

Come gestire le cardiopatie congenite dell'adulto?

Gabriella Agnoletti

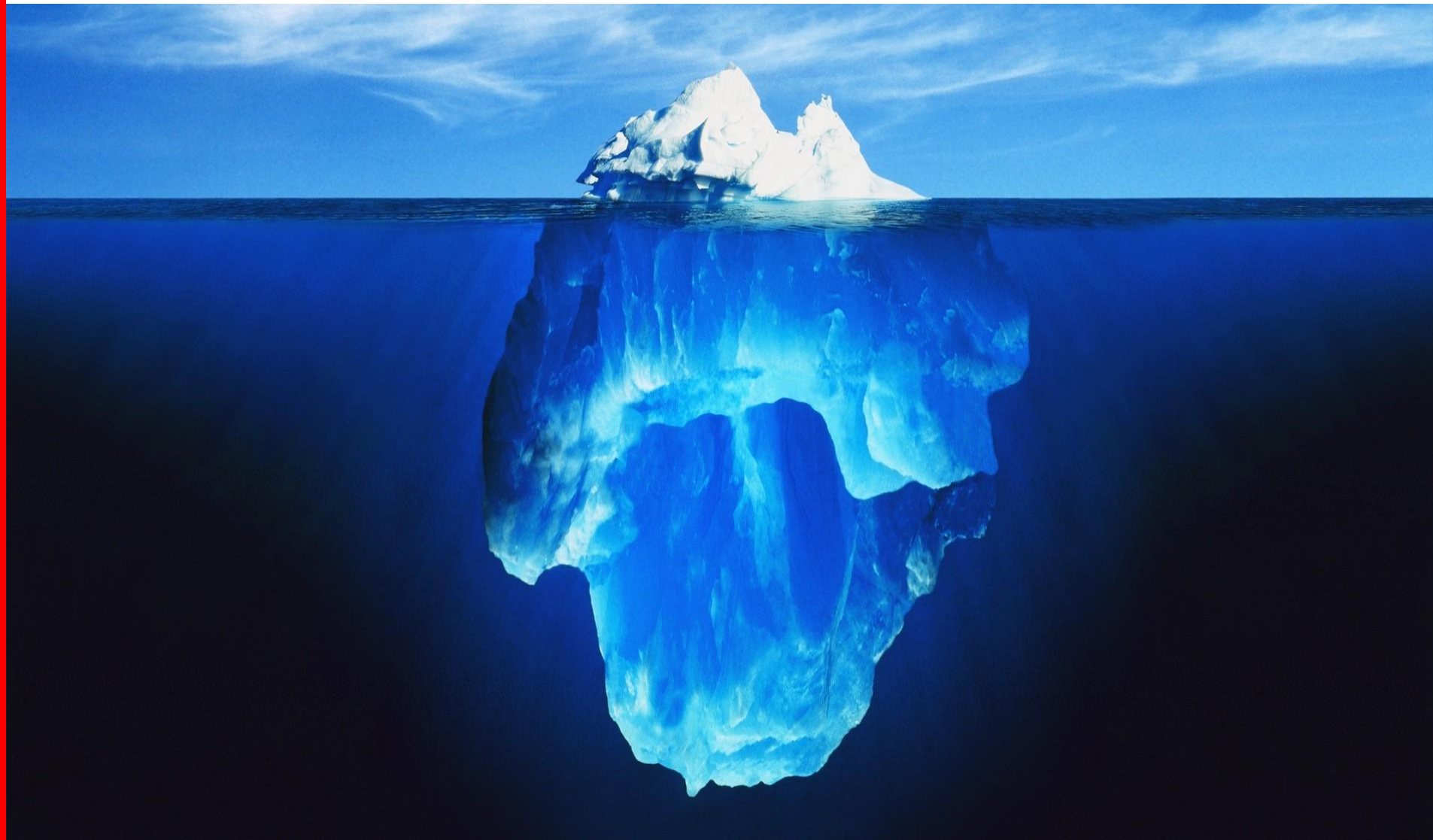
EMOCLINIC SYMPOSIUM
SULLE SPONDE DEL TICINO

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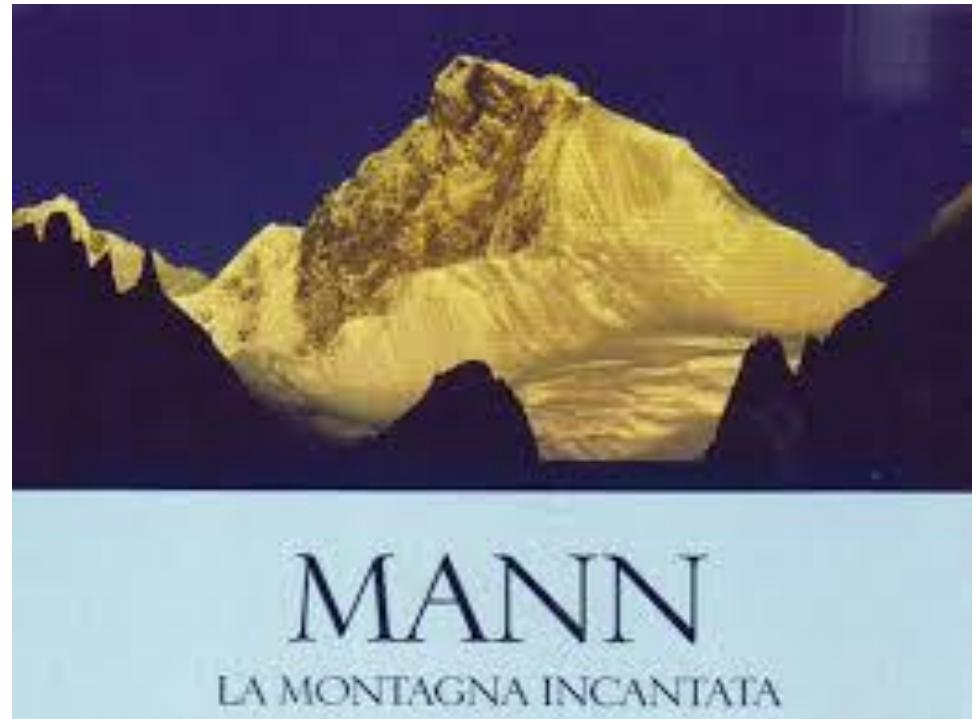


SULLE SPONDE DEL TICINO
CARDIOLOGIA IERI, OGGI E DOMANI
NOVARA, 7 E 8 GIUGNO 2018

Ieri...

