ESPERANÇA DE VIDA DELS PACIENTS ADULTS AMB CARDIOPATIES CONGÈNITES

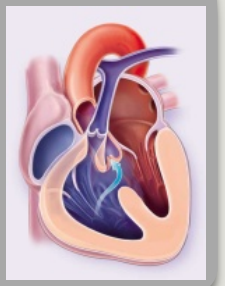
Berta Miranda Barrio
Unitat Integrada Vall d'Hebron-Hospital de Sant Pau de Cardiopaties Congènites de l'Adolescent i l'Adult
Servei Cardiologia

CAS CLÍNIC

- Pacient de 56 anys, treballa en la construcció
  - Exfumador
  - OH 3 UBES/dia
  - Probablement hipertens
  - Hipotiroidisme subclínic sense tractament

CARDIOPATIA REPARADA EN LA INFÀNCIA

DERIVACIÓ



→ Ingrés/18 pneumònia penumoccòccica amb empiema

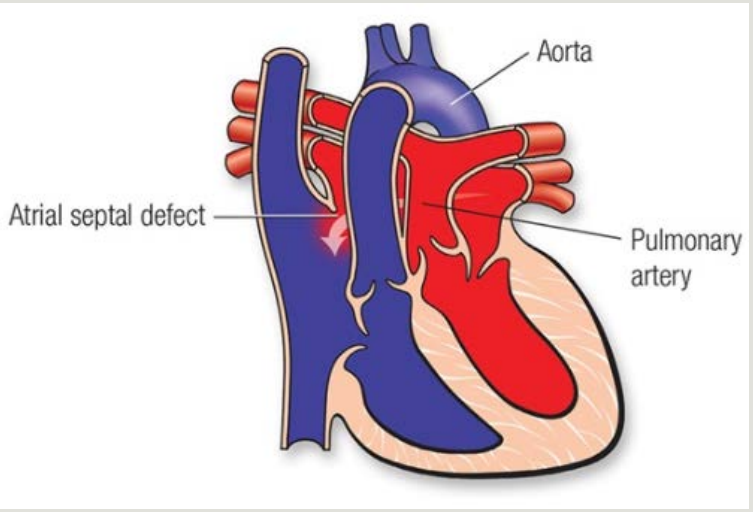
Vall d'Hebron Barcelona Campus Hospitalari

CAS CLÍNIC

Naixement / 1963

1 any per cianosis

12 anys / 1975



Centre hospitalari Madrid

Empitjorament clínic

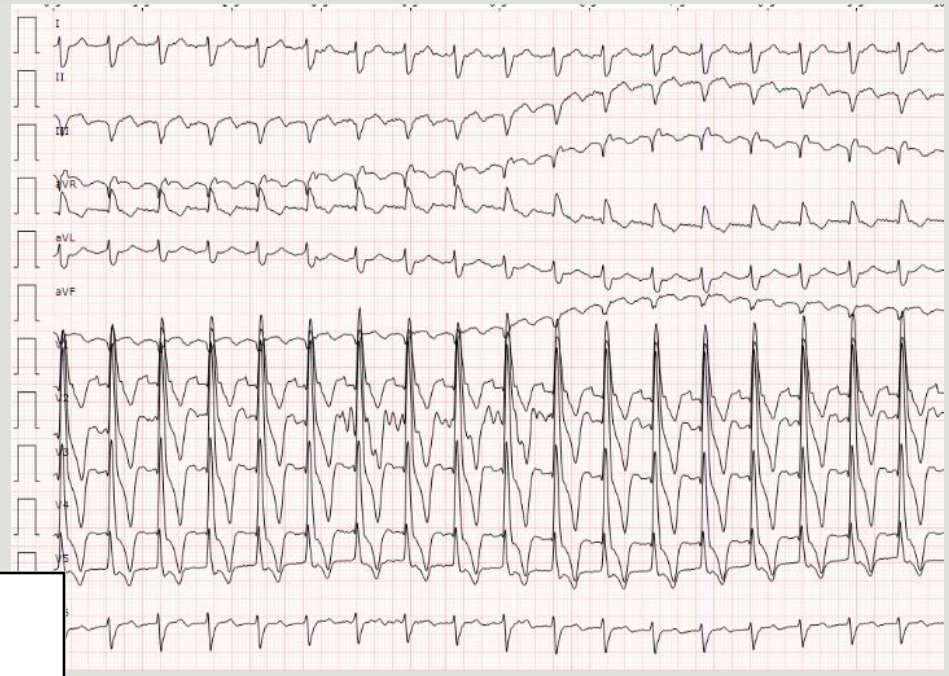
KT:
- TP y branques confluents
- PAPm 7mmHg
- CIA àmplia

PÈRDUA DE SEGUIMENT

Barcelona Campus Hospitalari

CAS CLÍNIC

UCCAA



Vall d'Hebron
Barcelona Campus Hospitalari

INTRODUCCIÓ

1940 1950 1960 1970 1980 1990 2000 2012

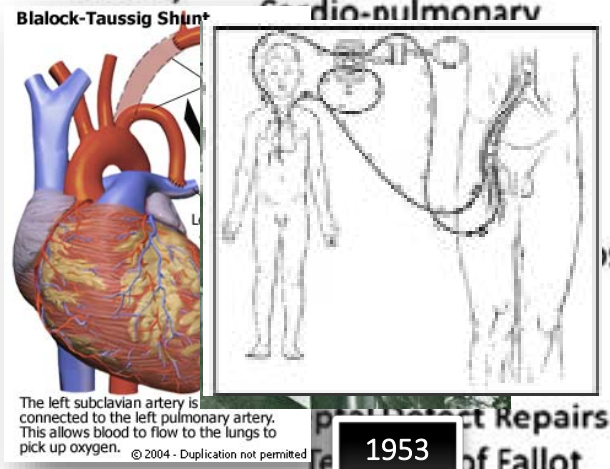
Blalock-Taussig Shunt;

Pediatric and Neonatal Cardiac Catheterization

Echocardiography

Prostaglandin E_1

Interventional Catheterization
Invasive Electrophysiology
Improved Peri-Operative Care
Randomized Clinical Trials



1953

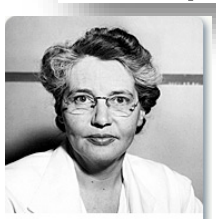
Balloon Valvuloplasty

Infant Surgery

Neonatal Surgery

Stage I Norwood
Arterial Switch Operation

Not Much New in Congenital Heart Surgery.....



Helen Taussig
Johns Hopkins University

Alfred Blalock
Johns Hopkins University

1944

Adult Heart Transplantation

Neonatal (and Pediatric) Heart Transplantation

Fontan-Kreutzer Procedure

11th Annual William J. Rashkind Memorial Lecture in paediatric cardiology – a short history of progress: Where we've been, where we are, where we're going. **G. Wernovsky. Card Young 2012.**

INTRODUCCIÓ

1940 1950 1960 1970 1980 1990 2000 2012

Blalock-Taussig Shunt; PA Band

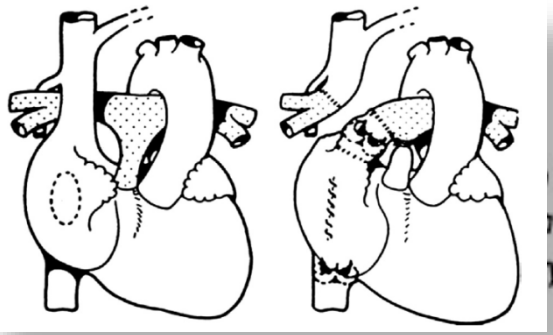
Echocardiography

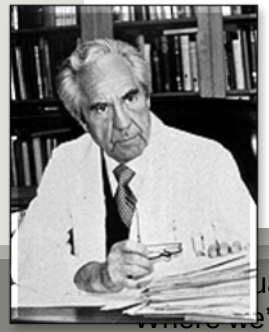
Prostaglandin E_1

Interventional Catheterization
Invasive Electrophysiology
Improved Peri-Operative Care
Randomized Clinical Trials

