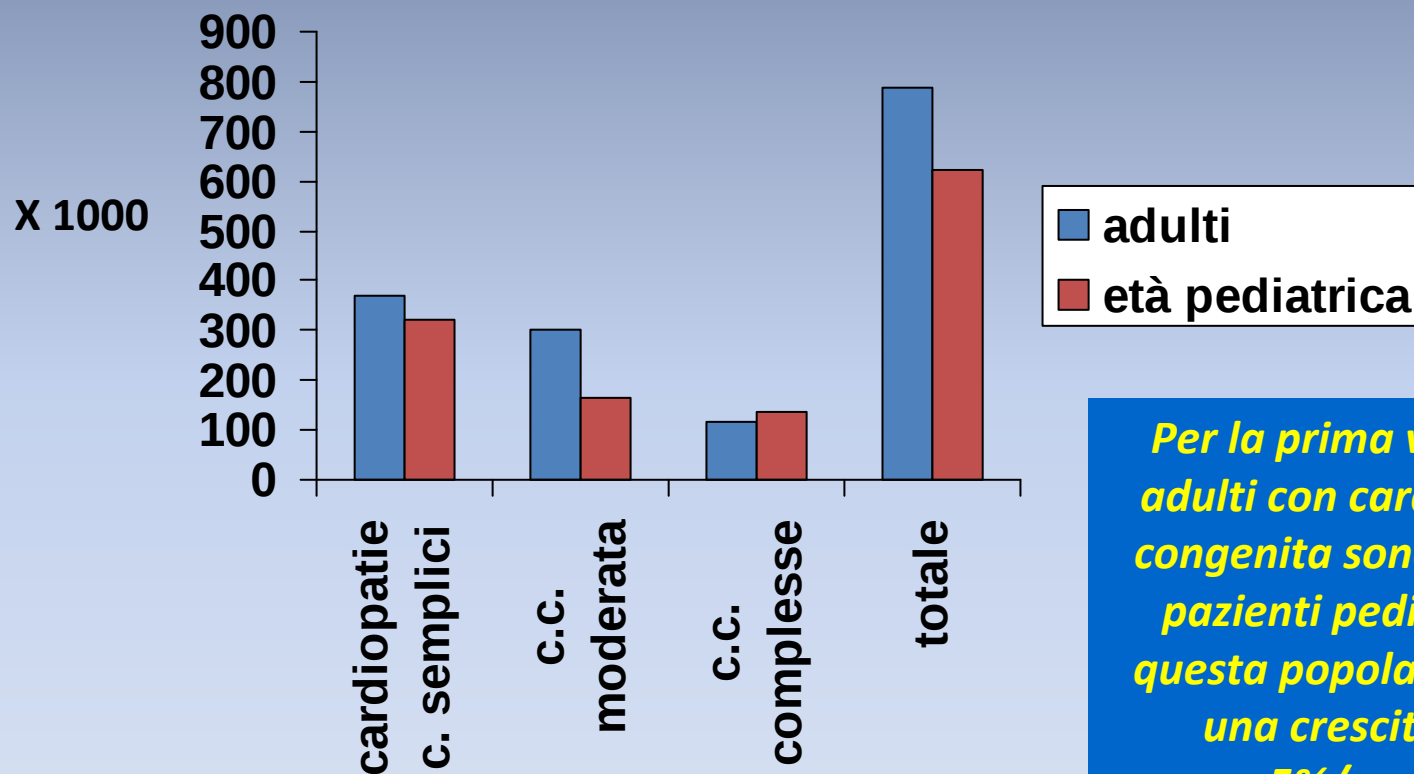


Acronimo GUCH

Grown-Up Congenital Heart disease

Stima dell'incidenza di cardiopatie congenite nell'anno 2000 negli USA



Per la prima volta gli adulti con cardiopatia congenita sono più dei pazienti pediatrici e questa popolazione ha una crescita del 5%/anno

Care of the Adult With Congenital Heart Disease. Presented at the 32nd Bethesda Conference, Bethesda, Maryland, October 2-3, 2000. J Am Coll Cardiol 2001;37:1161-98.

