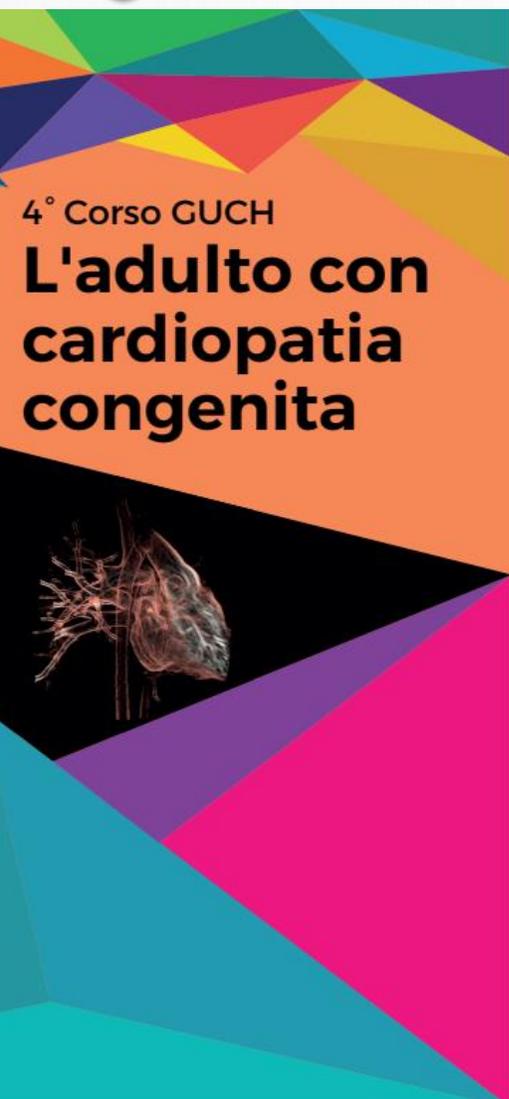


ULTIMO CORSO GUCH 2019

VIVERE CON UNA CARDIOPATIA CONGENITA

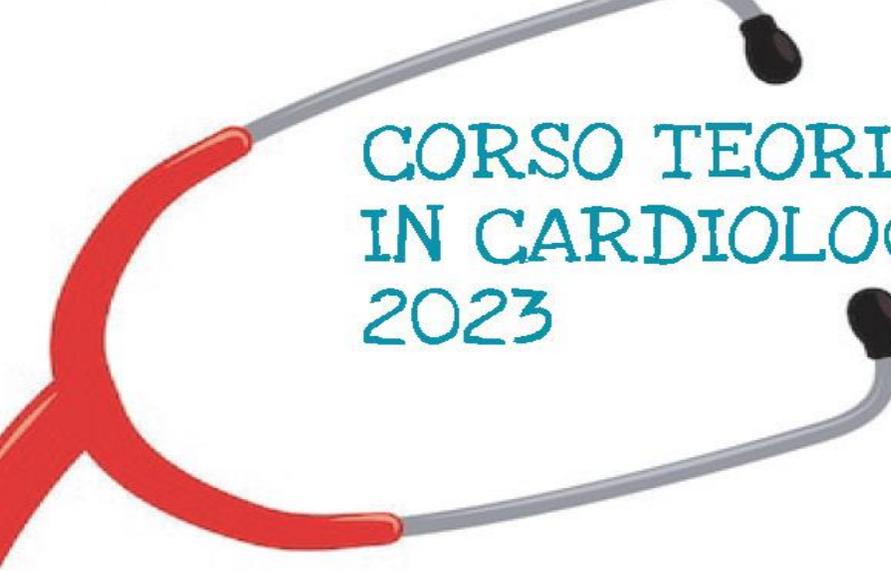


- Le CC sono M CRONICHE (ESC 2020)
- I pazienti vivono ma invecchiano prima
- I Registri
- PAH
- La Gravidanza
- Il Fallot
- La rete

- LATE FONTAN
- ARITMOLOGIA
- PAH
- ASSISTENZE CIRC
- TAVOLA ROTONDA

Cosa è cambiato?





CORSO TEORICO PRATICO IN CARDIOLOGIA PEDIATRICA 2023

I EDIZIONE
Aprile-Giugno

II EDIZIONE
Ottobre-Dicembre

RESPONSABILE SCIENTIFICO
Giuseppe Annoni

COMITATO SCIENTIFICO
Gabriella Agnoletti
Roberto Bordese
Giuseppe Mazza
Anna Maria Villar

SEDE
OSPEDALE INFANTILE REGINA MARGHERITA
Piazza Polonia 94 - TORINO

Corso di ecocardiografia clinica di II livello indirizzato a cardiologi dell'adulto, medici dello sport, pediatri, neonatologi e anestesisti che vogliono approfondire le loro competenze nelle cardiopatie congenite e nella gestione del neonato, del lattante e del bambino con problematiche cardiologiche congenite o acquisite. È richiesta una conoscenza dell'ecocardiografia di base e delle proiezioni ecocardiografiche standard.

Il corso, riservato a massimo 20 partecipanti per edizione, prevede un primo incontro teorico di una giornata in cui verranno trattate tutte le cardiopatie congenite semplici e una parte pratica di 40 ore complessive di frequenza del laboratorio di ecocardiografia, del reparto di cardiologia e neonatologia; un ultimo incontro teorico di mezza giornata chiuderà il corso.

Quota di iscrizione: 500 Euro + IVA comprensiva di materiale didattico, ticket lunch e conseguimento dei crediti ECM in seguito al superamento del test finale.

Accreditamento ECM: 50 crediti.



COMUNICARE, Torino

Tel. 011 660.42.84

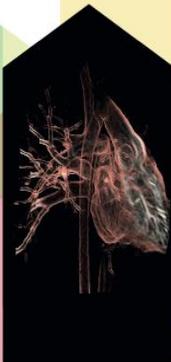
adesionicongressicomunicare@gmail.com

www.eventi-comunicare.it

VI CORSO GUCH

**Il paziente adulto
con cardiopatia
congenita**

TORINO
03 DICEMBRE 2022
NH TORINO CENTRO



IL PAZIENTE ADULTO CON CARDIOPATIA CONGENITA OGGI

G AGNOLETTI

GLI INGREDIENTI

- PERCHÈ NON CHIAMARLI PIU' GUCH
- CERTIFICAZIONE ACHD
- INVECCHIAMENTO DELLA POPOLAZIONE
- MALATTIA E MORTALITA
- TAVI
- TRAPIANTO (IN FONTAN)
- NUOVI SCORES
- DIVERSI?
- IL DIA



PERCHE' ACHD?



- NON SONO PIU' "BAMBINI CRESCIUTI"
- GUCH SOMMERVILLE FINE ANNI 80
- ISACHD 1992
- SPECIALISTI ACHD
- CERTIFICAZIONE ACHD (2 ANNI POST SPECIALITA)
- CENTRI ACHD: NAZIONALE: OGNI 3-10 ML, REGIONALE OGNI 2 ML



European Heart Journal (2010) 31, 2915–2957
doi:10.1093/eurheartj/ehq249

ESC GUIDELINES



ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC)

Endorsed by the Association for European Paediatric Cardiology (AEPC)

