Congenital Heart Disease: Not Just For Kids

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No disclosures

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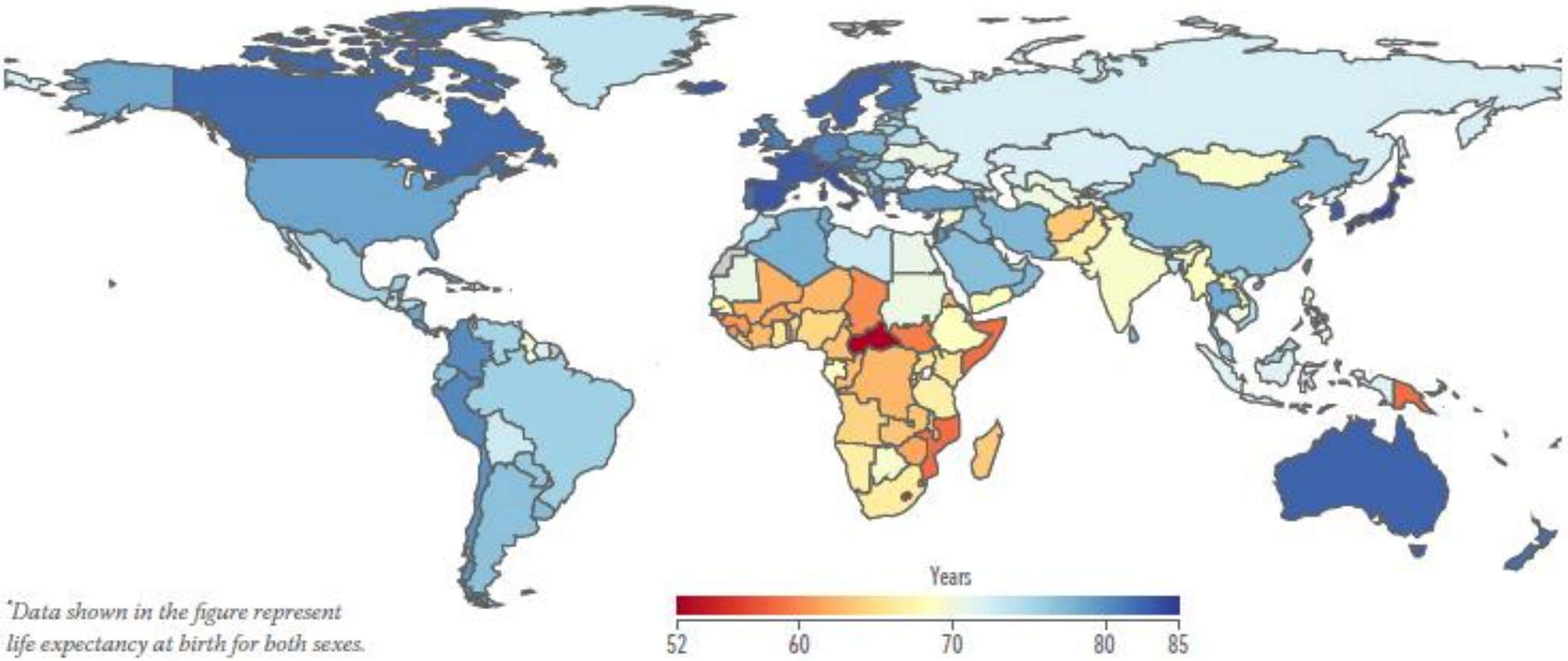
Scope of the specialty

- It includes all adults born with congenital heart disease and they can be:
  - Uncorrected,
    - ASD, PDA, TOF, UVH with balanced circulation
  - Palliated:
    - TOF who had only a BT shunt
    - VSD who had a PA band
    - Univentricular paliation
  - Corrected:
    - residual lesions, such as , VSD, PS etc…
    - Development of late failure, such as ventricular failure in atrial switch,
    - Late AI in TOF repair
    - Coronary obstruction after ASO

Patients Are Living Longer: Life Expectancy



*Data shown in the figure represent life expectancy at birth for both sexes.

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Data From CBD 2017

Because of major developments in cardiac surgery and interventional cardiology, worldwide > 1 million people with significant congenital heart disease reached adulthood over the last decades

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J THORAC CARDIOVASC SURG 1989;97:1-9

Honored Guest's Address

Do we really correct congenital heart defects?

J. Stark, FRCS, FACS, FACC, *London, England*

Corrective

Normal function achieved and maintained

Normal health during long term follow up

Further treatment is not necessary

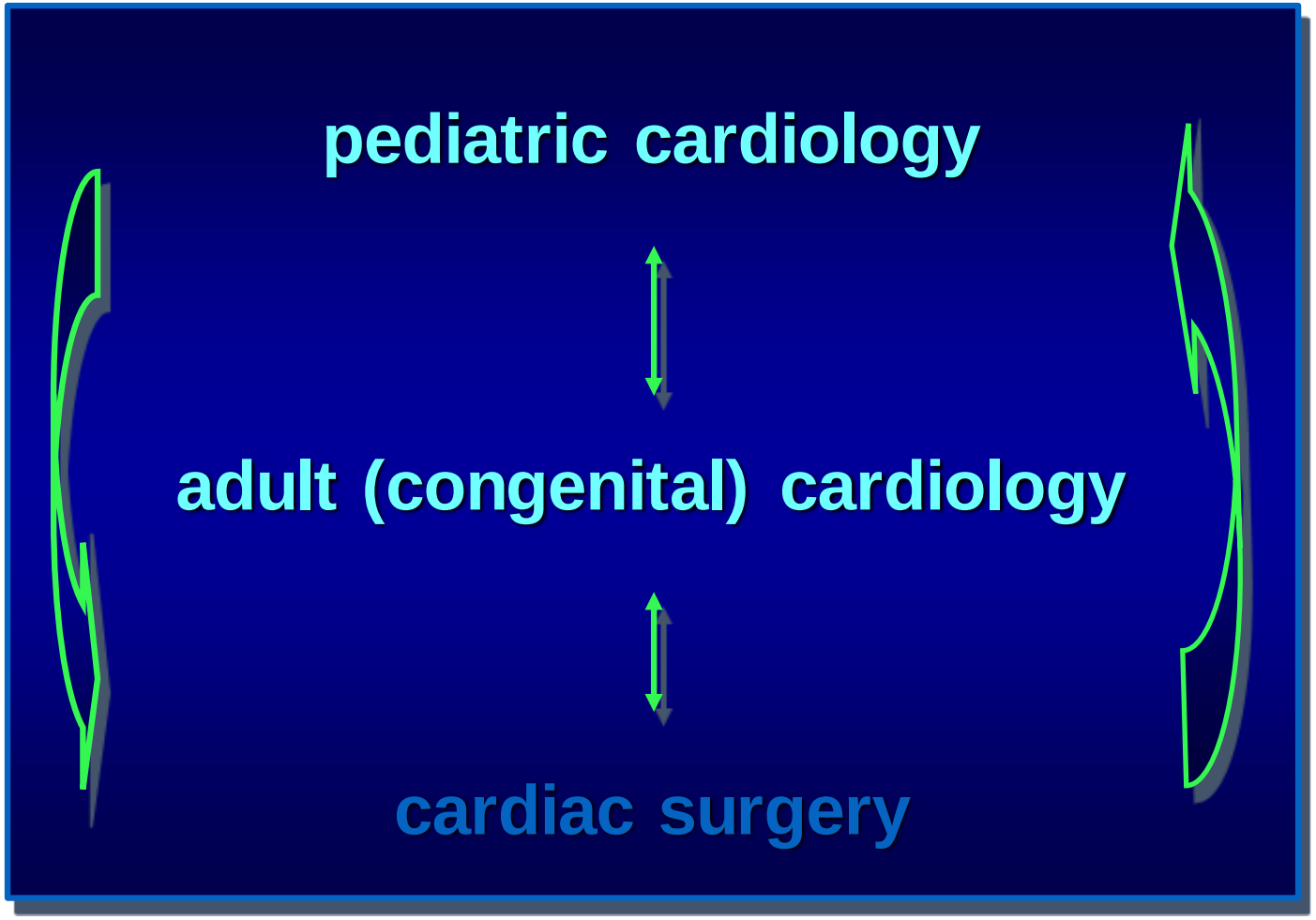
Congenital Heart Disease in Adults: Main Problems

