Risk Modeling in Congenital Heart Disease: Empiricism to Drive Change

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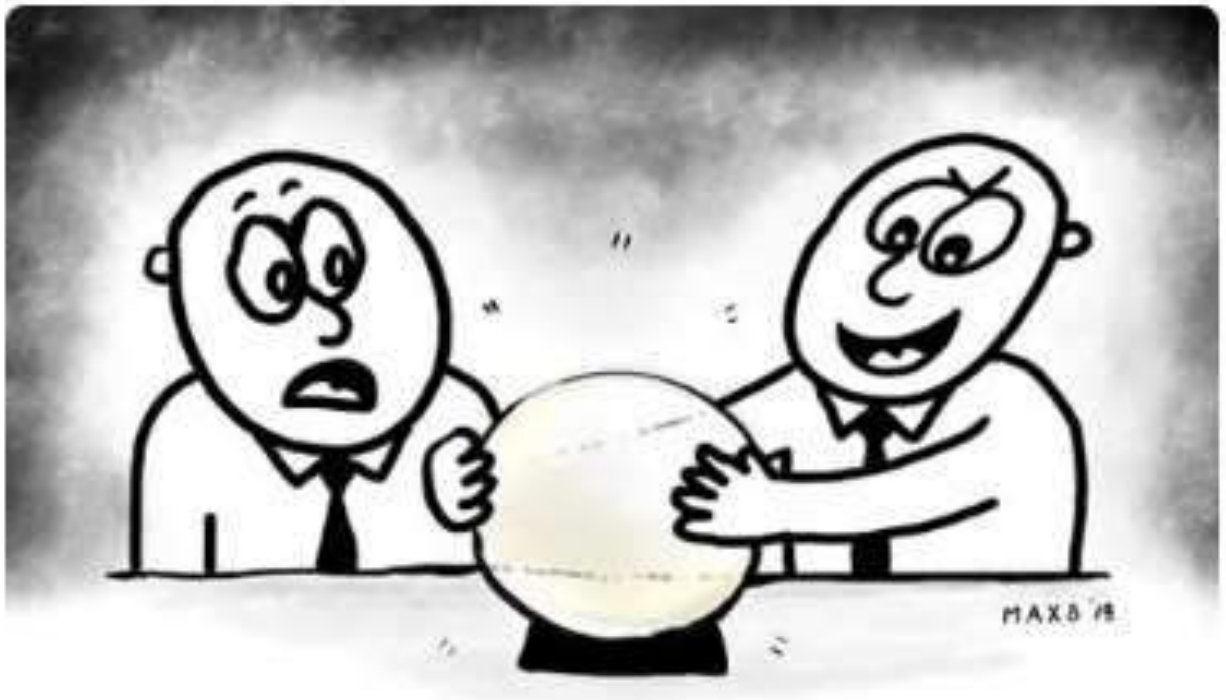
CHOP Meeting – February 2024

Disclosures: Edwards Lifesciences

CHOP Meeting – February 2024

Risk Models…..



- Let's see what our risk forecasting model has to say...

Developing (GOOD) Risk Models in CHS is HARD

- Imprecise definitions
- Heterogeneity in diagnoses and procedures
- Small numbers or patients
- Variation in practice/approach
- Competing (and often multiple) events
- Immortality bias
- Multi-modal data
- Longitudinal data
- Evolution of care/Introduction of New Therapies (era effects)

Multi-modal and Longitudinal Data

Optimizing evaluation in pediatric and young adult patients with Marfan syndrome: Novel longitudinal metrics to track growth of aortic structures

Salima Ahmed Bhimani, MD • Abdelrahman Rahmy, BS • Suzie Kim, BS • Justin B. Jin, MD • Kenneth Zahka, MD • Rukmini Komarlu, MD • Lars G. Svensson, MD, PhD • Amol Pande, PhD • Tara Karamlou, MD, MSc • the Members of Marfan Working Group * • Jeevanantham Rajeswaran, PhD • Vidayasagar Kalahasti, MD • Milind Desai, MD • Eric Roselli, MD • Miza Salim-Hammoud, MD • William C. Frankel, MD • Gosta Pettersson, MD, PhD • Eugene H. Blackstone, MD • Hani K. Najm, MD, MSc • Jared Klein, MD • Show less • Show footnotes