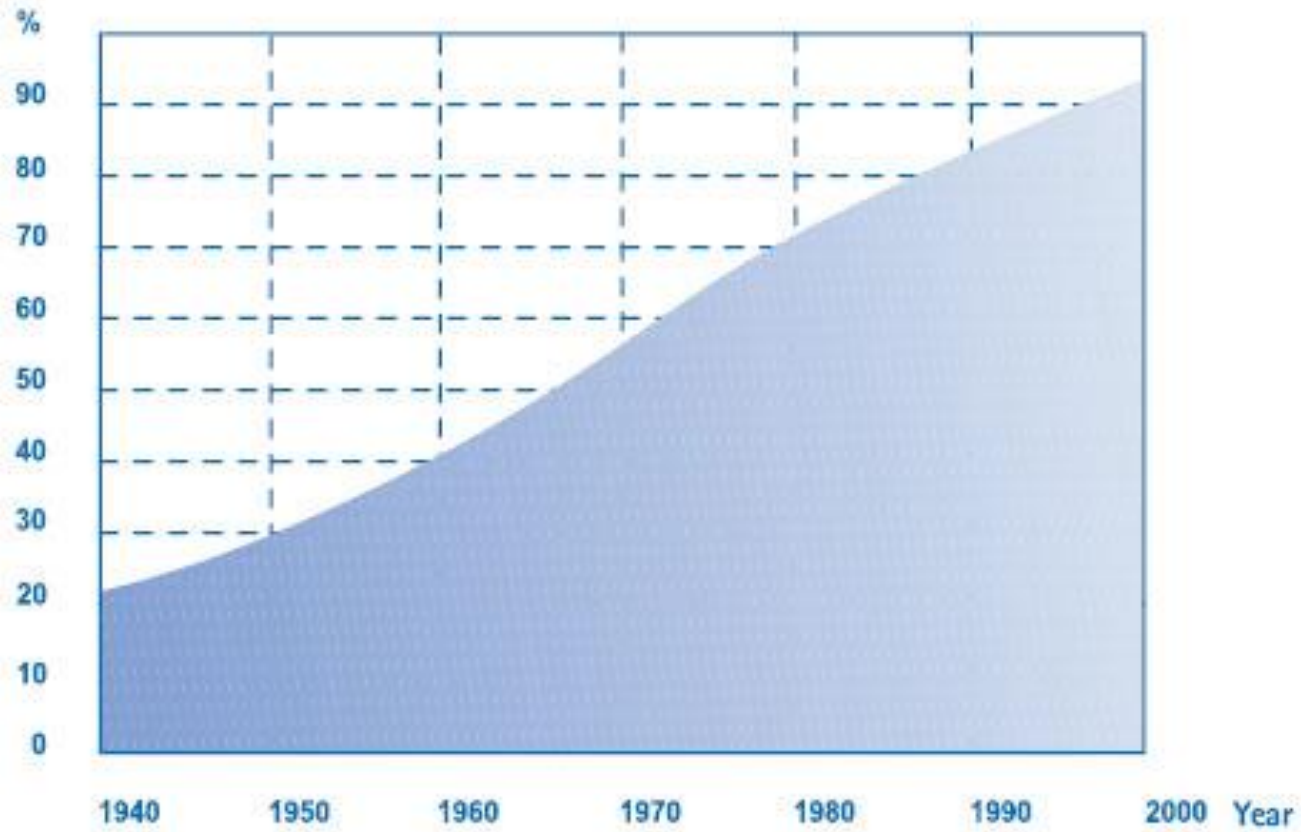


Oggi...



Sopravvivenza delle CC

Survival rate children with CHD

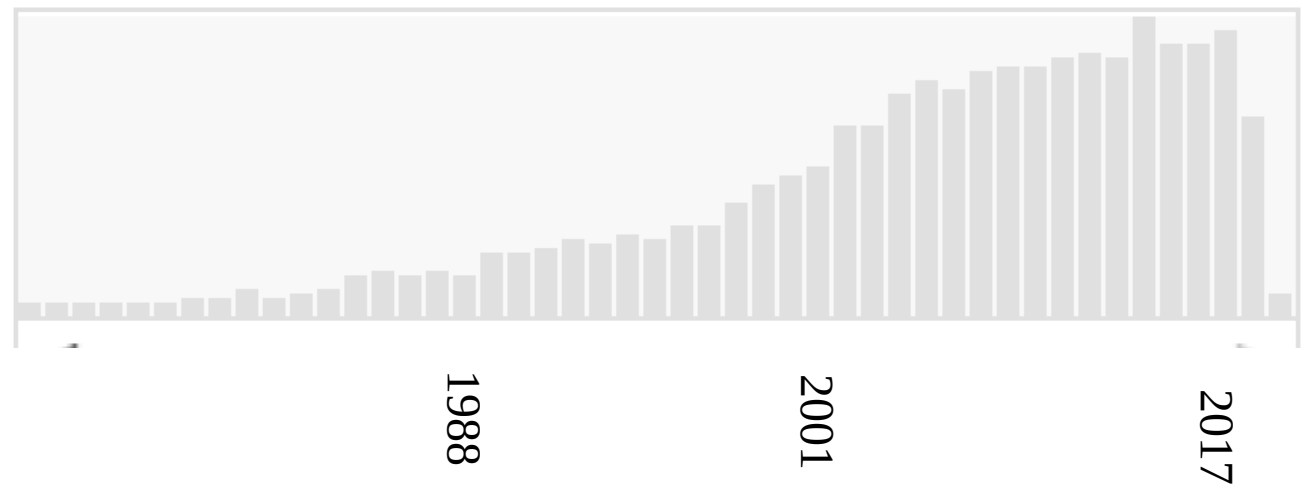


REGISTER ANGEBORENE HERZFEHLER

Pubmed : CHD Adults

- 2017:836
- 2016:736
- 2015:**912**
- 2014:887
- 2010:533
- 2000:202
- 1995:**156**

Results by year



CHI SONO QUESTI
SCONOSCIUTI?



A growing epidemic.....

Canadian Journal of Cardiology 30 (2014) S410–S419

Review

Adult Congenital Heart Disease: A Growing Epidemic

Pablo Ávila, MD, Lise-Andrée Mercier, MD, Annie Dore, MD, François Marcotte, MD, François-Pierre Mongeon, MD, MS, Reda Ibrahim, MD, Anita Asgar, MD, Joaquim Miro, MD, Gregor Andelfinger, MD, Blandine Mondésert, MD, Pierre de Guise, MD, Nancy Poirier, MD, and Paul Khairy, MD, PhD

Montreal Heart Institute Adult Congenital Center, Université de Montréal, Montreal, Quebec, Canada



100.000



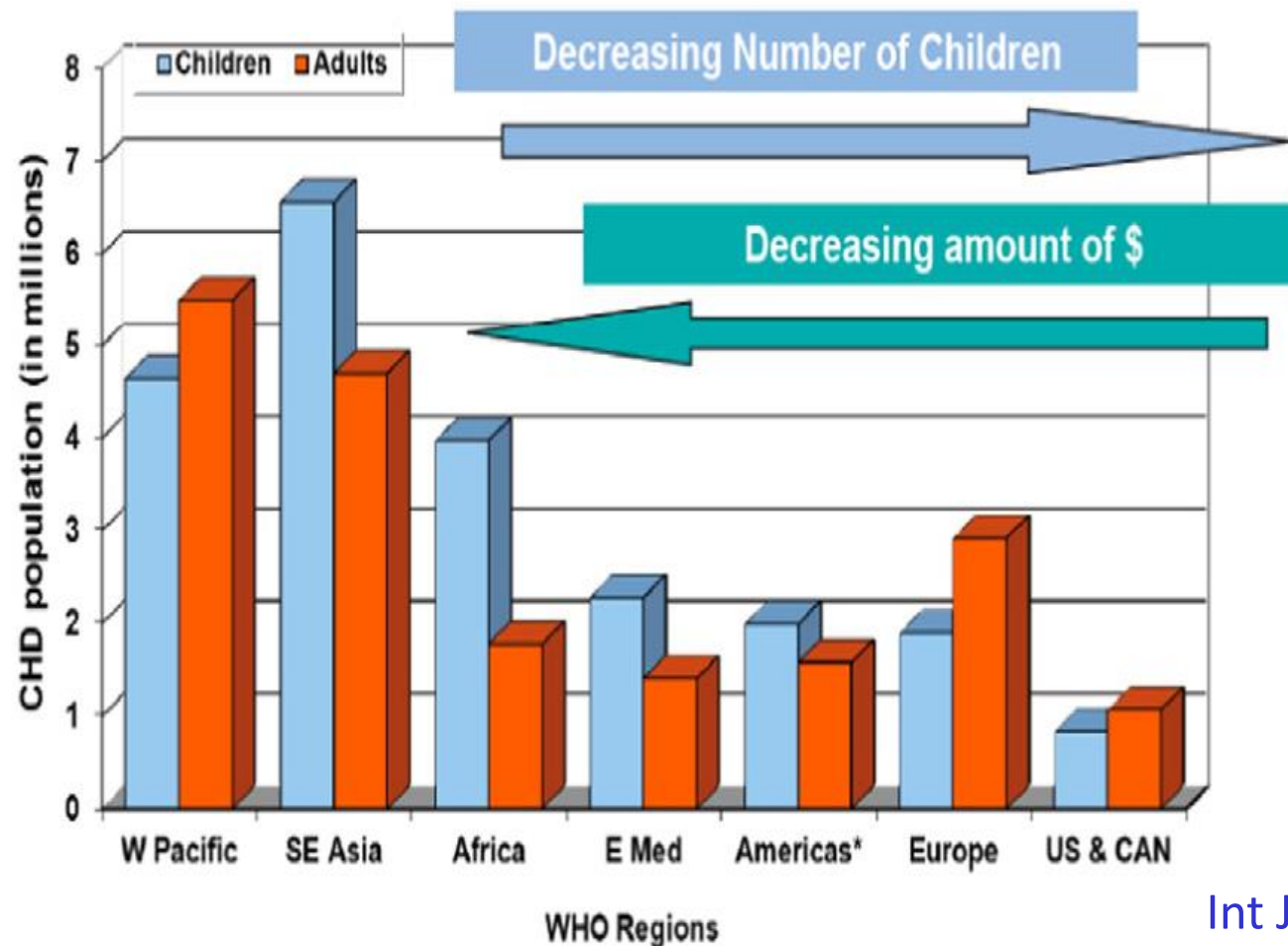
1 milione



1.8 milioni

Incremento stimato nei prossimi 15 anni pari a circa 5% l'anno

Dove sono i pazienti con CHD ?