1955

1958





Adult Heart Transplantation

Neonatal (and Pediatric) Heart Transplantation

Fontan-Kreutzer Procedure

Vall d'Hebron Barcelona Campus Hospitalari

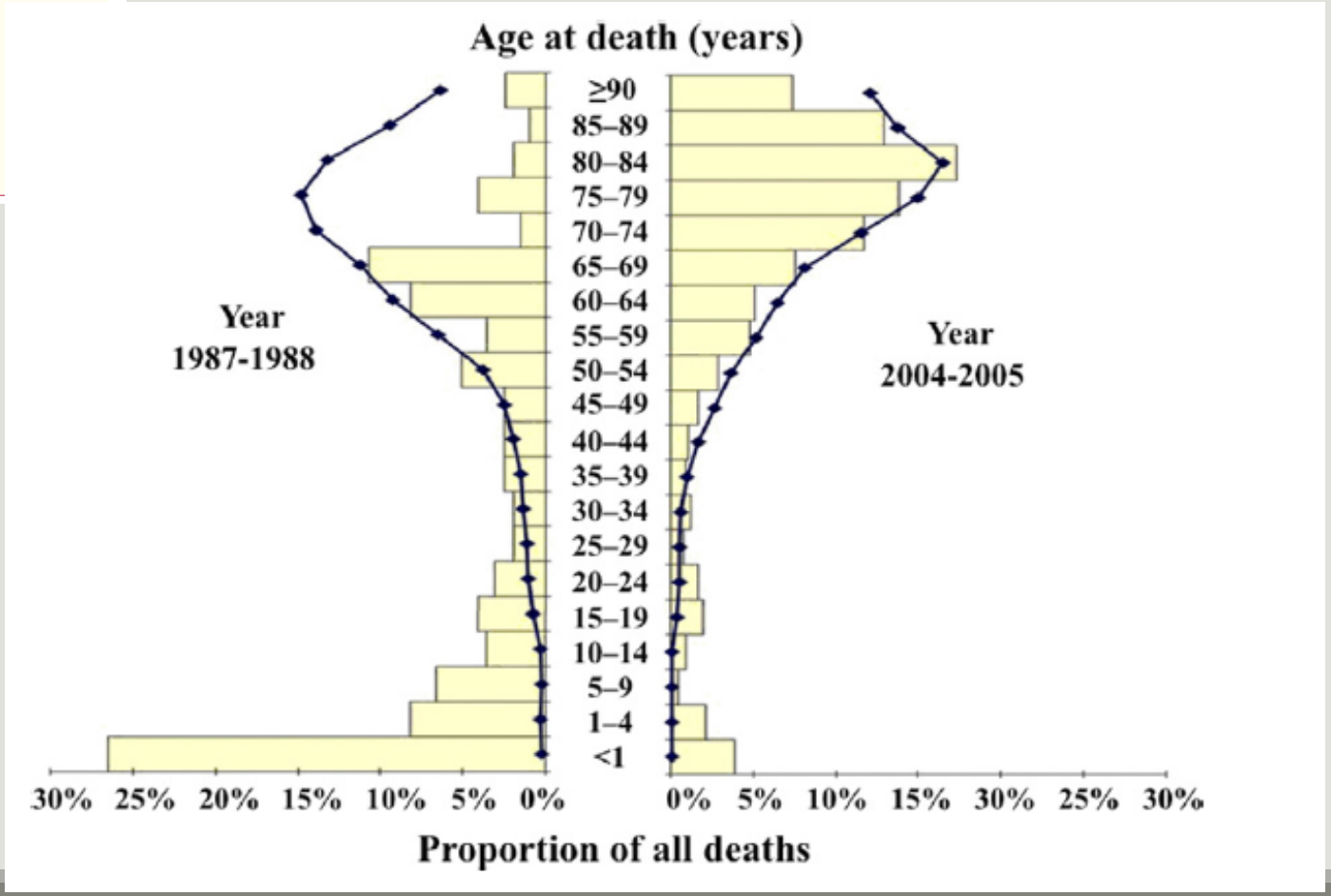
ual d Memorial Lecture in paediatric cardiology – a short history of progress: re, where we're going. **G. Wernovsky. Card Young 2012.**

CANVI EN LA SUPERVIVÈNCIA

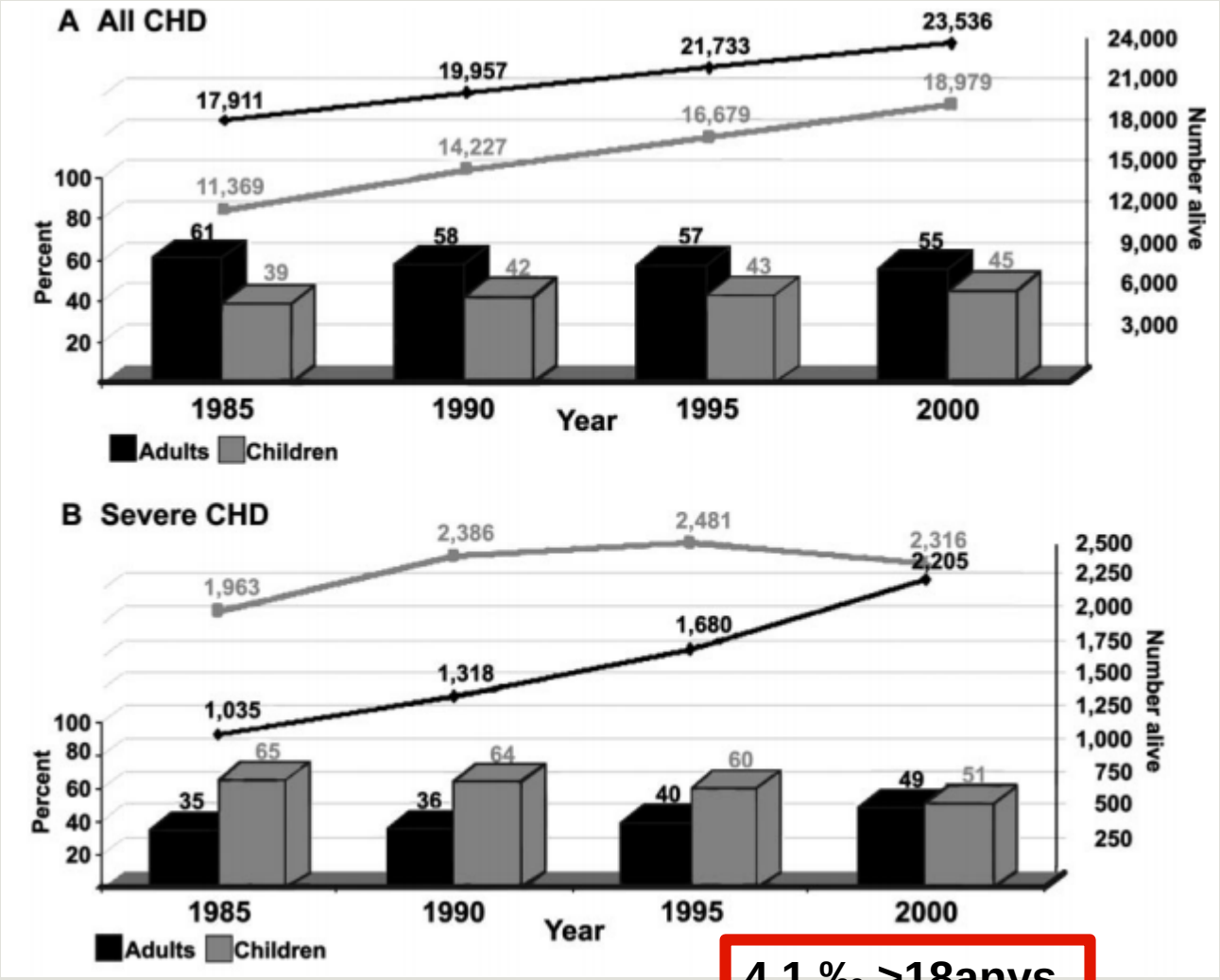
CONGENITAL HEART DISEASE

Changing Mortality in Congenital Heart Disease

Paul Khairy, MD, PHD,* Raluca Ionescu-Ittu, MSC,†§ Andrew S. Mackie, MD, SM,†
Michal Abrahamowicz, PHD,§ Louise Pilote, MD, MPH, PHD,‡§ Ariane J. Marelli, MD†
Montreal, Quebec, Canada