Cardiopatie congenite

Background

Raramente gli interventi correttivi delle cardiopatie congenite sono anche “curativi”

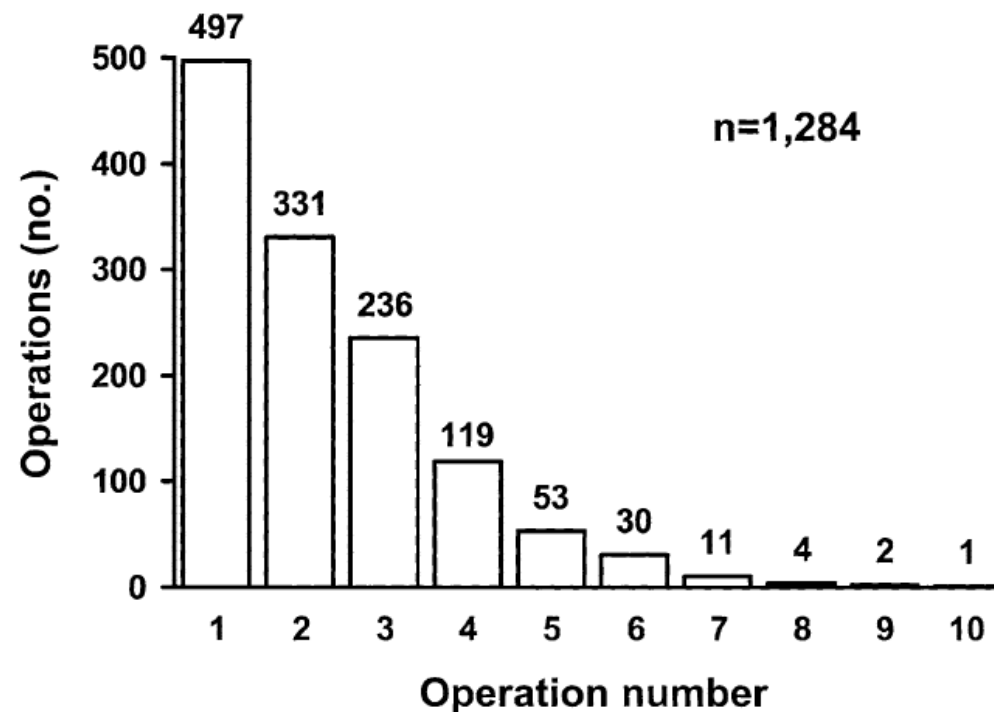


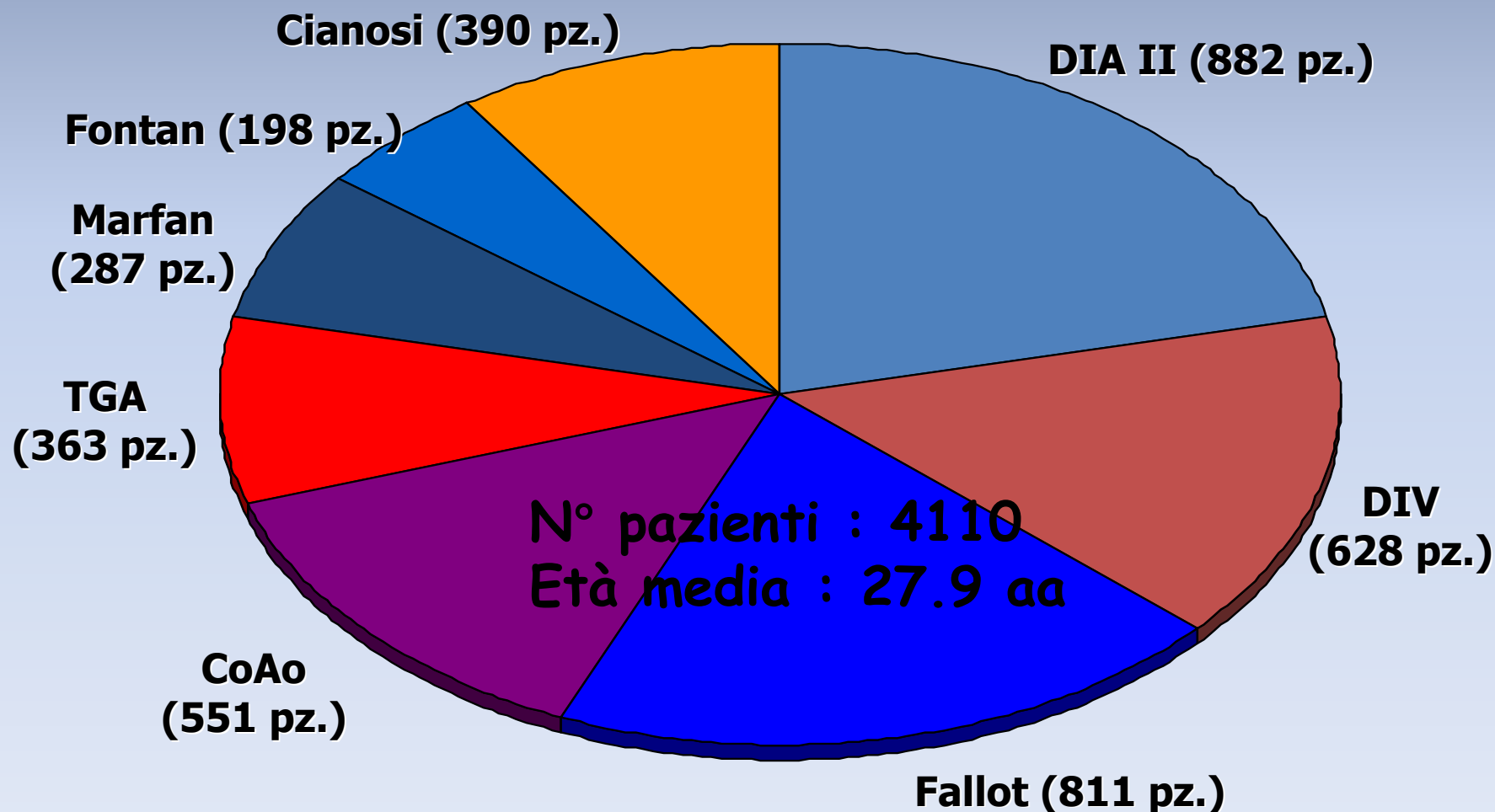
Figure 5. Cardiac operations performed at the Adult Congenital Heart Disease Clinic at Mayo Clinic from 1987 to 2003 (n = 1,284). More than one-third of the patients had operation number 3 or higher. no. = number of patients.