Downloaded from



ESC

European Society of Cardiology

European Heart Journal (2021) 42, 563–645
doi:10.1093/eurheartj/ehaa554

ESC GUIDELINES

2020 ESC Guidelines for the management of adult congenital heart disease

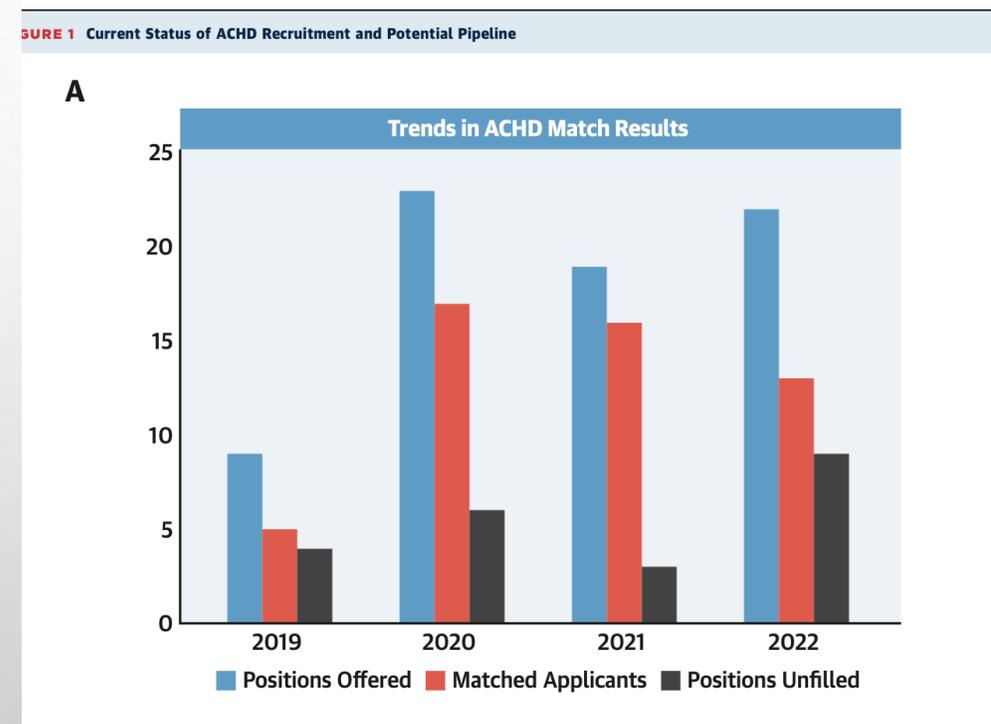
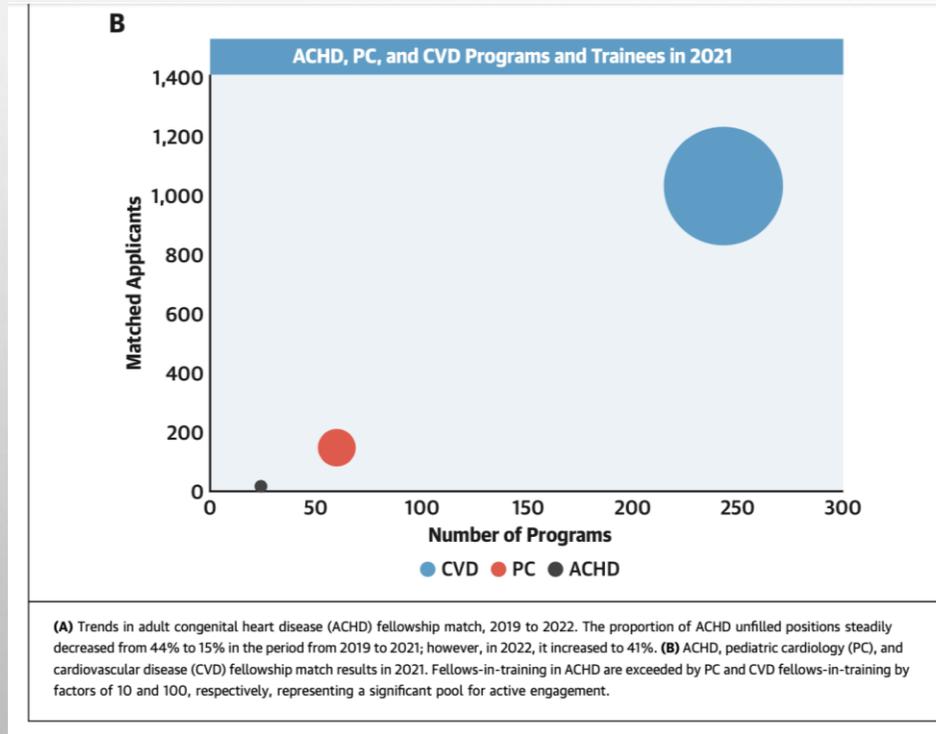
The Task Force for the management of adult congenital heart disease of the European Society of Cardiology (ESC)

Endorsed by: Association for European Paediatric and Congenital Cardiology

Downloaded from

ACHD TRAINING

- CHD: IL PIU' COMUNE DIFETTO CONGENITO (9/1000)
- IL 90% DI MORTALITA SI E TRASFORMATO NEL 90% DI SOPRAVVIVENZA
- ENORME MANCANZA DI SPECIALISTI

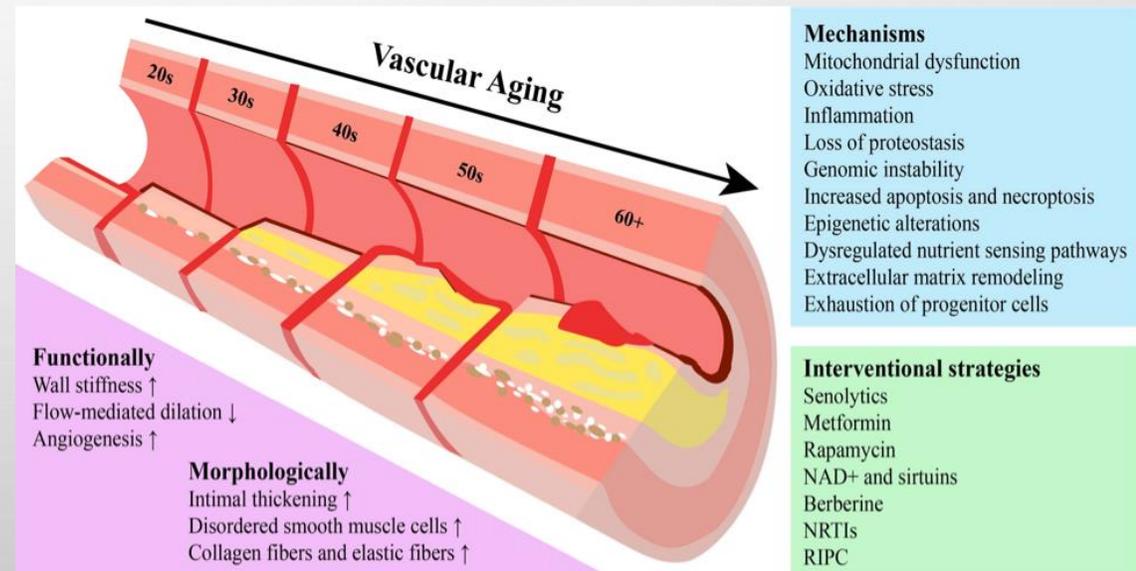
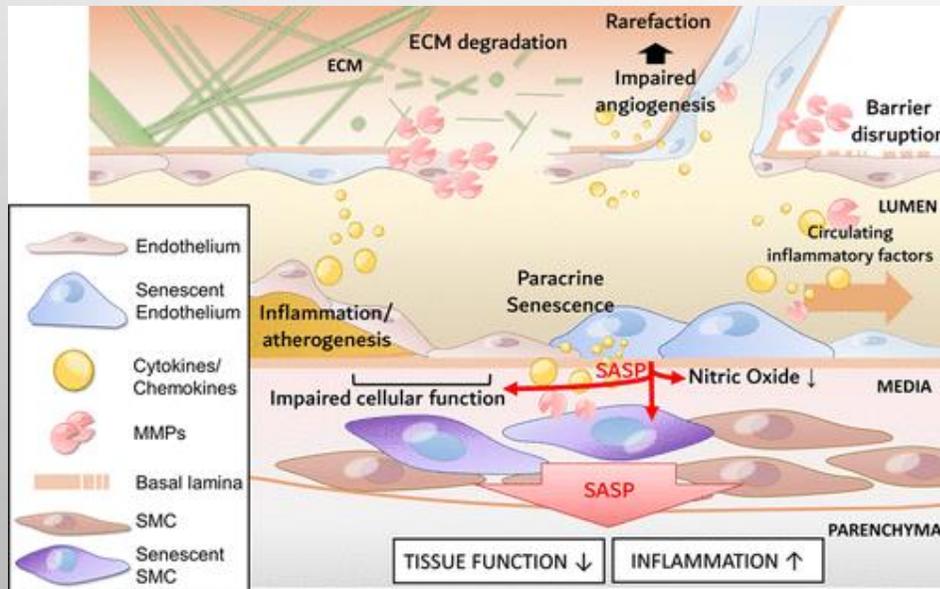


VASCULAR AGING

(EARLY VASCULAR AGING)

UN UOMO È VECCHIO QUANTO LE SUE ARTERIE

GLI ACHD INVECCHIANO
 HANNO PRECOCEMENTE LE MALATTIE DELLA VECCHIAIA
TUMORI-POLMONITE-M CARDIOVASCOLARI





ACCELERATED CARDIAC AGING

 **frontiers** | Frontiers in **Cardiovascular Medicine**

REVIEW
published: 26 May 2022
doi: 10.3389/fcvm.2022.892861



Accelerated Cardiac Aging in Patients With Congenital Heart Disease

*Dominga Iacobazzi, Valeria Vincenza Alvino, Massimo Caputo and Paolo Madeddu**

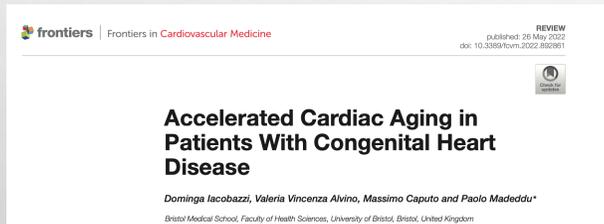
Bristol Medical School, Faculty of Health Sciences, University of Bristol, Bristol, United Kingdom

- **Cardiac aging**
- **Senescence**
- **Proinflammatory state**
- **Repeated stress**

CHF nel 25% of ACHD by the age of 30
NOT limited to complex CHD
Conventional treatment unsupported by clinical evidence

