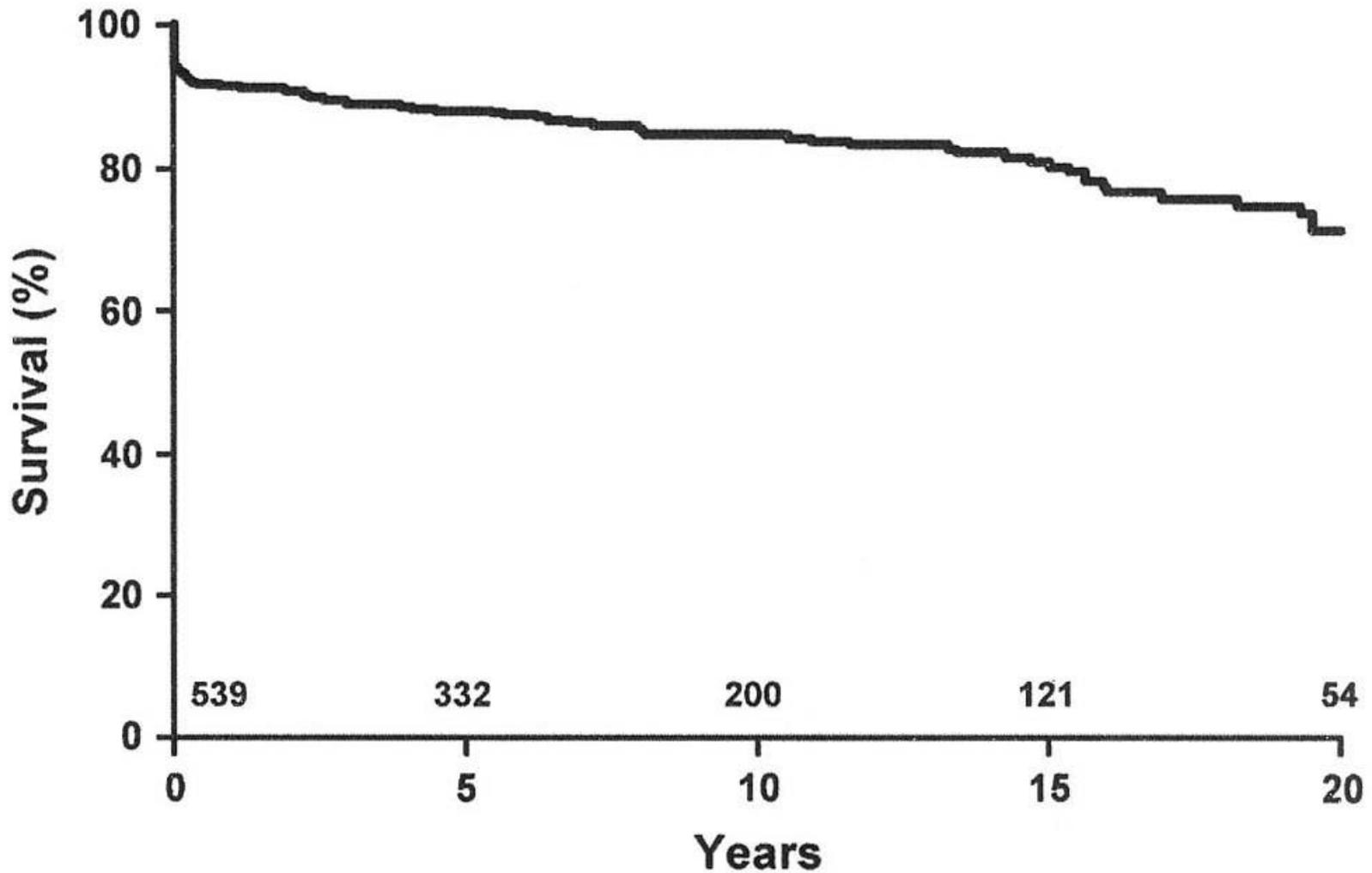
Ventilazione ?

