

# Chirurgia delle cardiopatie congenite nell'adulto

*Milano, 2012*

# Una nuova popolazione di pazienti



## Anni '70

- Solo il 10% dei bambini operati per CHD raggiungeva l'età adulta



## Anni '80

- Dal 1980 circa l' 85% dei bambini affetti da cardiopatie congenite CHD raggiunge l'adolescenza e l'età adulta

# Entità del fenomeno



## In USA nel 2000

- 800.000 GUCH
- 400 mila ogni anno richiedono un controllo in centri regionali
- Aumento spese da \$691 million to \$3.16 billion



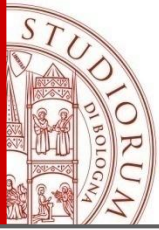
## In Europa nel 2000

- 1.000.000 di GUCH

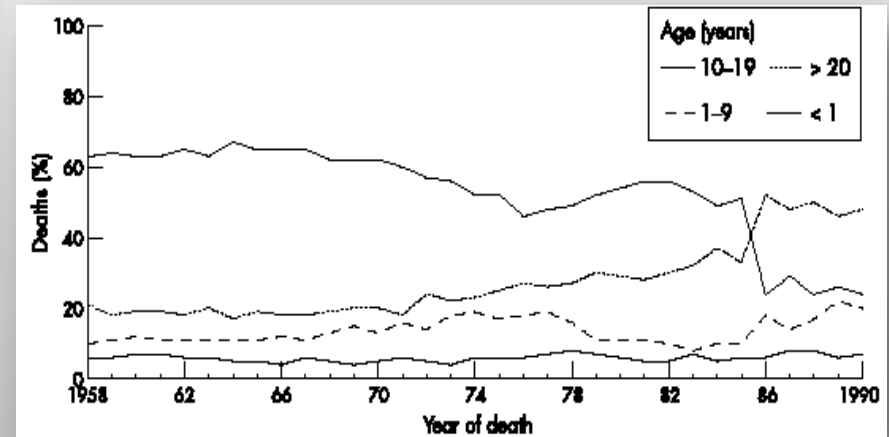
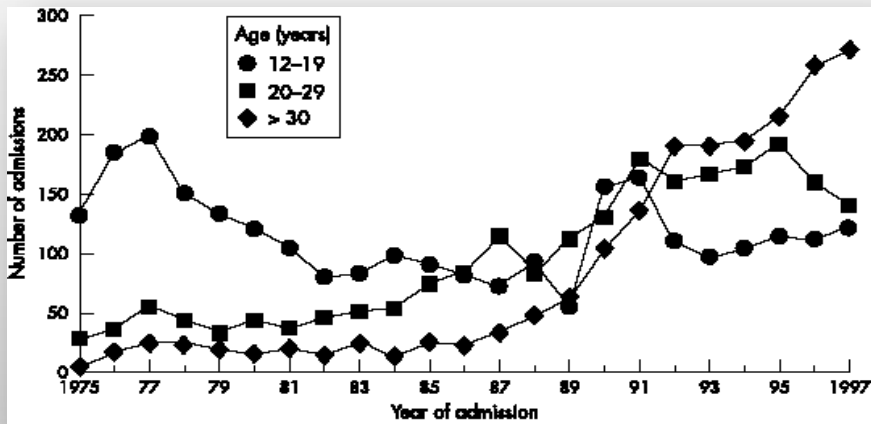