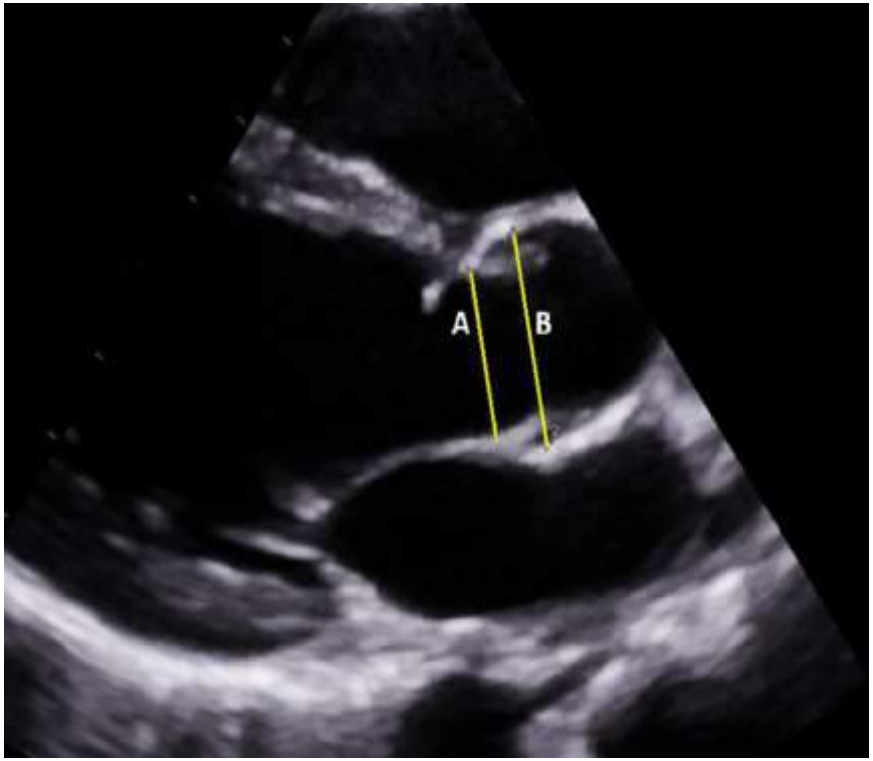
Published: March 15, 2022 • DOI: https://doi.org/10.1016/j.jtcvs.2021.12.056 •

Methods

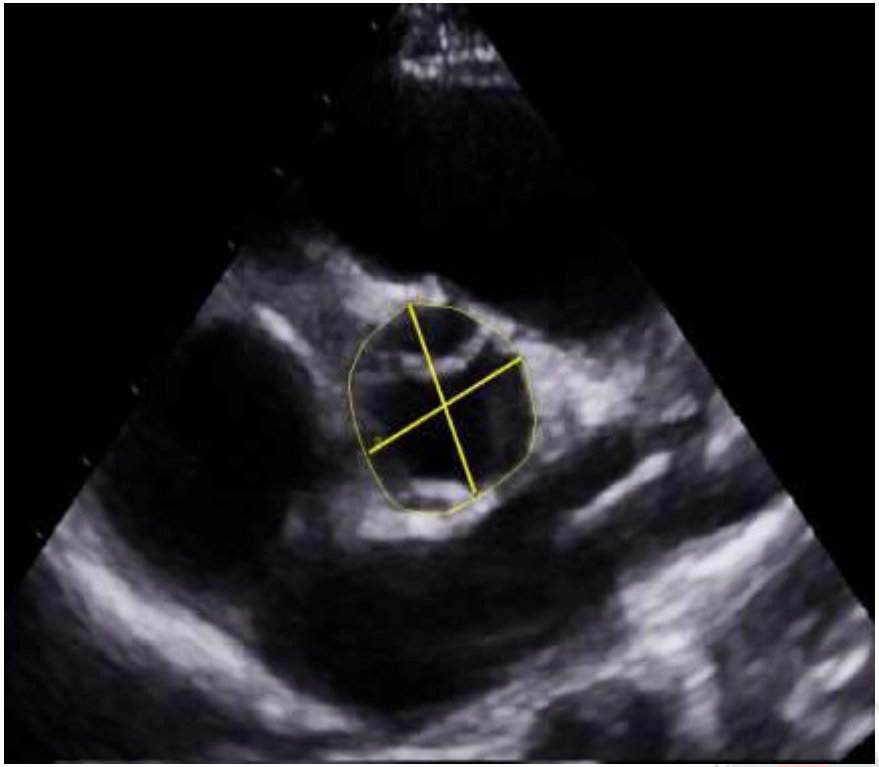
- Pediatric and young adults age < 26 yrs
- 2001 – 2020
- Inclusion criteria:
  - Genotype positive
  - Phenotype consistent with modified Ghent criteria
- Serial echocardiograms (N=809) underwent protocolized review
  - Aortic root cross-sectional area/Height (CSA/Ht)