- ***Proper*** **care**
- **after surgical/interventional management of CHD**
- **demands specific knowledge/expertise on:**
  - pathology and physiology
  - clinical symptoms
  - natural history
  - residual lesions, sequelae and complications

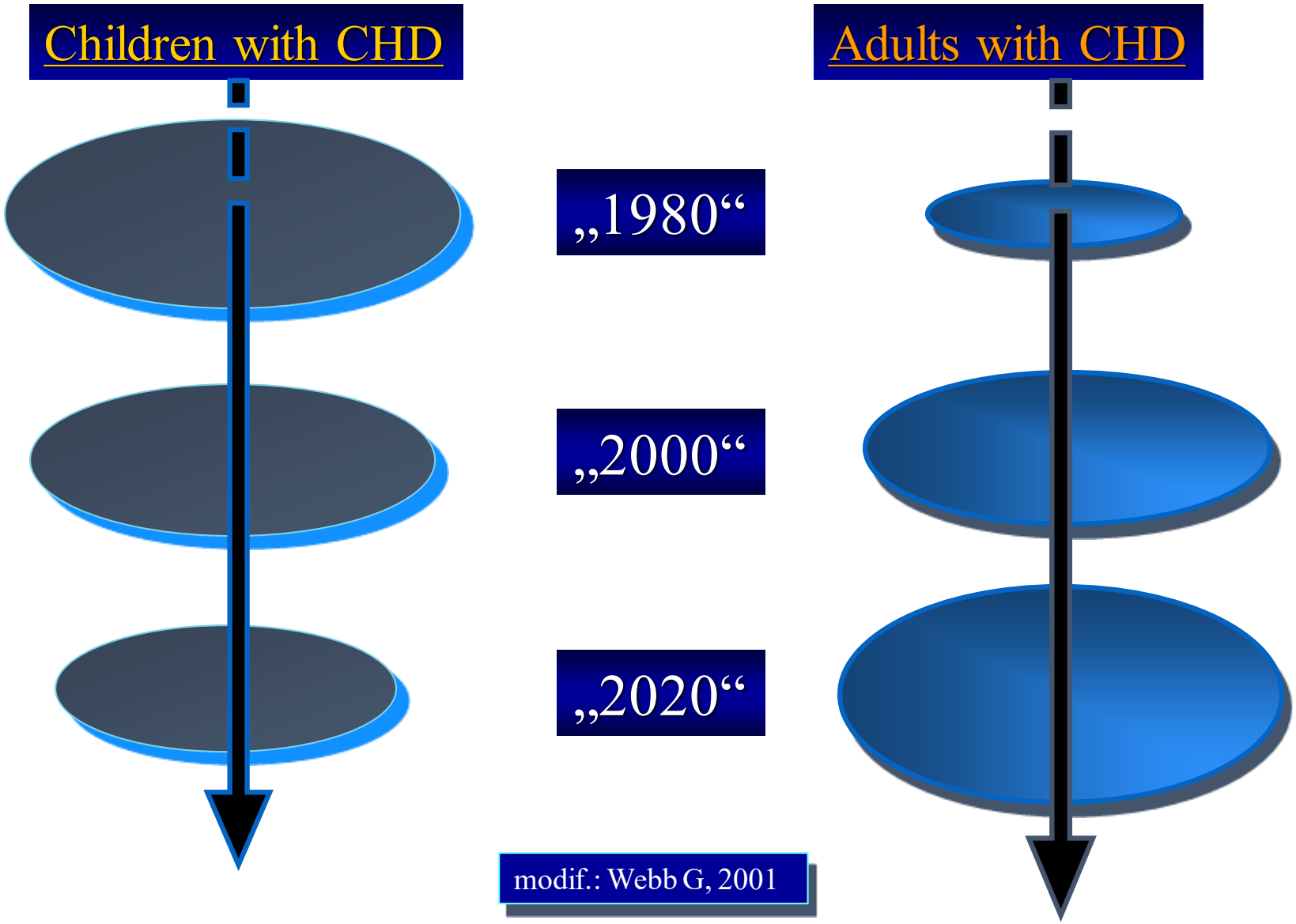
Congenital Heart Disease in Adults

Worldwide problem:

- **Lack of specialists with adequate qualification to manage adults with congenital heart disease**