## In Italia nel 2000

- 75.000 GUCH



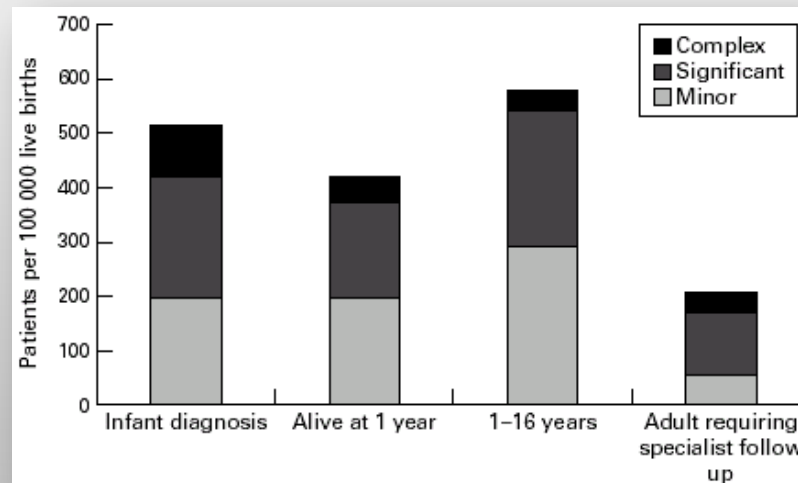
# I cambiamenti



Heart 2002;88(Suppl I):i1-i14

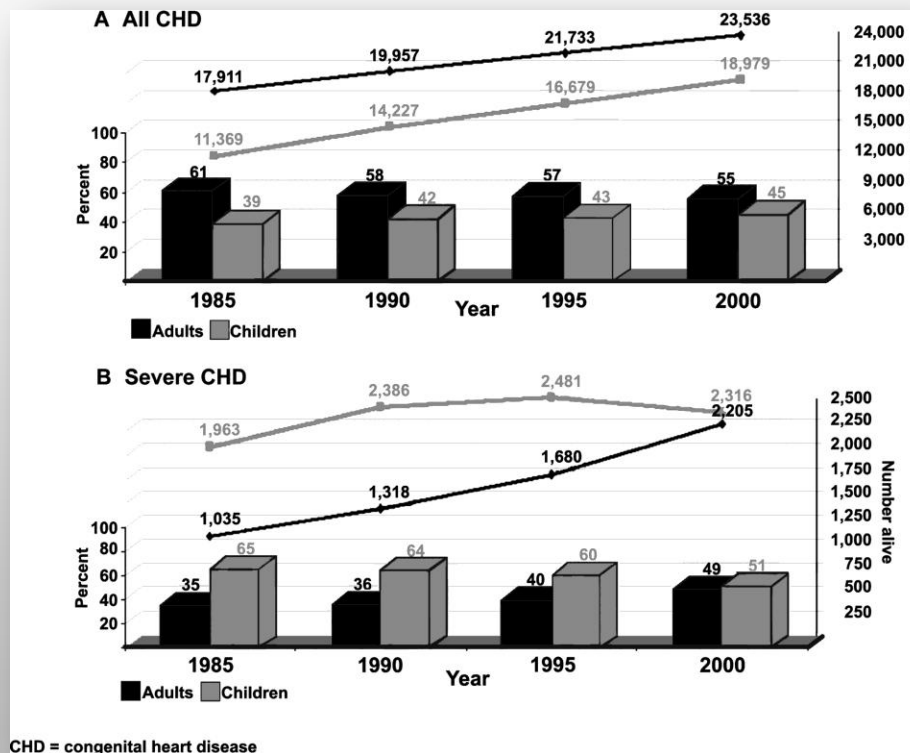
# Tipologia delle cardiopatie

- 20-25% Cardiopatie molto complesse
- 35-40% Complesse
- 35-40% Semplici

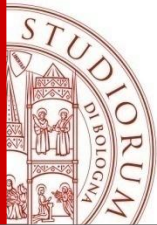


Heart 2002;88(Suppl I):i1-i14

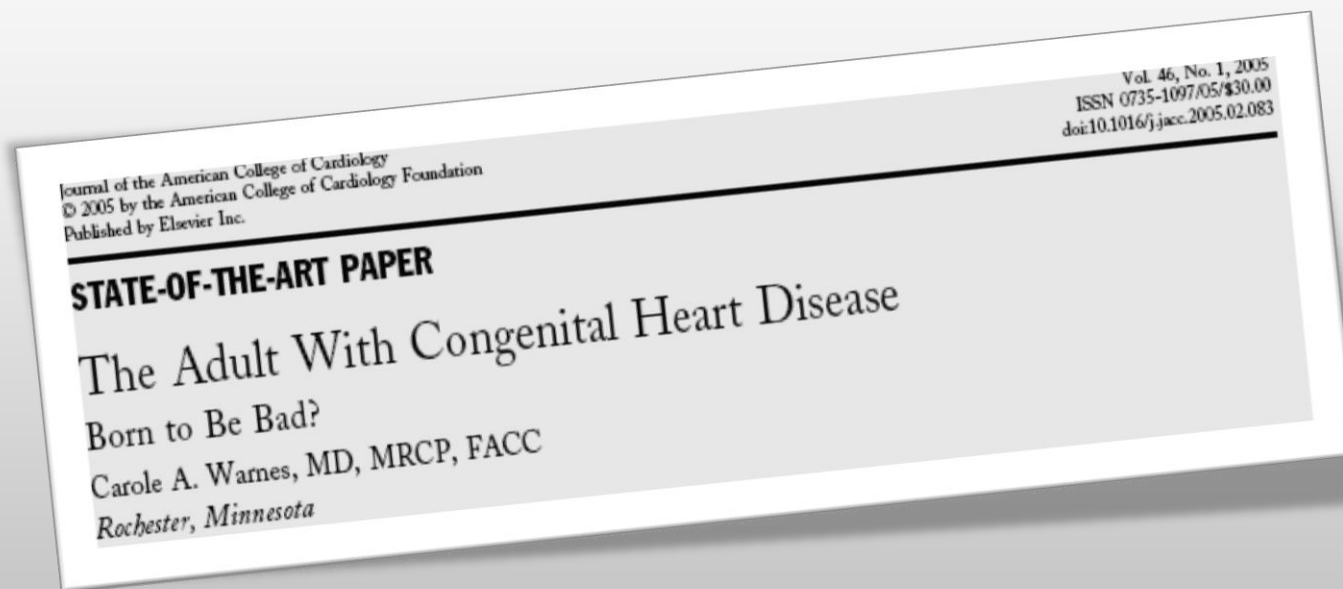
# Tipologia delle cardiopatie

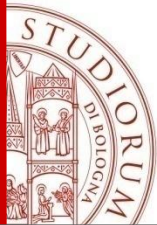


Marelli, A. J. et al. Circulation 2007;115:163-172



# Una prima domanda





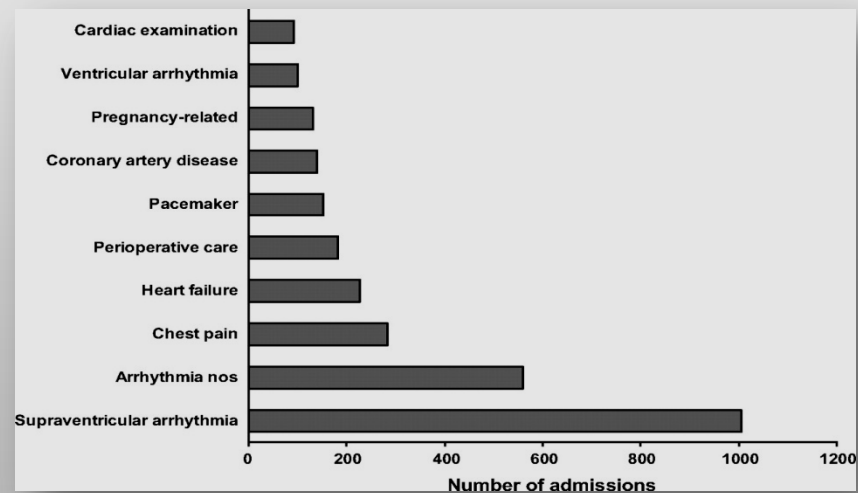
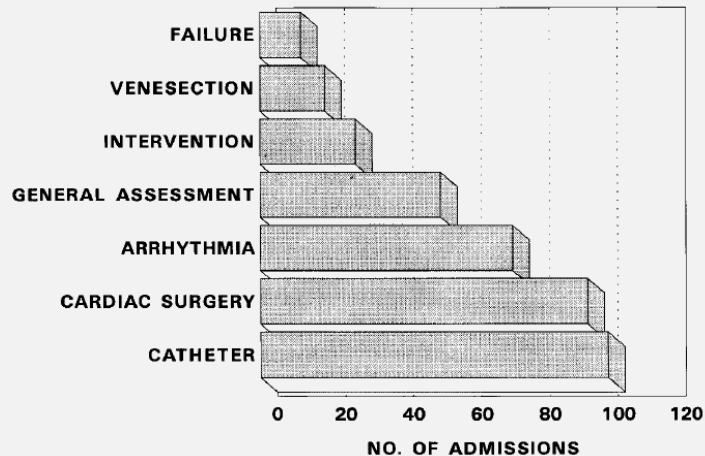
# Una prima risposta

*Annu. Rev. Med. 1997, 48:283-93  
Copyright © 1997 by Annual Reviews Inc. All rights reserved*

## MANAGEMENT OF ADULTS WITH CONGENITAL HEART DISEASE: An Increasing Problem

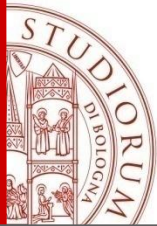
*Jane Somerville, MD, FRCP, FACC, FESC*

GUCH Unit, Royal Brompton Hospital, Sydney Street, London, SW3 6NP, United Kingdom



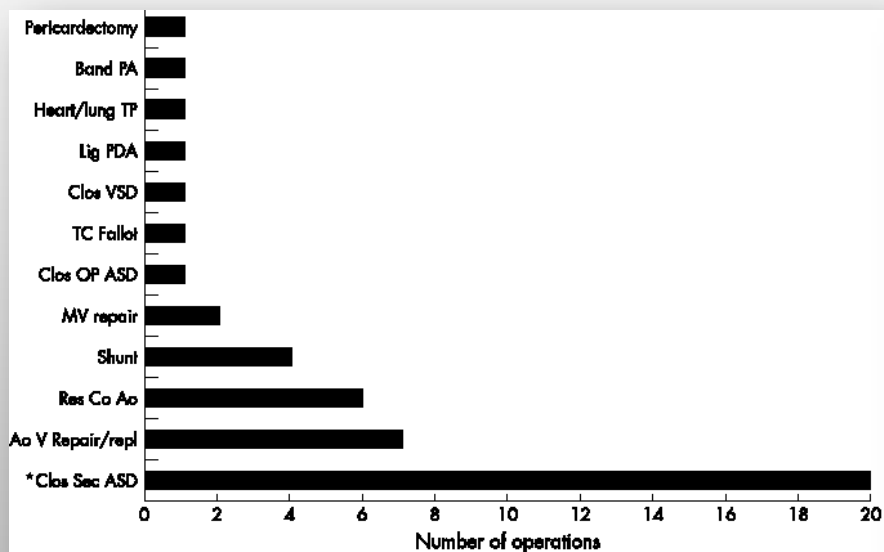
Verheugt C L et al. Heart 2010;96:872-878