CSA/Ht

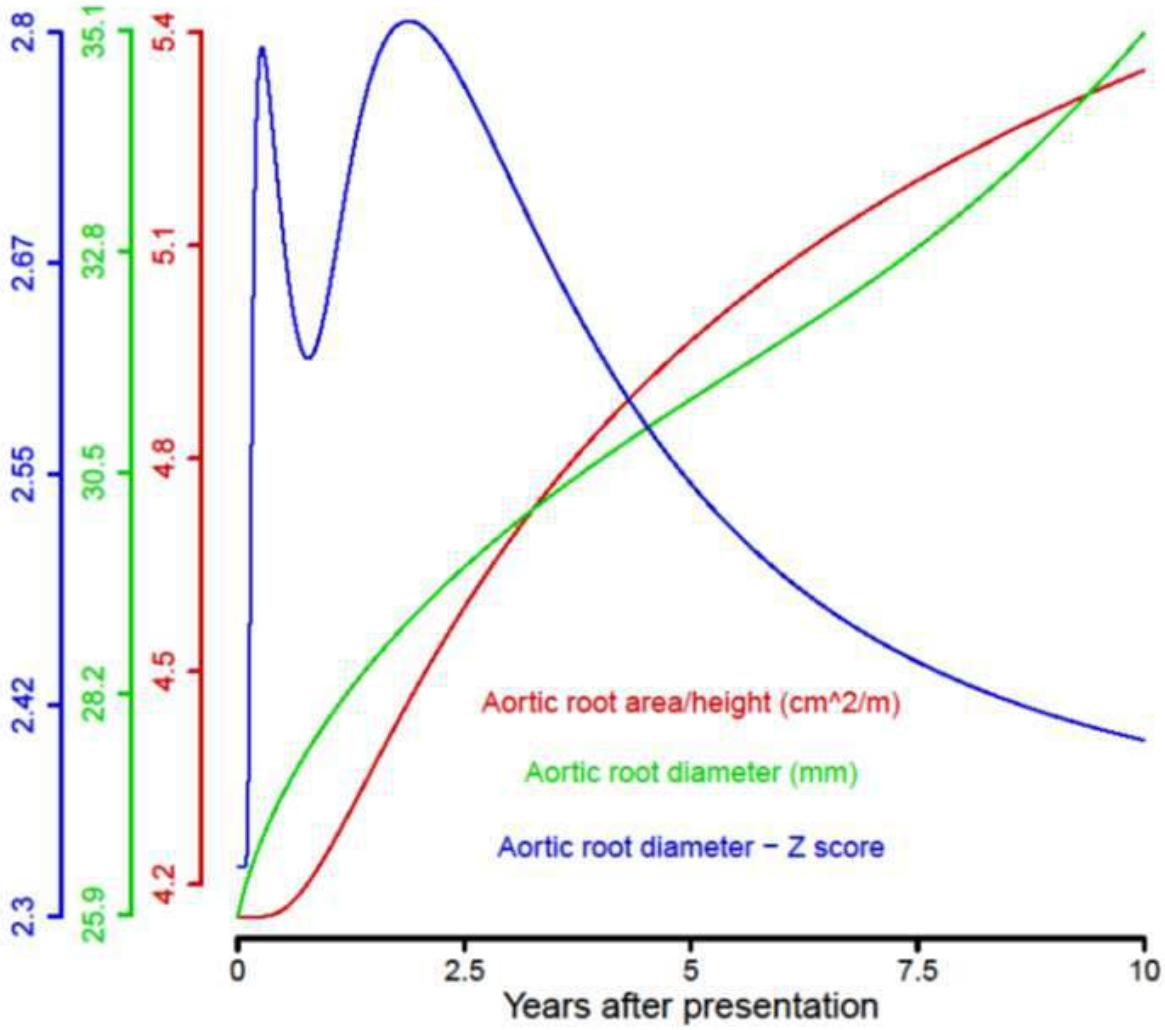
Parasternal long-axis view: leading-edge to leading-edge in early-mid systole