pediatric cardiology
adult (congenital) cardiology
cardiac surgery

The changing profile of congenital heart disease



Adult Congenital Heart Disease: A Growing Problem

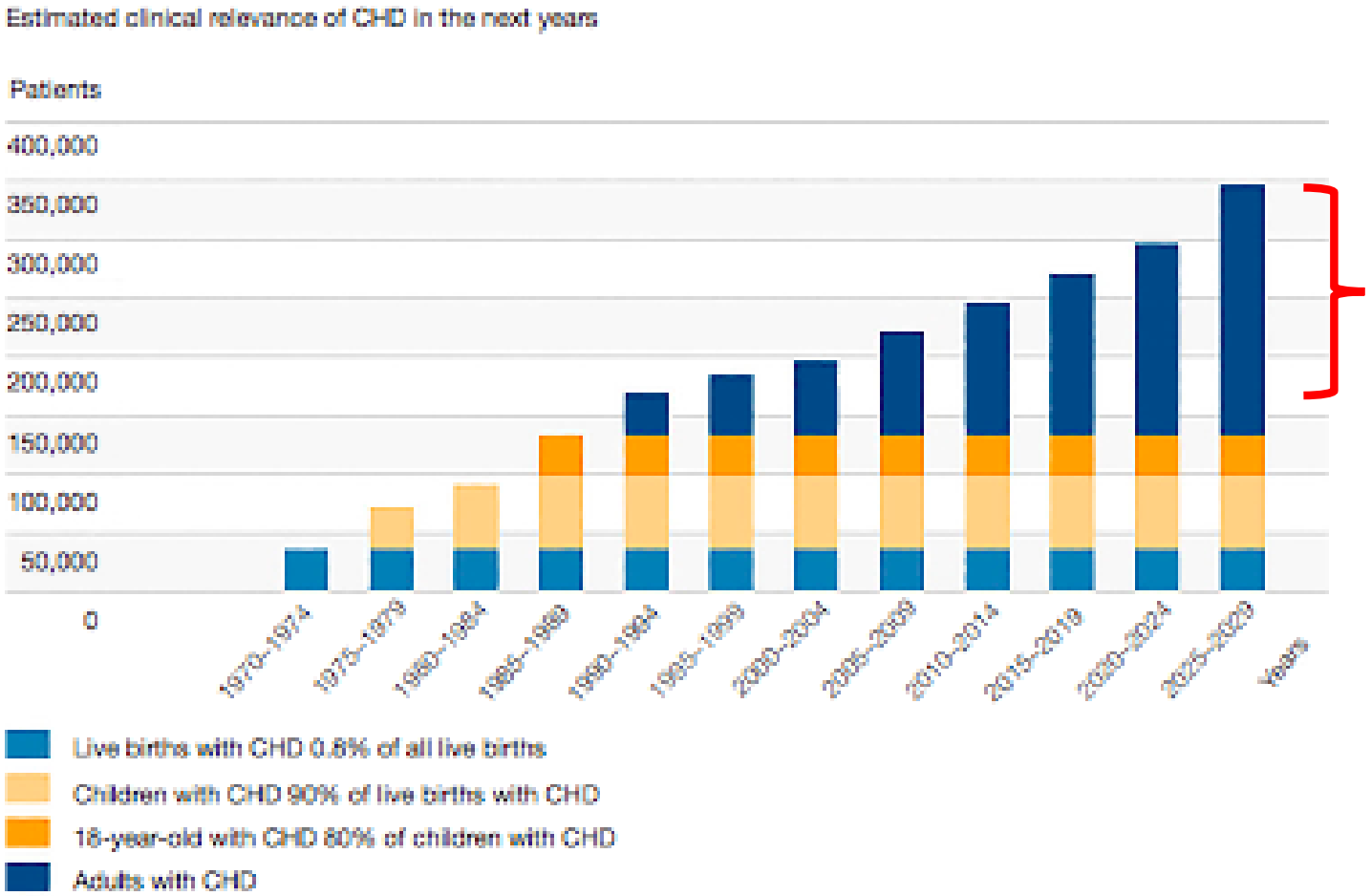
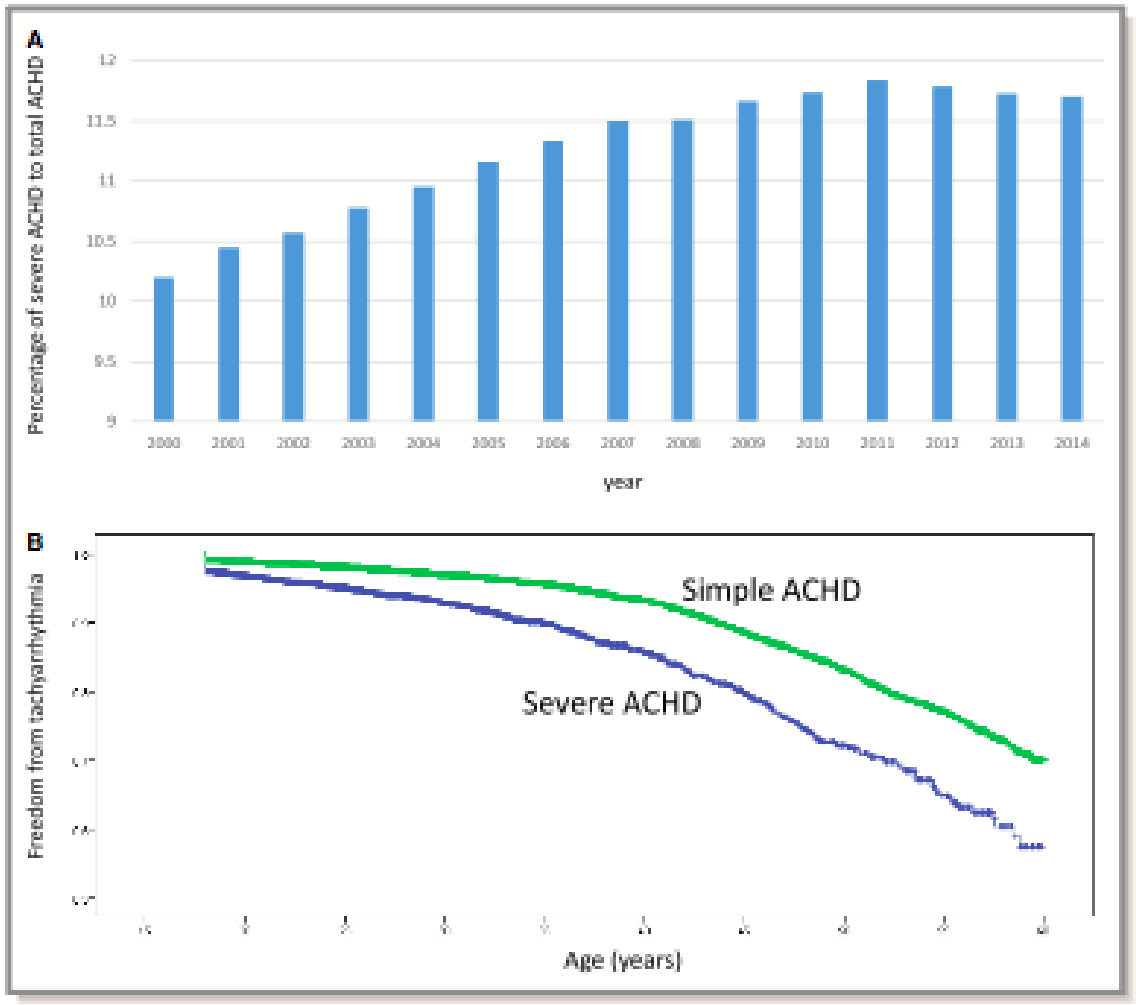


Image source: Competence Network for Congenital Heart Defects

Estimated ACHD population in 2025

Adult Congenital Heart Disease: A Growing Problem



ACHD patient population is growing…
and patients with severe ACHD are an increasing

Burden of disease (arrythmias, re-operations, expected comorbidities increase with age

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Unadjusted mortality rates for ACHD according to STS-CHSD coded procedure
Red circles: adults
Blue circles: Includes pediatric + adults

Note: Wide variation in mortality rates for adults compared to children

Estimating Mortality Risk for Adult Congenital Heart Surgery: An Analysis of The Society of Thoracic Surgeons Congenital Heart Surgery Database