The spectrum of adult congenital heart disease in Europe: morbidity and mortality in a 5 year follow-up period

The Euro Heart Survey on adult congenital heart disease

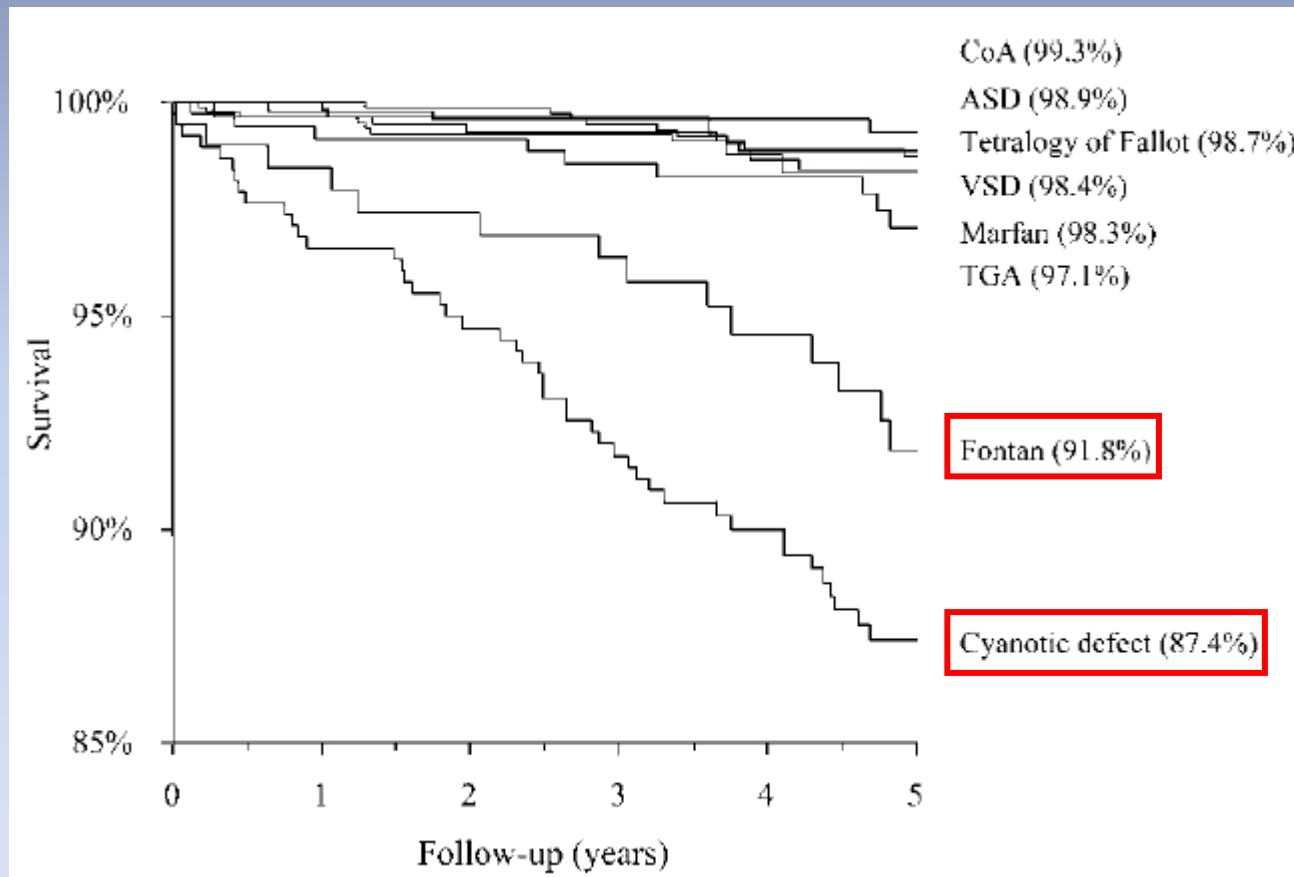
Peter Engelfriet¹, Eric Boersma², Erwin Oechslin³, Jan Tijssen¹, Michael A. Gatzoulis^{4,5}, Ulf Thilén⁶, Harald Kaemmerer⁷, Philip Moons⁸, Folkert Meijboom², Jana Popelová⁹, Valérie Laforest¹⁰, Rafael Hirsch¹¹, Luciano Daliento¹², Erik Thaulow¹³, and Barbara Mulder^{1*}