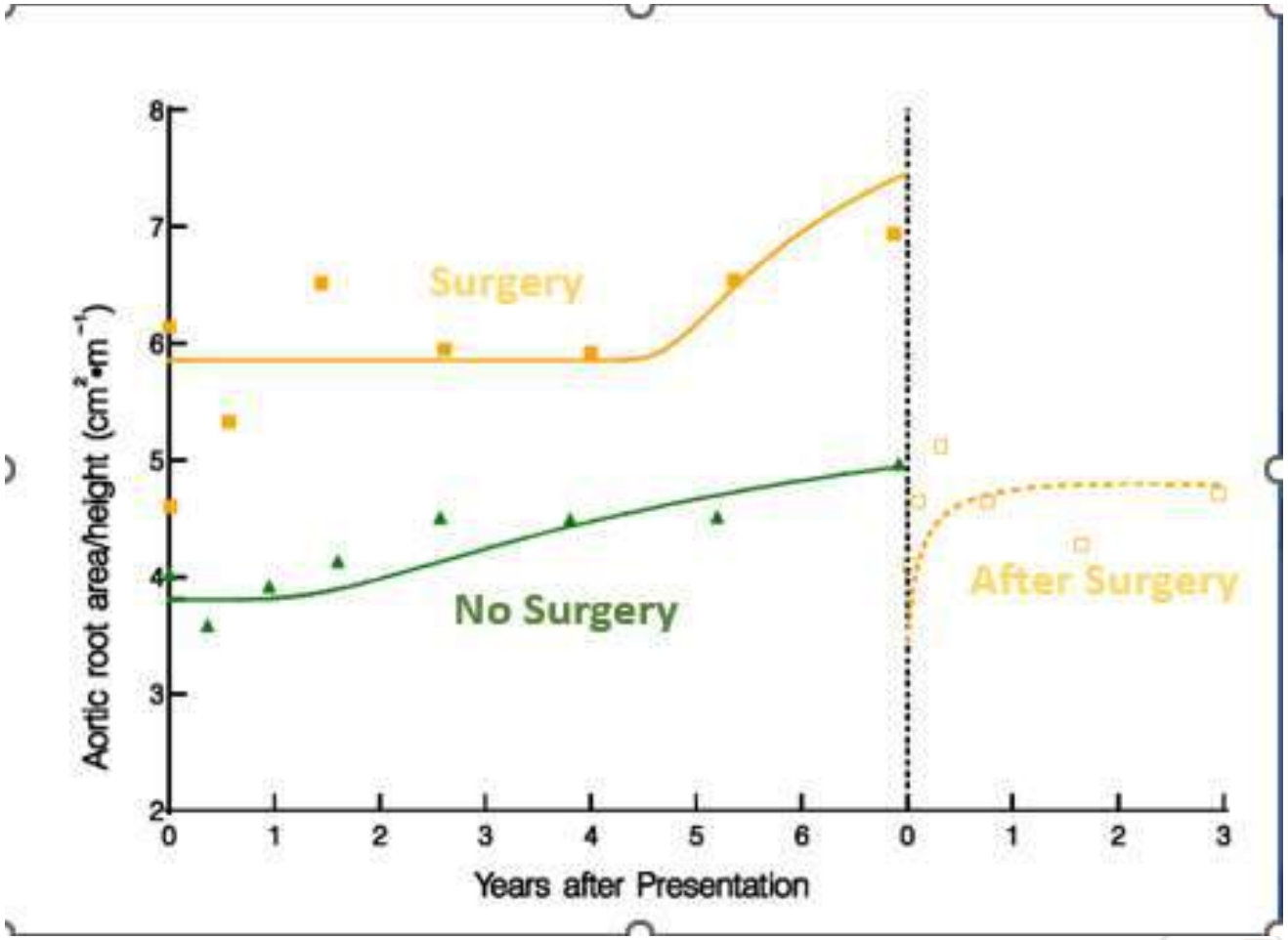
Planimetry: parasternal short-axis view- tracing the aortic root en face



Echo-derived Metrics of Aortic Root Growth



CSA/H Index as a Surgical Threshold



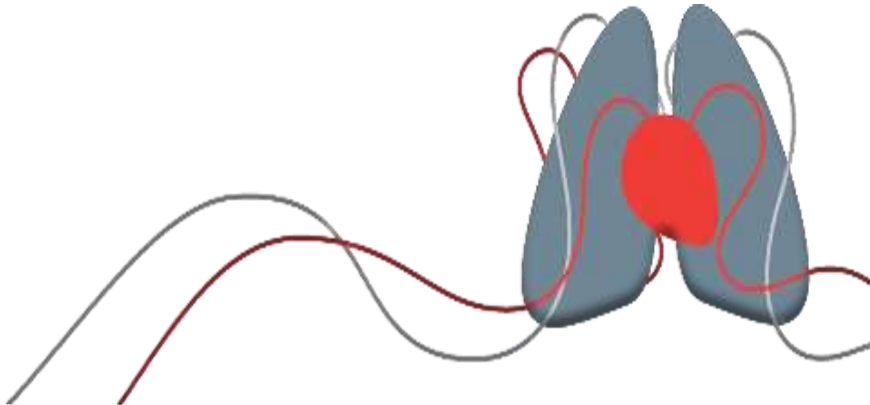
Modifying Longitudinal Growth Trajectories of Aortic Structures in Pediatric Marfan Syndrome: Impact of and Interactions Between Medications and Surgery

Tara Karamlou, MD, MSc;

Amol Pande, PhD; Salima Bhimani, MD; Justin Jin, MD; Kenneth Zahka, MD; Rukmini Komarlu, MD;