- Bambini ed adulti sintomatici per cianosi:

Chiusura DIA con device?

- Ablazione della via anomala (minori successi rispetto ai pazienti senza Ebstein).

- Nei pazienti più gravi: chirurgia



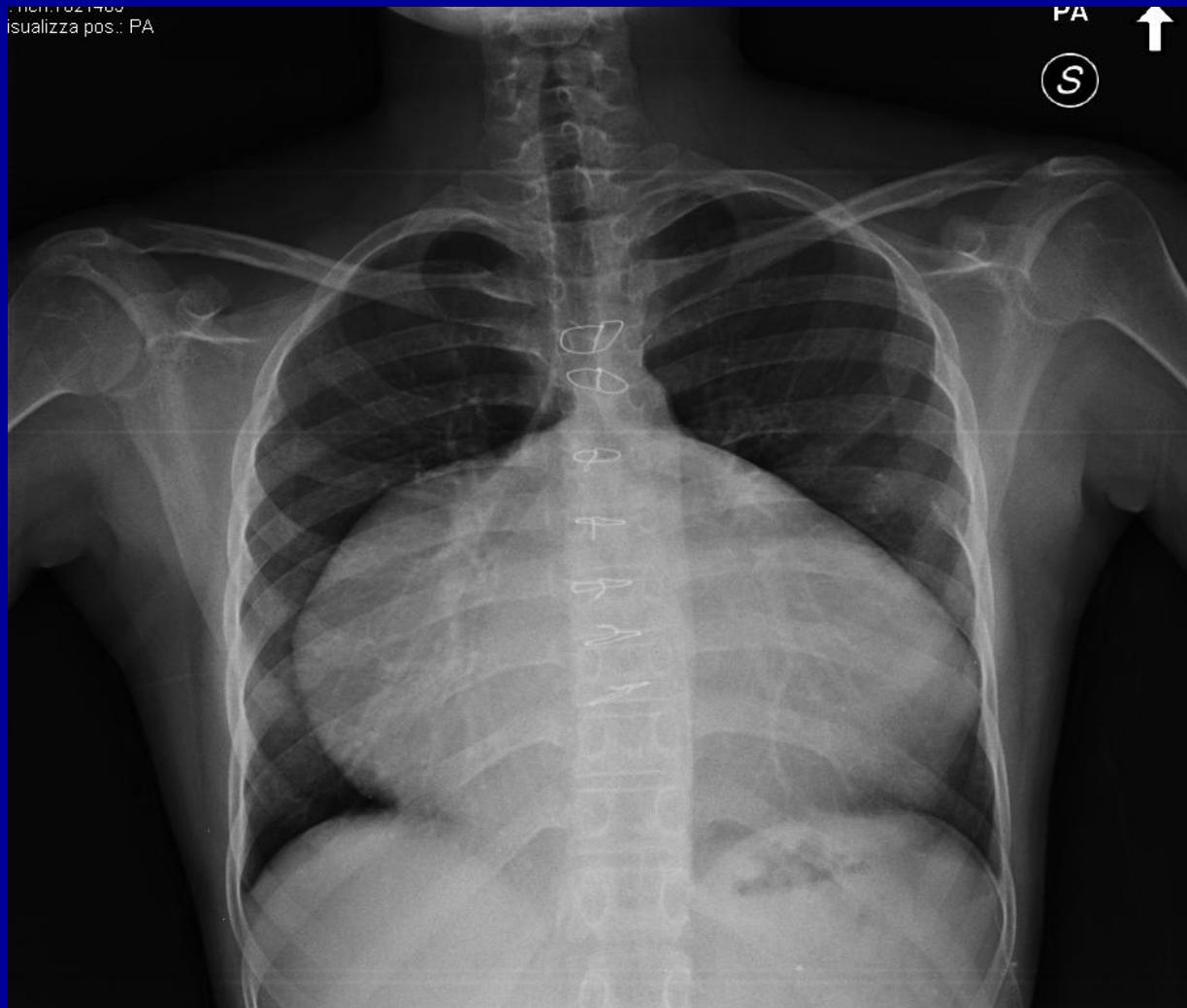
**Long-term survival for all 539 patients. Time 0 is the time of the first cardiac operation at the Mayo Clinic.**