Int J Cardio 2015

Fig. 1. The congenital heart disease (CHD) burden by WHO region indexed to regional population by age using a CHD prevalence of 12/1000 before and 4/1000 after age 18 years. This figure also illustrates countercurrent direction of resources and number of children with CHD. Where there are more children born and therefore more children with CHD likely to require services there are fewer resources allocated to CHD.

Popolazione NON PIU nuova



European Heart Journal (2014) 35, 673–679
doi:10.1093/eurheartj/ehu045



Congenital heart disease is no longer a paediatric specialty

Adults with congenital heart disease must be managed by a new generation of subspecialist cardiologists, says John Deanfield



(credit Sam Rogers)

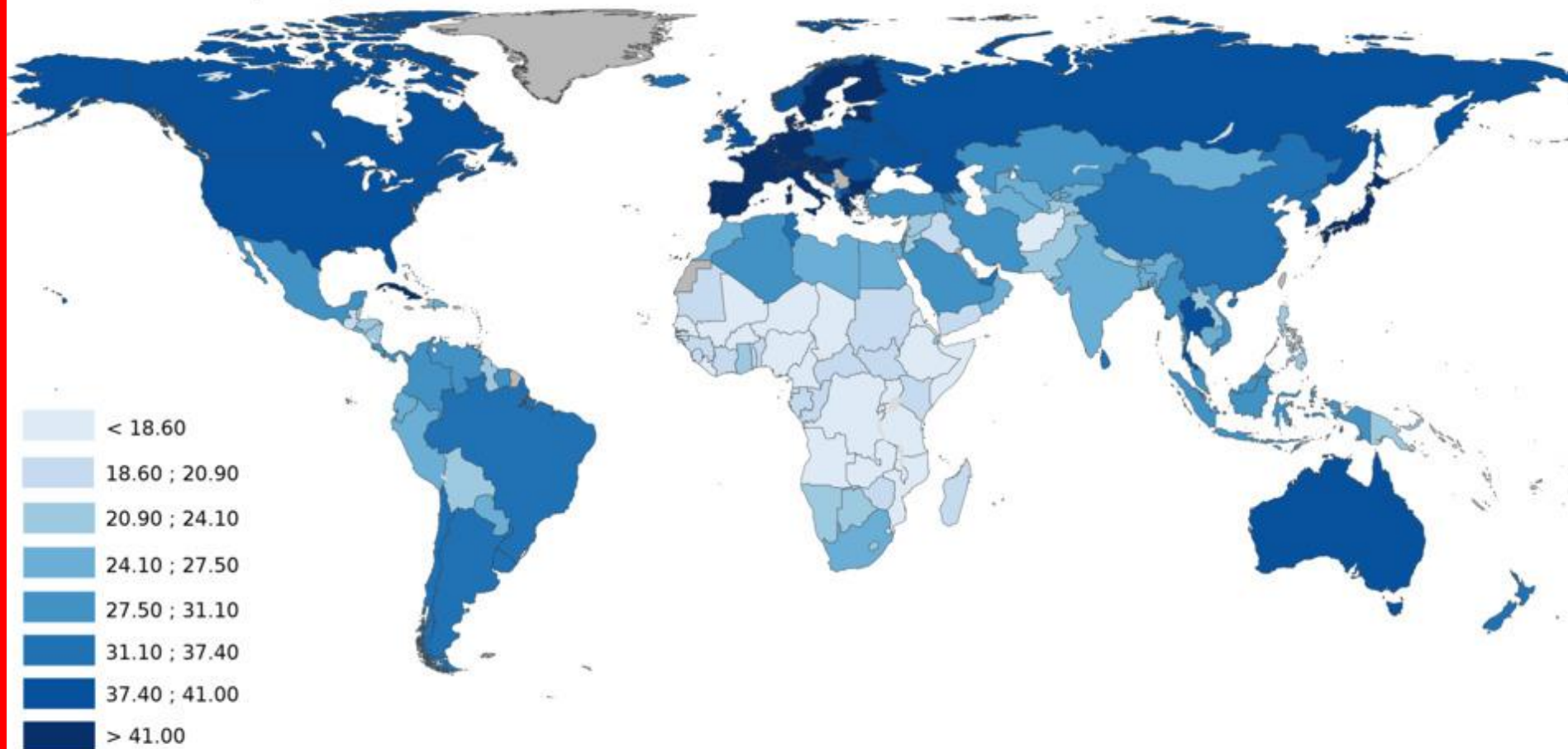
VERO, MA SOLO NEI PAESI CHE INVECCHIANO.....

Cala il numero degli italiani ma sale l'età media.
Sempre più vecchi, nuovo crollo delle nascite

La fotografia Istat: siamo quasi 100mila in meno rispetto a un anno fa. La presenza di stranieri aumenta del 12%

Eta' media nel mondo

Età media (anni)



Fonte : UNDP - 2015

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Dove sono i centri GUCH?