Lars G. Svensson, MD, PhD for the Members of the Marfan Working Group

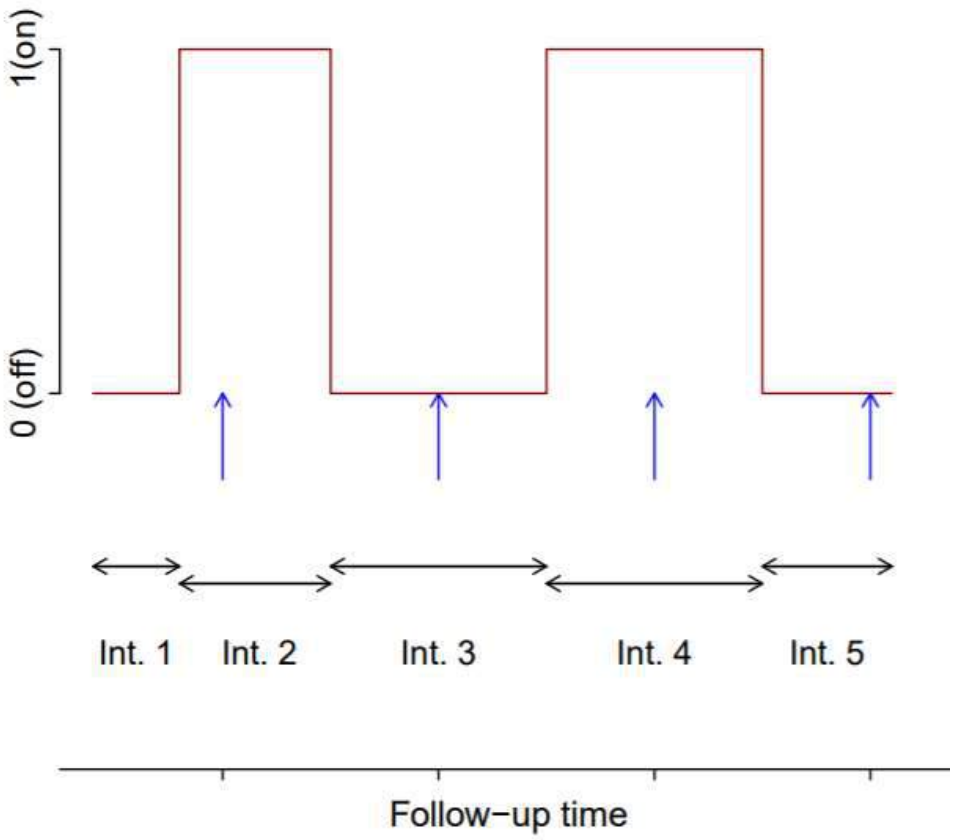
CHOP Meeting February 2024



Objective

To evaluate how the initiation and dosage adjustments of medications over time, accounting for surgical intervention, impacted aortic root growth in pediatric patients and young adults with Marfan syndrome

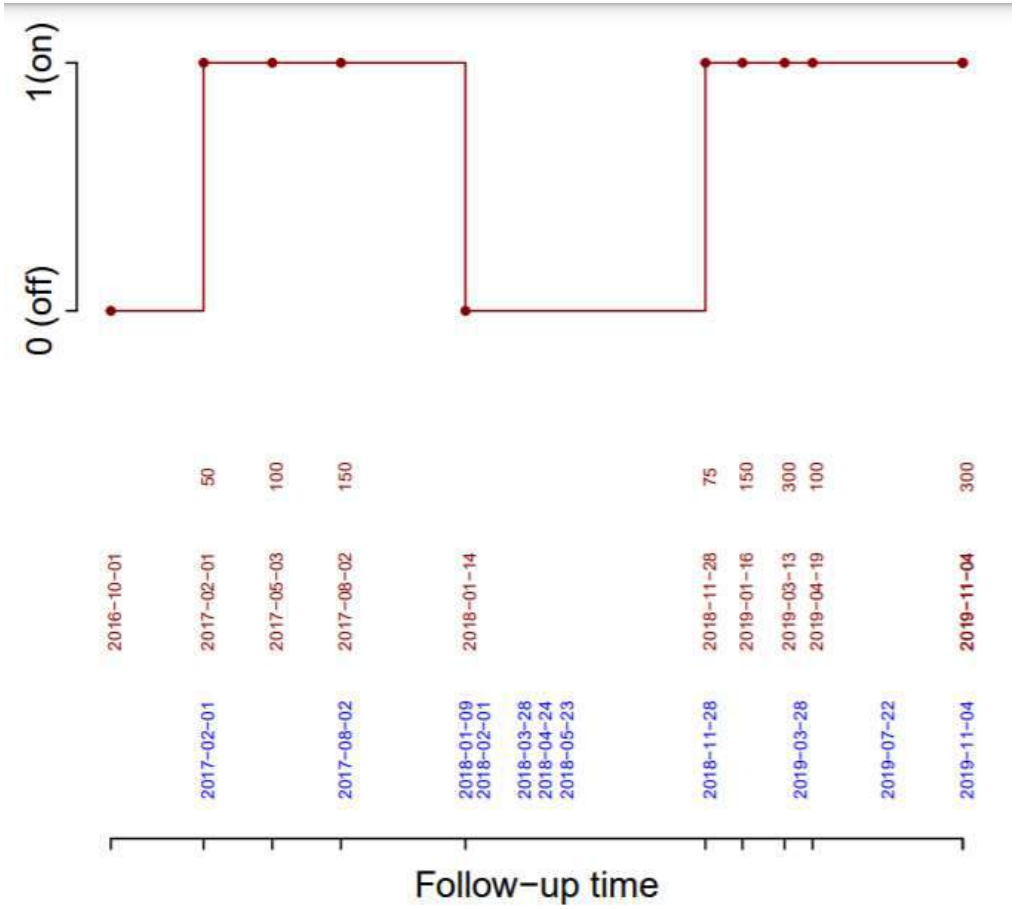
Time-Varying Longitudinal Data Related to Longitudinal Outcome