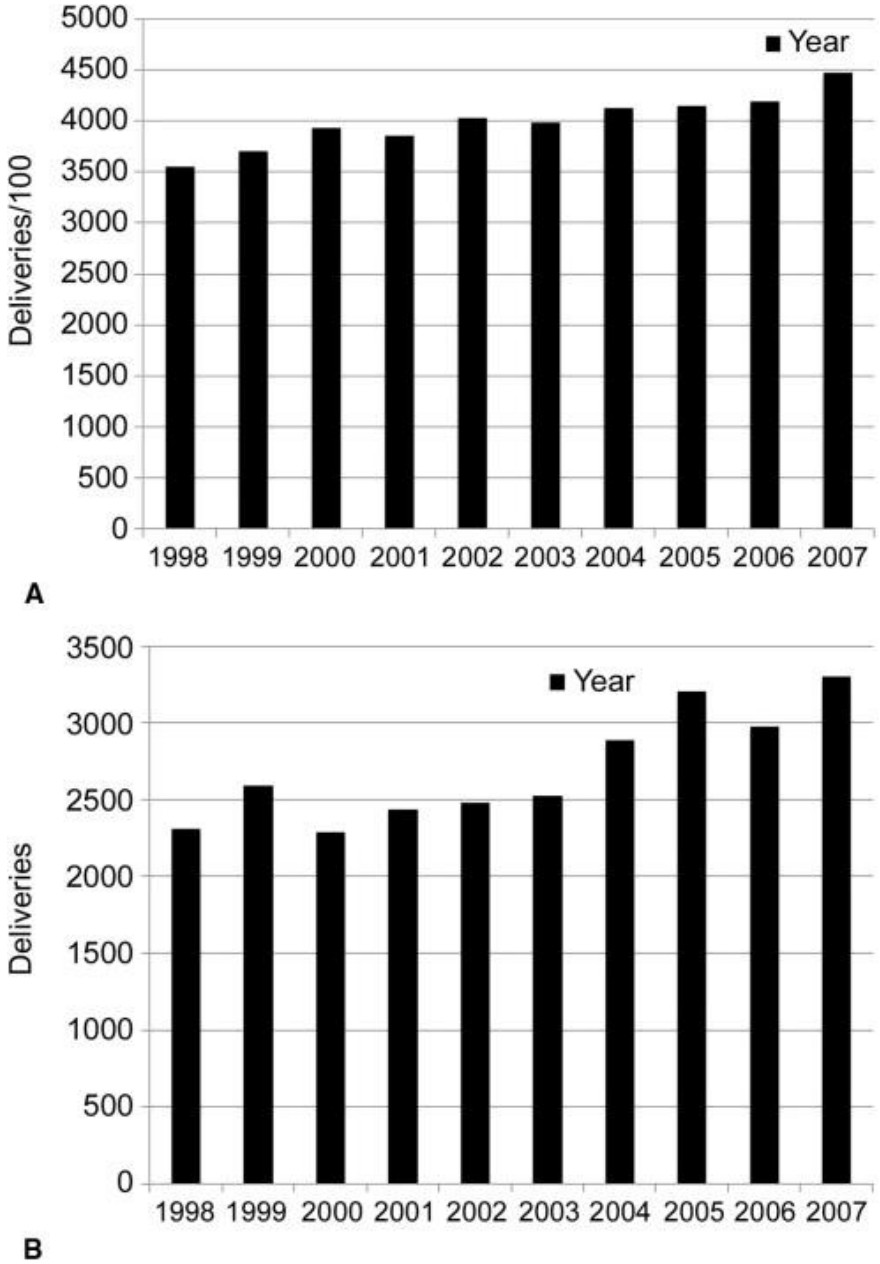
Stephanie M. Fuller, MD, MS, Xia He, MS, Jeffrey P. Jacobs, MD, Sara K. Pasquali, MD, MHS, J. William Gaynor, MD, Christopher E. Mascio, MD, Kevin D. Hill, MD, Marshall L. Jacobs, MD, Yuli Y. Kim, MD

The Annals of Thoracic Surgery
Volume 100, Issue 5, Pages 1728-1736 (November 2015)
DOI: 10.1016/j.athoracsur.2015.07.002

Growing Maternal Population with ACHD- Deliveries Increased by 26% over 9-year period

Caring for peripartum ACHD introduces additional complexities into the equation

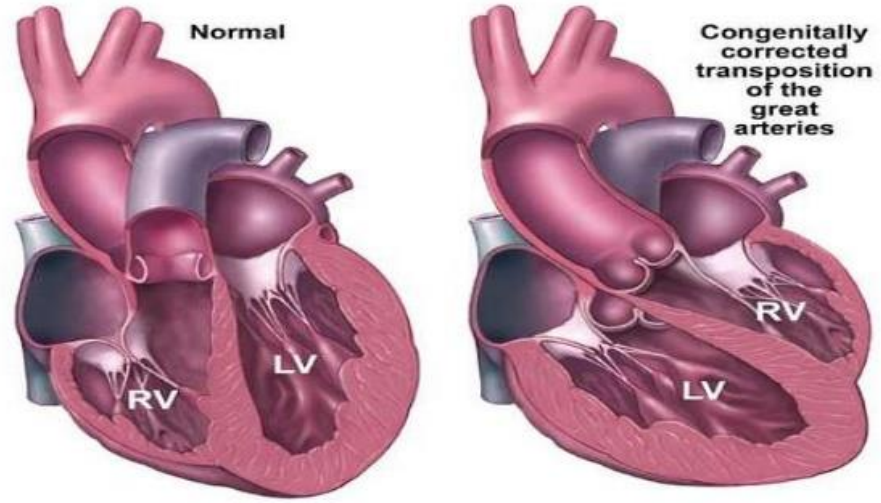
How can we leverage care delivery to provide highly specialized care to varied population with highly Individualized needs?

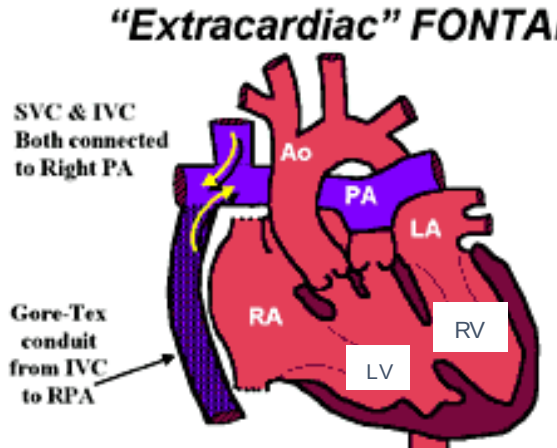


From Karamlou et al. Ann Thorac Surg 2011

A Clinical Question

- If you were given a choice between:
- ccTGA
- Fontan





Univentricular vs Biventricular Pathway

- Univentricular pathway provides satisfactory mid-term palliation but the long-term results are suboptimal.
- Long-standing systemic venous hypertension leads to morbidities like PLE, plastic bronchitis and chronic hepatic failure.
- Fontan Failure is a multi-system disease and poses difficult management issues.
- Two-ventricle Fontan does not confer a better outlook.

Factors associated with long-term mortality after Fontan procedures: a systematic review

Tarek Alsaied,[1] Jouke P Bokma,[2,3] Mark E Engel,[4] Joey M Kuijpers,[2,3] Samuel P Hanke,[1] Liesl Zuhlke,[5] Bin Zhang,[6] Gruschen R Veldtman[7]

► Additional material is published online only. To view please visit the journal online (http://dx.doi.org/10.1136/heartjnl-2016-310108).

For numbered affiliations see end of article.

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ABSTRACT

Background Despite an ageing Fontan population, data on late outcomes are still scarce. Reported outcome measures and determinants vary greatly between studies making comprehensive appraisal of mortality hazard challenging.

Methods We conducted a systematic review to

Systematic reviw. 28 studies, 6707 patients with an average follow-up time of 8.23±5.42 years and 1000 deaths. The late mortality in Fontan population was multimodal.

an average follow-up time of 8.23±5.42 years was identified. There were 1000 deaths. Causes of late death were reported in 697 cases. The five most common causes were heart/Fontan failure (22%), arrhythmia (16%), respiratory failure (15%), renal disease (12%)

circulation or at least 20 mortalities and/or transplants to maintain statistical power. We excluded studies that focused on short-term mortality within the 1st year of the Fontan procedure. We searched PubMed and the Cochrane Library for studies published in English with no restriction on date of publication. Search strategies for each database are

than one study included the same patients we excluded the studies with overlapping patients and we included only the most recent study that included these patients in the final analysis.