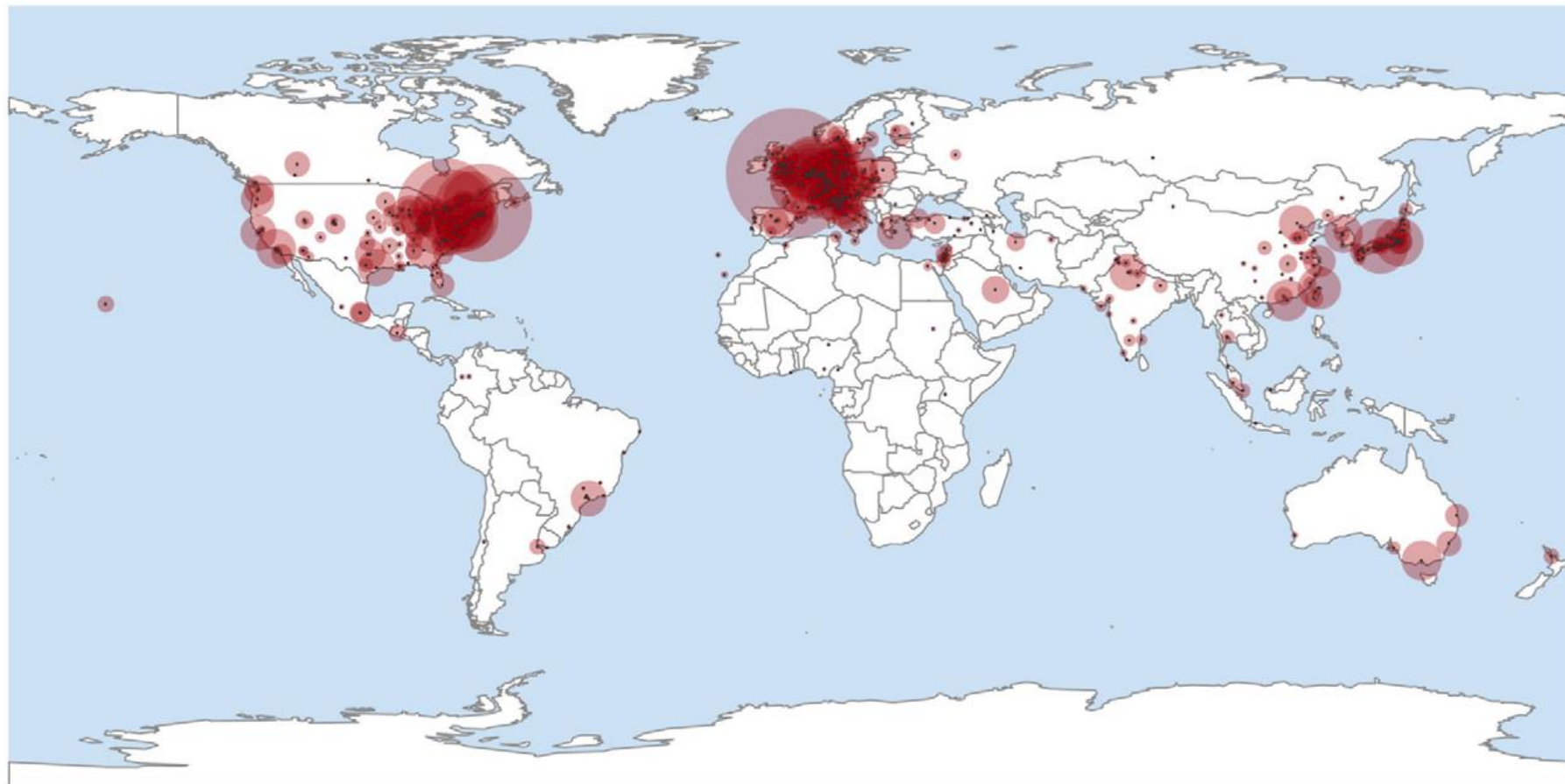
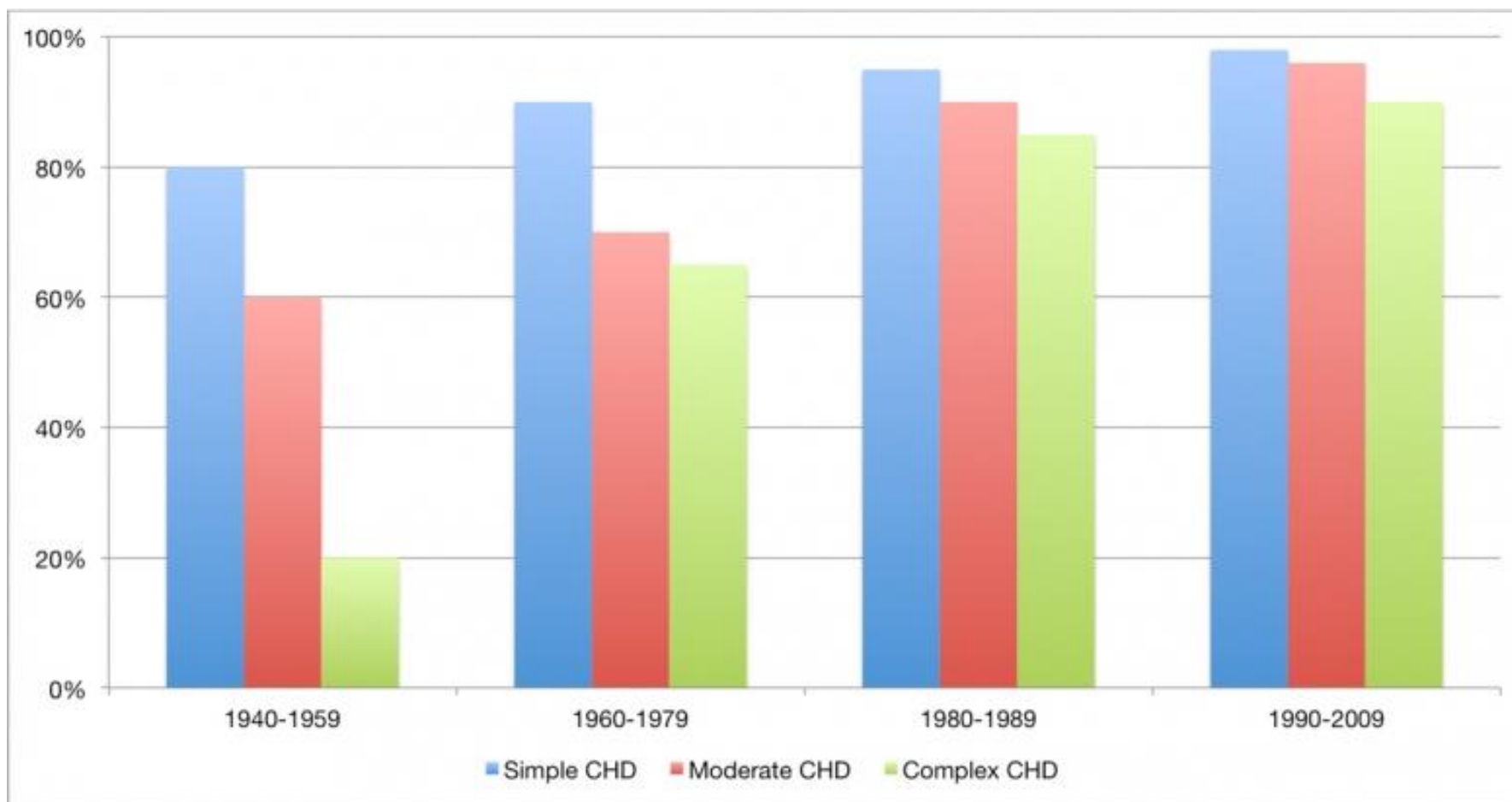
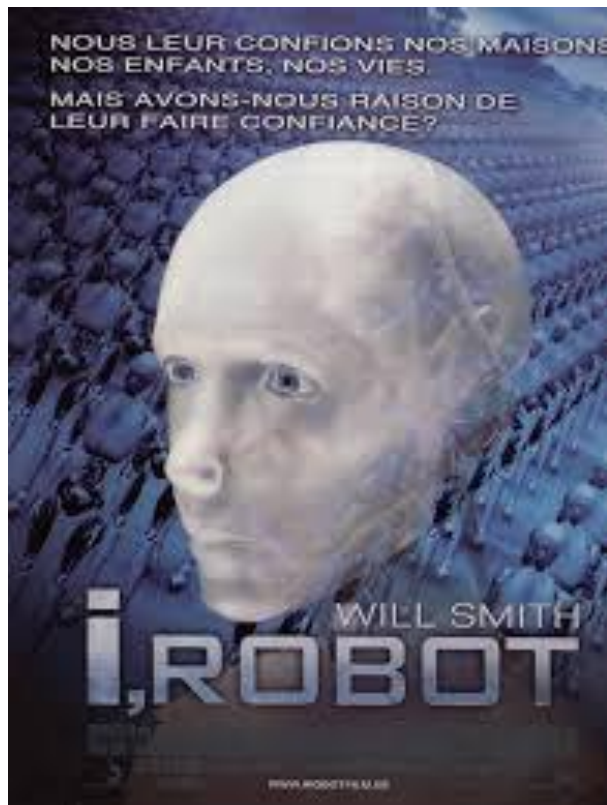


Fig. 3. ACHD centers worldwide. Black dots indicate the identified ACHD centers. The area of each pink circle is proportional to the center's impact factor sum (grouped by city) between 1995 and 2011.

Reproduced with permission from Kempny A et al. *Int J Cardiol* 2013; 168:5182-9 [27].