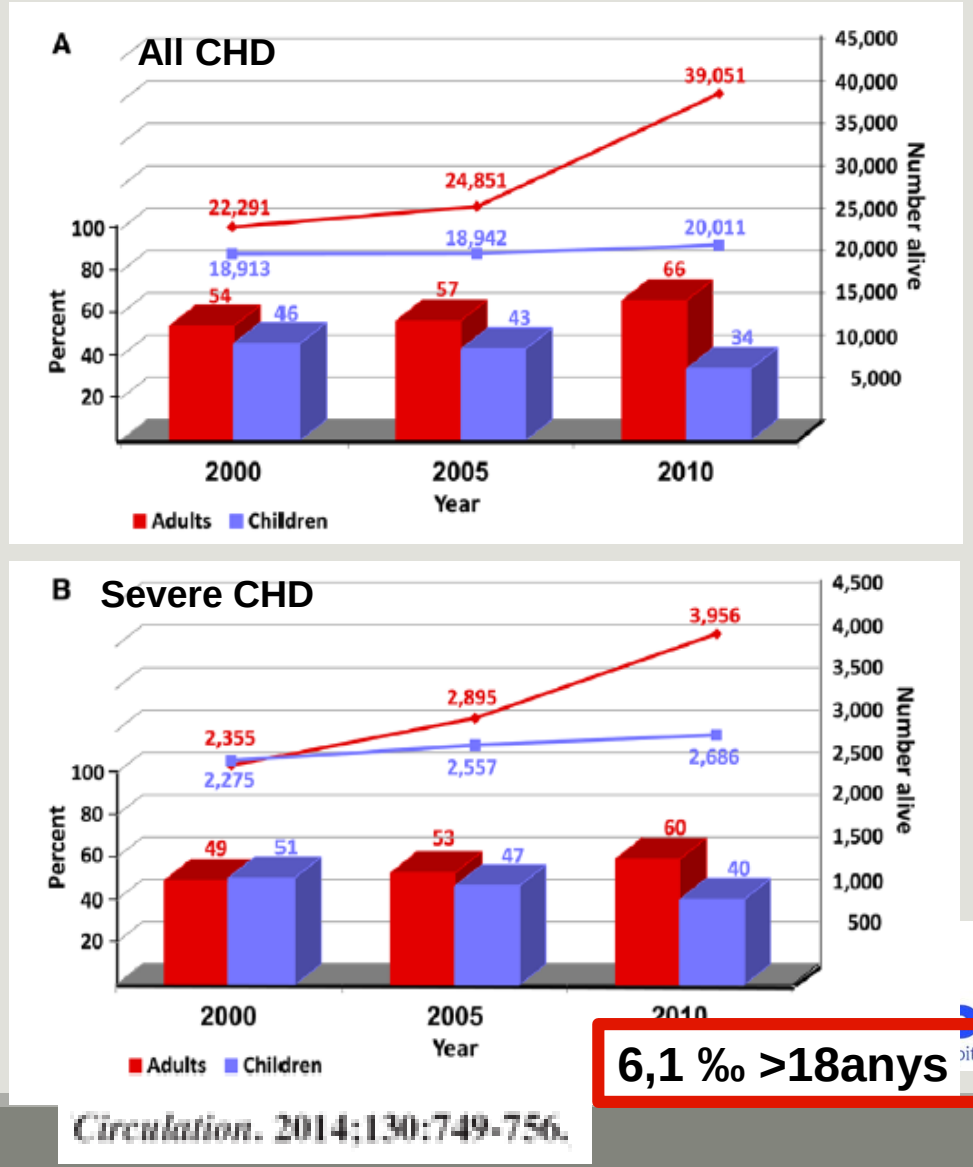


PREVALENÇA ACTUAL



4,1 ‰ >18anys

Circulation. 2007;115:163-172.



6,1 ‰ >18anys

Circulation. 2014;130:749-756.

PREVALENÇA ACTUAL

The prevalence of adult congenital heart disease, results from a systematic review and evidence based calculation

Teun van der Bom, MD, [a,b] Berto J. Bouma, MD, PhD, [a] Folkert J. Meijboom, MD, and Barbara J. M. Mulder, MD, PhD [a,b] Amsterdam and Utrecht, The Netherlands

Table I. Cross-sectional studies prevalence of CHD in ad

Author	Year study	Year estimate	Country	Age population
Marelli et al[3]	2007	2000	Canada	>15
Billet et al[22]	2008	2005	UK	>19
Videbaek et al[23]	2009	2009	Denmark	>15
CONCOR[19*]	2011	2011	The Netherlands	>18
Overall prevalence (weighted mean)				

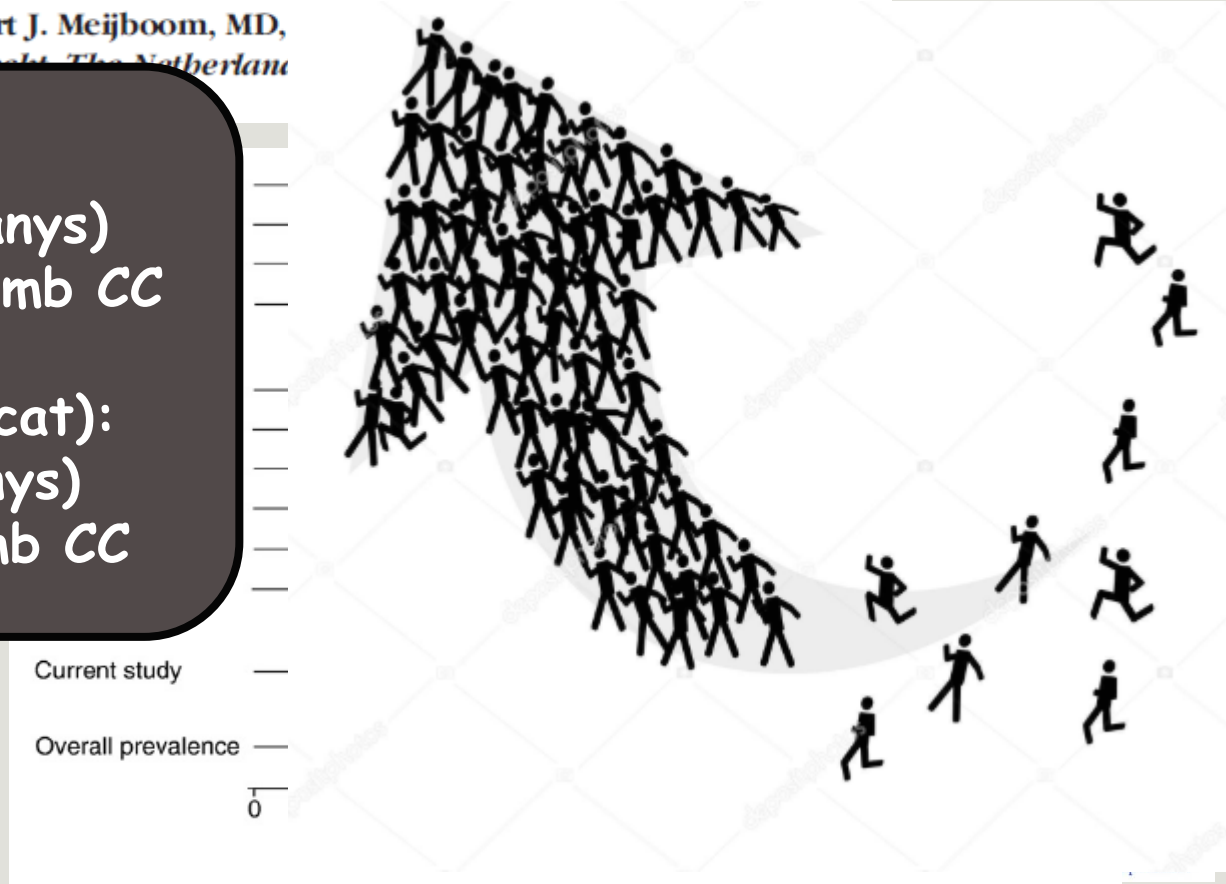
* CONCOR is still recruiting and therefore not included in the weighted m

Table II. Adult CHD prevalence

Author	Year study	Year estimate	Country	Pat... between	...estimate	...per million	...survival	...per million
Warnes et al[4]	2001	2000	USA	1940-1989	11-60	6200	57%	3560
Somerville[24]	2002	2010	UK	1940-1990	20-70	6000	55%	3300
Hoffman et al[25]	2003	2002	USA	1940-1989	16-60	8912	20%-44%	1770-3880
Fredriksen et al[26]	2007	2005	Norway	1940-1989	16-64	7800	62%	4810
Shiina et al[27]	2009	2007	Japan	1947-1992	16-60	10600	37%	3930
Daliento et al[28]	2011	2032	Italy	1982-2023	11-49	6200	63%	3907
Current study	2011	2008	The Netherlands	1940-1989	18-68	8979	36%	3228
Overall prevalence (mean)								3548