Eur Heart J 2005; 26: 2325-2333



The spectrum of adult congenital heart disease in Europe: morbidity and mortality in a 5 year follow-up period

The Euro Heart Survey on adult congenital heart disease



The spectrum of adult congenital heart disease in Europe: morbidity and mortality in a 5 year follow-up period

The Euro Heart Survey on adult congenital heart disease

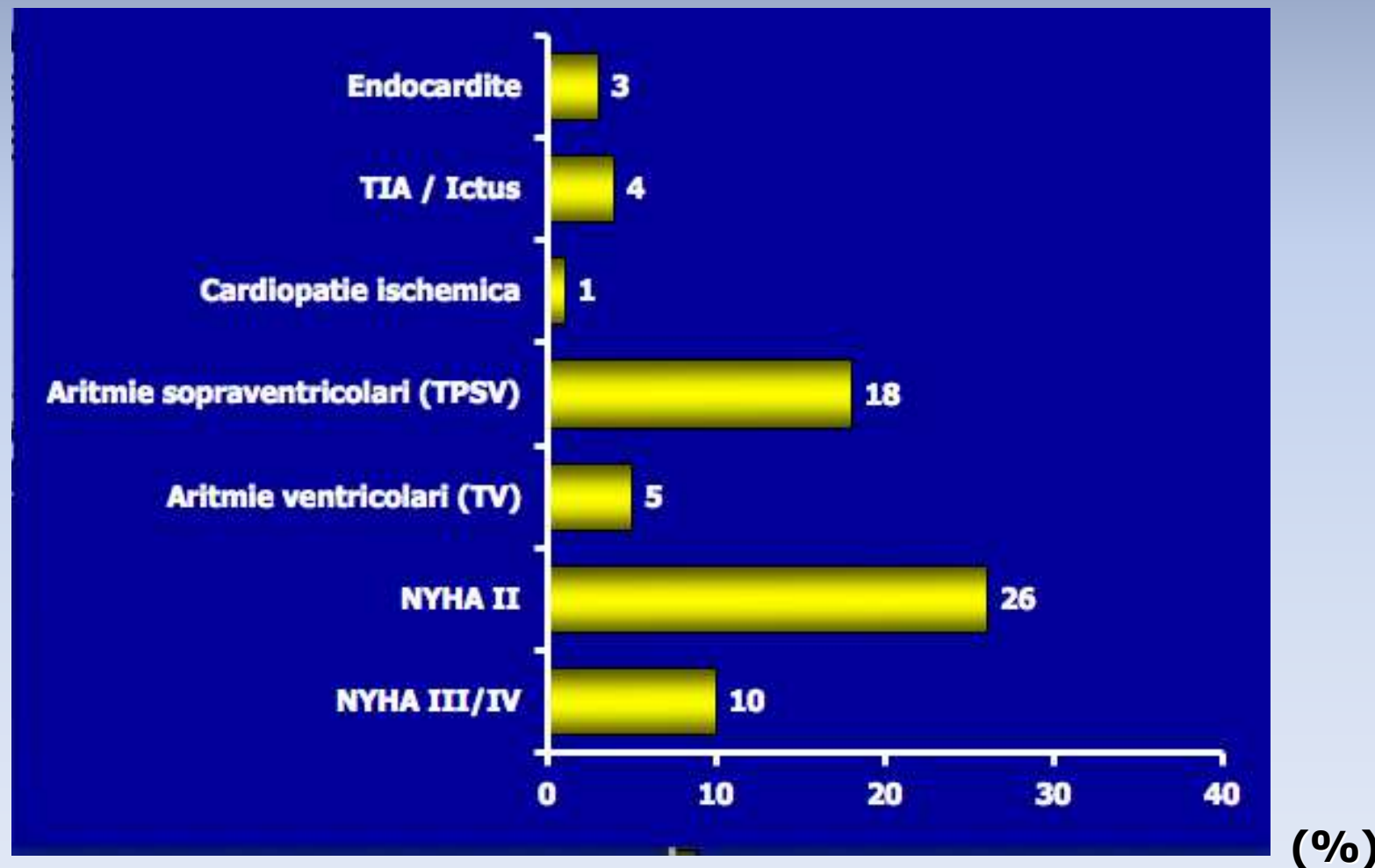
	<i>n</i>	Number of deaths		5 year mortality	
		Cardiovascular	Non-cardiovascular	Kaplan-Meier estimate (%)	95% CI (%)
ASD II	882	5	4	1.1	0.3, 1.9
VSD	628	6	2	1.6	0.5, 2.7
Tetralogy of Fallot	811	8	3	1.3	0.5, 2.1
CoA	551	2	1	0.7	0.0, 1.4
TGA	363	9	0	3.0	1.0, 4.9
Marfan syndrome	287	3	2	1.7	0.0, 3.3
Fontan circulation	198	12	4	8.2	4.0, 12.3
Cyanotic defect	390	44	10	12.6	9.1, 16.0
Overall	4110	89	26	3.0	2.4, 3.5