CHF DIVERSA E REFRATTARIA

- L'ETÀ E' IL MAGGIOR FATTORE DI RISCHIO DELLE M. CARDIOVASCOLARI



LA STORIA SI RIPETE

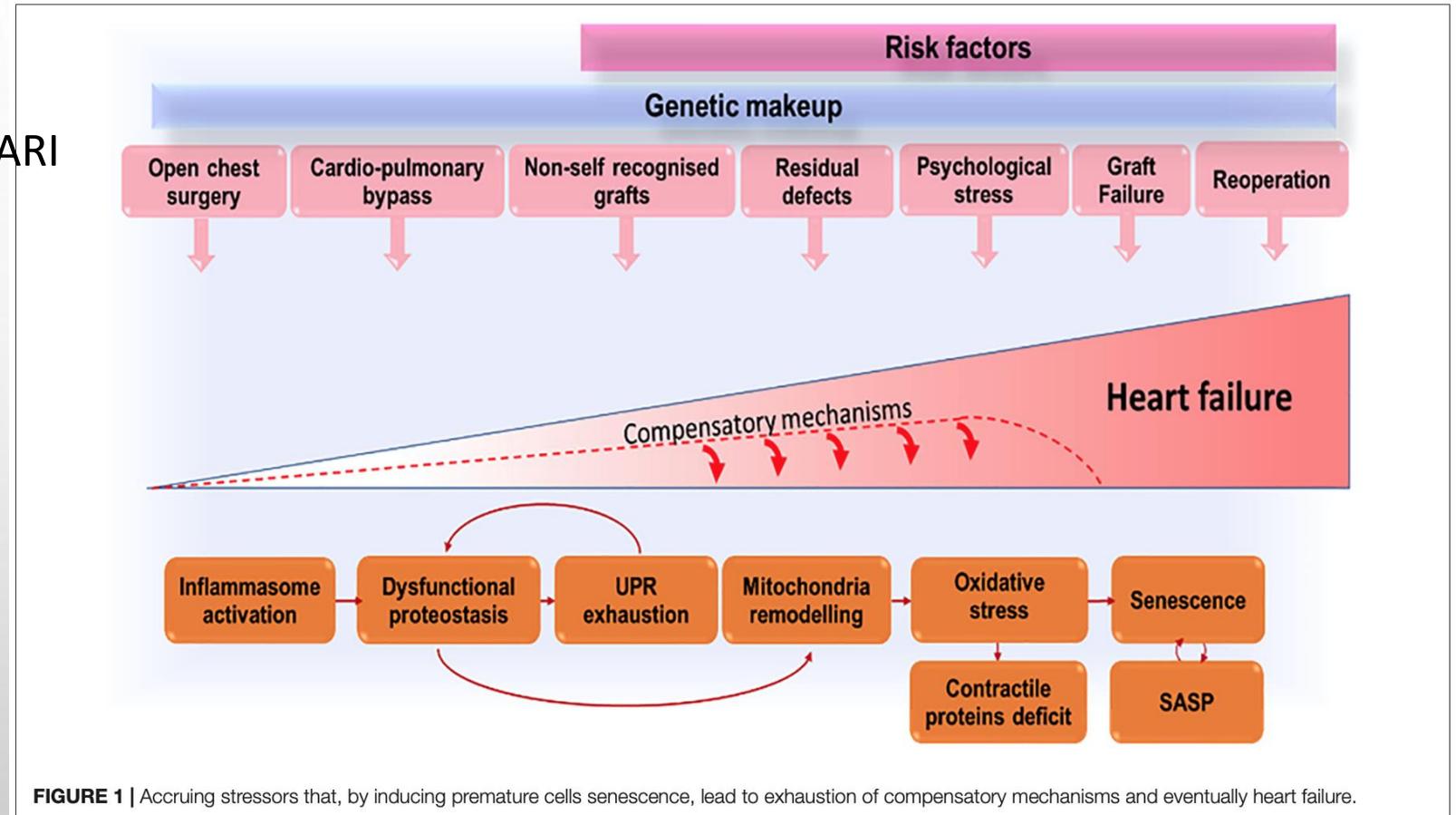


FIGURE 1 | Accumulating stressors that, by inducing premature cells senescence, lead to exhaustion of compensatory mechanisms and eventually heart failure.

VASCULAR AGING

Review Article

Vascular aging in adult congenital heart disease-a narrative review

Tomoaki Murakami

Cardiovascular Diagnosis and Therapy, Vol 12, No 4 August 2022

CIANOSI

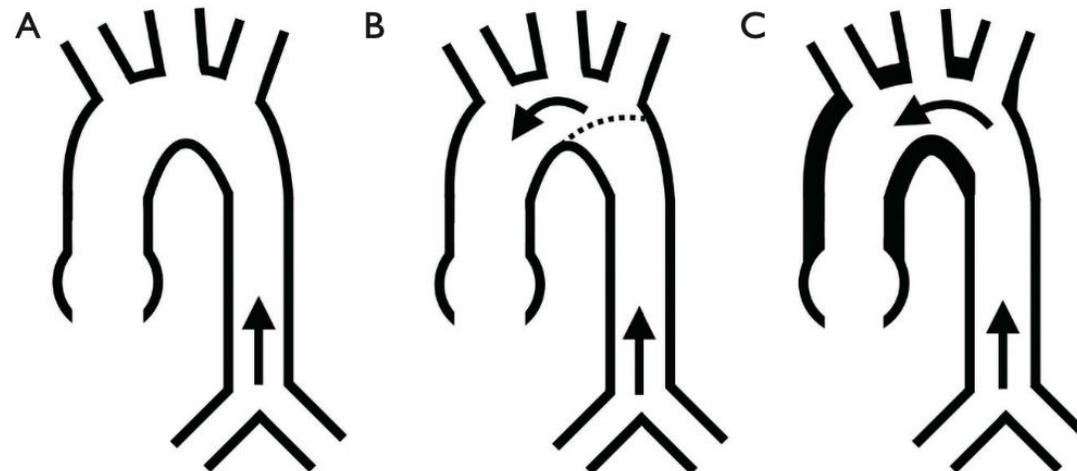


Figure 5 Possible origins of the reflected pressure wave. (A) Normal aorta. (B) Repaired aortic coarctation. (C) Cyanotic heart disease. Arrow indicates the reflected pressure wave.



NON SOLO CUORE

- M METABOLICA
- M ISCHEMICA
- AORTOPATIA
- IPERTENSIONE ARTERIOSA (**OVERLOAD, CIANOSI, SHUNTS, RV SISTEMICO**)
- M CEREBROVASCOLARE
- M RENALI (CIANOSI , CCH)
- AUMENTATA MORTALITA PER M ACQUISITE

Review Article



Vascular aging in adult congenital heart disease-a narrative review

Tomoaki Murakami

Department of Pediatrics, Sapporo Tokushukai Hospital, Sapporo, Japan

Correspondence to: Tomoaki Murakami, MD, PhD. Department of Pediatrics, Sapporo Tokushukai Hospital, 1-1, Higashi 1-chome, Ohyachi, Atsubetsu-ku, Sapporo 004-0041, Japan. Email: murat@seagreen.ocn.ne.jp.

Cardiovasc Diagn Ther 2022



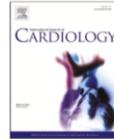
FRAILTY

International Journal of Cardiology 363 (2022) 30–39

Contents lists available at ScienceDirect

International Journal of Cardiology

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



Rationale, design and methodology of APPROACH-IS II: International study of patient-reported outcomes and frailty phenotyping in adults with congenital heart disease

APPROACH-IS-II



Fig. 2. Geographic distribution of the APPROACH-IS II participating centers. Yellow dots indicate centers that are participating in Part 1 and Part 2 of the study ($n = 21$). Pink dots indicate centers that are participating in Part 1 only ($n = 32$). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Gli studi multicentrici sovranazionali iniziano dopo gli anni 2000

E POICHE SONO FRAGILI E INVECCHIANO PRIMA MENO CHIRURGHI, PIU INTERVENTISTI

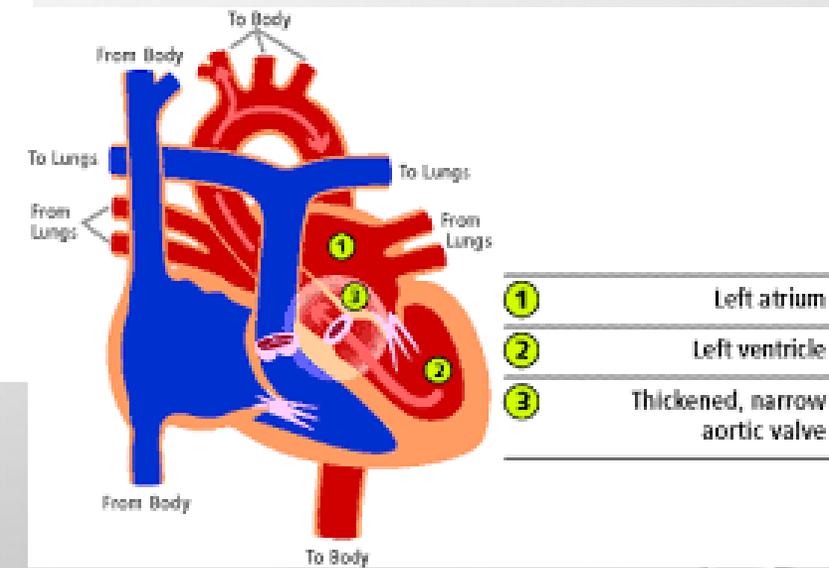
- TAVI E NON SOLO NEL PAZIENTE ACHD: BAMBINI ADOLESCENTI E GIOVANI ADULTI

Transcatheter and Surgical Aortic Valve Implantation in Children, Adolescents, and Young Adults With Congenital Heart Disease