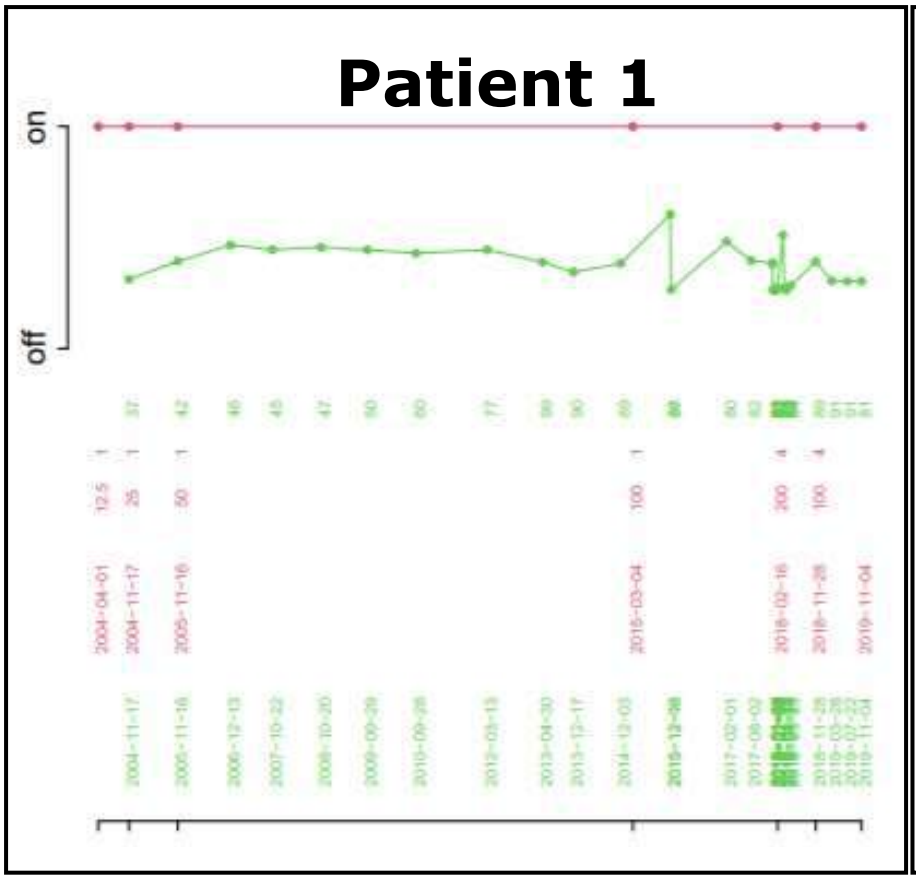
A common scenario without utility models!!