The spectrum of adult congenital heart disease in Europe: morbidity and mortality in a 5 year follow-up period

The Euro Heart Survey on adult congenital heart disease

Peter Engelfriet¹, Eric Boersma², Erwin Oechslin³, Jan Tijssen¹, Michael A. Gatzoulis^{4,5}, Ulf Thilén⁶, Harald Kaemmerer⁷, Philip Moons⁸, Folkert Meijboom², Jana Popelová⁹, Valérie Laforest¹⁰, Rafael Hirsch¹¹, Luciano Daliento¹², Erik Thaulow¹³, and Barbara Mulder^{1*}

Eur Heart J 2005; 26: 2325-2333

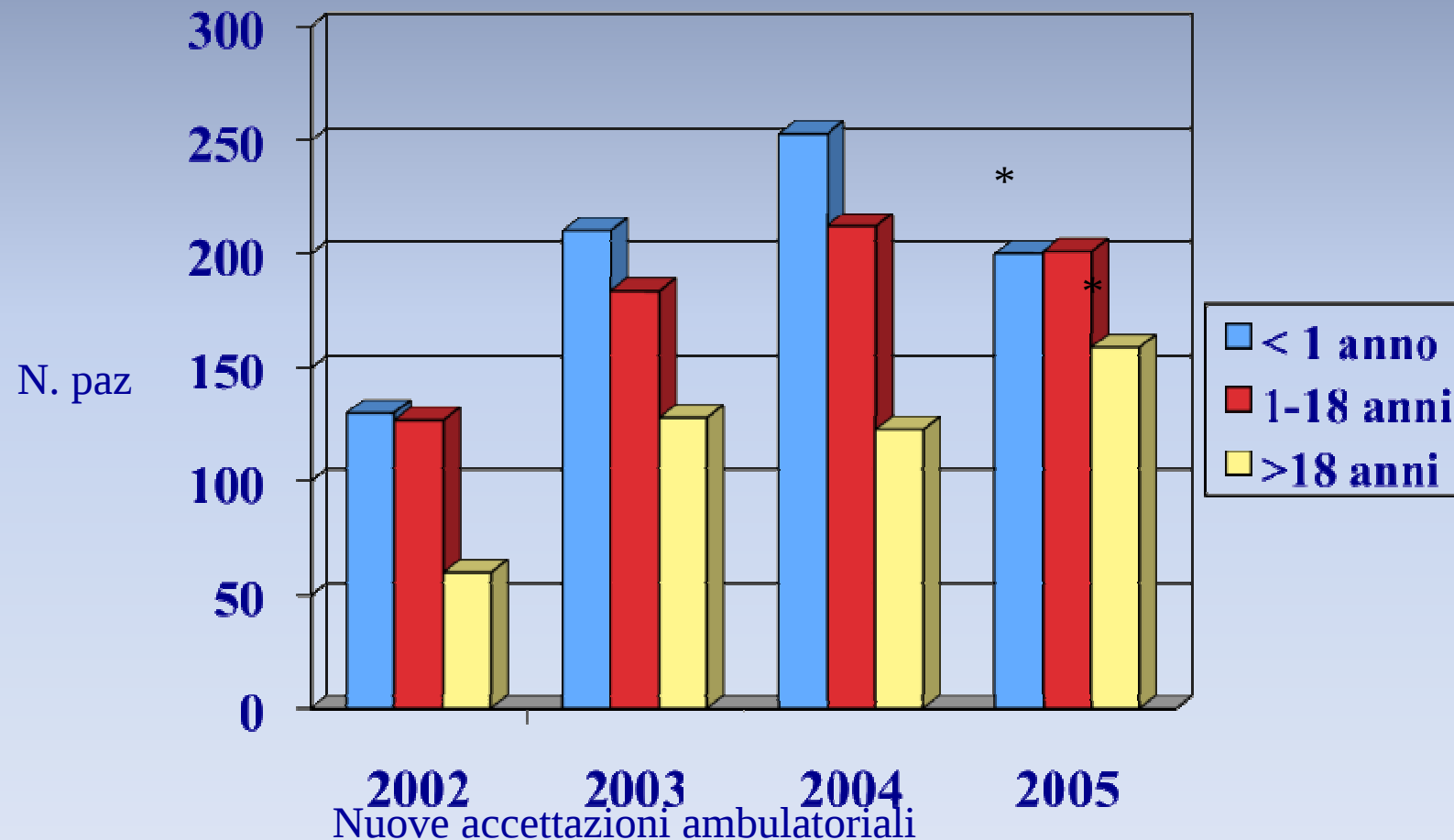


Cardiovascular emergencies

	Admissions	Percentage of total
Arrhythmia	66	53%
Supraventricular arrhythmias	58	47%
Ventricular tachycardia	4	3%
Symptomatic bradycardia	3	2%
ICD burst	1	1%
Acute congestive heart failure	24	19%
Syncope	3	2%
Cerebral ischemia	3	2%
Pacemaker problems	2	2%
Acute aortic dissection	2	2%
Pericardial tamponade/effusion	2	2%
Sudden cardiac death	1	1%
Total	103/124	83%

Endocarditis is classified as infectious emergency. *ICD*, Implantable cardioverter-defibrillator.

- In Italia si stima la presenza di circa 150.000 adulti con cardiopatie congenite di varia complessita'
- Tasso di crescita del 5% annuo



Consapevolezza delle dimensioni del problema GUCH

- 1994: l'ESC riconosce la necessita' di specialisti dedicati