# Ebstein's Anomaly Follow-up

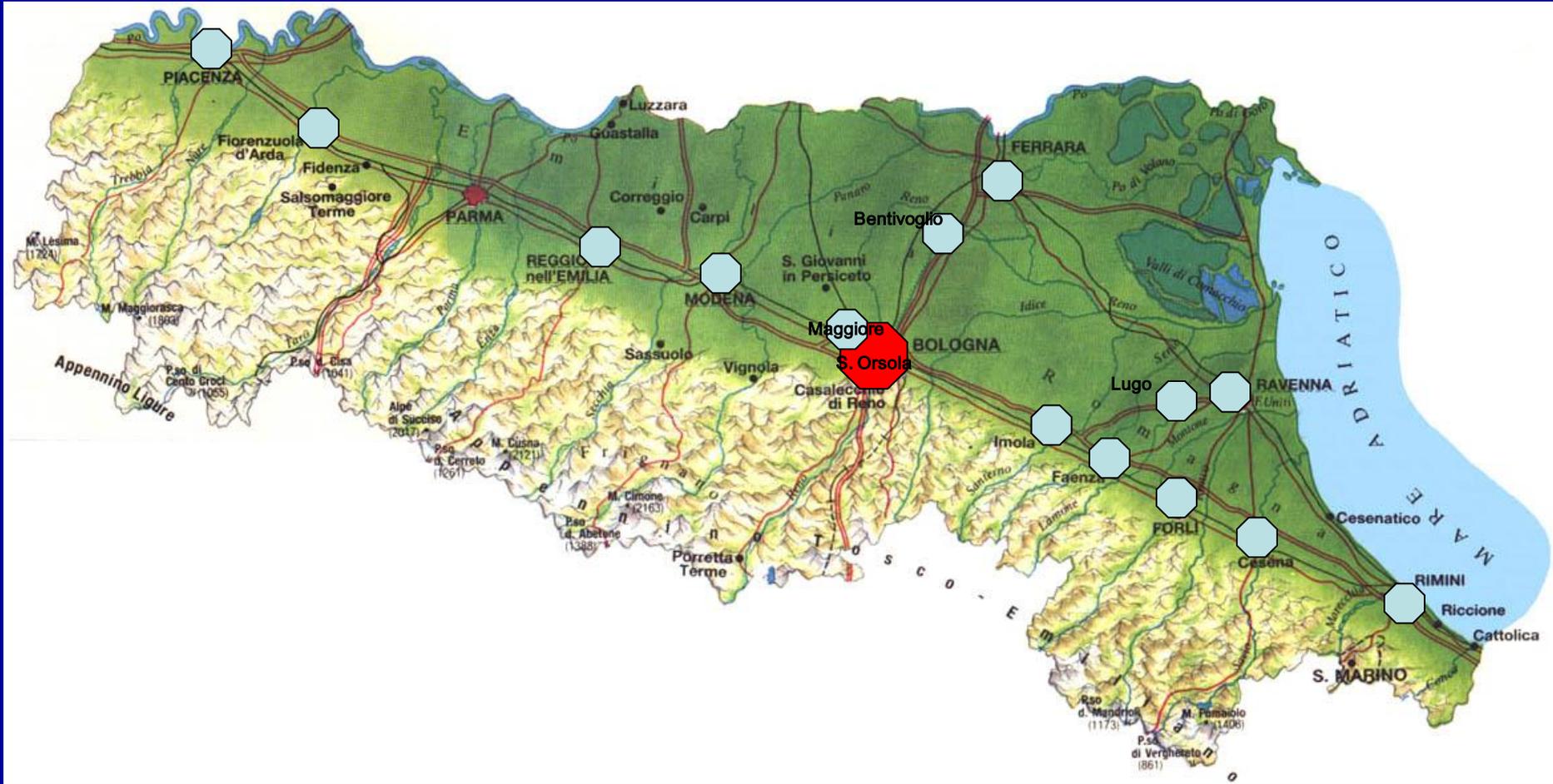
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- Regular follow-up (at least yearly) is required in all patients in specialized GUCH centers.
- Typical postoperative residual anomalies to look for are persisting or new TR, the usual complications after valve replacement, failure of RV or LV, residual atrial shunts, arrhythmias and higher grade heart blocks.