ESPANYA (INE):
38 milions adults (>18 anys)
114.000-231.800 adults amb *CC*

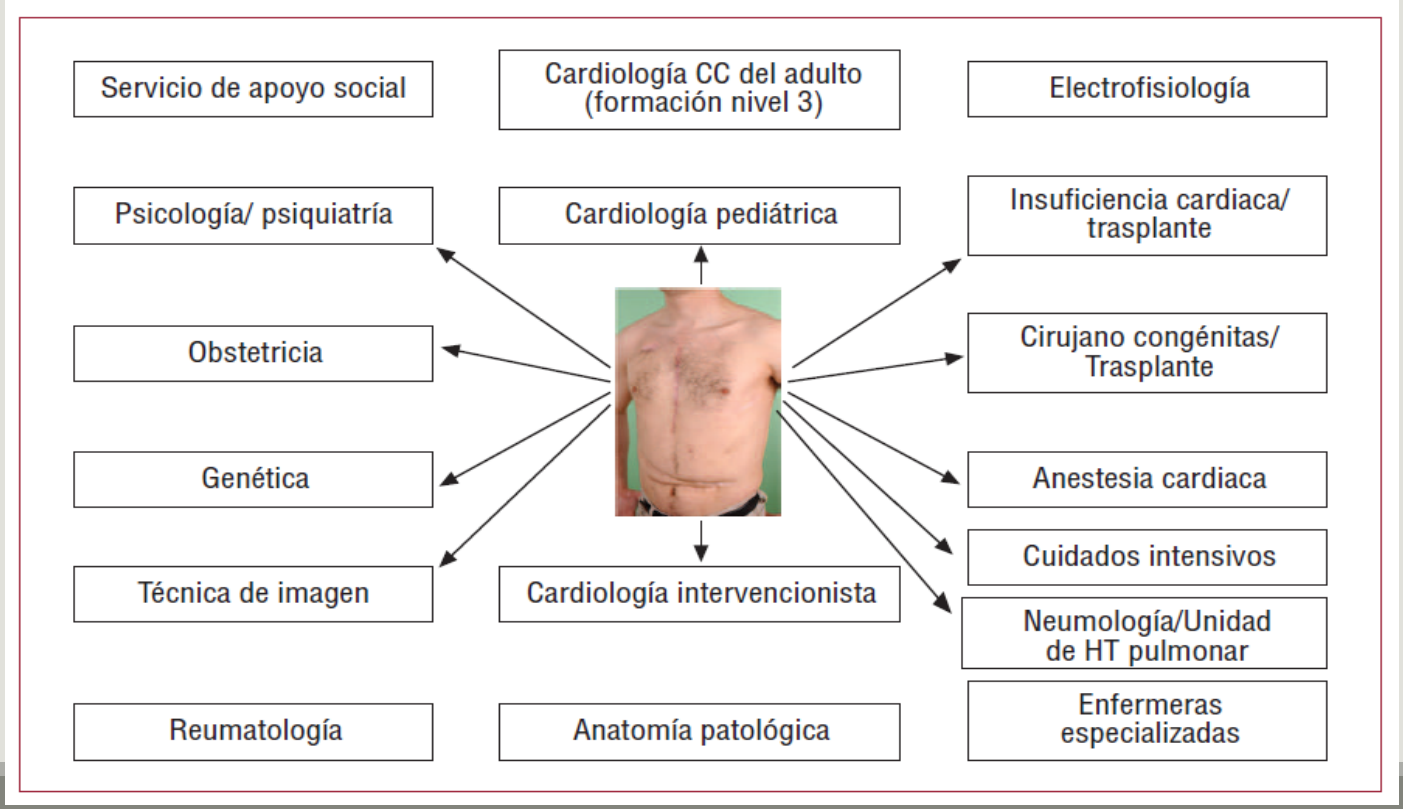
CATALUNYA 2017 (Idescat):
6 milions adults (≥19 anys)
18.000-36.600 adults amb *CC*



American Journal of Cardiology 2012. 164(4):568-75

ENVELLIMENT DE LA POBLACIÓ

- Major percentatge de FRCV clàssics i neoplàsies malignes
- Pacients que han sotmesos a PAL·LIACIONS / cirurgies REPARADORES (no CURATIVES)

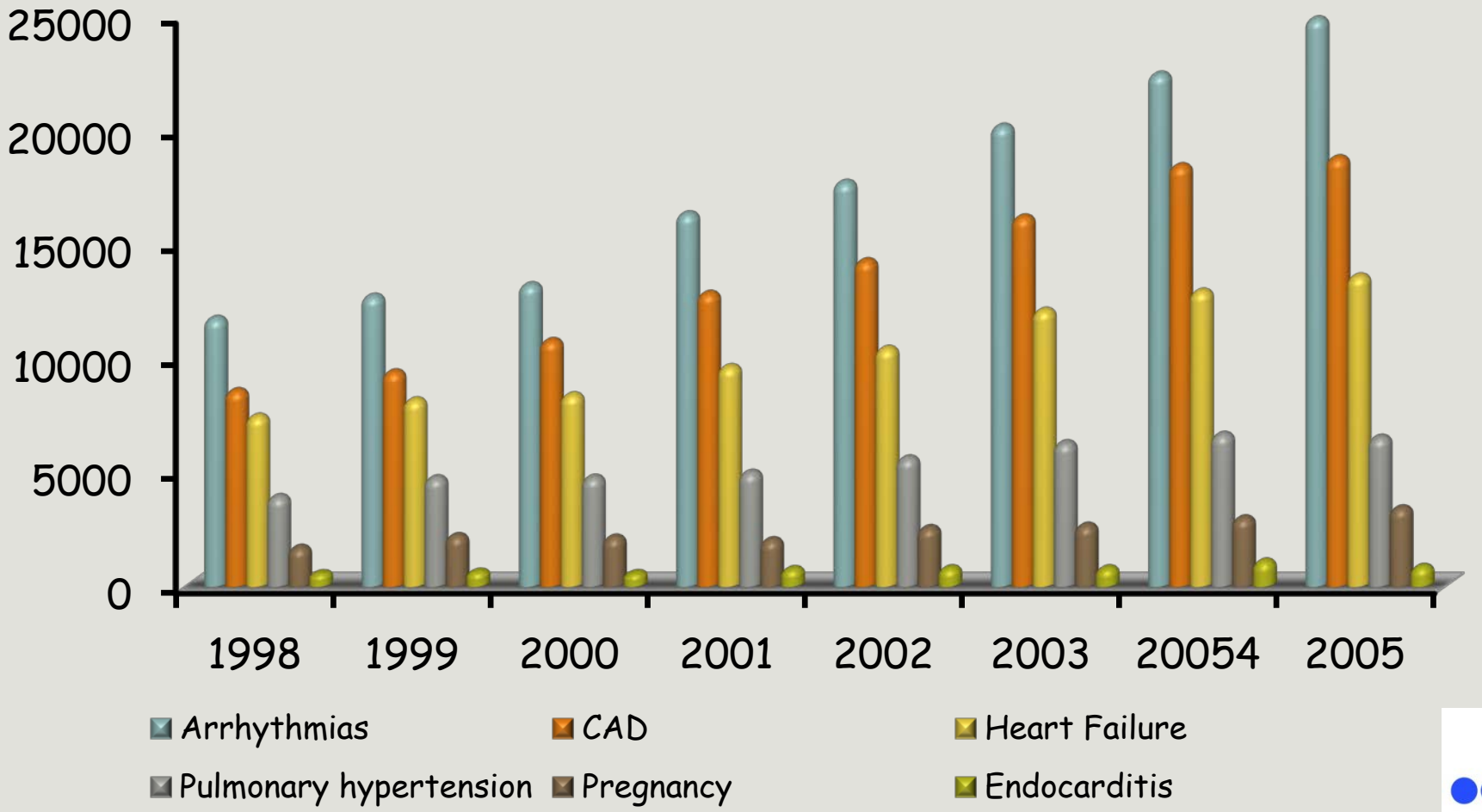


AUGMENT IMPORTANT DE L'ÚS DELS SERVEIS SANITARIS

Vall d'Hebron Barcelona Campus Hospitalari

Oechslin E.Modelos de asistencia sanitaria en Europa y America del Norte. Suplemento REC 2009..