The factors associated with long-term mortality

Surgical and Catheter-Based Reinterventions Are Common in Long-Term Survivors of the Fontan Operation

Tacy E. Downing, MD; Kiona Y. Allen, MD; David J. Goldberg, MD; Lindsay S. Rogers, MD; Chitra Ravishankar, MD; Jack Rychik, MD; Stephanie Fuller, MD; Lisa M. Montenegro, MD; James M. Steven, MD; Matthew J. Gillespie, MD; Jonathan J. Rome, MD; Thomas L. Spray, MD; Susan C. Nicolson, MD; J. William Gaynor, MD; Andrew C. Glatz, MD, MSCE

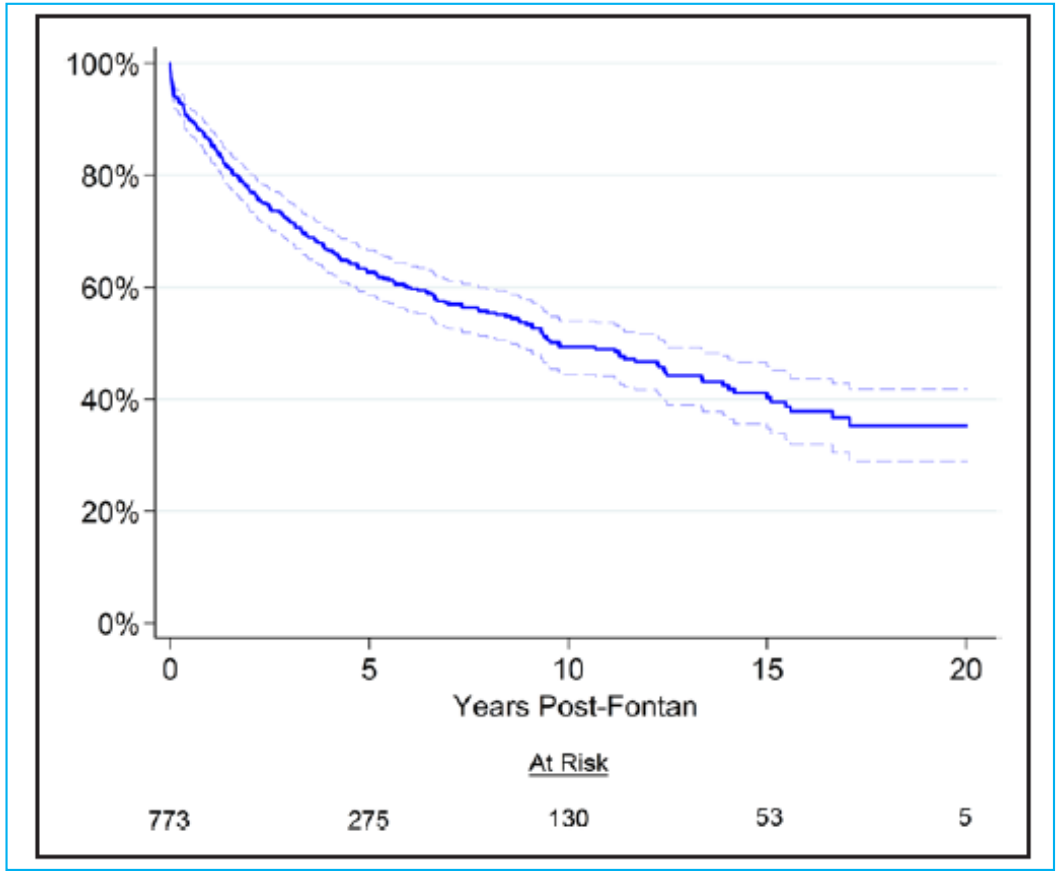
Background—There are limited follow-up studies examining surgical and catheter-based reinterventions in long-term survivors of the Fontan operation.

Methods and Results—All 773 patients who underwent Fontan at our institution between 1992 and 2009 were retrospectively reviewed. Current information regarding post-Fontan intervention was available for 70%. By 20 years after Fontan, 65% of patients had experienced either surgical or transcatheter intervention. The median time to first reintervention was 9.8 years. Freedom from reoperation was 69% at 15 years and 63% at 20 years. The most common operations were pacemaker placement and Fontan revision. Risk factors for pacemaker placement included systemic left ventricle (hazard ratio [HR], 2.2; *P*=0.006) and lateral tunnel Fontan (HR, 4.3; *P*=0.001). Freedom from interventional catheterization was 53% at

Conclusions—In this study of 773 Fontan survivors, two thirds of patients required surgical or catheter-based reintervention by 20 years. Families should be counseled that the Fontan is typically not the final stage of single-ventricle palliation.

(*Circ Cardiovasc Interv*. 2017;10:e004924. DOI: 10.1161/CIRCINTERVENTIONS.116.004924.)

Freedom from any intervention after Fontan Procedure.



Circulation Journal
Official Journal of the Japanese Circulation Society
http://www.j-circ.or.jp

REVIEW

The Adult With a Fontan: A Panacea Without a Cure?

– Review of Long-Term Complications –

Lauren Lastinger, MD; Ali N. Zaidi, MD

The univentricular heart includes a spectrum of complex cardiac defects that are managed by staged palliative surgical procedures, ultimately resulting in a Fontan procedure. Since 1971, when it was first developed, the procedure has undergone several variations. These patients require lifelong management, including a thorough knowledge of their anatomic substrate, hemodynamic status, management of rhythm and ventricular function, together with multi-organ evaluation. As these patients enter middle age, there is increasing awareness of long-term complications and mortality. This review highlights the concept behind the staged surgical palliations, the unique single ventricle physiology and the long-term complications in this complex cohort of patients. (*Circ J* 2013; **77**: 2672–2681)

Key Words: Congenital heart disease; Fontan procedure; Heart defects

Conclusions: Despite proper selection criteria, PLE, arrhythmias, thromboembolic concerns, liver dysfunction and ventricular failure lead to therapeutic and management challenges. Despite careful long-term follow-up, there remains continual but late attrition of patients

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Ann Thorac Surg. 2009 August ; 88(2): 558–564. doi:10.1016/j.athoracsur.2009.03.085.