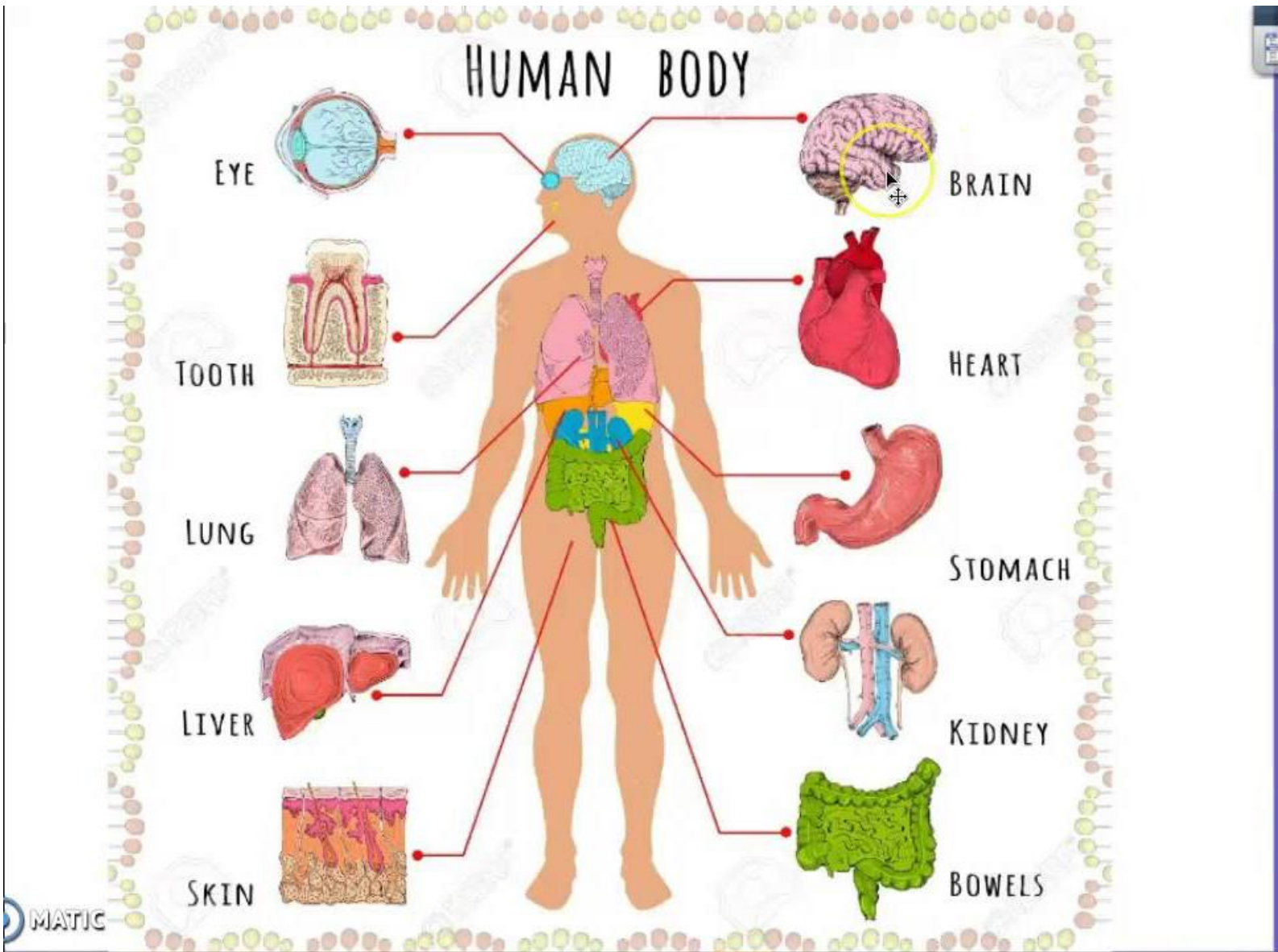


Complessi perche' diversi



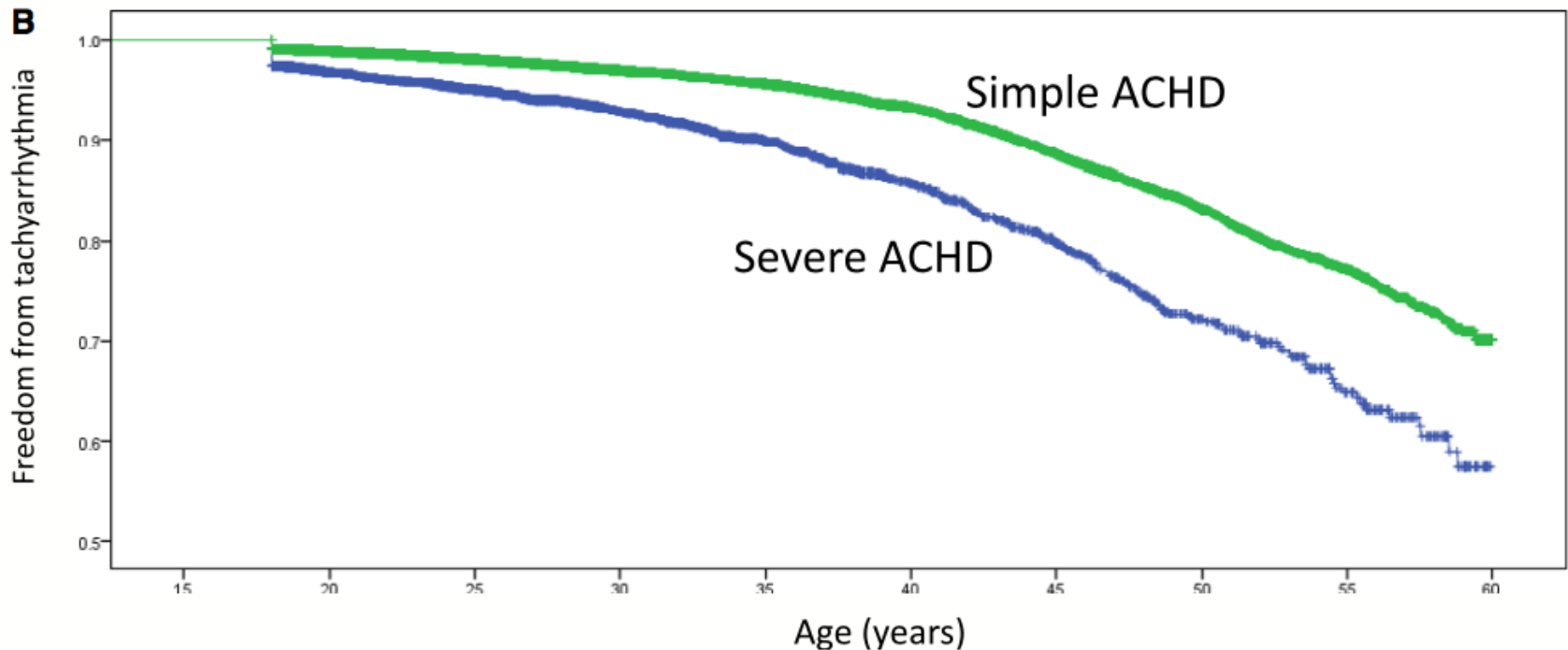
Complessi perche' diversi

- Aritmie: postincisionali
- IC: cicatrici, materiale protesico, fibrosi
- IP: soprattutto cardiopatie semplici....
- Endocardite: ostruttiva
- Problemi sociali e psicologici: malattia cronica.....



RITMO.....

Le aritmie sono la causa piu' comune di visite ospedaliere non programmate ed 1/3 dei ricoveri urgenti



Lo scompenso dei GUCH e' diverso...