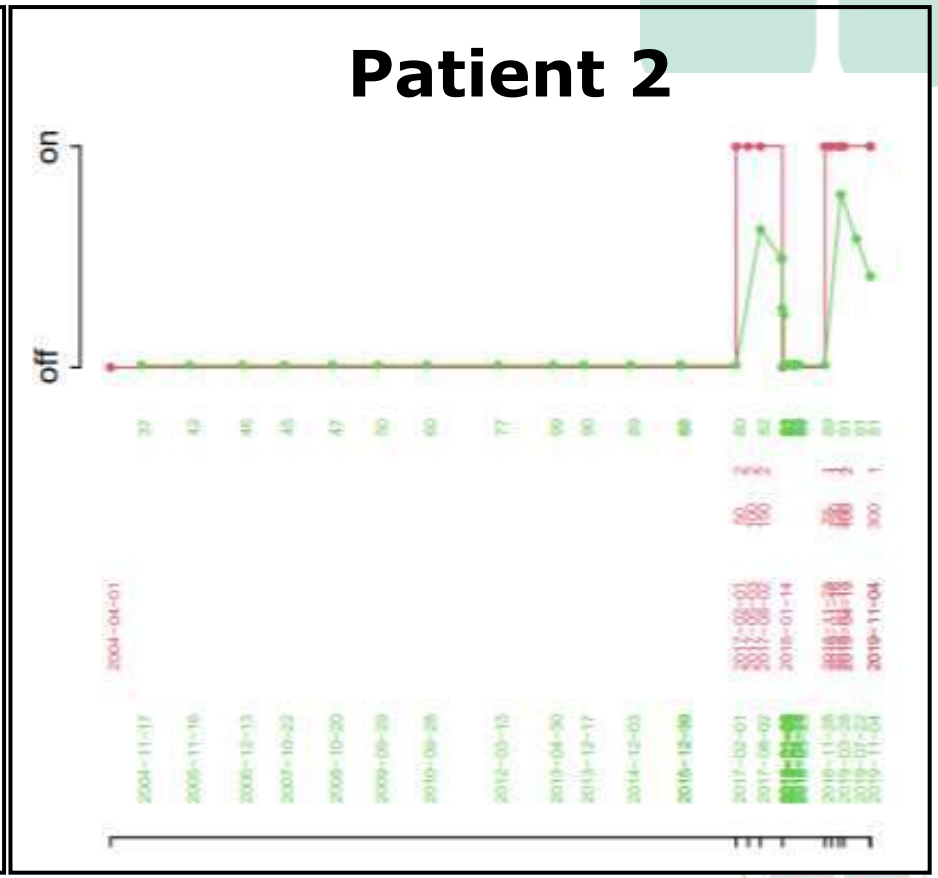
Pande A, et al

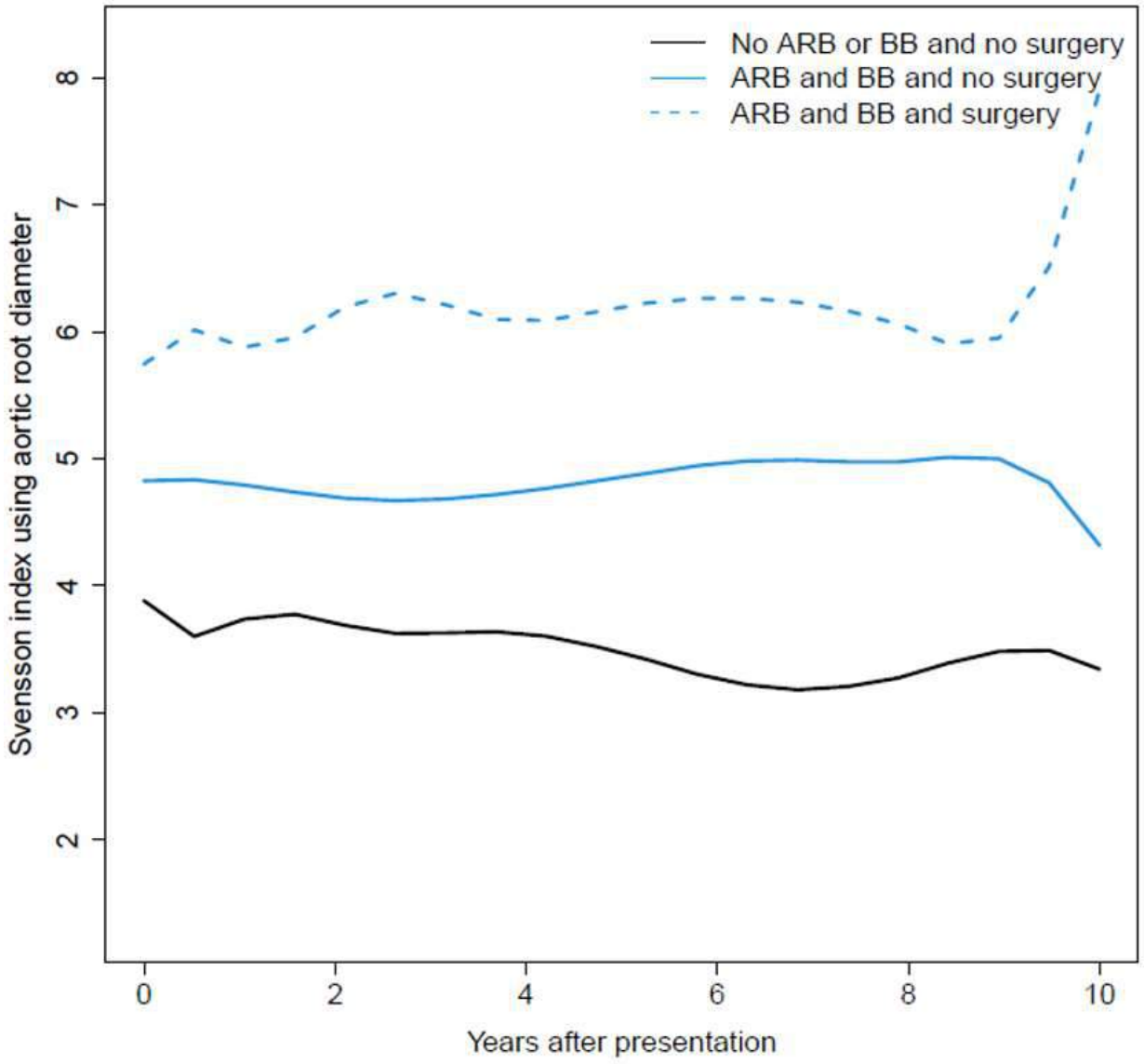
Clinical Scenario



Time-varying Medication Adjustments






No ARB or BB and no surgery
ARB and BB and no surgery
ARB and BB and surgery
Svensson index using aortic root diameter
2
3
4
5
6
7
8
0
2
4
6
8
10
Years after presentation