Evaluating Failing Fontans for Heart Transplantation: Predictors of Mortality

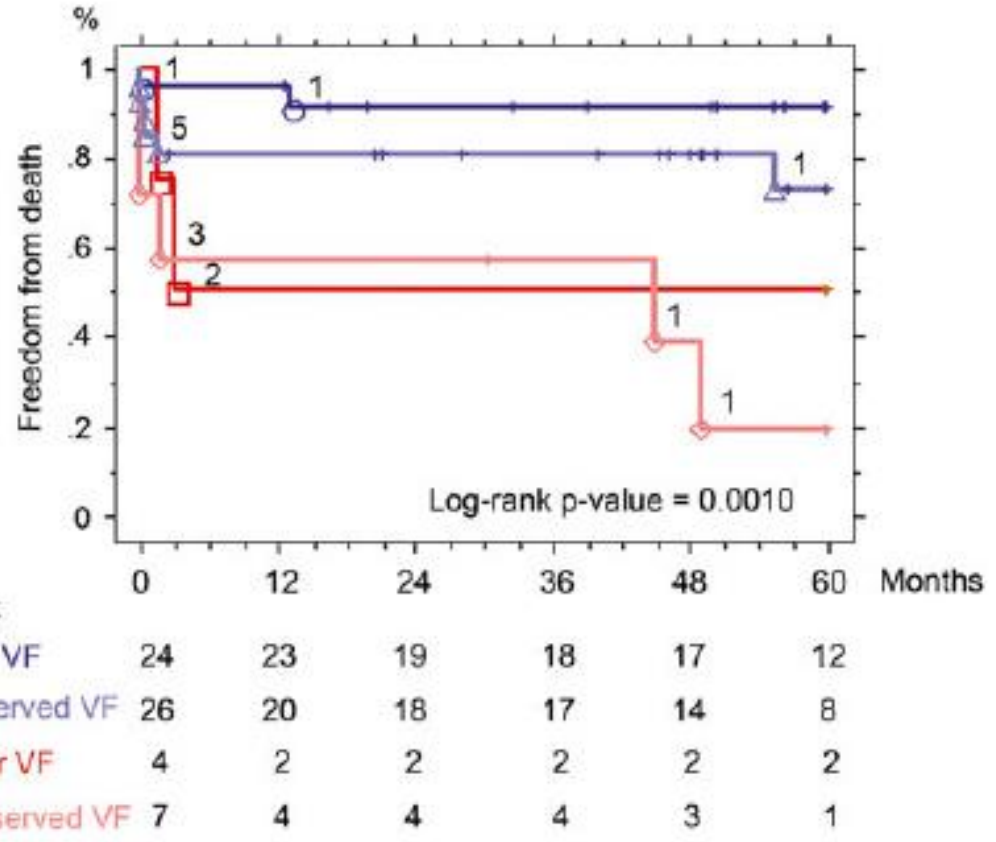
Eric R. Griffiths, M.D[1], **Aditya K Kaza, M.D**[1], **Moritz C. Wyler von Ballmoos, M.D**[1], **Hugo Loyola, B.S.**[1], **Anne Marie Valente, M.D**[2], **Elizabeth D. Blume, M.D**[2], and **Pedro del Nido, M.D**[1]

[1] Department of Cardiac Surgery, Children's Hospital Boston, Harvard Medical School Boston, MA

[2] Department of Cardiology, Children's Hospital Boston, Harvard Medical School Boston, MA

Those patients who presented with failed Fontan physiology but preserved ventricular function had greater than three fold risk of death within one year compared to those who presented with ventricular dysfunction (Actuarial survival of

Effect of timing & ventricular function on mortality after Fontan.



Patients at risk						
Late FF - poor VF	24	23	19	18	17	12
Late FF - preserved VF	26	20	18	17	14	8
Early FF - poor VF	4	2	2	2	2	2
Early FF - preserved VF	7	4	4	4	3	1

Figure 3: Kaplan–Meier 5-year freedom from death after orthotopic heart transplantation (OHT) in early and late Fontan failure (FF) stratified by pre-OHT ventricular function (VF) with the number of patients at risk.

Effect of PLE on Mortality after Fontan.

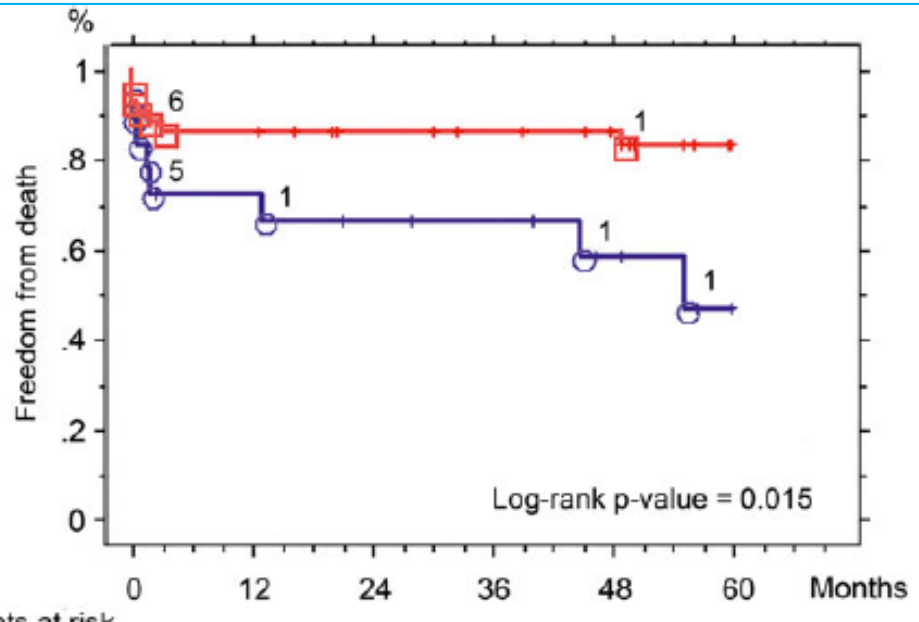


Figure 4: Kaplan–Meier 5-year freedom from death after orthotopic heart transplantation in protein-losing enteropathy (PLE) and non-PLE with the number of patients at risk.

The natural and unnatural history of the systemic right ventricle in adult survivors

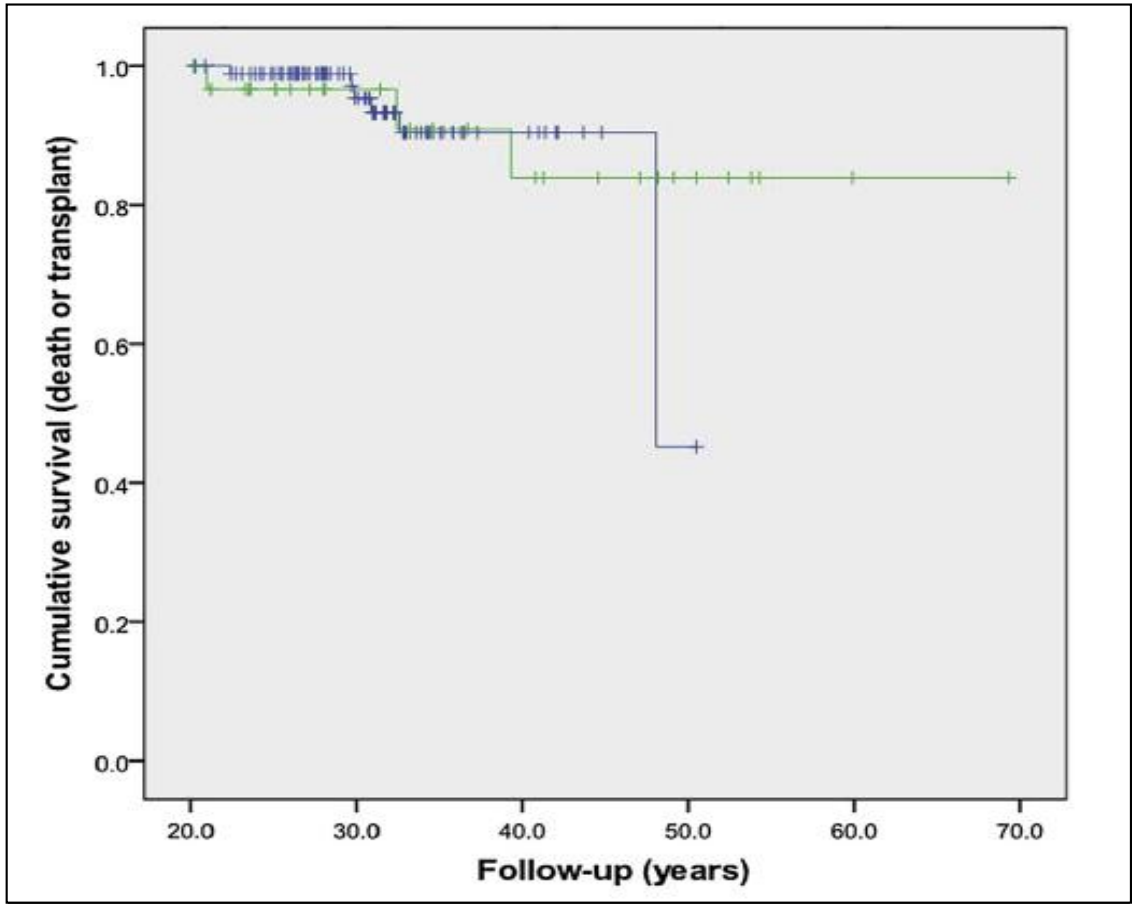
Richard Dobson, MBChB, MRCP, Mark Danton, MD, FRCS, Walker Nicola, PhD, MRCP, and Walker Hamish, MD, MRCP