ENVELLIMENT DE LA POBLACIÓ



Vall d'Hebron Barcelona Campus Hospitalari

Opotowsky AR, et al. J Am Coll Cardiol 2009;54:460-67

ENVELLIMENT DE LA POBLACIÓ

International Journal of Cardiology 228 (2017) 790–795

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Isolated heart transplant and combined heart-liver transplant in adult congenital heart disease patients: Insights from the united network of organ sharing☆

Elisa A Bradley [a,b,*,1], Krong-on Pinyoluksana [a,b,1], Melissa Moore-Clingenpeel [b,1], Yongjie Miao [b,1], Curt Daniels [a,b,1]

[a] The Ohio State University, 473 W 12th Avenue Suite 200, Columbus, OH 43210, USA
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Keywords:
Congenital heart disease
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Fontan
Combined heart-liver transplant

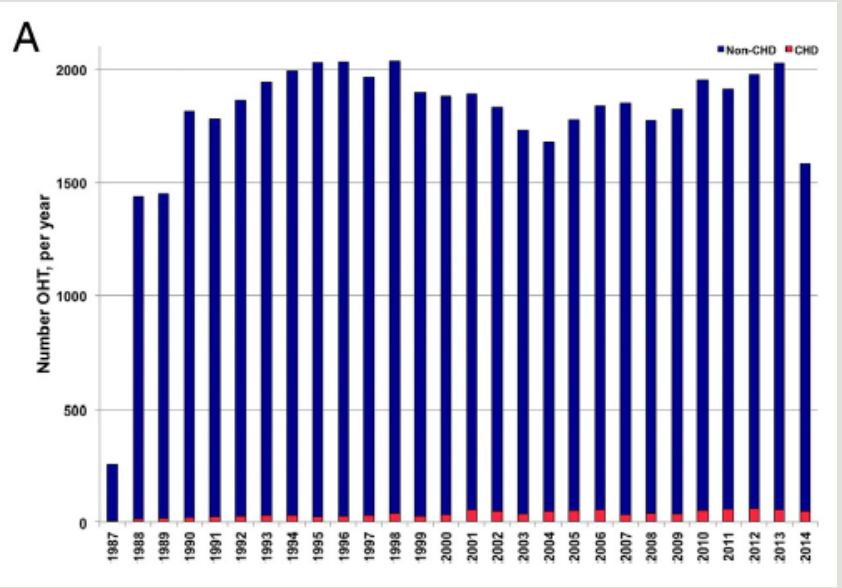
ABSTRACT

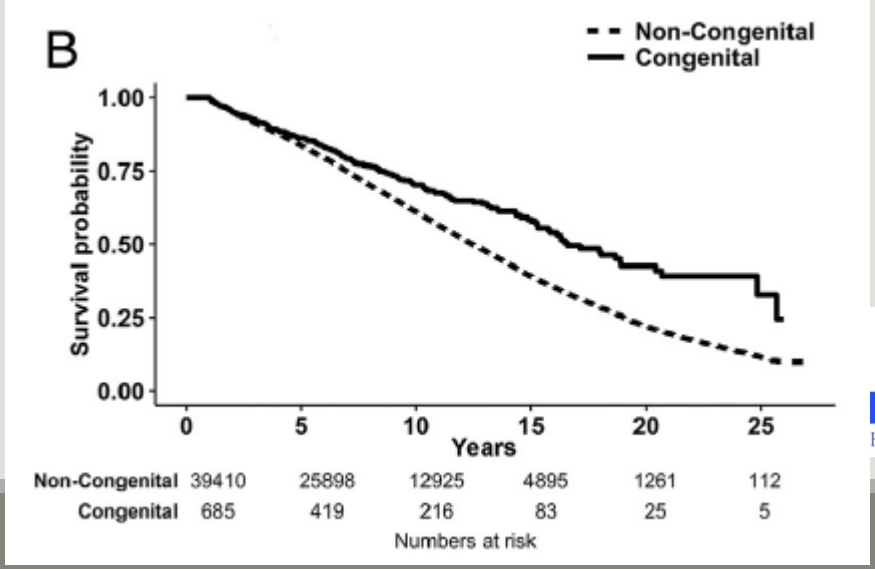
Background/method: The aging patient with severe congenital heart disease (CHD) faces many challenges: heart failure, arrhythmia, and in the Fontan patient, liver disease. Our goal was to define combined heart liver transplant (CHLT) and isolated orthotopic heart transplant (OHT) outcomes in U.S. adult CHD patients. The U.S. United Network for Organ Sharing (UNOS) thoracic and liver databases were queried for cardiac and CHD diagnoses, from inception-2014.

Results: In CHLT, CHD made up 22% of waitlist patients (non-CHD n = 262 vs. CHD n = 58), and 20% of transplanted patients (non-CHD n = 137 vs. CHD n = 27). Liver function tests in the non-CHD and CHD groups were similar and there was no difference in CHD and non-CHD survival (HR 0.93, CI: 0.36–2.38, p 0.48). In isolated OHT, CHD patients comprised 2% of those listed (non-CHD n = 74,080 vs. CHD n = 1599) and transplanted (non-CHD n = 48,985 vs. CHD n = 967) and had higher early (<1 year) mortality (HR 1.36, CI: 1.18–1.57, p < 0.0001), but better long-term survival (HR 0.66, CI: 0.57–0.76, p < 0.001) than non-CHD. Both groups benefitted from mechanical support when used (non-CHD HR 0.34, CI: 0.31–0.37 and CHD HR 0.14, CI: 0.03–0.58) and prior sternotomy had no effect on mortality in CHD (HR 0.63, CI: 0.15–2.58).

Conclusions: Survival of CHD patients undergoing CHLT is no different than in non-CHD, encouraging consideration of CHLT when clinically appropriate. Short-term mortality is higher in CHD (vs. non-CHD) patients undergoing OHT, regardless of prior cardiac surgery status. Modifications to CHD classification within UNOS would help better understand CHD CHLT and OHT outcomes.

E.A. Bradley et al. / International Journal of Cardiology 228 (2017) 790–795





ENVELLIMENT DE LA POBLACIÓ

Clin Transplant 2012 DOI: 10.1111/j.1399-0012.2012.01611.x

Clinical Transplantation

Transplantation for complex congenital heart disease in adults: a subanalysis of the Spanish Heart Transplant Registry

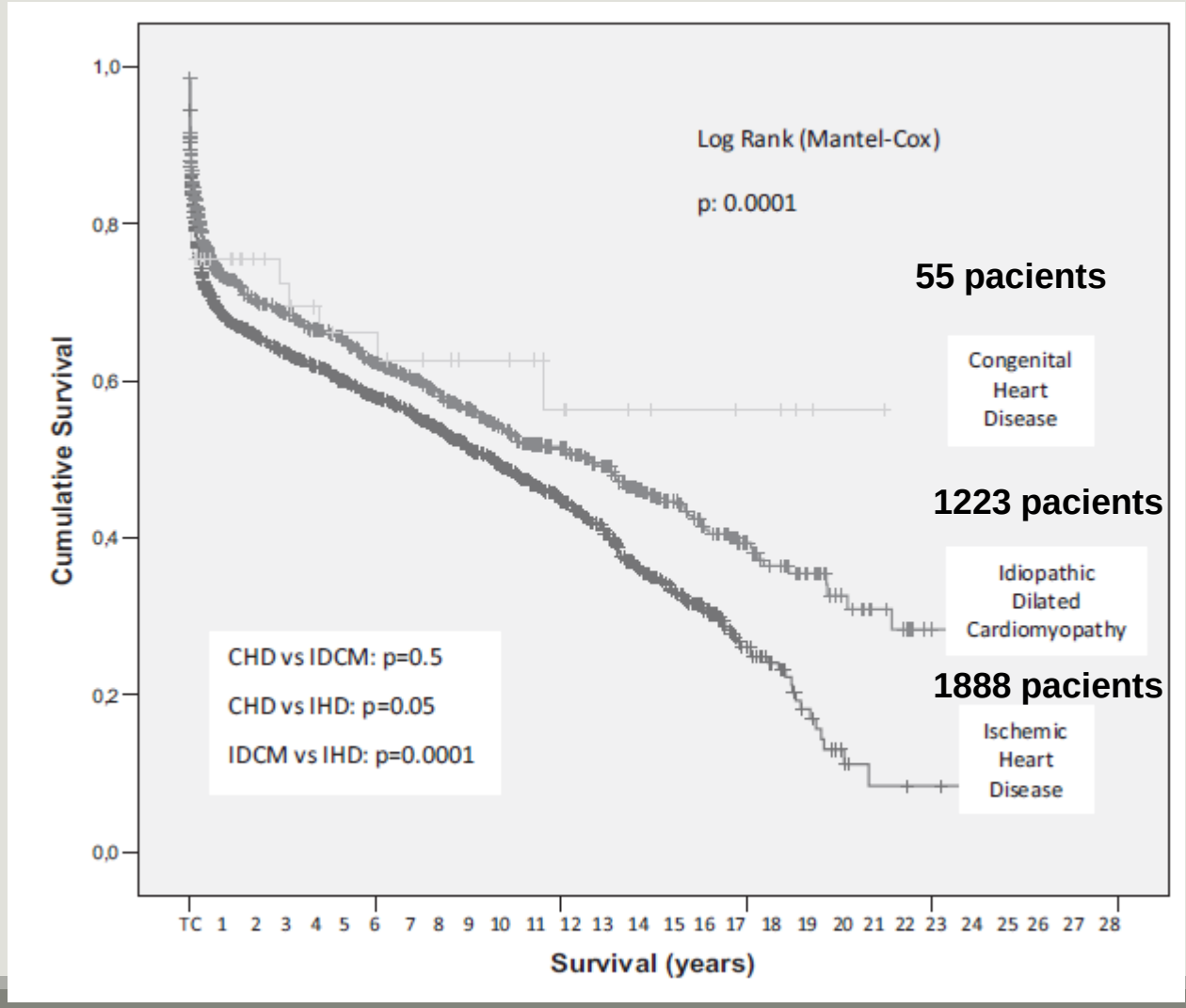
Paniagua Martín MJ, Almenar L, Brossa V, Crespo-Leiro MG, Segovia J, Palomo J, Delgado J, González-Vílchez F, Manito N, Lage E, García-Guereta L, Rodríguez-Lambert JL, Albert DC. Transplantation for complex congenital heart disease in adults: a subanalysis of the Spanish Heart Transplant Registry.
Clin Transplant 2012 DOI: 10.1111/j.1399-0012.2012.01611.x. © 2012 John Wiley & Sons A/S.