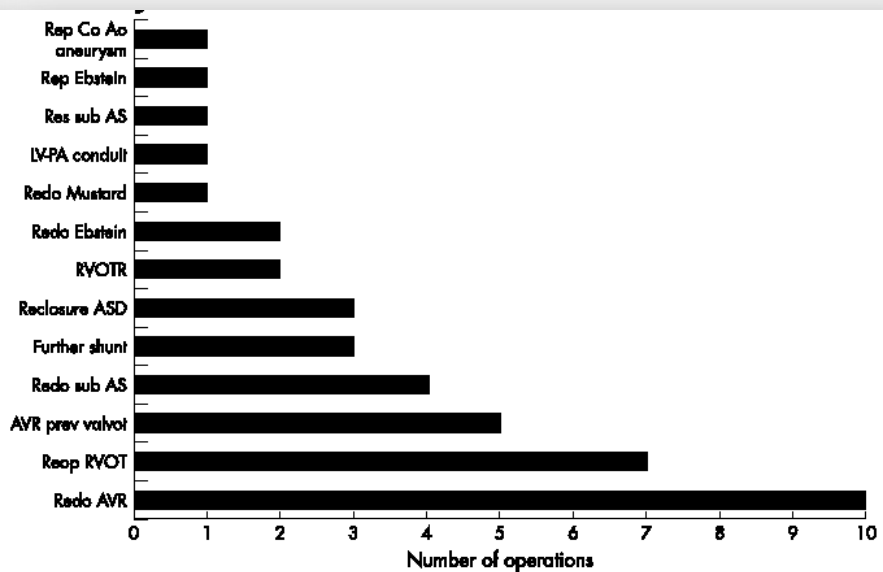


# Correggere è Curare ?

## Primo intervento



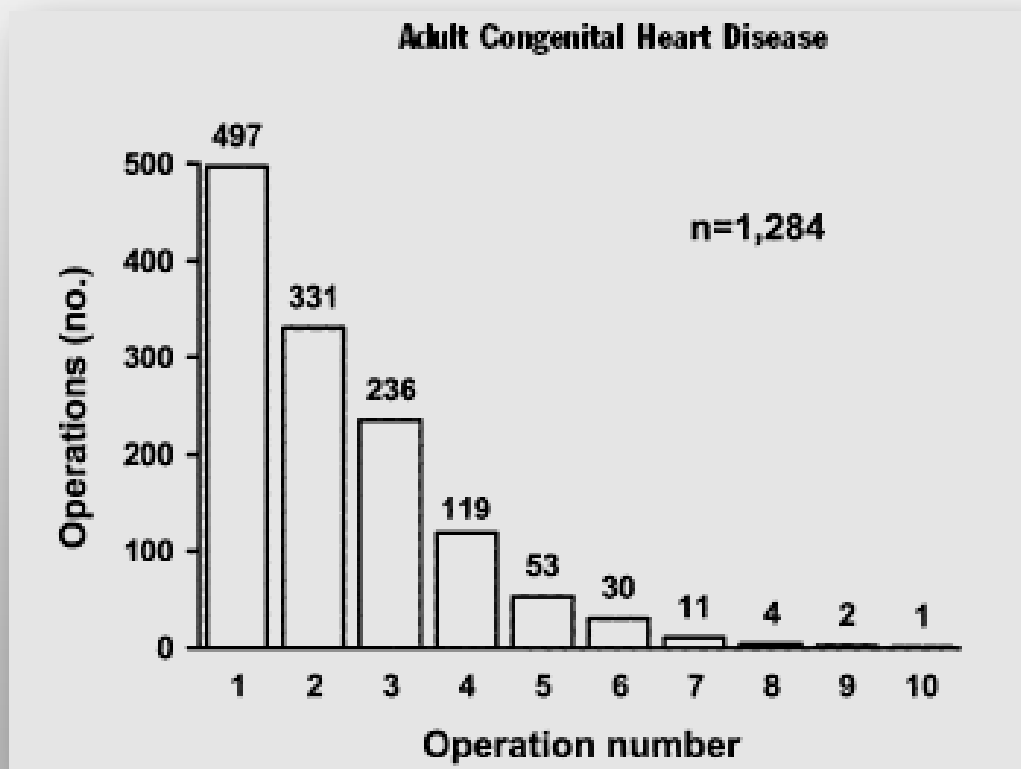
## Reintervento



Heart 2002;88(Suppl I):i1-i14

# I reinterventi

Il 20% dei GUCH rientra in ospedale per un reintervento

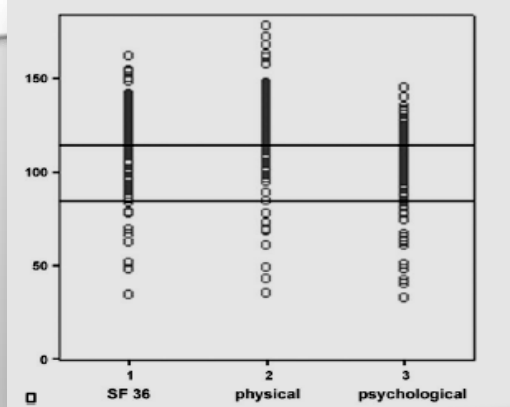


# La qualità di vita

## Quality of life of grown-up congenital heart disease patients after congenital cardiac surgery<sup>☆</sup>

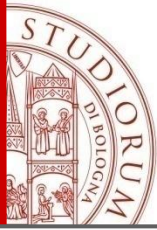
Ophélie Loup<sup>a</sup>, Catherina von Weissenfluh<sup>a</sup>, Brigitta Gahl<sup>a</sup>, Markus Schwerzmann<sup>b</sup>, Thierry Carrel<sup>a</sup>, Alexander Kadner<sup>a,\*</sup>

<sup>a</sup>Department of Cardiovascular Surgery, University Hospital Berne, Freiburgstrasse, 3010 Bern, Switzerland



**Congenital heart disease never goes away, even when it has been 'treated': the adult with congenital heart disease**

Aarti Hejmadi Bhat and David J. Sahn

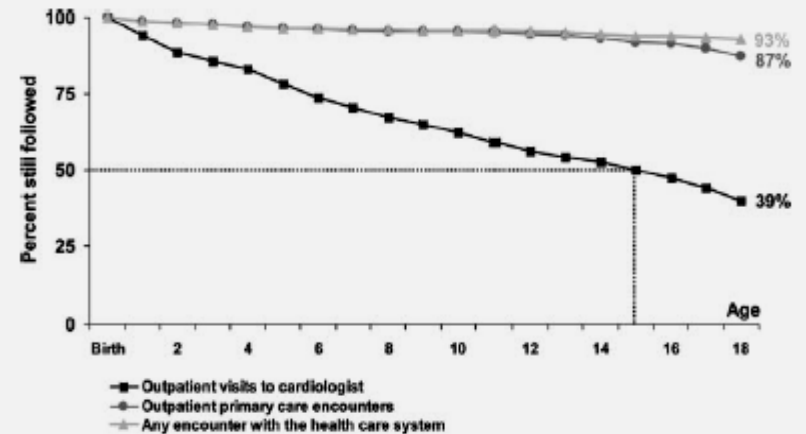
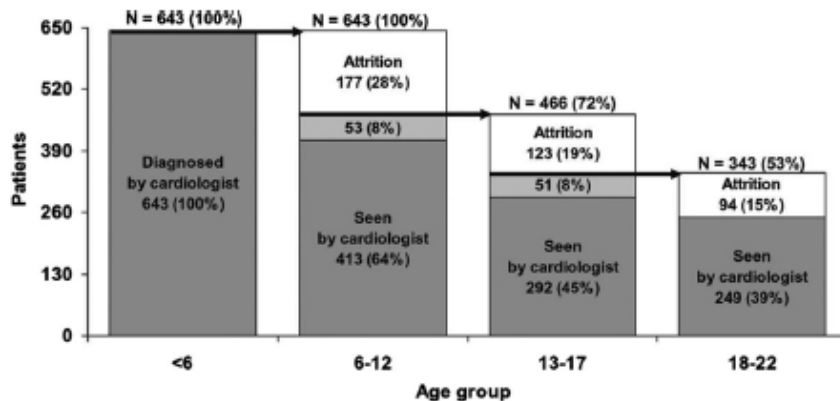


# Cosa non dovrebbe succedere...

## Children and Adults With Congenital Heart Disease Lost to Follow-Up: Who and When?

Andrew S. Mackie, Raluca Ionescu-Ittu, Judith Therrien, Louise Pilote, Michal Abrahamowicz and Ariane J. Marelli

*Circulation* 2009;120:302-309; originally published online Jul 13, 2009;  
DOI: 10.1161/CIRCULATIONAHA.108.830164





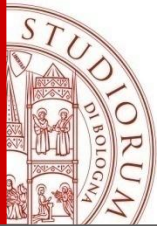
# La necessità di essere seguito

## **Relation Between Right Ventricular Structural Alterations and Markers of Adverse Clinical Outcome in Adults With Systemic Right Ventricle and Either Congenital Complete (After Senning Operation) or Congenitally Corrected Transposition of the Great Arteries**

Alessandro Giardini, MD<sup>a,\*</sup>, Luigi Lovato, MD<sup>b</sup>, Andrea Donti, MD<sup>a</sup>, Roberto Formigari, MD<sup>a</sup>,  
Guido Oppido, MD<sup>a</sup>, Gaetano Gargiulo, MD<sup>a</sup>, Fernando Maria Picchio, MD<sup>a</sup>,  
and Rossella Fattori, MD<sup>b</sup>