25 anni, Ebstein severo, FA, in lista per TX



# Una possibile soluzione organizzativa: l'Hub and Spoke

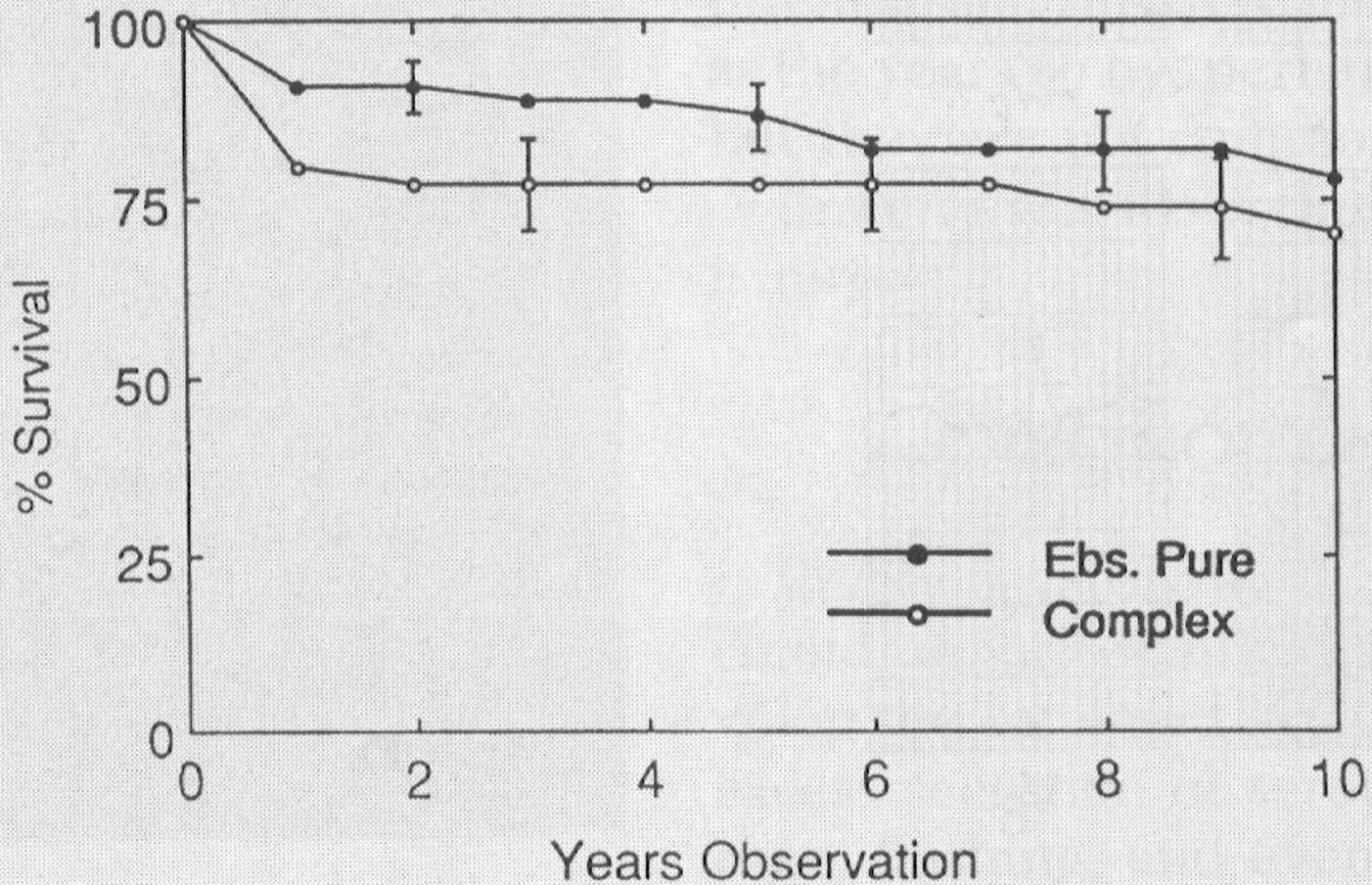


*Education, education, education!!!*



# Fontan Prognosis

- 10 year survival is 60-70%
- Late term complications include