European Heart Journal (2016) **37**, 1419–1427
doi:10.1093/eurheartj/ehv741

CURRENT OPINION

Treatment of heart failure in adult congenital heart disease: a position paper of the Working

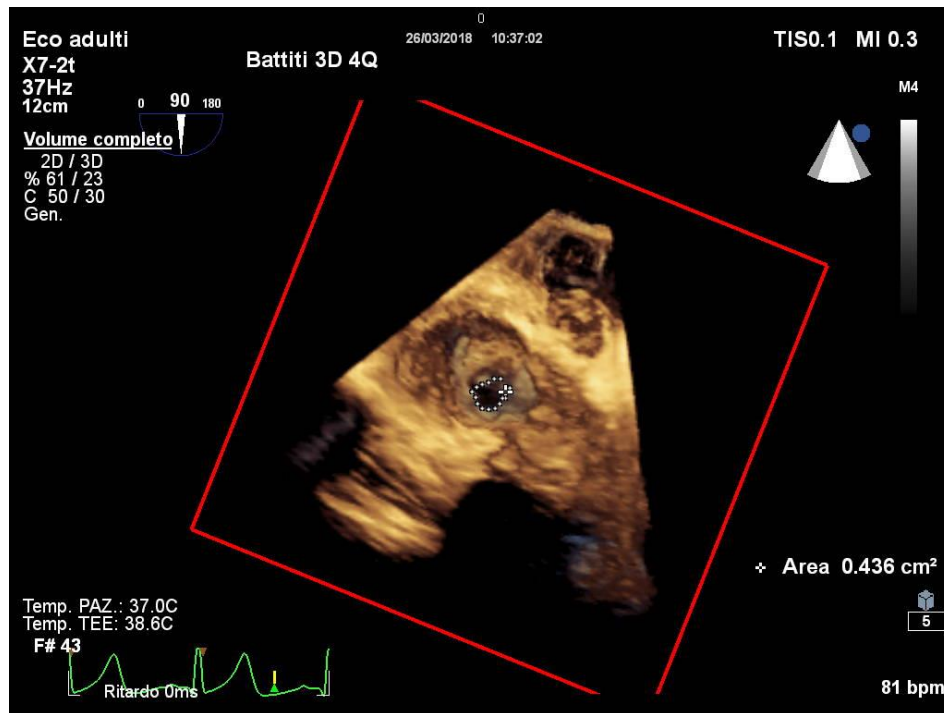
CONCOR: incidenza di ricoveri per scompenso cardiaco nei GUCH di 1.2/1000 pazienti all'anno: 10 volte piu' che nella popolazione generale di pari eta'

and Alexandra A. Frogoudaki

¹Congenital and Structural Cardiology, University Hospitals Leuven, Herestraat 49, B-3000 Leuven, Belgium; ²Department of Cardiology, Erasmus Medical Center Rotterdam, Rotterdam, The Netherlands; ³Department of Pediatric Cardiology, Saarland University Medical Center, Homburg, Germany; ⁴Deutsches Herzzentrum München, Munich, Germany; ⁵Department of Cardiology, King's College Hospital, London, UK; ⁶Department of Nursing, School of Health Sciences Cyprus University of Technology, Limassol, Cyprus; ⁷Advanced

Endocardite

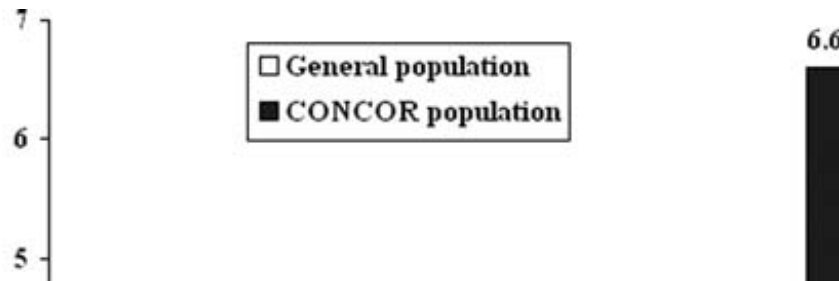
I pazienti GUCH hanno un rischio aumentato di endocardite
1 / 1000 pazienti ha 1 episodio di endocardite all'anno
(20/30 volte maggiore che nella popolazione generale)



Endocardite su
Melody

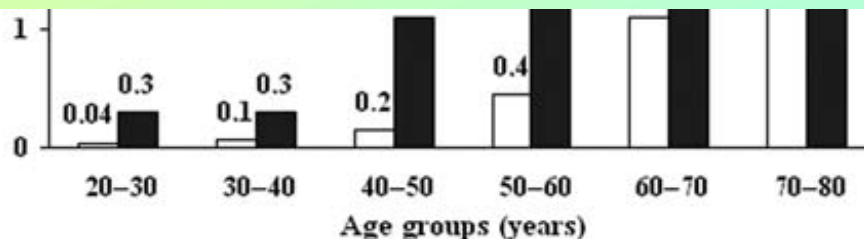
Swiss Med Wkly 2017

Mortalità nei pazienti GUCH CONCOR: >15000 pts



- 30 decessi all'anno
- età mediana: 48.8 aa
- 77% cardiovascolare

L' IC e' una delle due cause principali di decesso.
 Con la morte improvvisa (15- 25% dei decessi)
 costituisce quasi la meta' della mortalita' tardiva
 totale





ISACHD Fellowship Information now on-line

ISACHD Semi-Annual Meeting



We are relocating our ISACHD semiannual meetings to where our members most frequently gather. Hence there will be **no ISACHD meeting in Orlando at ACC** in March, but we look forward to welcoming you to a business meeting to update you on our Society and its exciting developments at the **28th International Symposium on Adult Congenital Heart Disease in Toronto, June 8-9, 2018, Marriott Downtown, Eaton Centre**.
Registration and call for abstracts are

ISACHD Update

Newsflash, February, 2018
 Newsflash, November 16, 2017
 November 2017
 November 2016
 July 2016
 March 2016
 November 2015
 May 2015
 December 2014
 October 2014
 May 2014
 February 2014
 December 2013
 September 2013
 June 2013
 April 2013
 Click for the latest ISACHD news

ISACHD Endorsed Meetings



April 20-21, 2018, Münster, Germany

ACHD FEATURED REVIEWS

Last update April 19, 2017

Body mass index in adult congenital heart disease.
Commentary from Dr. Tabitha Moe (Phoenix)

Pregnancy outcomes in patients with pulmonary arterial hypertension associated with congenital heart disease.
Commentary from Dr. Tabitha Moe (Phoenix)

Value of Cardiovascular Magnetic Resonance Imaging in Noninvasive Risk Stratification in Tetralogy of Fallot.
Commentary from Dr. Maan Johkadar (Atlanta)

Limited Ventricular Preload is the Main Reason for Reduced Stress Reserve after Atrial Baffle Repair.
Commentary from Dr. W. Aaron Kay (Indianapolis)

National In-Hospital Outcomes of Pregnancy in Women with Single Ventricle Congenital Heart Disease.
Commentary from Dr. Blanche Cupido (Cape Town, South Africa)



Contents lists available at ScienceDirect
International Journal of Cardiology
journal homepage: www.elsevier.com/locate/ijcard



Review

The care of adults with congenital heart disease across the globe: Current assessment and future perspective
A position statement from the International Society for Adult Congenital Heart Disease (ISACHD)

Gary Webb^{a,1}, Barbara J. Mulder^{b,1}, Jamil Aboulhosn^c, Curt J. Daniels^d, Maria Amalia Elizari^e, Gu Hong^f, Eric Horlick^g, Michael J. Landzberg^h, Ariane J. Marelliⁱ, Clare P. O'Donnell^j, Erwin N. Oechslin^g, Dorothy D. Pearson^h, Els P.G. Pieper^k, Anita Saxena^l, Markus Schwerzmann^m, Karen K. Stoutⁿ, Carole A. Warnes^o, Paul Khairy^{p,*}

Int J Cardio 2015

Founded in 1992, the International Society for Adult Congenital Heart Disease (ISACHD) is the leading global organization of professionals dedicated to pursuing excellence in the care of adults with CHD worldwide.