- 1996: linee-guida Canadian Cardiovascular Society
- 1999: Task force ESC (WG22)
- 2000: 32nd Bethesda Conference (ACC)
- 2003: linee-guida ESC
- 2008: linee guida ACC/AHA
- 2010: *aggiornamento linee guida ESC*



Organisation of care

Transition of care

Age 16–18, involving patient, parents, paediatric and adult cardiologist, specialist nurse. Topics should include understanding of disease, future prospects, follow-up visits, medication, endocarditis prophylaxis, insurance, exercise and sports, career planning, pregnancy and contraception (stepwise approach)

Levels of care

- 1) Exclusively at specialist centre
- 2) Shared care between adult cardiac unit and specialised centre
- 3) Nonspecialised care

Specialist centre

Serving population of 5–10 million with trained GUCH specialist. Special needs are best covered by a multidisciplinary team in a specialised centre (catheterisation and electrophysiology lab, congenital cardiac surgery, transplantation, MRI, CT, OB/Gyn)

Identificazione ed educazione del paziente

- Molti GUCH non ricevono controlli regolari e non conoscono i benefici che potrebbero riceverne
- Solo il 37 - 47% dei GUCH hanno una transizione efficace tra Cardiologo Pediatra e specialista GUCH
- Il 27% dei GUCH non effettua controlli dopo i 18 anni di età'