## **Usefulness of Cardiopulmonary Exercise to Predict Long-Term Prognosis in Adults With Repaired Tetralogy of Fallot**

Alessandro Giardini, MD<sup>a,\*</sup>, Salvatore Specchia, MD<sup>b</sup>, Theresa Ann Tacy, MD<sup>c</sup>,  
Gloria Coutsoumbas, MD<sup>b</sup>, Gaetano Gargiulo, MD<sup>a</sup>, Andrea Donti, MD<sup>a</sup>, Roberto Formigari, MD<sup>a</sup>,  
Marco Bonvicini, MD<sup>a</sup>, and Fernando Maria Picchio, MD<sup>a</sup>



# Le tecniche diagnostiche

Pediatr Radiol (2010) 40:885–894  
DOI 10.1007/s00247-010-1622-x

REVIEW

## Post-operative cardiac lesions after cardiac surgery in childhood

Phalla Ou • Laurence Iserin • Oliver Raisky •  
Pascal Vouhe • Francis Brunelle • Daniel Sidi •  
Damien Bonnet

KARDIOL MED (2011) 112:000–000  
DOI 10.1007/s11547-007-0171-3

CARDIAC RADIOLOGY  
CARDIORADIOLOGIA

Role and effectiveness of cardiovascular magnetic resonance in the diagnosis, preoperative evaluation and follow-up of patients with congenital heart diseases

*Ruolo ed utilità clinica della RM nella diagnosi, nella valutazione pre-operatoria e nel follow-up delle cardiopatie congenite*

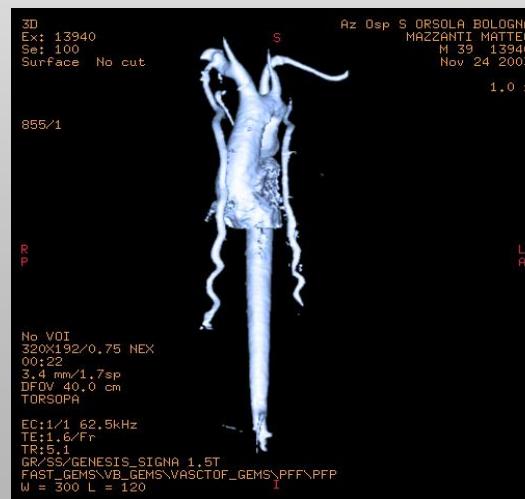
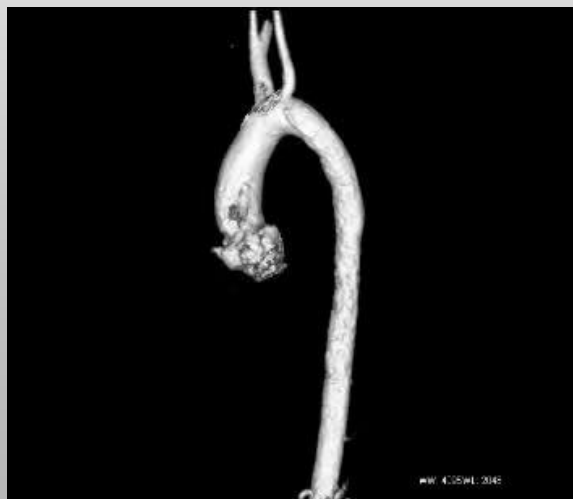
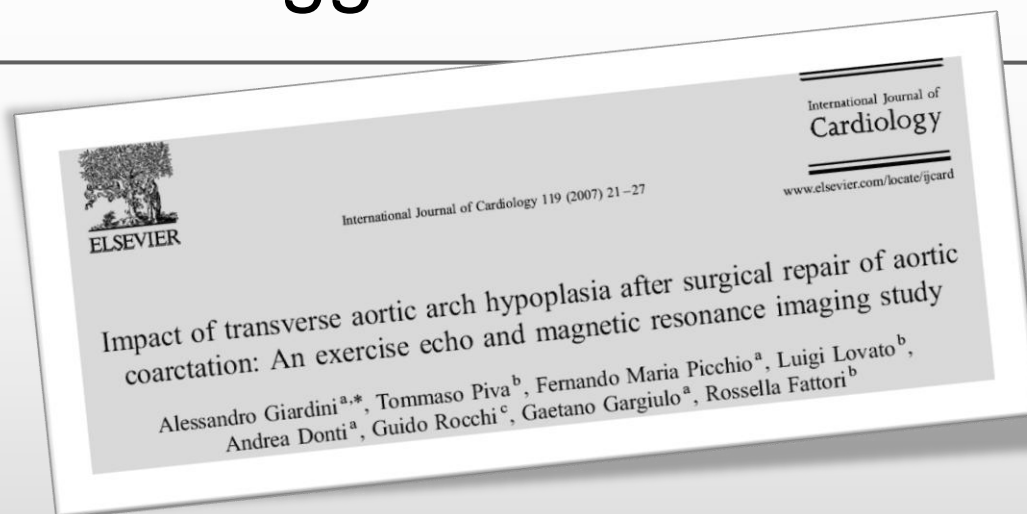
L. Lovato<sup>1</sup> • A. Giardini<sup>2</sup> • C. La Palombara<sup>1</sup> • V. Russo<sup>1</sup> • V. Gostoli<sup>1</sup> • G. Gargiulo<sup>3</sup>  
E.M. Picchio<sup>2</sup> • R. Fattori<sup>1</sup>



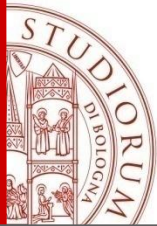
# Informazioni sempre più complete



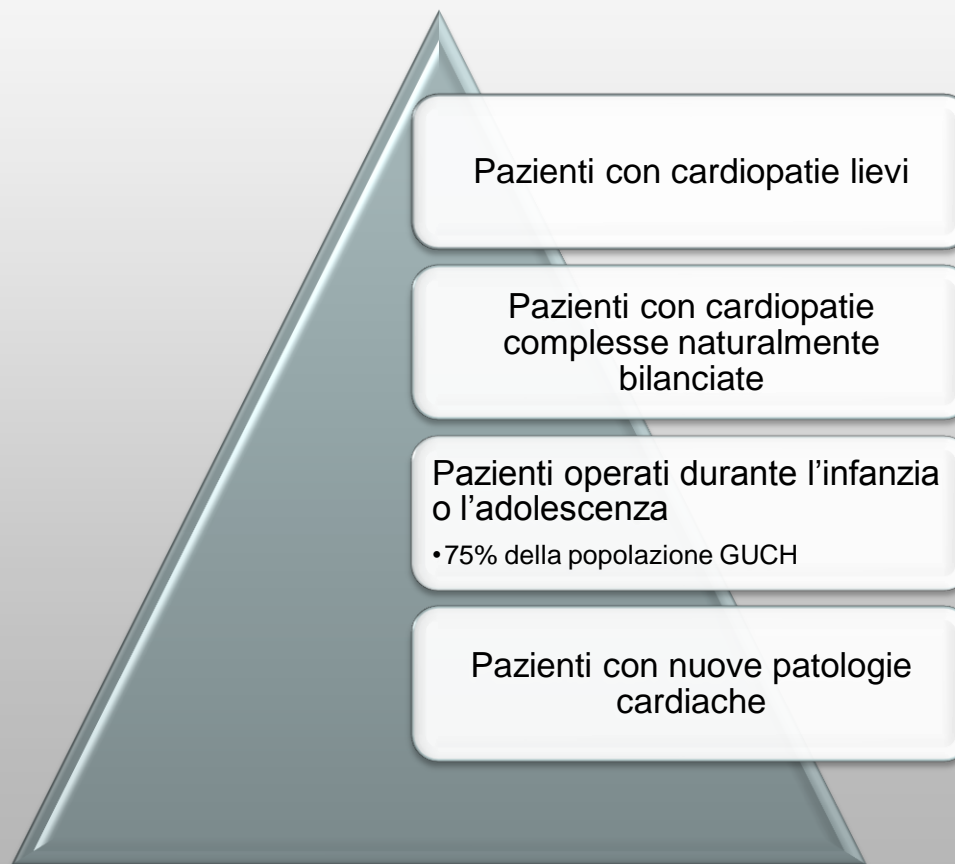
# Correggere è Curare?