Dwight M. Robertson, DO^{a,b,*}, Dana M. Boucek, MD^a, Mary Hunt Martin, MD^a, Robert G. Gray, MD^a,
Eric R. Griffiths, MD^c, Aaron W. Eckhauser, MD^c, Zhining Ou, MS^d, Linda M. Lambert, MSN-cFNP^a,
Richard V. Williams, MD^a, and S. Adil Husain, MD^c

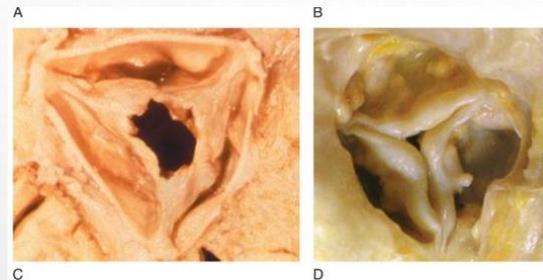
Am J Cardiol 2022



SAVR VS TAVI

ESPERIENZA SALT LAKE

- 30 SAVR E 17 TAVI
- ETA' 10-21 ANNI
- FU 3.8 E 1.5 AA
- OUTCOME SIMILE IN TERMINI DI LIBERTA' DA STROKE ENTRO 6 MESI, RIOSPEDALIZZAZIONE ENTRO 30 GG E DECESSO
- PIU LUNGA DEGENZA IN SAVR, PIU RISCHIO DI REINTERVENTO IN TAVI



Edwards valve
(Sapien 3)



Medtronic valve
EvolutR

TRAPIANTO CARDIACO



ORIGINAL CONTRIBUTION

Observations and Single-Center Outcomes in Orthotopic Heart Transplant for Patients With Adult Congenital Heart Disease: A Call for Equity and Parity

Joshua Rezkalla^{a*}, Megan Kamath^b, and Leigh Reardon^c

^aDepartment of Medicine-Pediatrics, David Geffen School of Medicine, University of California Los Angeles, Los Angeles, California; ^bAhmanson/UCLA Cardiomyopathy Center, Los Angeles, California; and ^cAhmanson/UCLA Adult Congenital Heart Disease Center, Los Angeles, California

- Selezione
- Criteri non uniformi
- Timing (messa in lista tardiva)
- Disfunzione multiorgano
- Indicazioni all'assistenza meccanica
- Carezza di chirurghi congenitalisti

4

REZKALLA, KAMATH, REARDON ET AL

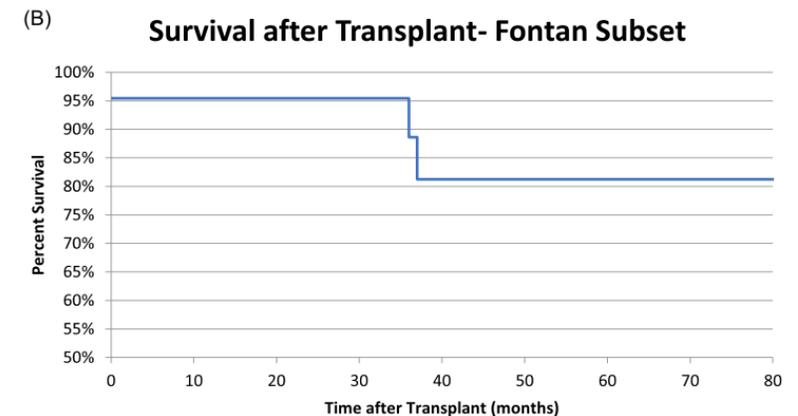
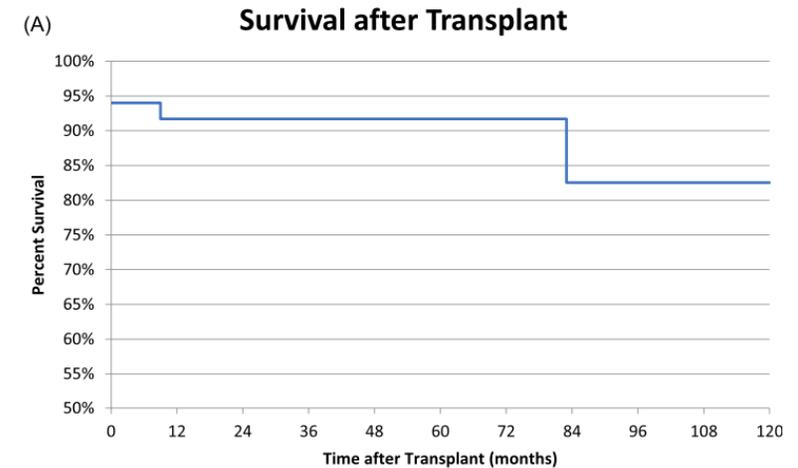


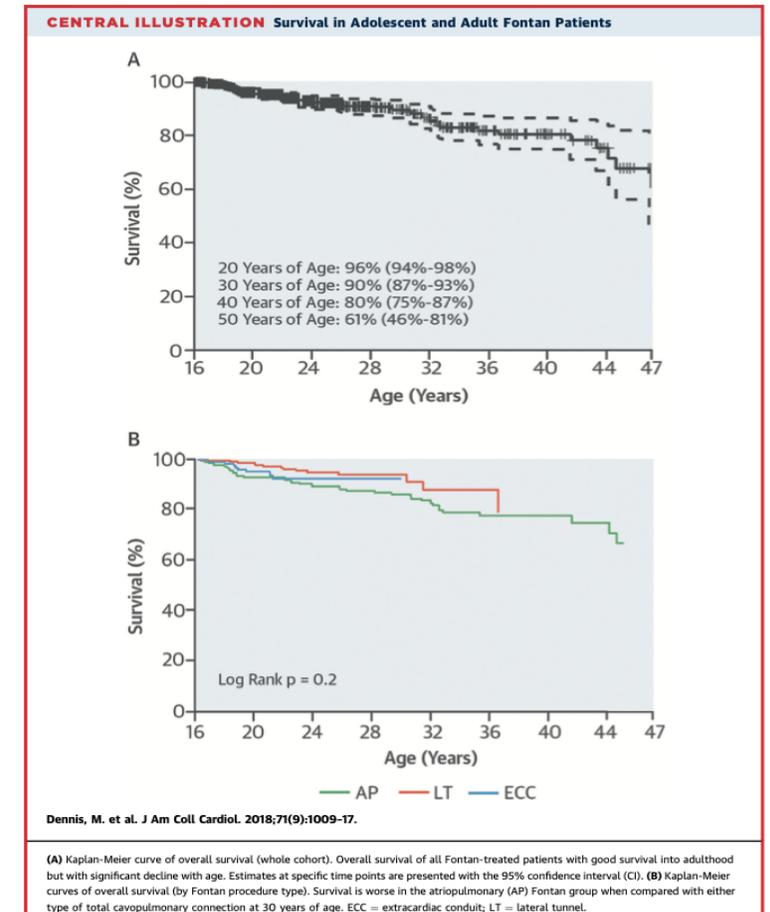
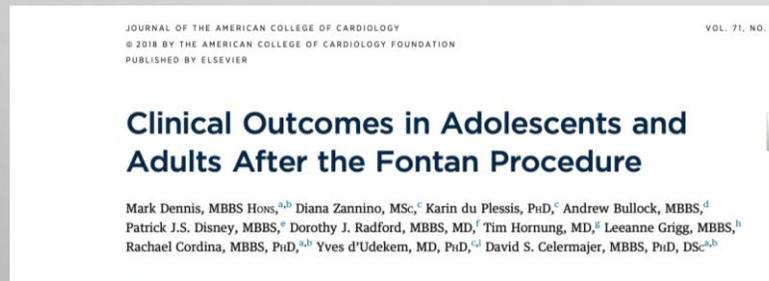
Figure 1. Kaplan Meier survival of ACHD patients undergoing heart transplantation +/- other organ transplantation as needed for (A) all ACHD patients and (B) patients palliated by the Fontan circulation.

TRAPIANTO IN FONTAN

ANZFR

201 atriopulmonari
482 cavopolmonari totali

- PRIMA FONTAN 1971
- PRIMA DCPT (MARCELLETTI) 1990
- SOPRAVVIVENZA AI 30 AA 90%, AI 40 AA: 80%
- IL 60% HA AVUTO UN EVENTO GRAVE ENTRO I 40 ANNI
- E QUINDI TRAPIANTO ????



(8%) had a pulmonary embolism. Only 1 patient had multiple thromboembolic events. The median age at reintervention was 8 years (range 1.7 to 15.9 years), and 51 (7%) patients required a second reintervention (median age 9.8 years [range 3.1 to 15.7 years]). PLE occurred in 11 patients (2%) at a median age of 12 years (range 3.4 to 15.3 years).