  - Atrial flutter or fibrillation
  - Right atrial thrombus
  - Obstruction of the Fontan circuit
  - Ventricular dysfunction
  - Protein losing enteropathy

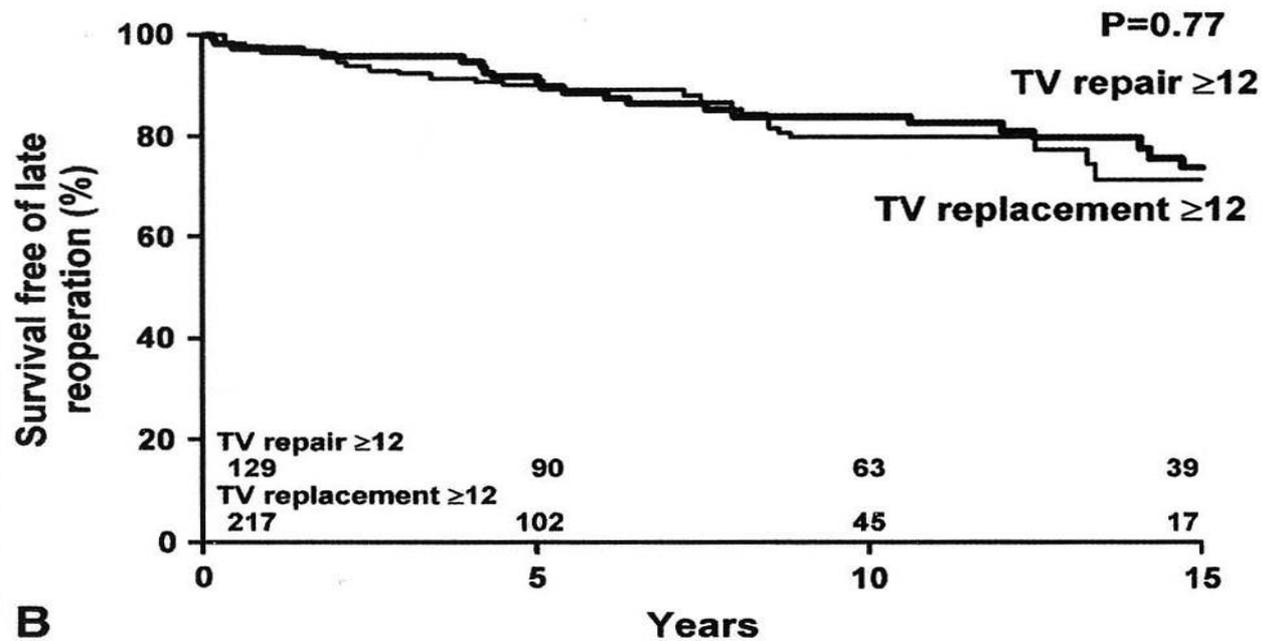
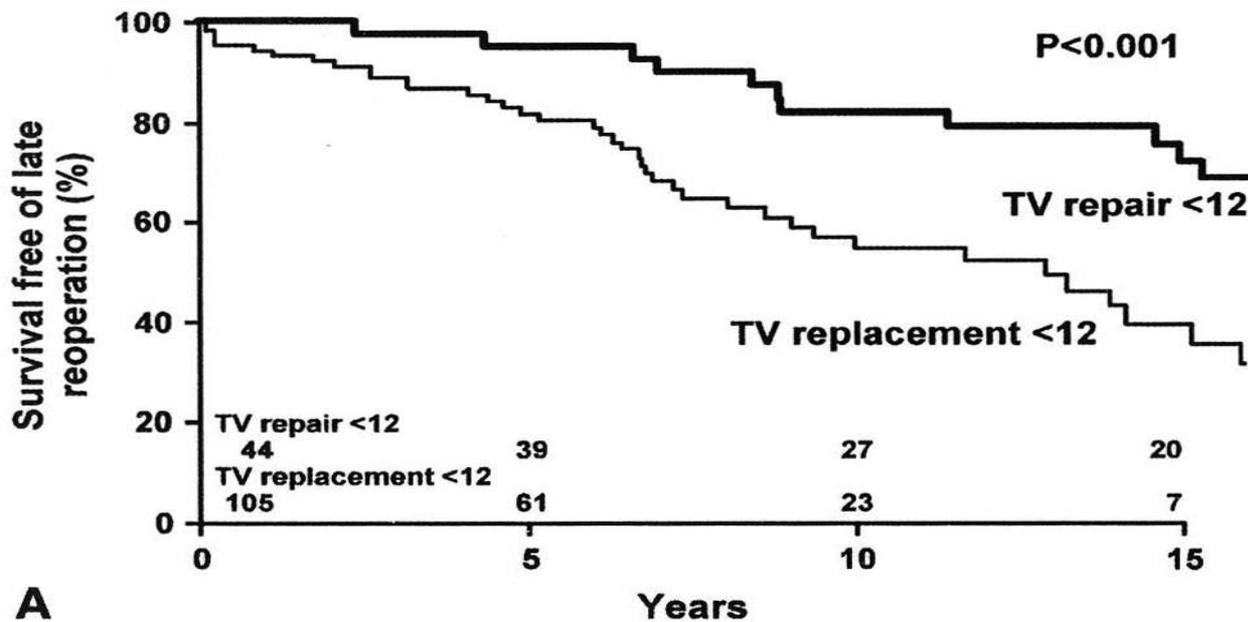