# Una nuova popolazione



# Come dovrebbe essere



Centro cure  
primarie



Cardiologo



Centro regionale

- 2 milioni di ab
- Cardiologo pediatra
- Cardiologo Adulto
- Aritmologo
- Team Eco

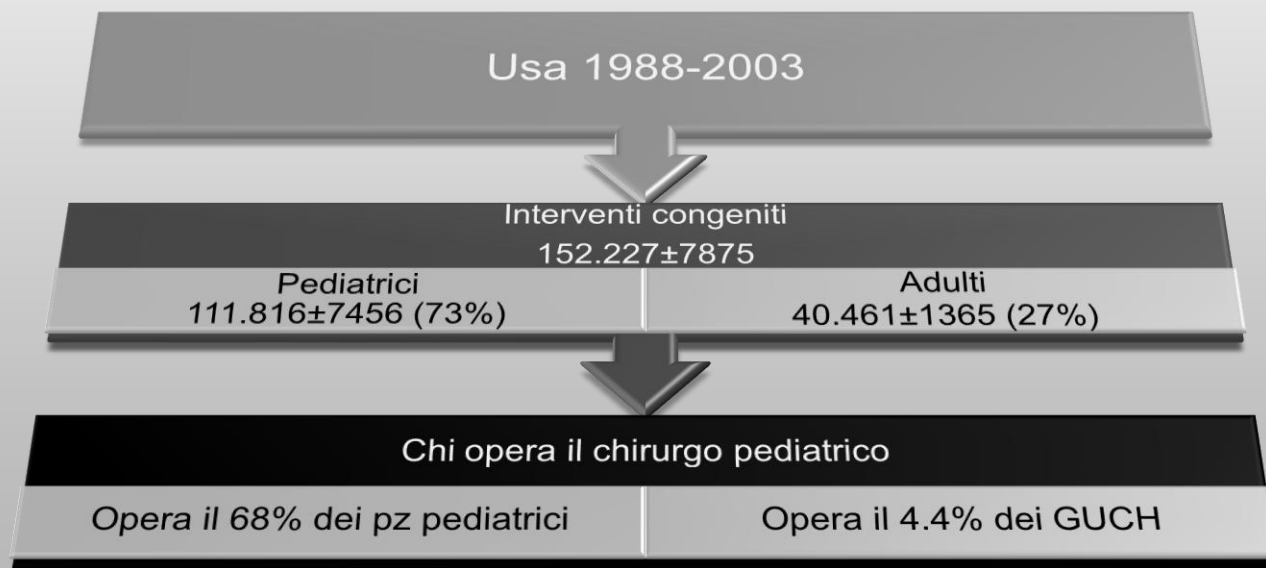


Centro Nazionale

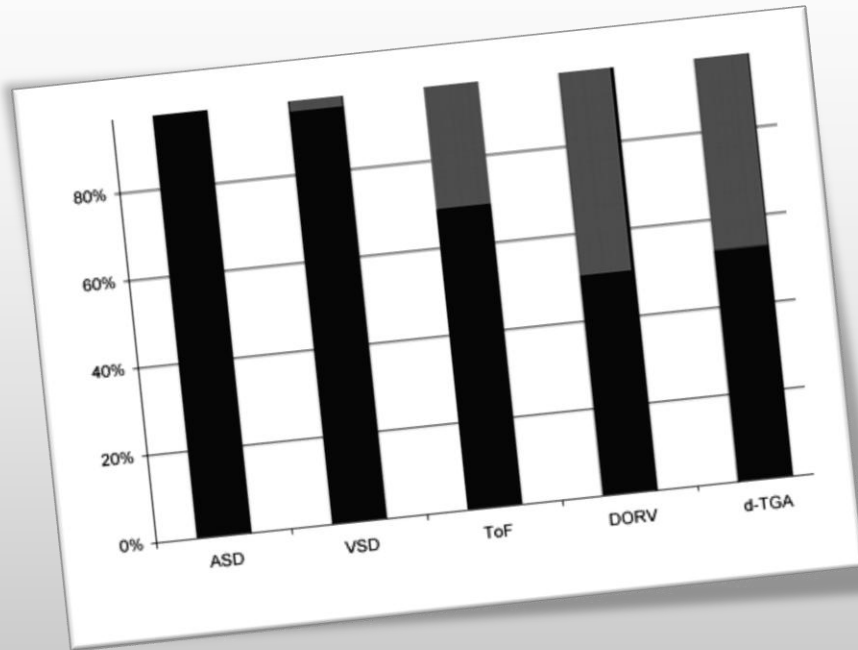
- 3-10 milioni di ab.
- Cardiochirurgia  
Pediatrica
- Cardiochirurgia Adulti
- Cardiologia Pediatrica  
ed Adulti III livello
- Centro Trapianti

# Chi deve operare

**National Practice Patterns for Management of Adult Congenital Heart Disease: Operation By Pediatric Heart Surgeons Decreases In-Hospital Death**  
Tara Karamlou, Brian S. Diggs, Thomas Person, Ross M. Ungerleider and Karl F. Welke  
*Circulation* 2008;118:2345-2352; originally published online Nov 7, 2008;

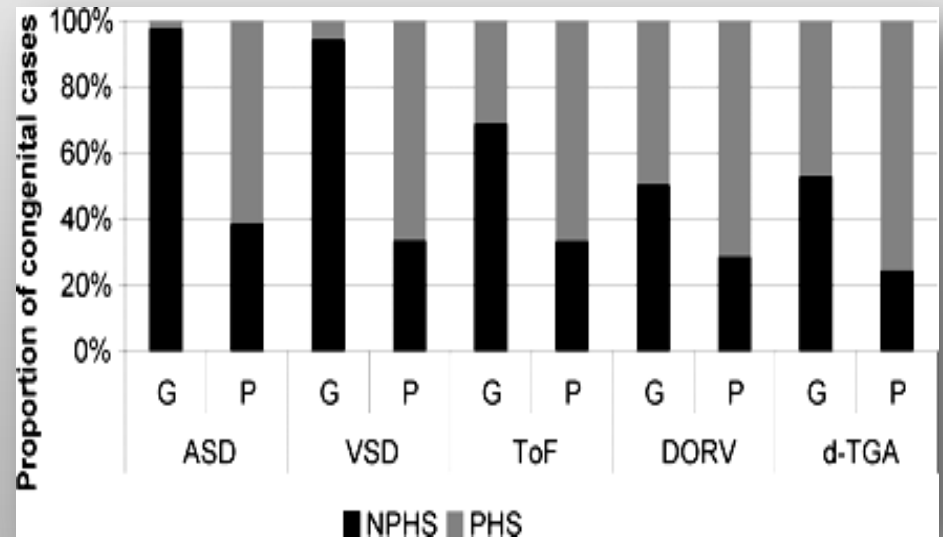


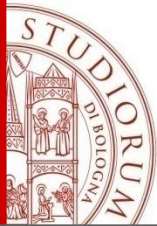
# Questione non definita



GUCH

Pediatrici e GUCH





# Una possibile indicazione ?

**Table 4. Multivariable Factors Associated With In-Hospital Mortality**

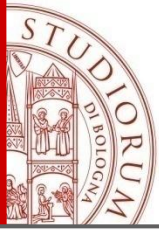
Variable	Odds Ratio	95% CI	P
Pediatric heart surgeon as dichotomous variable			
Nonpediatric heart surgeon	4.50	2.12–9.53	<0.0001
Female gender	1.32	1.05–1.64	0.0150
Diagnosis of VSD	2.71	1.75–4.20	<0.0001
Ostium primum ASD	0.18	0.06–0.55	0.0026
Coarctation of the aorta	0.19	0.09–0.41	<0.0001
Ostium secundum ASD	0.18	0.12–0.29	<0.0001
Congestive heart failure	6.65	2.90–15.24	<0.0001
PVD	1.89	1.17–3.07	0.0099
Renal failure	2.79	1.43–5.44	0.0027
Complicated diabetes mellitus	3.31	1.68–6.52	0.0005