Reid GJ Pediatrics 2004

Sopravvivenza dei pazienti persi al follow-up

Centro per le cardiopatie congenite nell'adulto di Monaco (Germania)

- Più' del 68% dei pazienti con diagnosi o chirurgia in tale sede **non ha visite di follow-up regolari** (nessun controllo negli ultimi 5 anni)
- Dei pazienti che ricevevano un follow-up di qualche genere:
 - 25% seguiti dal medico curante
 - 42% seguiti da medico internista
 - 16% seguiti da un cardiologo
 - 9% seguiti da un cardiologo pediatrico
 - 8% seguiti da uno specialista in GUCH

I pazienti non seguiti da centro GUCH → prognosi peggiore

Organisation of care

Transition
of care

Age 16–18, involving patient, parents, paediatric and adult cardiologist, specialist nurse. Topics should include understanding of disease, future prospects, follow-up visits, medication, endocarditis prophylaxis, insurance, exercise and sports, career planning, pregnancy and contraception (stepwise approach)

Levels of
care

- 1) Exclusively at specialist centre
- 2) Shared care between adult cardiac unit and specialised centre
- 3) Nonspecialised care

Specialist
centre

Serving population of 5–10 million with trained GUCH specialist. Special needs are best covered by a multidisciplinary team in a specialised centre (catheterisation and electrophysiology lab, congenital cardiac surgery, transplantation, MRI, CT, OB/Gyn)

Table 6. Types of Adult Patients With Simple Congenital Heart Disease*

Native disease
Isolated congenital aortic valve disease
Isolated congenital mitral valve disease (e.g., except parachute valve, cleft leaflet)
Isolated patent foramen ovale or small atrial septal defect
<u>Isolated small ventricular septal defect (no associated lesions)</u>
Mild pulmonic stenosis
Repaired conditions
Previously ligated or occluded ductus arteriosus
<u>Repaired secundum or sinus venosus atrial septal defect without residua</u>
Repaired ventricular septal defect without residua

*Those patients can usually be cared for in the general medical community.

Modified from Connelly MS, et al. Canadian Consensus Conference on Adult Congenital Heart Disease, 1996. Can J Cardiol 1998;14:395–452.

Table 5. Types of Adult Patients With Congenital Heart Disease of Moderate Severity*

Aorto-left ventricular fistulae
Anomalous pulmonary venous drainage, partial or total
Atrioventricular canal defects (partial or complete)
<u>Coarctation of the aorta</u>
Ebstein's anomaly
Infundibular right ventricular outflow obstruction of significance
Ostium primum atrial septal defect
<u>Patent ductus arteriosus (not closed)</u>
Pulmonary valve regurgitation (moderate to severe)
Pulmonic valve stenosis (moderate to severe)
Sinus of Valsalva fistula/aneurysm
Sinus venosus atrial septal defect
Subvalvar or supravulvar aortic stenosis (except HOCM)
Tetralogy of Fallot
<u>Ventricular septal defect with</u>
Absent valve or valves
Aortic regurgitation
Coarctation of the aorta
Mitral disease
Right ventricular outflow tract obstruction
Straddling tricuspid/mitral valve
Subaortic stenosis

*These patients should be seen periodically at regional adult congenital heart disease centers. Modified from Connelly MS, et al. Canadian Consensus Conference on Adult Congenital Heart Disease, 1996. Can J Cardiol 1998;14:395–452.
HOCM = hypertrophic obstructive cardiomyopathy.

Table 4. Types of Adult Patients With Congenital Heart Disease of Great Complexity*

Conduits, valved or nonvalved
Cyanotic congenital heart (all forms)
<u>Double-outlet ventricle</u>
<u>Eisenmenger syndrome</u>
<u>Fontan procedure</u>
Mitral atresia
Single ventricle (also called double inlet or outlet, common or primitive)
Pulmonary atresia (all forms)
Pulmonary vascular obstructive diseases
<u>Transposition of the great arteries</u>
Tricuspid atresia
Truncus arteriosus/hemitruncus
Other abnormalities of atrioventricular or ventriculoarterial connection
not included above (i.e., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

*These patients should be seen regularly at adult congenital heart disease centers.

Modified from Connelly MS, et al. Canadian Consensus Conference on Adult Congenital Heart Disease, 1996. Can J Cardiol 1998;14:395–452.

ESC Guidelines 2010

Organization of care

pulmonary arterial hypertension (PAH)]. Many experts therefore feel that the best care is provided when most GUCH patients are seen once in a specialist centre, and recommendations are then made with regard to follow-up intervals and level of care on an individualized basis. A well working network of specialist centres with general adult care is of critical importance.