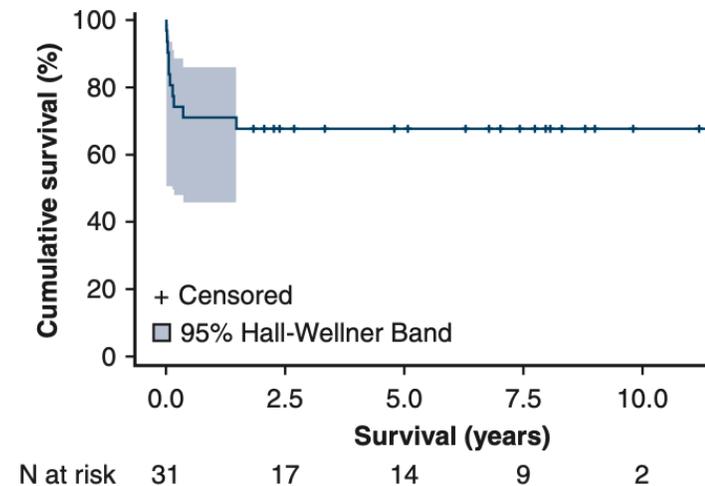
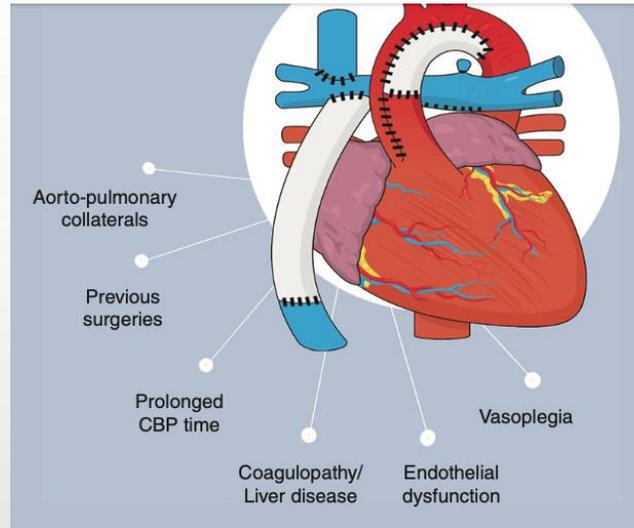
TRAPIANTO IN FONTAN

CONGENITAL: FONTAN

Improving outcomes for transplantation in failing Fontan—what is the next target?

Check for updates

Barbara Cardoso, MD,^a Andras Kelecsenyi, MD,^b Jonathan Smith, MB ChB, MRCP (UK), FRCA,^{b,c} Katrijn Jansen, MD,^{a,c} Fabrizio De Rita, MD,^{a,c,d} Mohamed Samy Nassar, MD, FRCS,^{a,c,d,e} and Louise Coats, MBBS, PhD^{a,c}

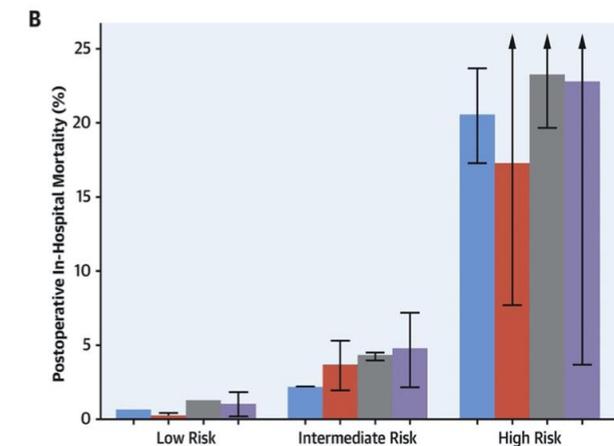
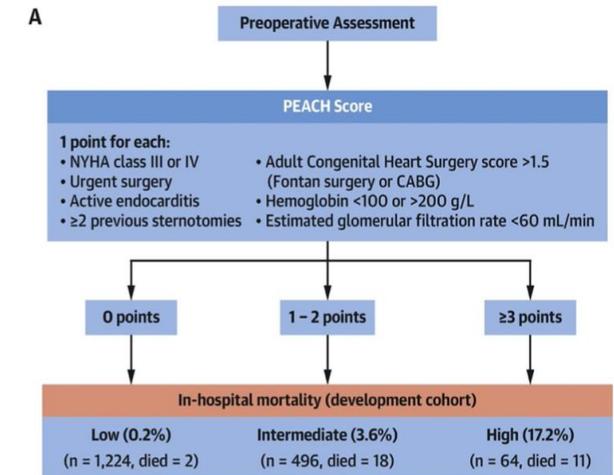


- Sopravvivenza a 30 gg, 1 e 5aa : 81%, 71%, e 67%.
- Prima causa di morte: sanguinamento intraoperatorio
- 77.4% danno renale postoperatorio CHE NECESSITA TRAPIANTO

SCORES DI RISCHIO

- ACAP
- GUCH
- ACHS
- EUROSCORE I & II
- PEACH

CENTRAL ILLUSTRATION: The PEACH Score: Risk Score Calculation, Predicted and Observed In-Hospital Mortality



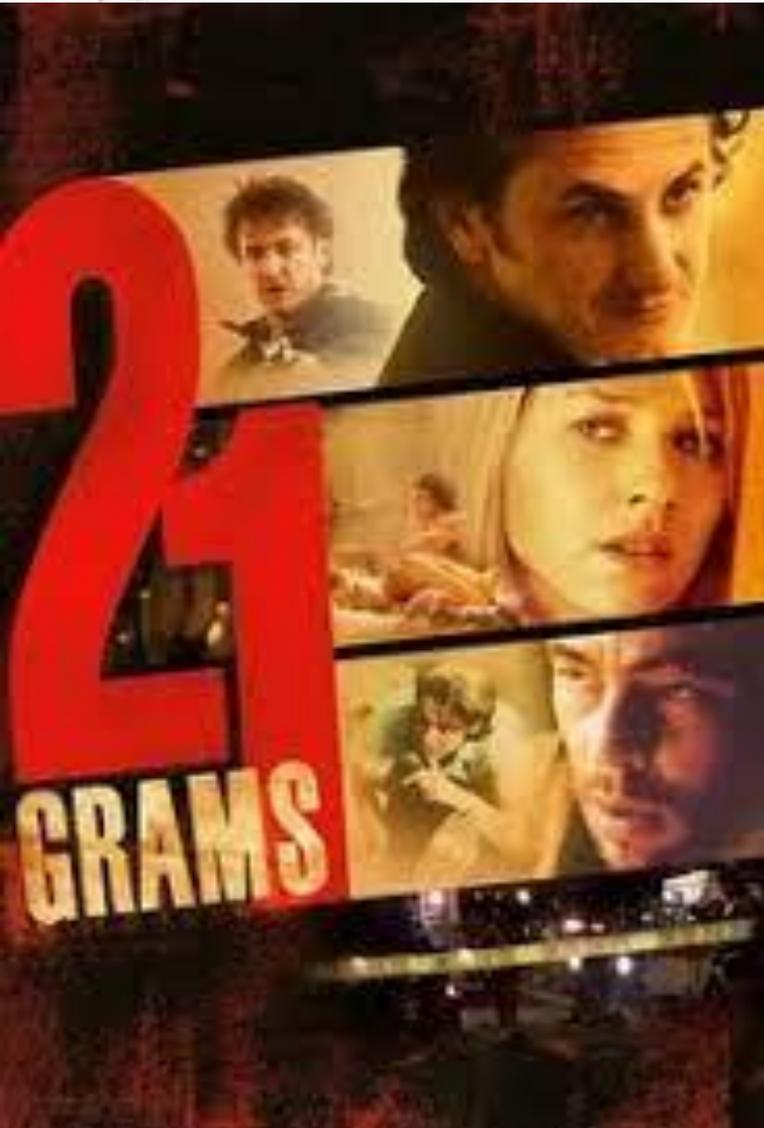
Expert Review
Mortality in Adult Congenital Heart Disease: Analysis of Outcomes and Risk Stratification

Table 3
A Brief Comparison of Major Perioperative Risk Models in Adult Congenital Heart Disease

Score System	Score Basis	Advantages	Disadvantages
PEACH Score	Patient- and procedure-specific factors specifically validated factors related to patients with ACHD and procedures in an ACHD cohort	Specifically validated in ACHD population, easy to calculate, validated against an external cohort	Recently developed, not yet validated by other groups, excludes some likely important factors due to lack of data, limited number of events in cohort
EuroSCORE I & II	Patient-specific factors based on an adult cardiac surgery cohort	Based on large dataset	The design of the score specifically excluded patients with ACHD. Most procedures used to create the dataset are not common for the ACHD population
ACHS Score	Procedure-specific risk score	Simple to calculate	Ignores patient-specific risk factors
GUCH Score	Super-score of other pediatric CHD risk models		Not yet externally validated. Complicated to calculate
ACAP Score	Anatomy and physiology of the patient	Outperforms ACHS score, simple to calculate	Ignores other patient-specific risk and procedure-specific risk factors. Has yet to be externally validated

Abbreviations: ACHD, Adult Congenital Heart disease; ACHS, Adult Congenital Heart Surgery; GUCH, Grown-Ups with Congenital Heart disease; PEACH, Perioperative Adult Congenital Heart Disease; EuroSCORE, European System for Cardiac Operative Risk Evaluation.