**Table 5. Multivariable Factors Associated With Increased LOS and Total Hospital Charges**

Variable	Regression Coefficient±SE	P
Multivariable factors associated with increased LOS, d		
Lower annual percentage of pediatric heart cases	3.17±0.67	<0.001
Male gender	0.65±0.24	0.006
VSD	2.98±1.06	0.005
Congestive heart failure	34.2±4.8	<0.001
Renal failure	8.08±2.53	0.001

## Mortalità

- Chirurghi Adulti 4.84%
- Chirurghi Pediatrici 1.87%



# Cardiopatìa semplice

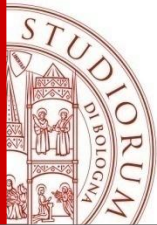


Alma Mater Studiorum  
University of Bologna

S.Orsola-Malpighi Hospital

Pediatric Cardiac Surgery and GUCH Unit  
Prof. G.D.Gargiolo

ASD, PAPVC, pulmonary artery dilatation and ... other things



# I GUCH, una sfida per il team

## Problemi

### Funzionalità miocardica

- Cardioplegia
- Conduzione della CEC

### Risternotomie

- Cannulazione femorale
- Cannulazione succlavia

### Conduzione della CEC

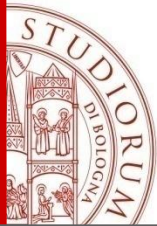
- Ultrafiltrazione
- Collaterali sistemico-polmonari
- Bassi flussi
- Perfusioni cerebrali

### Gestione del sangue

- Autotrasfusioni
- Piastrine
- Ultrafiltrazione
- Aprotinina

### Conduzione dell'anestesia

- Farmaci differenti in base alla patologia
- Corretto monitoraggio del paziente



# Pazienti ad alto rischio

Rischio medio 8 – 10 %

**Chirurgia più complessa**

**Rischio aumentato rispetto alle correzioni fatte nell'infanzia**

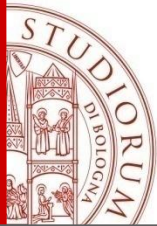
**Rischio proporzionale al numero di reinterventi**

**Il rischio aumenta con l'aumentare dell'età**

**Maggior rischio**

- di aritmie post-operatorie,
- di sanguinamento,
- di depressione psichica





# Risultati

## Midterm results of surgical intervention for congenital heart disease in adults: An Italian multicenter study

Massimo A. Padalino, MD,<sup>a</sup> Simone Speggorin, MD,<sup>a</sup> Giulio Rizzoli, MD,<sup>a</sup> Giancarlo Crupi, MD,<sup>b</sup> Vladimiro L. Vida, MD,<sup>a</sup>  
 Massimo Bernabei, MD,<sup>c</sup> Gaetano Gargiulo, MD,<sup>d</sup> Alessandro Giamberti, MD,<sup>e</sup> Francesco Santoro, MD,<sup>f</sup>  
 Carlo Vosa, MD,<sup>g</sup> Giuseppe Pacifico, MD,<sup>g</sup> Raffaele Calabrò, MD,<sup>g</sup> Luciano Daliento, MD,<sup>h</sup> and Giovanni Stellin, MD<sup>a</sup>

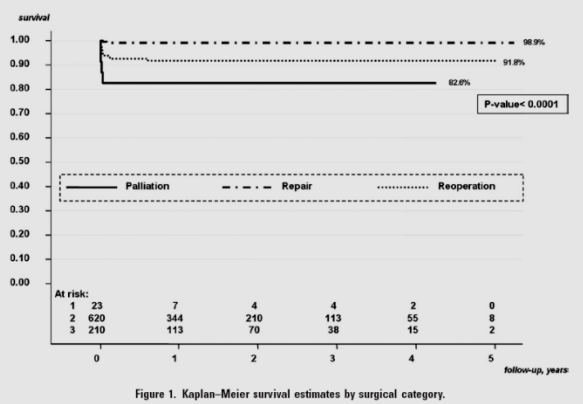


Figure 1. Kaplan-Meier survival estimates by surgical category.

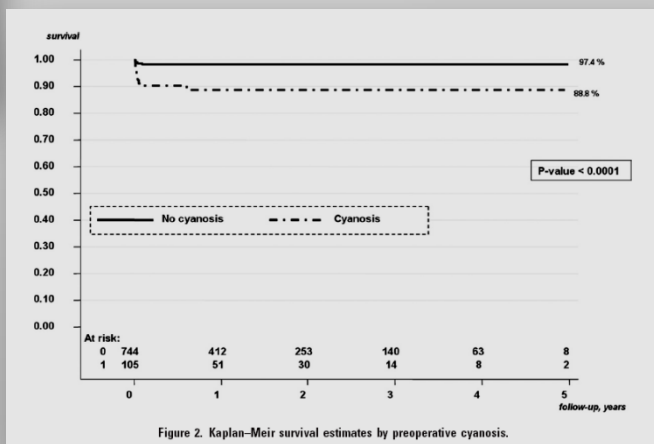
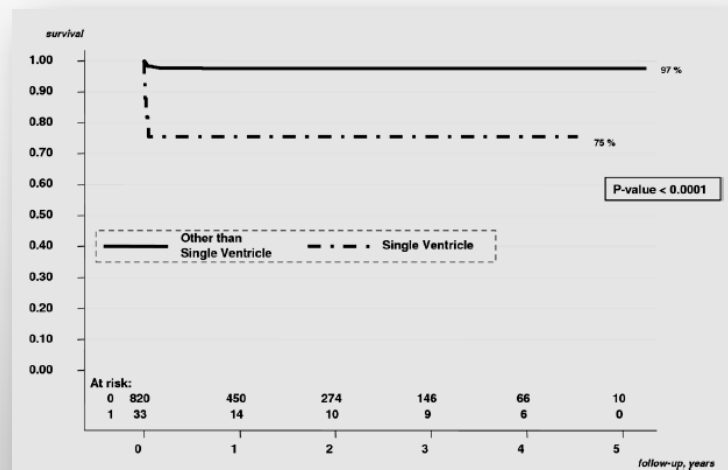
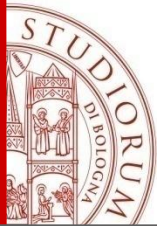


Figure 2. Kaplan-Meier survival estimates by preoperative cyanosis.



# Ruolo della chirurgia

## Primo intervento

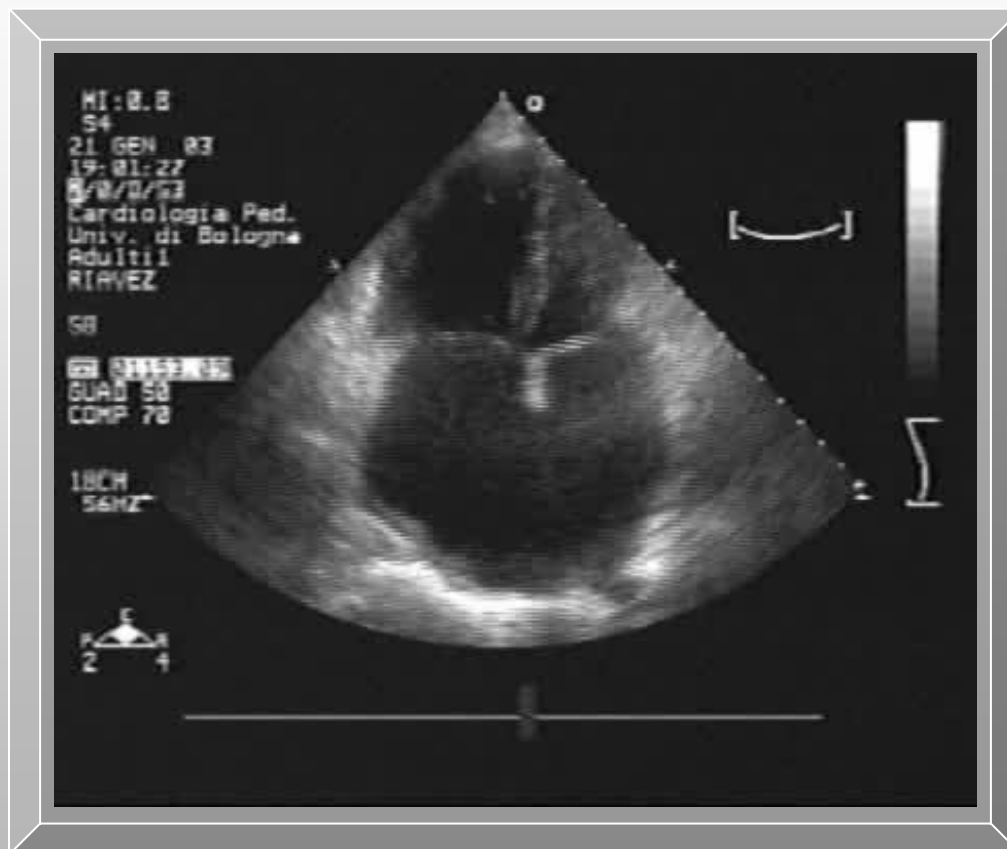
- CHD non scoperta prima
- CHD considerata non severa
- CHD severa bilanciata