Objective: The study objective was to evaluate long-term trends in morbidity and mortality in a national cohort of adult patients with a systemic right ventricle due to the atrial switch for transposition of the great arteries or congenitally corrected transposition of the great arteries.

Methods: We performed a retrospective cohort study from a baseline of 18 years, including life table and Kaplan–Meier analysis for probability of death/transplant, arrhythmia, surgical or percutaneous intervention, and permanent pacemaker insertion.

Results: A total of 97 adults with transposition of the great arteries–atrial switch (Mustard procedure in 80/Senning procedure in 17) and 32 adults with congenitally corrected transposition of the great arteries survived. The median ages at latest follow-up were 29 and 34 years, respectively. At 40 years of follow-up, freedom from death or transplant was 0.90 for those with transposition of the great arteries–atrial switch and 0.84 for those with congenitally corrected transposition of the great arteries ($P = .833$). Freedom from arrhythmia at 40 years of follow-up was 0.51 for those with transposition of the great arteries–atrial switch and 0.93 for those with congenitally corrected transposition of the great arteries ($P = .007$). Freedom from intervention at 40 years of follow-up was 0.33 for those with transposition of the great arteries–atrial switch after initial repair and 0.53 for those with congenitally corrected transposition of the great arteries ($P = .938$). Freedom from pacemaker insertion at 40 years of follow-up was 0.77 for those with transposition of the great arteries–atrial switch and 0.62 for those with congenitally corrected transposition of the great arteries ($P = .161$).

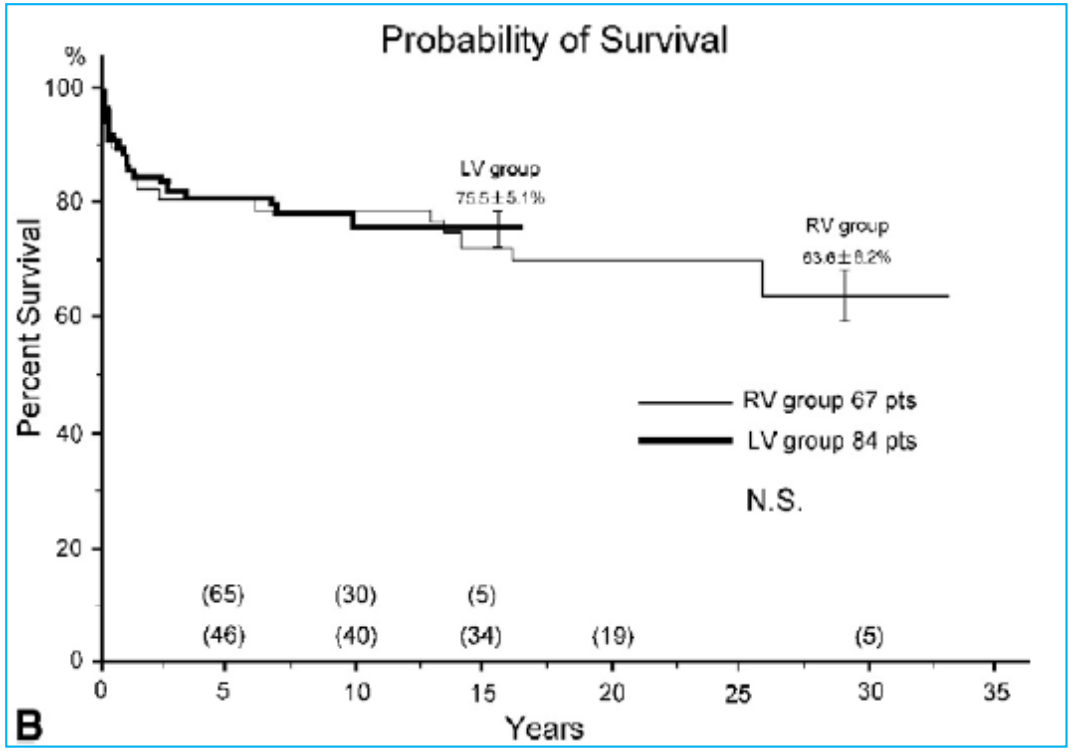
Long-term survival with systemic RV



Outcomes of definitive surgical repair for congenitally corrected transposition of the great arteries or double outlet right ventricle with discordant atrioventricular connections: Risk analyses in 189 patients

Toshiharu Shin'oka, MD, PhD, Hiromi Kurosawa, MD, PhD, Yasuharu Imai, MD, PhD, Mitsuru Aoki, MD, PhD, Masakuni Ishiyama, MD, PhD, Takahiko Sakamoto, MD, PhD, Shinka Miyamoto, MD, Kyoko Hobo, MD, and Yuki Ichihara, MD

Survival with Systemic RV vs Systemic LV in patients with discordant great vessels.



Perspective

Overview of adult congenital heart transplants

Roosevelt Bryant III[1,2], David Morales[1,2]

[1]Division of Cardiovascular Surgery, The Heart Institute, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA; [2]Department of Surgery, The University of Cincinnati College of Medicine, Cincinnati, OH, USA

Correspondence to: Roosevelt Bryant III, MD. Associate Professor of Surgery, The University of Cincinnati Medical Center, Surgical Director, Heart Transplant Program, Cincinnati Children's Hospital Medical Center, 3333 Burnett Avenue, Cincinnati, OH, USA. Email: roosevelt.bryantiii@cchmc.org.

Patients with systemic RV seems to merge well with the rest of the population of ACHD requiring cardiac replacement therapy. This is in marked contrast to the Fontan failure patients who have almost universally been noted to have adverse outcome as compared to the rest of ACHD population and non-ACHD population.