IL PESO DELL'ANIMA



Where Adults with Congenital Heart Disease Die: Insights from the CDC-WONDER Database



ESC

European Society
of Cardiology

European Heart Journal (2022) 00, 1–10
<https://doi.org/10.1093/eurheartj/ehac484>

FASTTRACK CLINICAL RESEARCH

Congenital heart disease

Last year of life of adults with congenital heart diseases: causes of death and patterns of care

Liesbet Van Bulck ^{1,2}, Eva Goossens ^{1,3}, Lucas Morin ^{4,5}, Koen Luyckx ^{6,7},
Fouke Ombelet ^{1,8,9}, Ruben Willems ¹⁰, Werner Budts ^{11,12},
Katya De Groote ¹³, Julie De Backer ¹⁴, Lieven Annemans ¹⁰,
Stéphane Maniette ¹⁵, Michèle de Hosson ¹⁴, Arianne Marelli¹⁶,



ESC

European Society
of Cardiology

European Heart Journal (2022) 00, 1–3
<https://doi.org/10.1093/eurheartj/ehac492>

EDITORIAL .CODAC consortium

A view from the end: what the last year of life can teach us about palliative care on the adult congenital heart disease journey

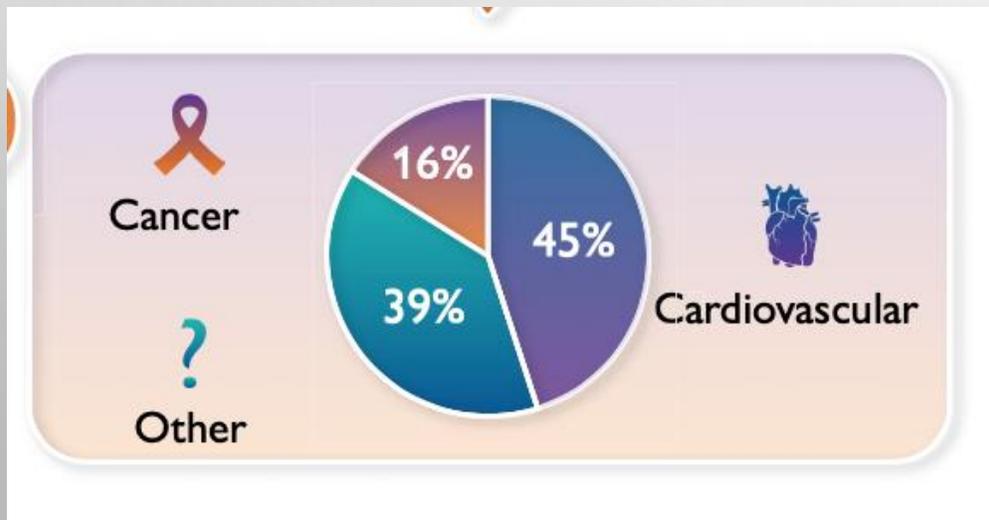
Jill M. Steiner ^{1*} and James N. Kirkpatrick²

¹Division of Cardiology, Department of Medicine & Cambia Palliative Care Center of Excellence, University of Washington, 1959 NE Pacific St., HSB C502, Box 356422 Seattle, WA 98195, USA; and ²Division of Cardiology, Department of Medicine & Department of Bioethics and Humanities, University of Washington, 1959 NE Pacific St., Seattle, WA 98195, USA

MORTALITÀ

DOVE SONO GLI SPECIALISTI ACHD?

- GLI ACHD MUOIONO IN OSPEDALE (64% VS 46% -POPOLAZIONE GENERALE)
- 390 PTS: 45% CAUSA CARDIOVASCOLARE [EUR J CARDIOL22 \(BELGA\)](#)
- ULTIMO ANNO DI VITA OSPEDALIZZAZIONE: 87%, 78% DEA, 19% ICU, 17% CURE PALLIATIVE DI CUI IL 4% DEI DECESSI PER CAUSA CARDIOVASCOLARE



	% of patients who used service ≥ 1		Cardiovascular	Cancer
	Last year	Last month		
GP visit	97%	70%	↔	
Hospitalization	87%	70%		↑
ED visit	78%	51%	↔	
CHD physician encounter	28%	11%	↑	
Cardiovascular procedure	25%	11%	↑	
ICU admission	19%	15%	↔	
Specialist palliative care	17%	13%		↑

ESC European Society of Cardiology
European Heart Journal (2022) 00, 1–10
<https://doi.org/10.1093/eurheartj/ehac484>

FASTTRACK CLINICAL RESEARCH
Congenital heart disease

Last year of life of adults with congenital heart diseases: causes of death and patterns of care

Liesbet Van Bulck ^{1,2}, Eva Goossens ^{1,3}, Lucas Morin ^{4,5}, Koen Luyckx ^{6,7}, Fouke Ombelet ^{1,8,9}, Ruben Willems ¹⁰, Werner Budts ^{11,12}, Katya De Groot ¹³, Julie De Backer ¹⁴, Lieven Annemans ¹⁰, Stéphane Moniotte ¹⁵, Michèle de Hosson ¹⁴, Arianne Marelli ¹⁶, Philip Moons ^{1,17,18*}, and BELCODAC consortium

LA PARADOSSALE FORZA DELLA FRAGILITÀ



ACHD

PIU' UMANI DEGLI UMANI?

PUBBLICAZIONE ESC SU COVID INCLUDONO PARTI SCRITTE DAI PAZIENTI

Graphical Abstract

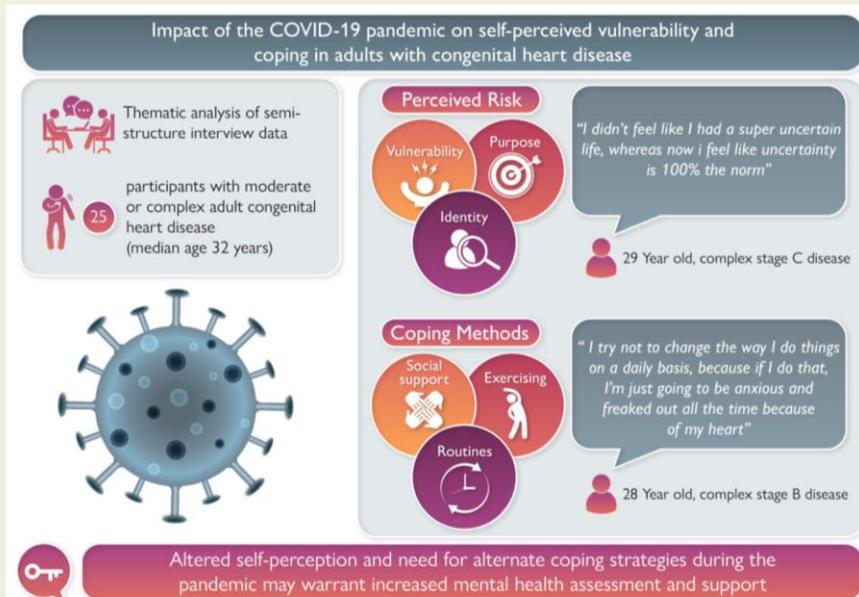


Table 2 Key participant quotations

COVID affected participants' sense of vulnerability

[Previously], when I thought of myself as a vulnerable population, I thought of it more as because I was pregnant not, 'Oh, and you have this heart condition.' Which is strange, ... a double whammy. When I had a heart appointment during my pregnancy, one of the cardiologists said, 'Yeah, it's good that you're being extra cautious because you are a vulnerable population, because of your heart condition.' I was like, 'Oh, yeah.'—32 years old, moderate Stage C disease

Before COVID, I was just like, 'Oh, I have this job, it's just a job to me, but I'm getting by and I'm reasonably happy.' Then COVID happened, and then we all kind of had to... confront the idea that we could get a terrible disease and die. I think, when I was kid, having to go through sort of a realization that I could die, it had some positive effect. It had some self-actualization in, it granted me some immediacy. I would say that that's what that did for me now, is, I feel like I do actually need to do something.—25 years old, moderate Stage B disease

My family was told to prepare for this, prepare for that. 'He may not make it to this age and that age'. So I've been preparing the whole time through my life because my mother shared this with me... And I don't know if that helped or hurt me, but I'm not going to sit here and pass my life in fear. I'm aware of the COVID... but I try not to live my life thinking, 'Oh, well I got to be super cautious in this and that'. So I try to live my life, and go through my daily routine, without thinking of it too much.—44 years old, moderate Stage D disease

I am one of the numbers of person that would totally die if I got it. I respect that deeply. And I've changed my life a little bit for that, but no more than anybody that was already expecting to die tomorrow... I laugh with my friends, like, 'Wouldn't it be weird if I lived all this extra time, and then this stupid thing got me?' It's a waste of a defeat of all these other, bigger diseases in my life I've conquered.—37 years old, complex Stage C disease

It's funny because I never saw my heart condition as ... It never caused that much unsettling uncertainty. There was always uncertainty, but it was enough able to be put in the back of my head. I didn't feel like I had a super uncertain life whereas now I feel like uncertainty is 100% the norm.—29 years old, complex Stage C disease

COVID impacted use of existing coping strategies

ACHD

PIU' UMANI DEGLI UMANI?