## Reinterventi

- Correzione definitiva
- Altre procedure palliative
- Correzione delle sequele post-intervento
- Riparazione di nuove lesioni (coronarie-valvole-aorta)

# Cardiopatía in storia naturale

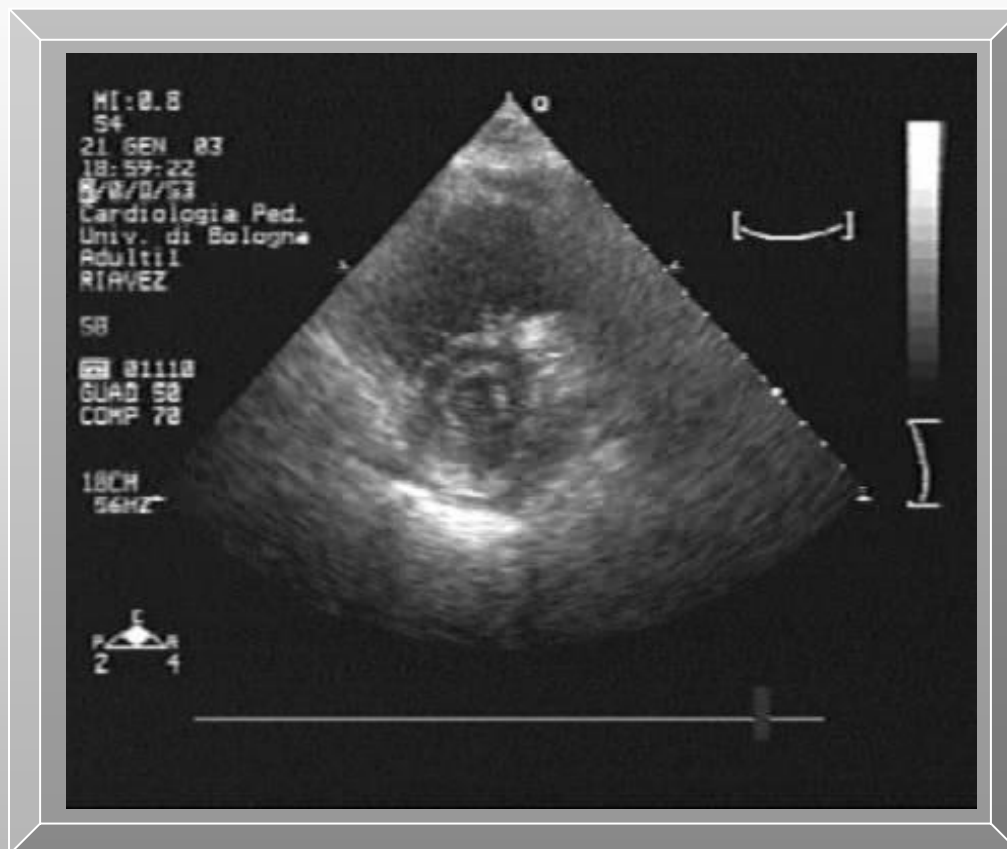
## CHD “semplici”



Paziente di anni 67 con diagnosi di “semplice” DIA ostium secundum...

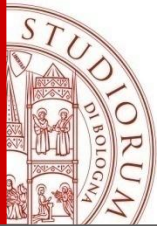
# Cardiopatía in storia naturale

## CHD "semplici"



...sezioni dx dilatate, Vsx restrittivo, ... cat. Cardiaco...PAP media 26 mmHg





# Chirurgia

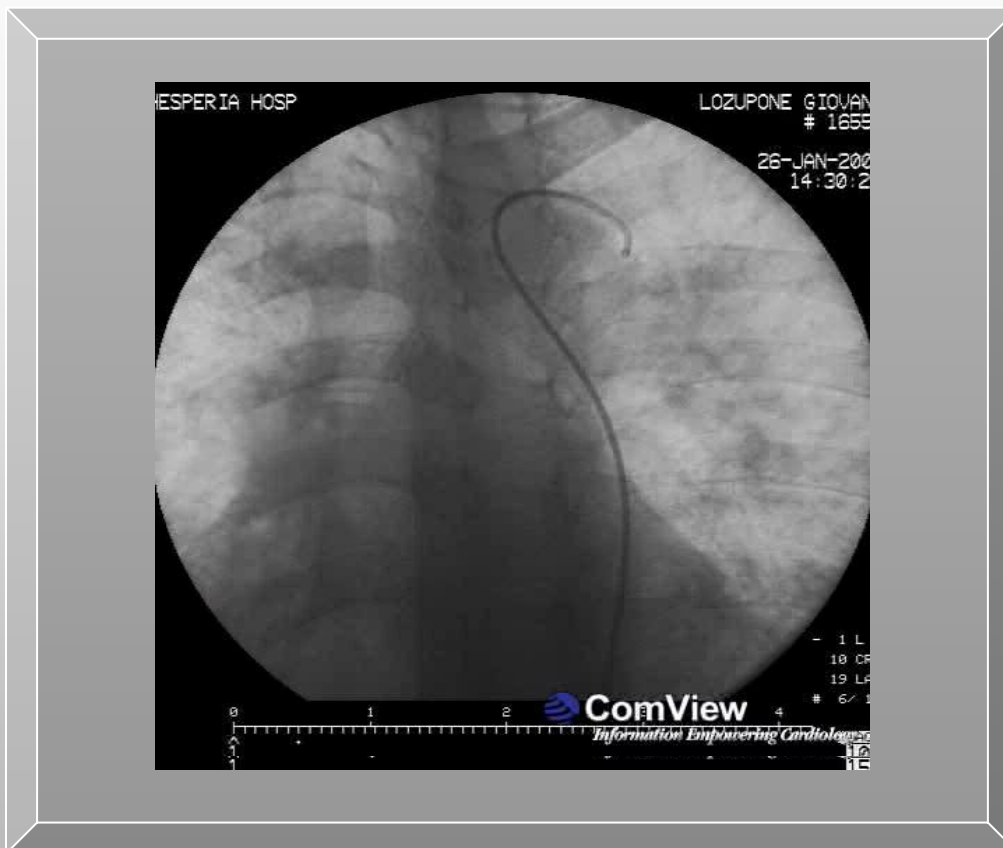
---

## CHD “semplici”

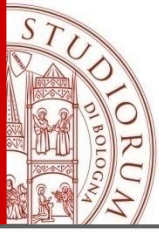
- Intervento “semplice” di chiusura DIA con patch
- Maze operation
- Plastica Tricuspidale
- Decorso post-operatorio complicato da ipertensione polmonare e bassa portata, pz svezzata dal respiratore con NO.
- Degenza 55 gg

# Cardiopatie palliate

## CHD “complesse”



Tetralogia di Fallot: 1974 palliazione con BT classica sx  
2002 dopo 28 anni e 17 anni di danza professionistica...cianosi marcata e segni di  
scompenso