The need for specialized care for patients with Grown-up Congenital Heart Disease

The pressing need for subspecialisation and specialized units for the care of Grown-up Congenital Heart Disease (GUCH) patients is discussed by Günter Breithardt with illustration from the system initiated in Germany

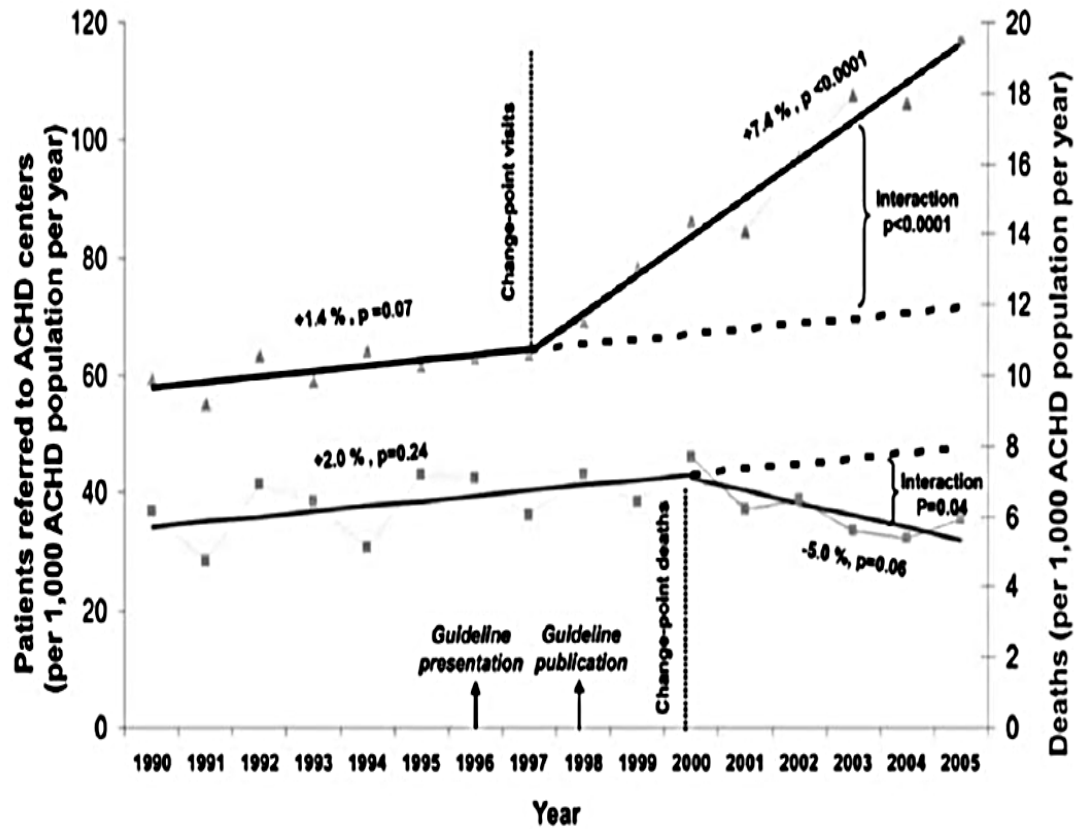
Why are adults with CHD disease
different from other cardiac
condition?

The adult with congenital heart disease gets lost and feels lost

A structured **transition programme** with a defined transfer of care from the paediatric to the adult care environment is important to avoid lapses of care in today's adolescents with CHD.

Centri GUCH

L' Assistenza **dedicata** al paziente GUCH migliora la sopravvivenza e la qualità di vita



Mylotte et al. *Circulation*. 2014;129:1804-1812.

GUCH unit

ROCK principle

Impresa impossibile?

- **R**iconoscerei bisogni
- **O**rganizzare le cure e affrontare le sfide
- **C**entralizzare le cure e gli specialisti
- **K**nowledge specialisti, sottospecialisti



Recommendations for starting a grown up
congenital heart disease (GUCH) unit

Braz J Cardiovasc Surg 2015

ACHD:

Elettrofisiologi, interventisti, cardiocirurghi
CHD, specialisti in IC e trapianto, internisti.

Presenza di almeno:

Pneumologo, gastroenterologo, epatologo,
ematologo, nefrologo, endocrinologo,
ostetrico/ginecologo, genetista, psichiatra,
psicologo, dentista/chirurgo, servizi sociali

Recommendations for organization of care for adults with congenital heart disease and for training in the subspecialty of ‘Grown-up Congenital Heart Disease’ in Europe: a position paper of the Working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology

Helmut Baumgartner^{1*}, Werner Budts², Massimo Chessa³, John Deanfield⁴, Andreas Eicken⁵, Johan Holm⁶, Laurence Iserin⁷, Folkert Meijboom⁸, Jörg Stein⁹, Andras Szatmari¹⁰, Pedro T. Trindade¹¹, and Fiona Walker¹², for the Working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology

Il centro GUCH

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

Organizzazione delle cure

National GUCH centres (Centre of maximum medical care) with all the necessary facilities for the complete medical care of patients concerned, including heart surgery.

Regional GUCH-specialised medical practices and clinics

Patient medical care by adult or paediatric cardiologists, who have undergone or are undergoing further training (GUCH-certified), outpatient departments in the centres for special questions and problems (e.g. rhythmology, pregnancy, etc.), close co-operation with centres of maximum patient care.

Medical care by general practitioners (basic medical care)

Medical care of all patients, provided by general practitioners, general practitioners who are internal specialists, or paediatric and juvenile doctors. Care of all patients in co-operation with GUCH-specialised medical practices.

Fig. 1. Recommendations for the structuring of medical care of GUCH patients.

In CANADA Si considera ragionevole...

Centro NAZIONALE: per una popolazione di **3-10 milioni**

Centro REGIONALE: per una popolazione di **2 milioni**

E inoltre NECESSARIO

Che i cardiologi del centro GUCH acquisiscano
a certificate of competence (an additional qualification)
for “Congenital heart disease in adulthood”,



Table 3 Training in subspecialty grown-up congenital heart disease: specific recommendations

Outpatients [majority with great or moderate complexity (see Supplementary material online, Table S2 and S3)]	≥ 400
Inpatients	≥ 200
Echocardiography in CHD	
TTE performed	≥ 250
TEE performed	≥ 50
CMR/CT in CHD	
Interpreted	≥ 50
Heart catheterization in CHD	
Interpreted	≥ 30
Catheter intervention in CHD	
Participated	≥ 20
Electrophysiology in CHD	
Participated	≥ 5

Lo specialista GUCH

Eur Heart J 2014

Training di 24 mesi:

18 in un centro GUCH accreditato, 6 in una cardiologia generale con ICU, CHF, aritmologia.. per i cardiologi pediatri e 6 mesi di cardiologia pediatrica (centro ad alto volume) per i cardiologi dell'adulto

Imparare a lavorare insieme

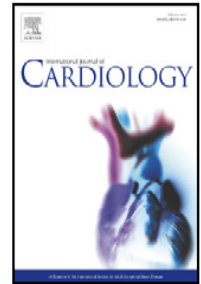
- Anatomia cardiaca



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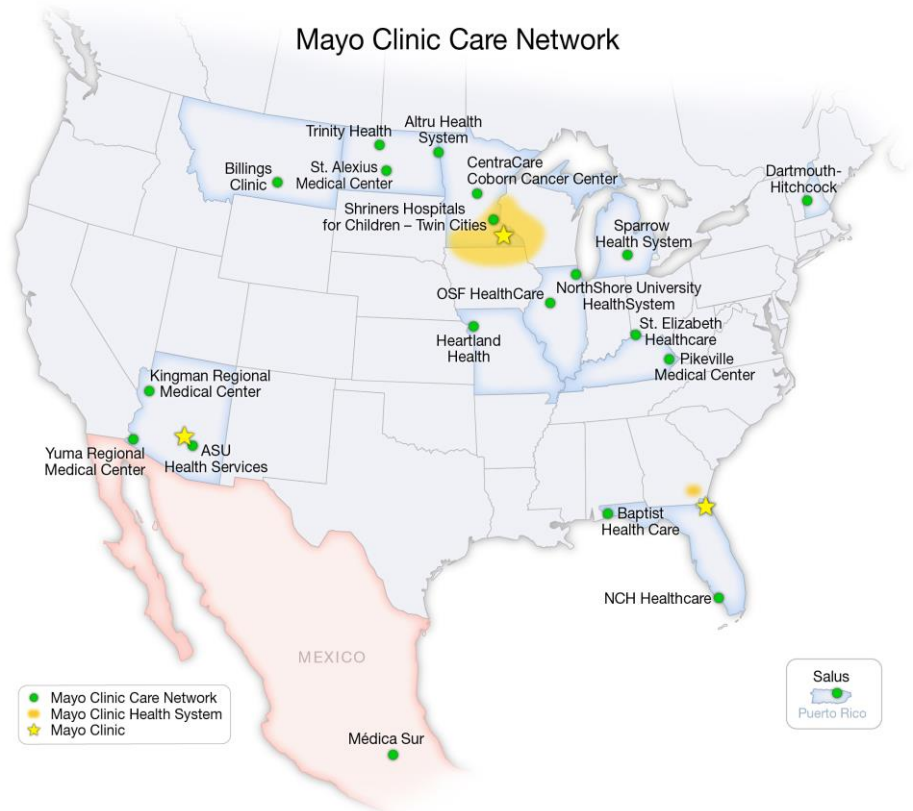
journal homepage: www.elsevier.com/locate/ijcard