Life table comparing Ebstein's disease as a primary and secondary diagnosis

# **Anomalia di Ebstein**

## **Cosa cercare**

- ▣ I tre lembi**
- ▣ Come si attaccano e se sono trattenuti (tethering)**
- ▣ Il grado di displasia**
- ▣ L'estensione del lembo anteriore**



# Indications for Intervention in Ebstein's Anomaly

## Indications for surgical repair

- In patients with more than moderate TR and symptoms (NYHA class > II or arrhythmias) or deteriorating exercise capacity measured by CPET.
- If there is also an indication for tricuspid valve surgery, then ASD/PFO closure should be performed surgically at the time of valve repair.
- Surgical repair should be considered regardless of symptoms in patients with progressive right heart dilation or reduction of RV systolic function and/or progressive cardiomegaly on chest x-ray.

## Indications for catheter intervention

- Patients with relevant arrhythmias should undergo electrophysiologic testing followed by ablation therapy, if feasible, or surgical treatment of the arrhythmias in the case of planned heart surgery.
- In the case of documented systemic embolism likely caused by paradoxical embolism, isolated device closure of ASD/PFO should be considered.
- If cyanosis (oxygen saturation at rest < 90%) is the leading problem, isolated device closure of ASD/PFO may be considered but requires careful evaluation before intervention.

Class <sup>a</sup>	Level <sup>b</sup>
I	C
I	C
IIa	C
I	C
IIa	C
IIb	C

a = class of recommendation. b = level of evidence.

ASD = atrial septal defect; CPET = cardiopulmonary exercise testing; NYHA = New York Heart Association; PFO = patent foramen ovale; RV = right ventricle; TR = tricuspid regurgitation; VSD = ventricular septal defect.