# Chirurgia

---

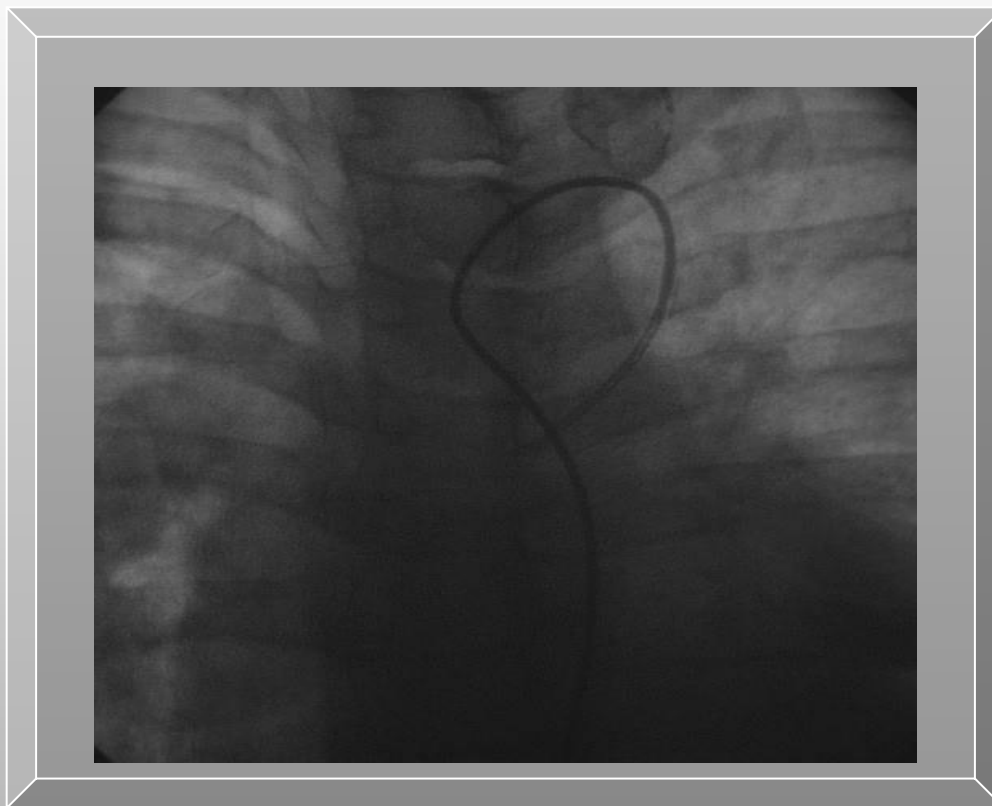
## CHD “complesse”

- Correzione completa
- Homograft Vdx-AP
- Chiusura BT
- Decorso post-operatorio non complicato
- Attualmente paziente asintomatico



# Il desiderio di maternità...

CHD “molto complesse”



Donna di 32 anni con Trasposizione dei grossi vasi, stenosi polmonare e DIV multipli. Cianosi marcata e dilatazione Vdx

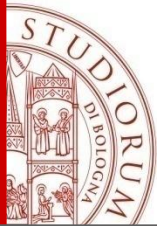
1977

Waterston

-----

1979

BT a sx



# Chirurgia

---

## CHD “molto complesse”

- Correzione completa secondo Rastelli
- Homograft Vdx-AP
- Chiusura DIV accessori
- Plastica aortica in sede di Waterston
- Chiusura BT
- Decorso post-operatorio non complicato

# Storia naturale delle correzioni

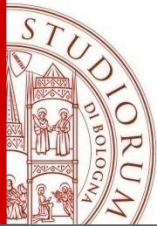
## CHD “molto complesse”



Paziente di 32 aa  
diagnosi di ToF,  
CATCH 22.

1980 Correzione completa con patch trans-anulare

2003 Insufficienza polmonare massiva – Vdx dilatato – Vsx dilatato – FEsx ridotta – Insufficienza aortica severa e dilatazione Ao Asc. 7 cm



# Chirurgia

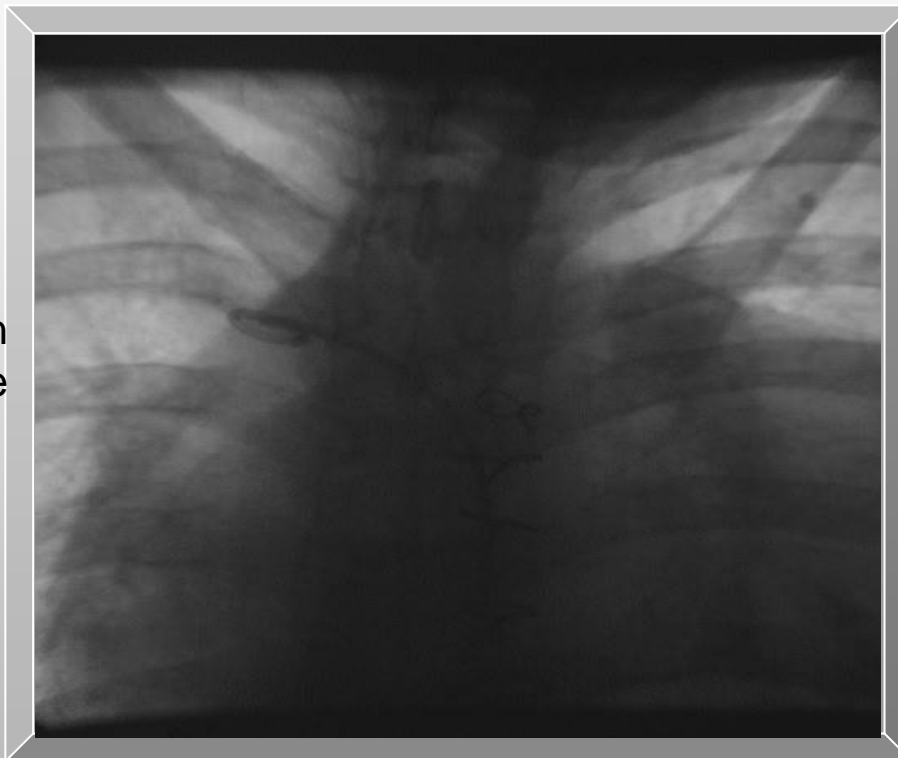
---

## CHD “molto complesse”

- Interposizione di homograft Vdx-AP
- Sostituzione aorta ascendente con protesi composita (tubo valvolato)
- Reimpianto coronarico
- Decorso post-operatorio non complicato
- Paziente attualmente asintomatico

# Storia naturale Fontan

CHD “molto complesse”



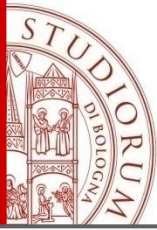
Paziente di 22 anni con diagnosi di AT, TGV e stenosi polmonare.

1986 Fontan classica

1998- 2001 Scopenso cardiaco dopo episodi di FFA

# Anastomosi cavo-polmonare totale





# Chirurgia

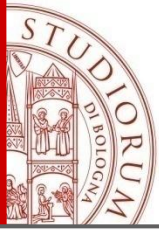
## CHD “molto complesse”

- Riconversione in cavo-polmonare totale extra-c.
- Plastica APsx
- Maze operation
- Impianto elettrodo PM epicardico definitivo
- Decorso post-operatorio non complicato
- Ritmo sinusale

# Coartazione aortica



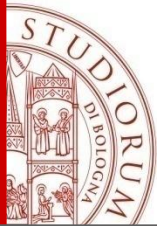




# Complicanze degli interventi

---





# Considerazioni finali

La chirurgia nei GUCH deve essere considerata necessaria e utile, in quanto modifica la storia naturale della cardiopatia congenita, migliora le condizioni cliniche e lo stato psico-fisico del paziente, e deve essere effettuata in Centri dedicati.

Cardiologia Adulti

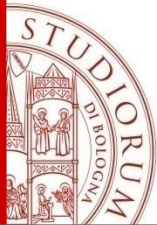
Cardiochirurgia Adulti

Anest.- Rianim adulti

Cardiologia Pediatrica

Cardiochirurgia Pediatrica

Anest.- Rianim pediatrica



ALMA MATER STUDIORUM  
UNIVERSITÀ DI BOLOGNA

**Prof. Gaetano Gargiulo**

Cardiochirurgia Pediatrica e dell'Età Evolutiva

[gaetano.gargiulo@unibo.it](mailto:gaetano.gargiulo@unibo.it)

*[www.unibo.it](http://www.unibo.it)*