*.....even simple defects may require specialist care under certain circumstances
(e.g. ASD with pulmonary arterial hypertension)*

Organisation of care

Transition of care

Age 16–18, involving patient, parents, paediatric and adult cardiologist, specialist nurse. Topics should include understanding of disease, future prospects, follow-up visits, medication, endocarditis prophylaxis, insurance, exercise and sports, career planning, pregnancy and contraception (stepwise approach)

Levels of care

- 1) Exclusively at specialist centre
- 2) Shared care between adult cardiac unit and specialised centre
- 3) Nonspecialised care

Specialist centre

Serving population of 5–10 million with trained GUCH specialist. Special needs are best covered by a multidisciplinary team in a specialised centre (catheterisation and electrophysiology lab, congenital cardiac surgery, transplantation, MRI, CT, OB/Gyn)

Table 2. Personnel and Services Recommended for Regional ACHD Centers

Type of Service	Personnel/Resources
Cardiologist specializing in ACHD	One or several 24/7
Congenital cardiac surgeon	Two or several 24/7
Nurse/physician assistant/nurse practitioner	One or several
Cardiac anesthesiologist	Several 24/7
Echocardiography*	Two or several 24/7
● Includes TEE, intraoperative TEE	
Diagnostic catheterization*	Yes, 24/7
Noncoronary interventional catheterization*	Yes, 24/7
Electrophysiology/pacing/AICD implantation*	One or several

Table 2. Personnel and Services Recommended for Regional ACHD Centers

Type of Service	Personnel/Resources
Exercise testing	● Echocardiography ● Radionuclide ● Cardiopulmonary ● Metabolic
Cardiac imaging/radiology*	● Cardiac MRI ● CT scanning ● Nuclear medicine

Table 2. Personnel and Services Recommended for Regional ACHD Centers

Type of Service	Personnel/Resources
Multidisciplinary teams	● High-risk obstetrics ● Pulmonary hypertension ● Heart failure/transplant ● Genetics ● Neurology ● Nephrology ● Cardiac pathology ● Rehabilitation services ● Social services ● Vocational services ● Financial counselors
....ematologia	
....ortopedia	

Gestione del cardiopatico congenito in età adulta

Necessaria collaborazione tra discipline diverse

Educazione precoce del paziente e della sua famiglia circa diagnosi, stile di vita e follow-up da seguire

Obiettivi da perseguire:

- Stabilire una rete di centri regionali
- Incoraggiare la formazione di specialisti
- Coordinare registri locali o nazionali
- Condivisione dei dati
- Facilitare la ricerca

Gestione del cardiopatico congenito in età adulta

- ✓ Medico di base
- ✓ Centri di cardiologia pediatrica dovrebbero individuare centro per GUCH
- ✓ Cardiologo dell'adulto, cardiochirurgo e unità di PS dovrebbero avere stretto rapporto con il centro GUCH
- ✓ Centro specializzato GUCH (1/5-10 milioni di abitanti) in genere regionale
- ✓ Centri "satelliti" con riferimento regionale

Inoltre

- ✓ necessità di un team integrato di ostetrici, anestesisti, cardiologi dell'adulto per l'assistenza in gravidanza e negli interventi di chirurgia non cardiaca
- ✓ Database per registrare attività, outcome e audit dei risultati per facilitare la ricerca

Com'è la situazione a Parma:

- ❖ Il pz GUCH esegue i suoi controlli ambulatoriali presso la SSD di Cardiologia Pediatrica
- ❖ Un eventuale ricovero viene effettuato presso U.O. Complessa di Cardiologia adulti. (cateterismo cardiaco studio emodinamico invasivo)
- ❖ Casi particolari presso altri reparti adulti come Infettivologia, Medicina interna, Nefrologia, ecc.. previa discussione collegiale del caso
- ❖ Collaborazione tra cardiologo pediatra e medico del reparto ospitante
- ❖ Meeting periodici con gli aritmologi della Cardiologia dell'adulto (SEF, ablazioni transcatetere,...)
- ❖ RMN

Paziente di 28 anni F.M.

2002 Intervento cardiocirurgico c/o Ospedale San Donato Milanese di connessione dello sbocco venoso anomalo polmonare con l'atrio sinistro dopo creazione di DIA in corrispondenza della fossa ovale mediante interposizione di tubo vascolare di dacron 14 mm ed ampliamento della vena cava superiore con patch di pericardio eterologo.