- LINE GUIDA PH 2022: INCLUDONO ASSOCIAZIONI DI PAZIENTI

New standards for PH centres have been presented and, for the first time, patient representatives were actively involved in developing these guidelines.



EUROPEAN RESPIRATORY JOURNAL
ERS OFFICIAL DOCUMENTS
M. HUMBERT ET AL.

2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

Developed by the task force for the diagnosis and treatment of pulmonary hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)

Endorsed by the International Society for Heart and Lung Transplantation (ISHLT) and the European Reference Network on rare respiratory diseases (ERN-LUNG)

PAH : 4-6% di ACHD

L'uomo non è che un fucello,
il più debole della natura,
ma è un fucello che pensa.
Blaise Pascal

Aforismario

ACHD

PIU' UMANI DEGLI UMANI?

- **LE CC SONO M CRONICHE: TUTTE**
- PERSONE A CUI E STATO DATO IL BENESSERE FISICO, LA POSSIBILITA DI ESSERE PARTE ATTIVA DELLA SOCIETA, SPESSO LA VITA
- CONTINUANO AD AVER BISOGNO DI REINTERVENTI E CURE
- DISAGIO PSICOLOGICO
- DIFFICOLTA' NEL MONDO DEL LAVORO



IL DIA E ANCORA UNA CHD SEMPLICE?

«QUESTO PAZIENTE HA UN DIA, LO POSSO GESTIRE DA SOLO»

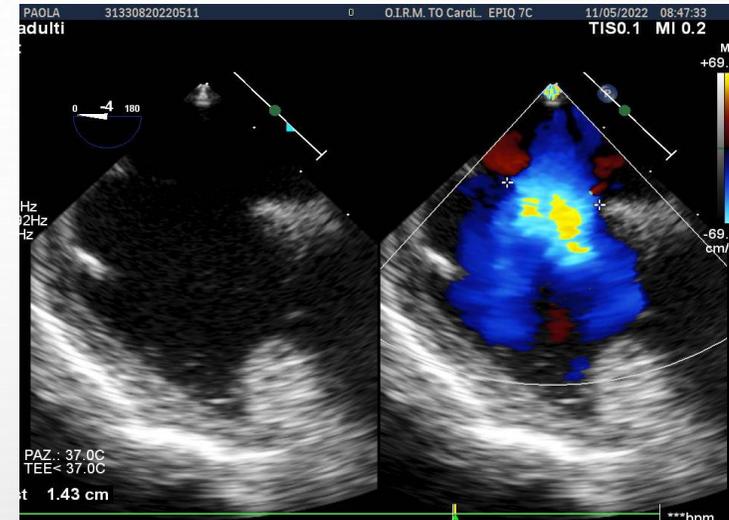
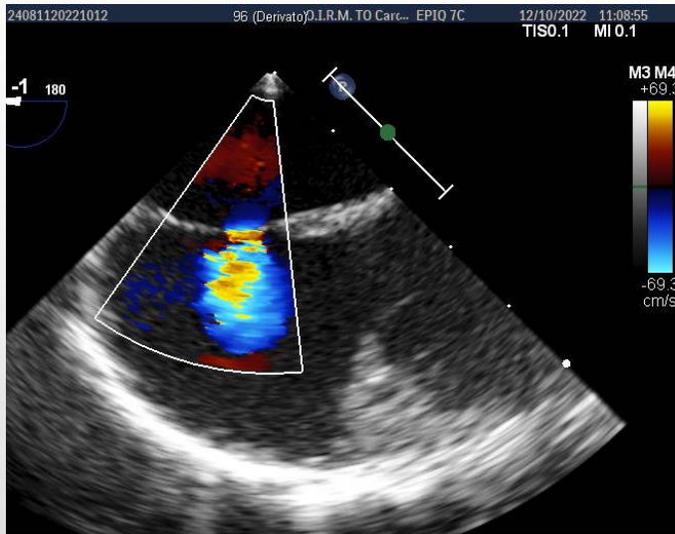
Rischio aumentato di

- **FA e stroke** (Heart2015, Front Cardiovasc Med 2022)
- **polmonite** (Am J Cardiol 2014)
- **tumore** (Eur Heart J 2018)
- **problemi psichiatrici** (Congen Heart Dis 2019, Am J Cardiol 2020)



IL DIA E ANCORA UNA CHD SEMPLICE?

DIA PICCOLO E GRANDE

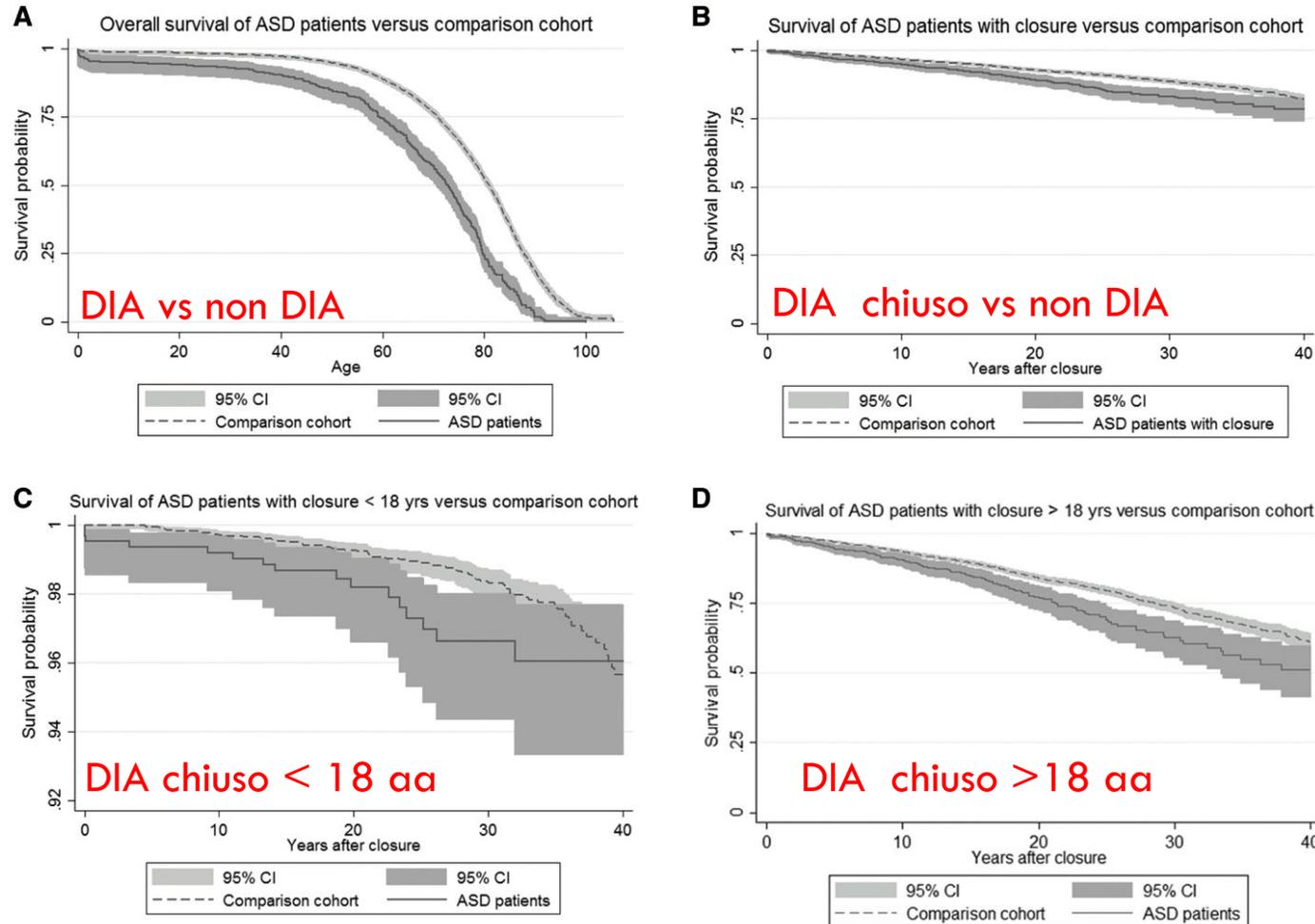


I pz con DIA piccolo in storia naturale:
stesse problematiche dei pz con DIA ampio

Danish national registry

Int J cardiol 2019

MORTALITA' DEI PAZIENTI CON DIA



Mortalita' piu' alta, sia che il DIA sia stato chiuso o meno
Eur Heart J. 2018

Figure 1 (A) Survival in years in atrial septal defect (ASD) patients without closure compared with the comparison cohort with age as an underlying time scale. (B) Survival in ASD patients after closure compared with the comparison cohort with years after closure as an underlying time scale. (C) Survival in patients with closure before the age of 18 compared with the comparison cohorts with years after closure as an underlying timescale.