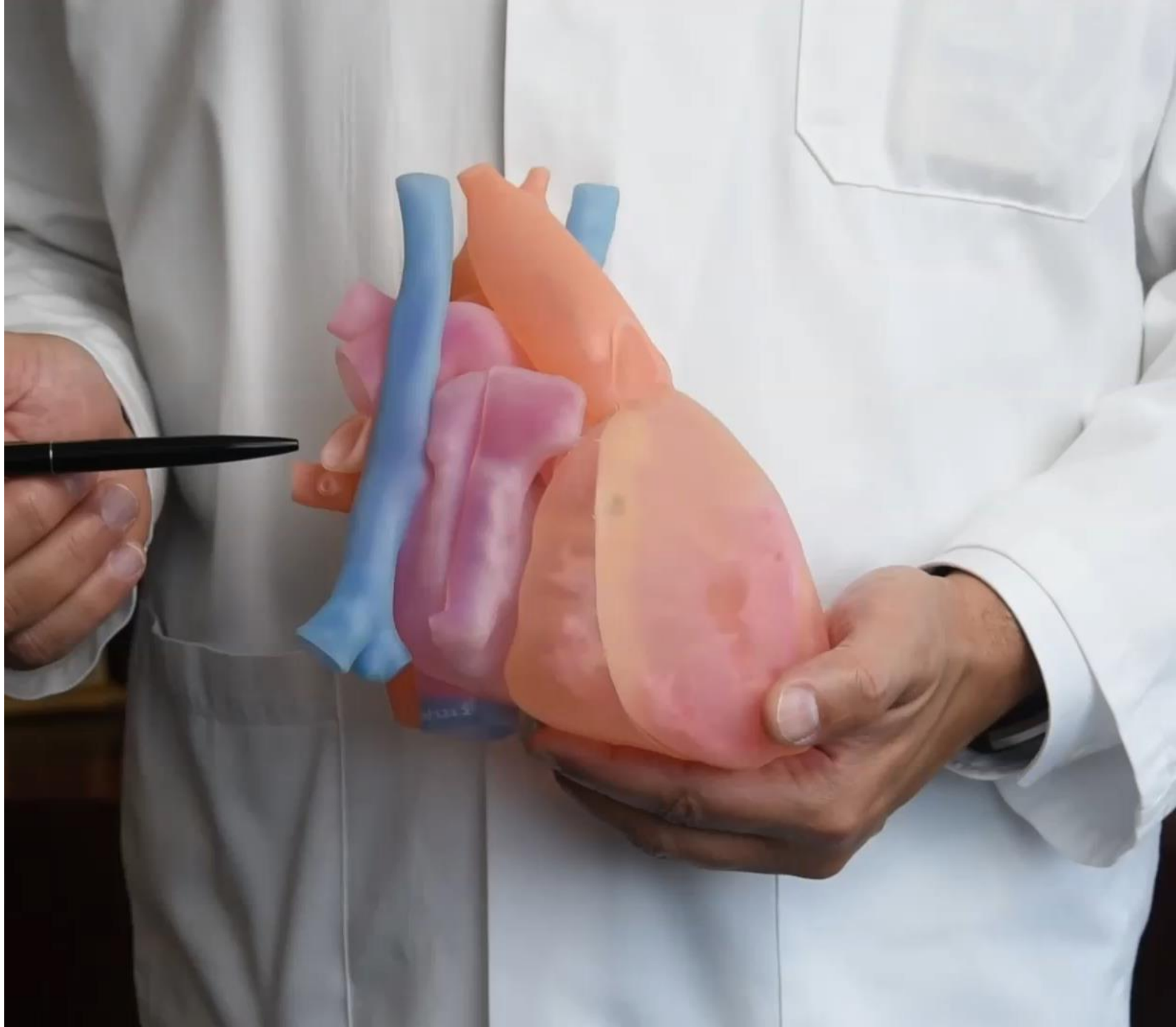
Effect of Systemic RV on outcome of HTx.

From 216 patient in INTERMAC Registry, ACHD patients requiring mechanical circulatory support as bridge to transplantation had survival similar to non-ACHD patients and that ventricular morphology had no adverse effect on the mortality of these patients

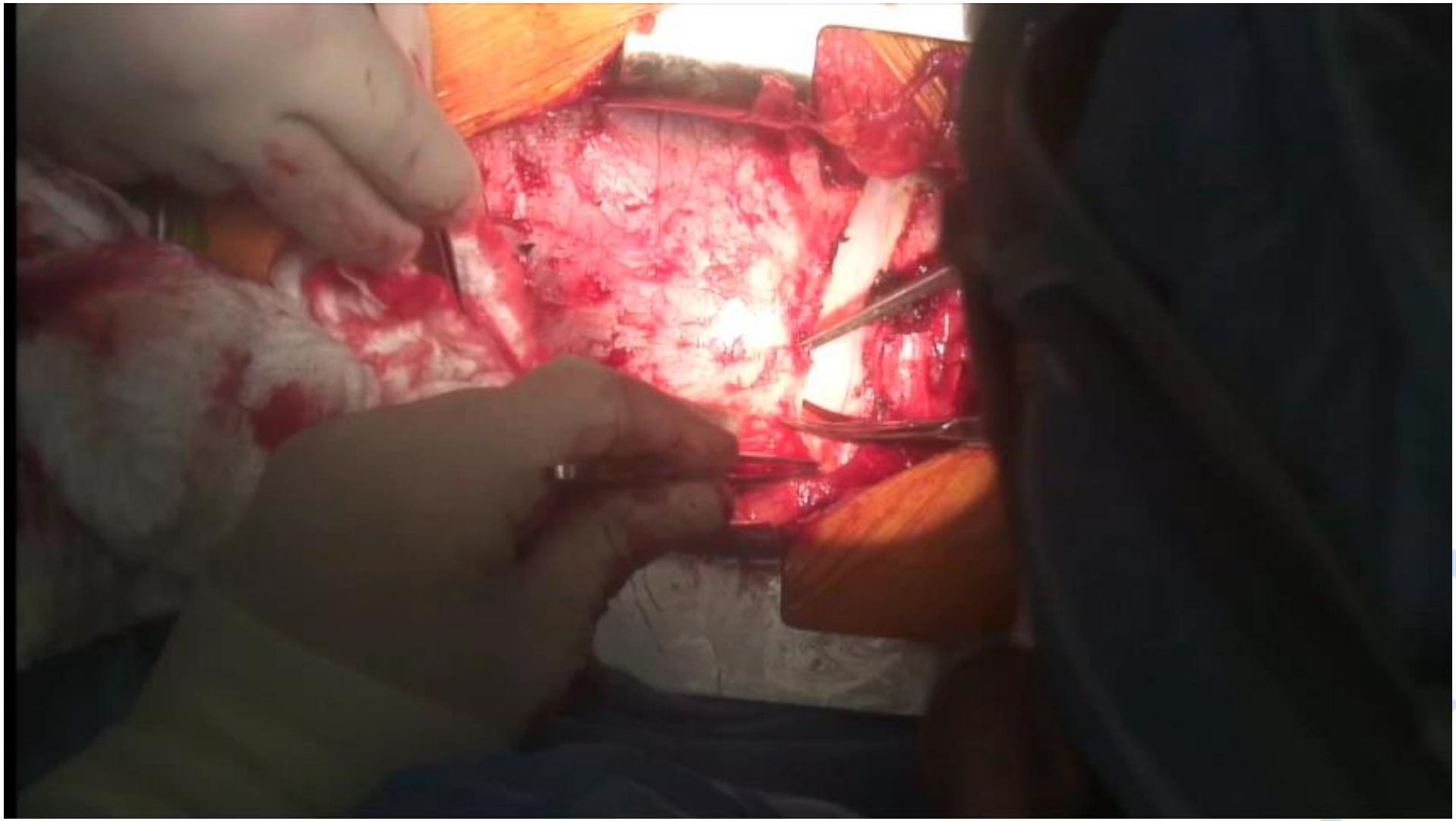
Our Hypothesis

A biventricular repair with CCTGA physiology will provide better palliation than Fontan physiology and a failing systemic right ventricle will result in better transplant candidacy and better long-term outcome.

John



Biventricular Conversion



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