Abstract: Background: Congenital heart diseases (CHDs) have high infant mortality in their severe forms. When adulthood is reached, a heart transplant (HTx) may be required. Spanish adult population transplanted for CHD was analyzed and compared with the most frequent causes of HTx and between different subgroups of CHD.
Materials and Methods: A total of 6048 patients (HTx 1984–2009) were included. Pediatric transplants (<15 yr), combined transplants, reHTx, and HTx for heart diseases other than idiopathic dilated cardiomyopathy (IDCM) and ischemic heart disease (IHD) were excluded. Total patients included: 3166 (IHD = 1888; IDCM = 1223; CHD = 55). Subgroups were studied as follows: (1) single ventricle with pulmonary stenosis (n = 18), (2) single ventricle with tricuspid atresia and Glenn/Fontan surgery (n = 10), (3) congenitally corrected transposition of the great vessels (TGV) or with switch atrial surgery (n = 10), and (4) CHD with right ventricle overload (n = 17).
Results: Survival probability was different between groups (p = 0.0001). Post hoc analysis showed some differences between groups (CHD vs. IHD, p = 0.05; CHD vs. IDCM, p = 0.5; IHD vs. IDCM, p = 0.0001). Early mortality was different between CHD subgroups (group 1 = 19%, group 2 = 40%, group 3 = 0%, group 4 = 29%; p < 0.001); however, overall mortality did not show differences between subgroups (p = 0.5).
Conclusions: The percentage of Spanish adult HTx patients for CHD is low (1%). The survival curve is better than for other HTx causes (IHD). Nevertheless, early mortality was higher, particularly in some subgroups (Fontan).

María J. Paniagua Martín[a], Luis Almenar[b,c], Vicenç Brossa[d], Marisa G. Crespo-Leiro[a], Javier Segovia[e], Jesús Palomo[f], Juan Delgado[g], Francisco González-Vílchez[h], Nicolás Manito[i], Ernesto Lage[j], Luis García-Guereta[k], José L. Rodríguez-Lambert[l], Dimpna C. Albert[m]

[a]Department of Cardiology, Hospital Universitario A Coruña, A Coruña, [b]Spanish Heart Transplantation Registry, Heart Failure and Transplantation Unit, Spanish Society of Cardiology, [c]Department of Cardiology, Hospital Universitario y Politécnico La Fe, Valencia, [d]Department of Cardiology, Hospital Santa Cruz y San Pablo, Barcelona, [e]Department of Cardiology, Clínica Puerta de Hierro de Majadahonda, Madrid, [f]Department of Cardiology (Adults), Hospital Gregorio Marañón, Madrid, [g]Department of Cardiology, Hospital 12 de Octubre, Madrid, [h]Department of Cardiology, Hospital Marqués de Valdecilla, Santander, [i]Department of Cardiology, Hospital de Bellvitge, Barcelona, [j]Department of Cardiology, Hospital Virgen del Rocío, Sevilla, [k]Department of Pediatric Cardiology, Hospital La Paz, Madrid, [l]Department of Cardiology, Hospital Universitario Central de Asturias, Oviedo [m]Department of Pediatric Cardiology, Hospital Vall d'Hebron, Barcelona, Spain

Key words: adulthood – congenital – heart –

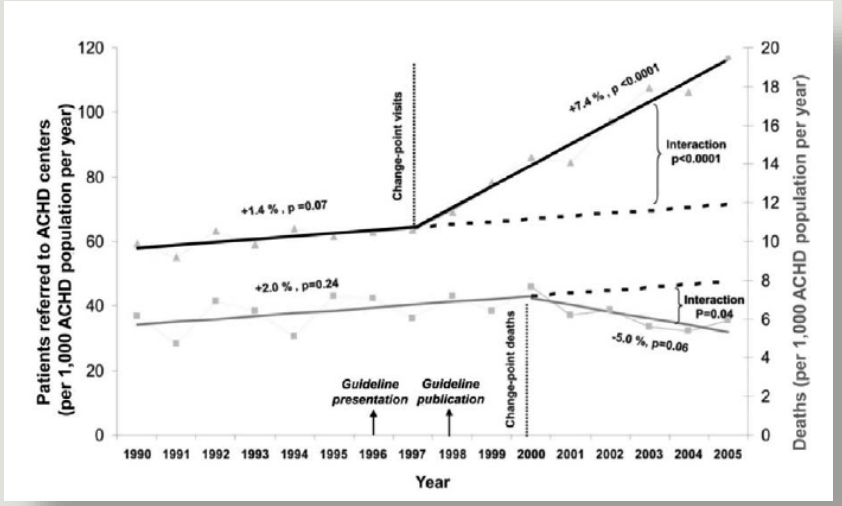


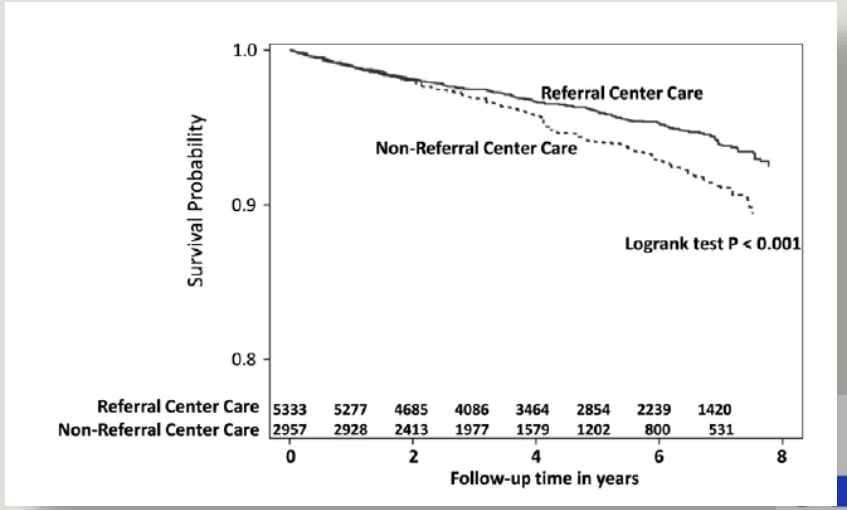
NECESSITAT D'ATENCIÓ ESPECIALITZADA

Specialized Adult Congenital Heart Disease Care

The Impact of Policy on Mortality

Darren Mylotte, MD; Louise Pilote, MD, MPH, PhD; Raluca Ionescu-Ittu, PhD; Michal Abrahamowicz, PhD; Paul Khairy, MD, PhD; Judith Therrien, MD; Andrew S. Mackie, MD, SM; Ariane Marelli MD, MPH