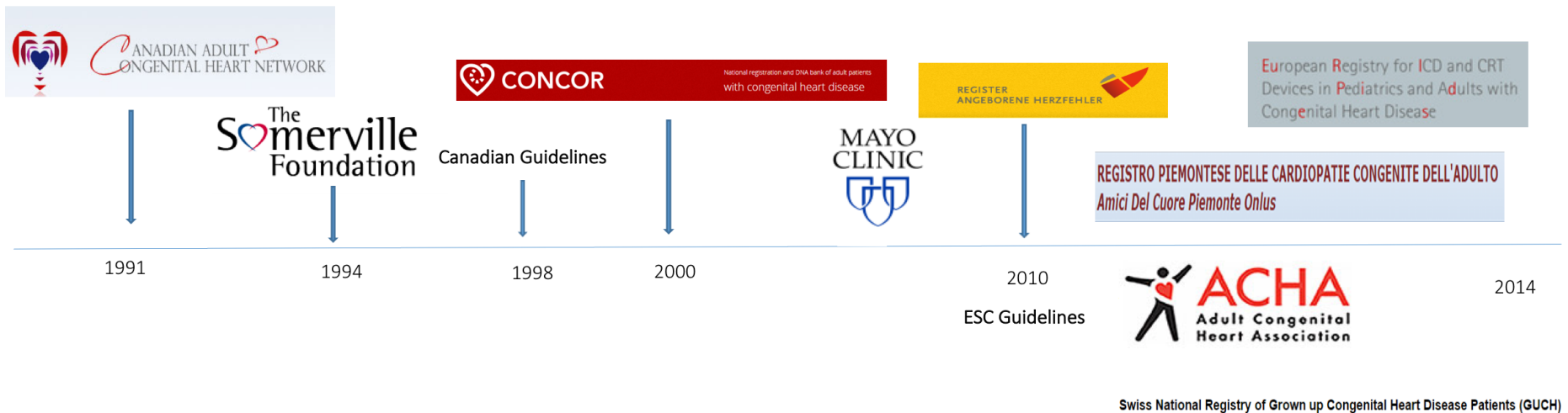
Recommendations for improving the quality of the interdisciplinary medical care of grown-up with congenital heart disease (GUCH)★

- E altro....

La multidisciplinarieta' non basta.....



Reti e Registri



- Danimarca
- Grecia..

REGISTRO PIEMONTESE DELLE CARDIOPATIE CONGENITE DELL'ADULTO



HOME LA RETE I CENTRI CONTATTI LOG IN

Amici Del Cuore

RETE GUCH
GROWN UP CONGENITAL HEART

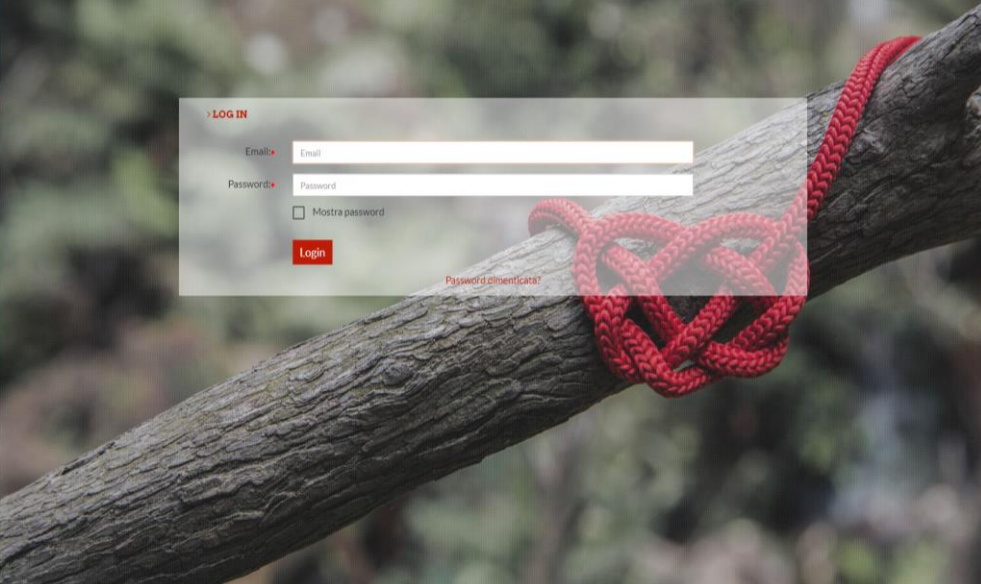
Sono già 91 le strutture che partecipano al progetto!

ENTRA NELLA RETE! 

Amici del cuore
LOG IN



GUCH
Piemonte



LOG IN

Email

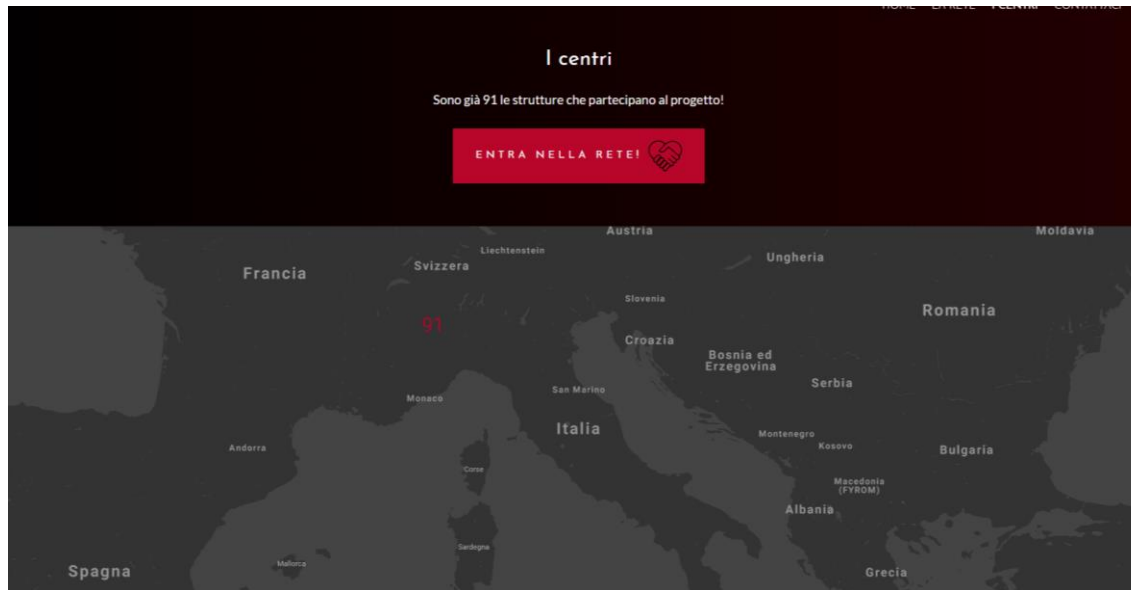
Password

☐ Mostra password

Login

Password dimenticata?

GUCH Piemonte



- Inseriti 910 pazienti
- 30 pazienti PAH: aggiustamento terapeutico
- Corsi/incontri di formazione
- 1 pubblicazione

TAKE HOME message

- Più'

- Si

-

*MULTIDISCIPLINARIETA
LAVORO DI SQUADRA
NECESSITA DI RETI E
REGISTRI*