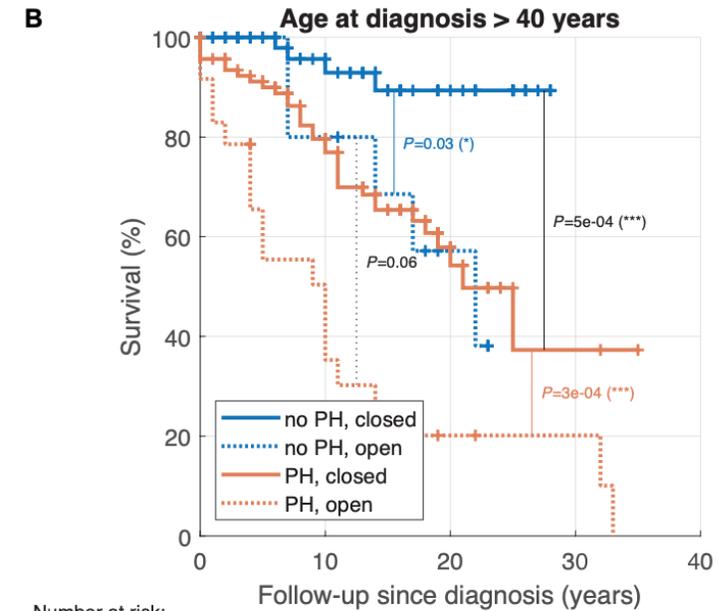
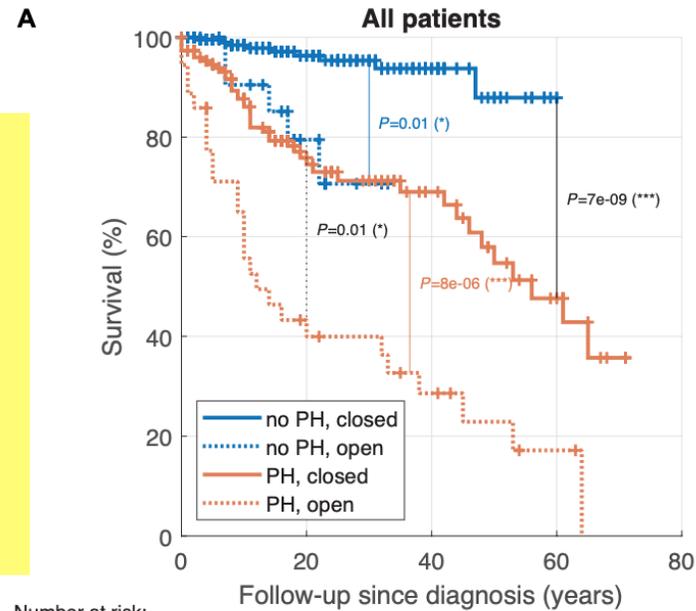
CHIUSURA VS STORIA NATURALE

Rubáčková Popelová et al.

Survival in Atrial Septal Defect

TUTTI

DIAGNOSI TARDIVA



Number at risk:

	0	20	40	60	80
no PH, closed	217	166	110	62	33
no PH, open	24	19	9	2	0
PH, closed	150	109	56	38	27
PH, open	36	21	13	11	7

Number at risk:

	0	10	20	30	40
no PH, closed	67	35	13	0	0
no PH, open	11	8	3	0	0
PH, closed	92	59	16	2	0
PH, open	24	10	3	2	0

FIGURE 1 | Kaplan-Meier survival analysis of atrial septal defect (ASD) patients stratified by pulmonary hypertension (PH) and ASD closure; all patients (model A). **(A)** Patients of all ages. **(B)** Patients diagnosed at the age above 40. ASD, atrial septal defect; MR, moderate or severe mitral regurgitation; NYHA, New York Heart Association class; PH, pulmonary hypertension.

Mortalita' e incidenza di CHF piu' alta nei pz con

- DIA aperto
- Diagnosi tardiva
- PH

BLU: NO PH

ROSSO: PH

ARRIVA IL METAVERSO

Virtual reality and augmented reality: imaging or imagining?

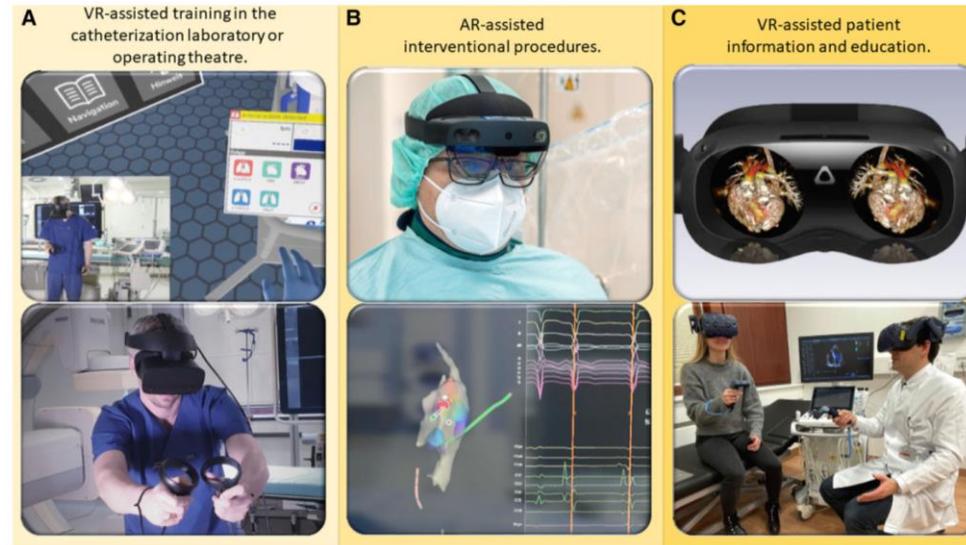
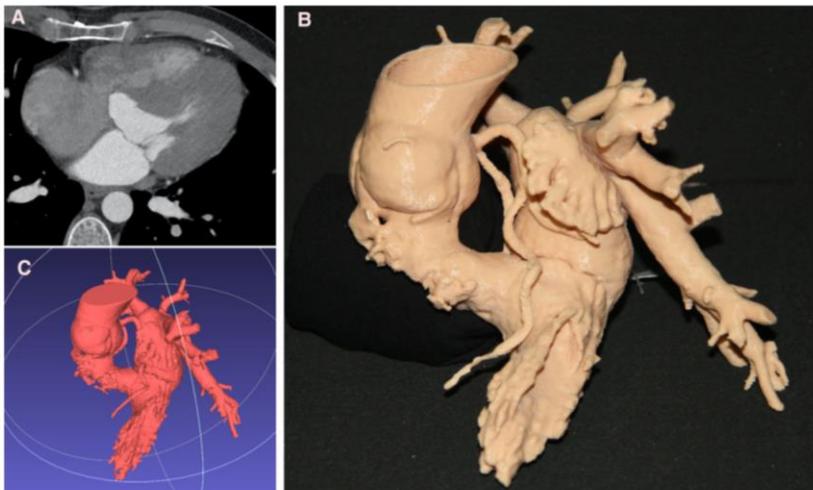
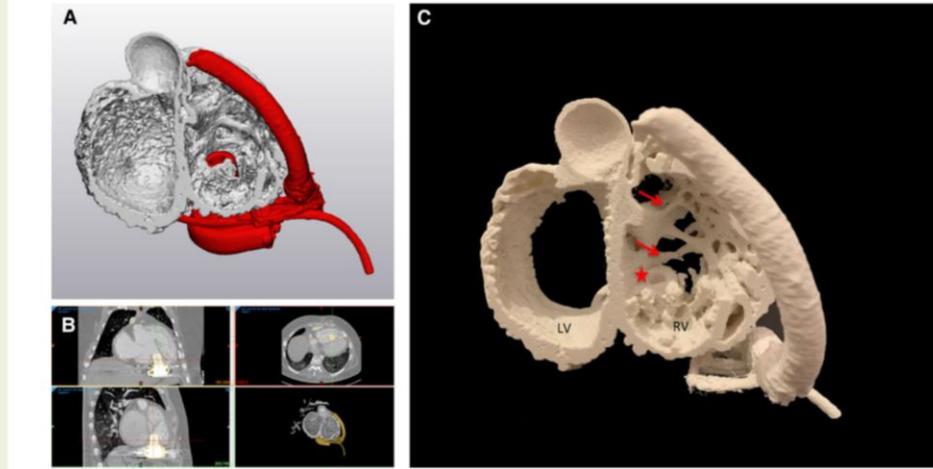


Figure 7 (A) Virtual reality-assisted training the catheterization laboratory or operating theatre. (B) Augmented reality-assisted interventional procedures; (C) virtual reality-assisted patient information and education.



Three-dimensional printing, holograms, computational modelling, and artificial intelligence for adult congenital heart disease care: an exciting future