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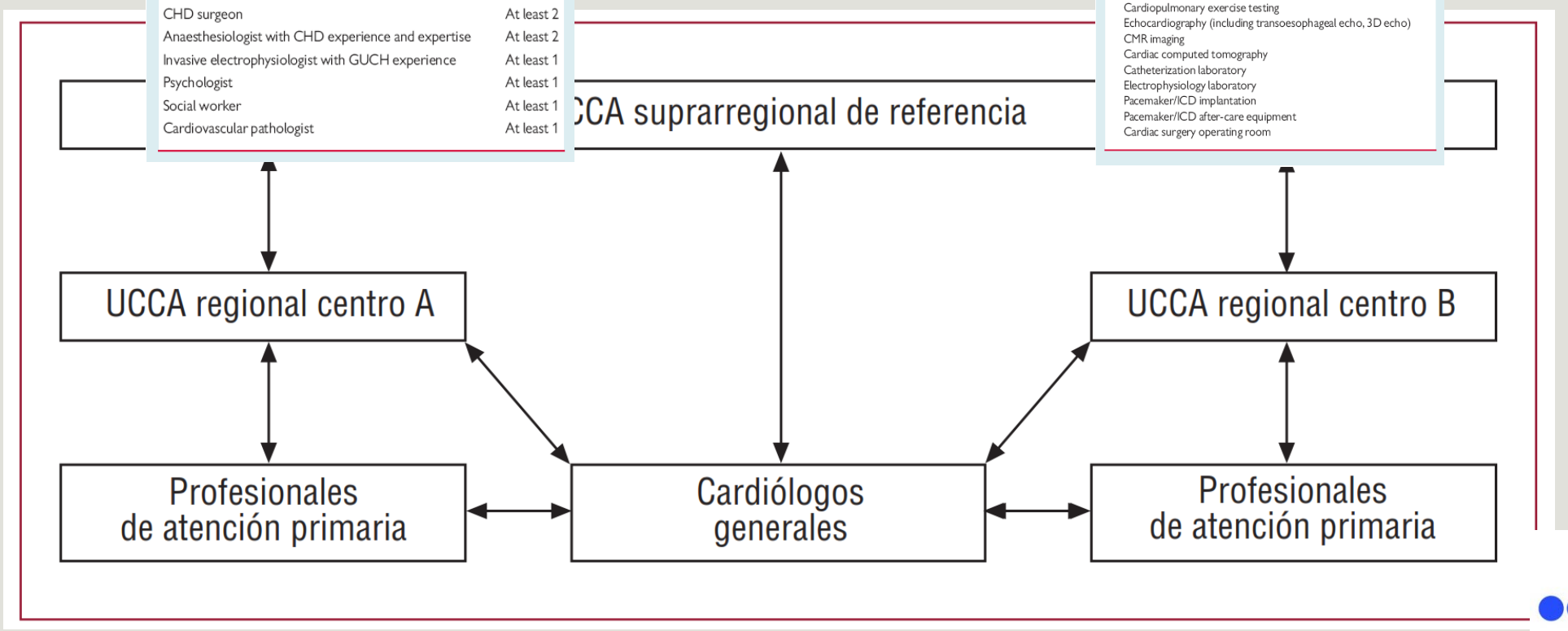
RECOMANACIÓ ORGANITZACIÓ UNITATS D'ATENCIÓ

Table 1 Staff requirements of a specialist grown-up congenital heart centre

Adult/paediatric cardiologist with GUCH certification	At least 2
GUCH imaging specialist (echo, CMR, CT)	At least 2
Congenital invasive cardiologist	At least 2
CHD surgeon	At least 2
Anaesthesiologist with CHD experience and expertise	At least 2
Invasive electrophysiologist with GUCH experience	At least 1
Psychologist	At least 1
Social worker	At least 1
Cardiovascular pathologist	At least 1

Table 2 Equipment requirements of specialist grown-up congenital heart disease centres

- ECG
  - Holter monitoring
  - Stress ECG
  - Ambulatory blood pressure monitoring
  - Event recorder
  - Cardiopulmonary exercise testing
  - Echocardiography (including transoesophageal echo, 3D echo)
  - CMR imaging
  - Cardiac computed tomography
  - Catheterization laboratory
  - Electrophysiology laboratory
  - Pacemaker/ICD implantation
  - Pacemaker/ICD after-care equipment
  - Cardiac surgery operating room



Oechslin EN. REC Suplemento Cardiopatías Congénitas 2009.
Baumgartner et al. Recomendations and organization of care ...EurHJ 2014.
Warnes et al. AHA Guidelines forthe management of ACHD. Circ 2008.
Lanzberg et al. Task Fore 4. Bethesda Conference. JACC 2001
Pla director Cardiopaties Congènites. Generalitat Catalunya 2008.

ORGANITZACIÓ NACIONAL

Unidades CSUR
Unidades Nivell II.

JM Oliver et al. RECC 2019 (accepted)

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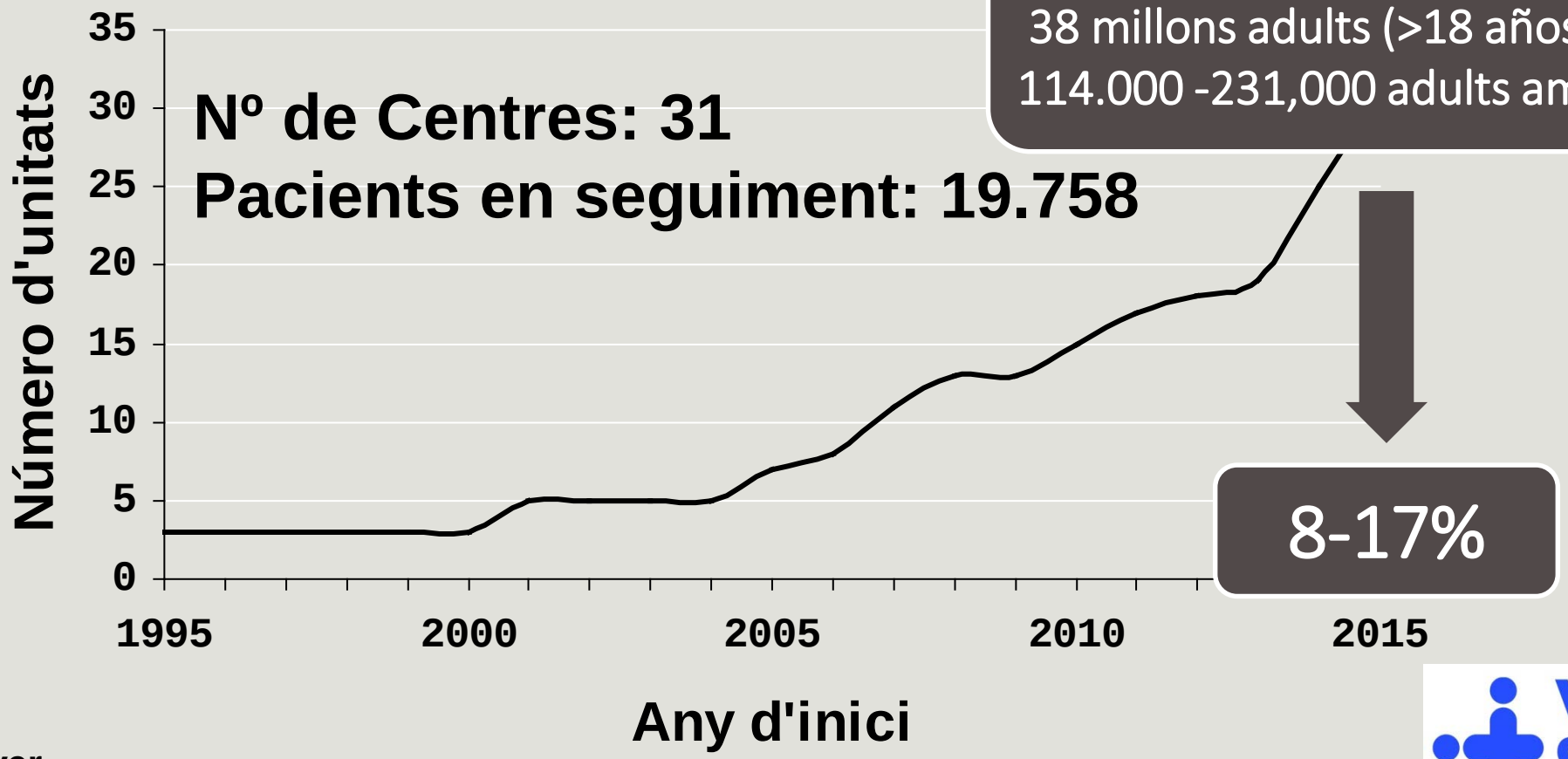
Barcelona Campus Hospitalari

ORGANITZACIÓ NACIONAL

ESPANYA (INE)
38 millons adults (>18 años)
114.000 -231,000 adults amb CC



8-17%

Gentilesa Dr. JM Oliver

Vall d'Hebron
Barcelona Campus Hospitalari

PÈRDUES EN EL SEGUIMENT

Lapse of care as a predictor for morbidity in adults with congenital heart disease

Elizabeth Yeung, Joseph Kay, Genie E. Roosevelt, Mary Brandon, Anji T. Yetman *

International Journal of Cardiology 125 (2008) 62–65

- 158 pacients inclosos
- 63% pèrdua ≥2 anys en transfer
- Mitjana pèrdua: 10 anys (2-50)
- Principal motiu: desconeixement necessitat seguiment

Table 4
Factors associated with need for urgent intervention: univariate analysis

Factor	Odds ratio	95% Confidence interval	*P* value
Lapse of medical care	3.1	1.5, 6.8	0.003
Symptoms on presentation	4.3	2.1, 8.6	<0.001
Additional diagnosis made	9.7	3.8, 24.6	<0.001

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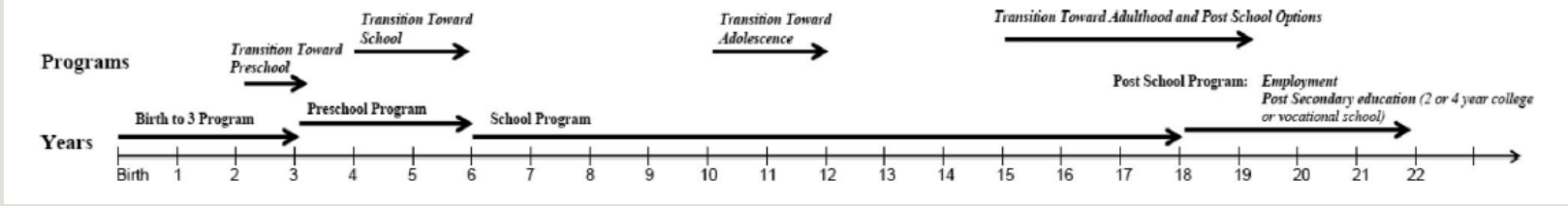
MILLORES EN LA TRANSICIÓ

AHA Scientific Statement

Best Practices in Managing Transition to Adulthood for Adolescents With Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues

A Scientific Statement From the American Heart Association

Craig Sable, MD, FAHA, Co-Chair; Elyse Foster, MD, FAHA, Co-Chair;
Karen Uzark, PhD, PNP, FAHA, Co-Chair; Katherine Bjornsen, BSN, ARNP;
Mary M. Canobbio, RN, MN, FAHA; Heidi M. Connolly, MD; Thomas P. Graham, MD, FAHA;
Michelle Z. Gurvitz, MD, MS; Adrienne Kovacs, PhD, CPsych; Alison K. Meadows, MD, PhD;
Graham J. Reid, PhD, CPsych; John G. Reiss, PhD; Kenneth N. Rosenbaum, MD;
Paul J. Sagerman, MD, MS; Arwa Saidi, MB, BCh; Rhonda Schonberg, MS; Sangeeta Shah, MD;
Elizabeth Tong, MS, RN, CPNP, FAHA; Roberta G. Williams, MD, FAHA;
on behalf of the American Heart Association Congenital Heart Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, Council on Clinical Cardiology, and Council on Peripheral Vascular Disease



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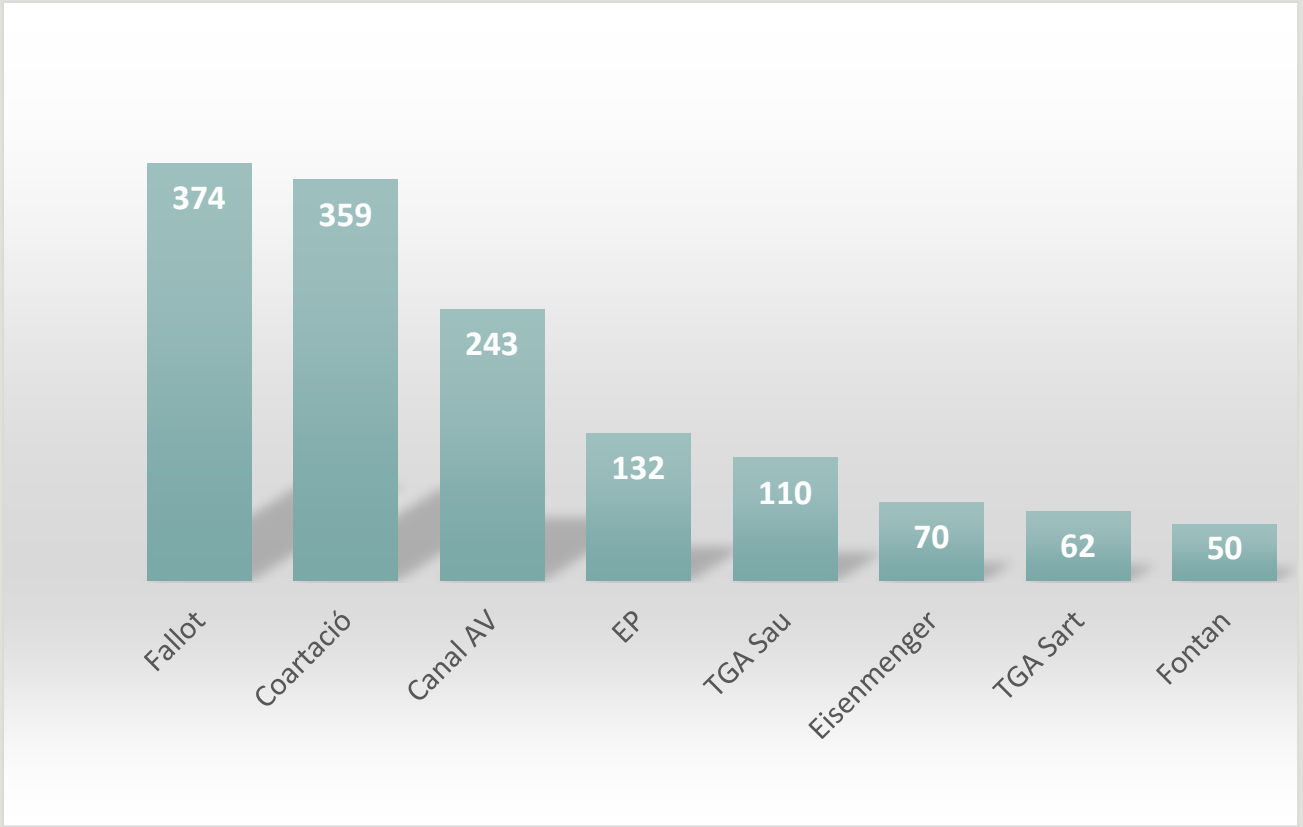
(*Circulation*. 2011;123:1454-1485.)

LA NOSTRA UCCAA

- 4176 pacients (51% homes)
- Edat mitja 36 anys (17-79)
- Mortalitat últims 10 anys:
  - 258 → 28 morts sobtades
- Transplantaments:
  - 8 cardiacs
  - 3 cardio-pulmonar
  - 1 cardio-hepàtic



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CONCLUSIONS

- Les millores en el diagnòstic, tractaments quirúrgic i mèdic, així com de l'esperança de vida en la població general han contribuït a la millor supervivència dels pacients amb CC
- Actualment existeix un major percentatge de pacients amb CC adults que infants
- Pacients que requereixen d'un maneig integral d'una patologia d'elevada complexitat médica → atenció multidisciplinar + consum recursos
- És imprescindible establir una adequada xarxa d'assistència i fer esforços per evitar les pèrdues en el seguiment → pitjor pronòstic
- Les principals causes de morbi-mortalitat actuals arritmiques i ICC → transplantament

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