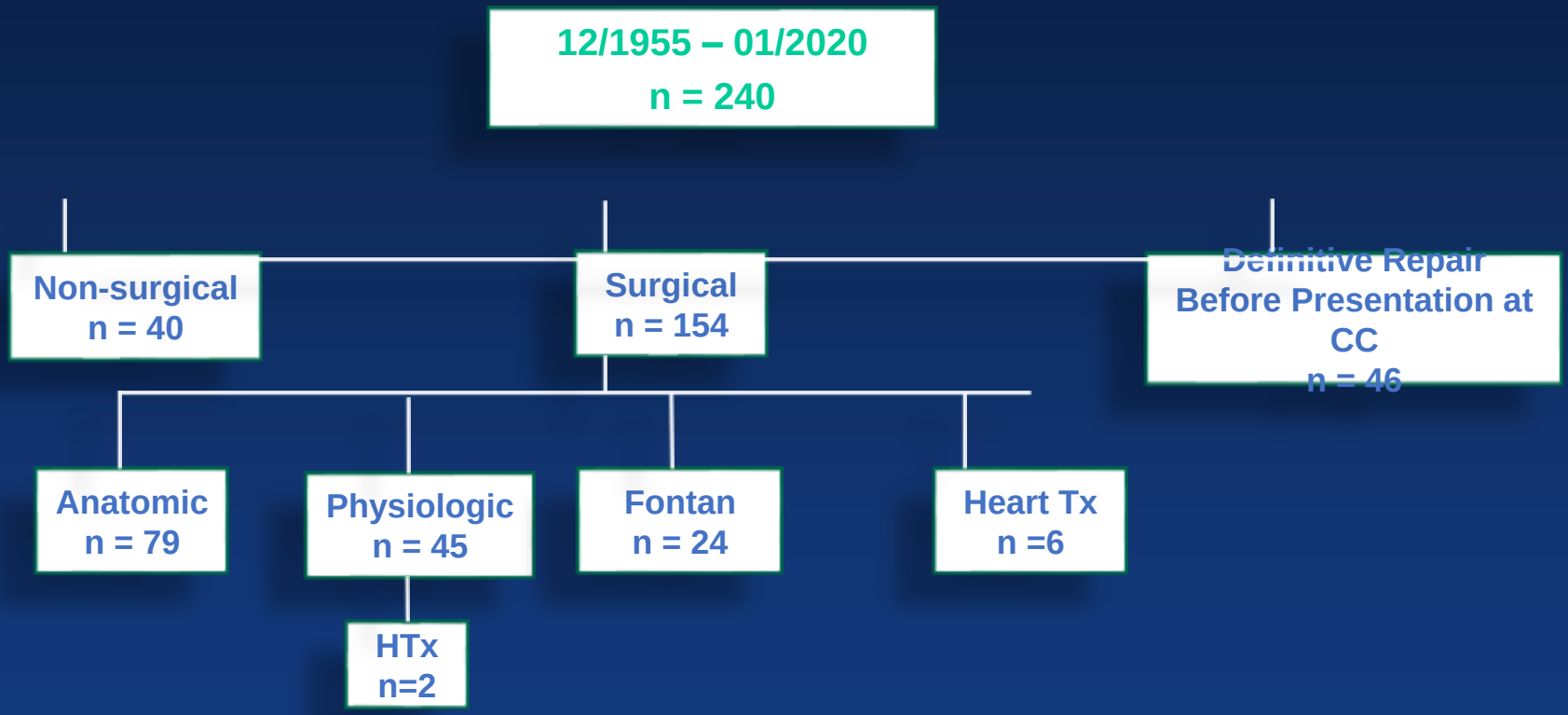
Heterogeneous Diagnoses and Procedures
Variable Approaches
Competing and Repeated Events

Outcomes of Treatment Pathways in 240 Patients with Congenitally Corrected Transposition of Great Arteries

Paola A. Barrios, Aisha Zia, Gosta Pettersson, Hani K. Najm, Jeevanantham Rajeswaran, Salima Bhimani, Jared Klein, Elizabeth Saarel, Rukmini Komarlu, Naseeb G. Shaheen, Eugene H. Blackstone, Leah J. Lee, Tara Karamlou

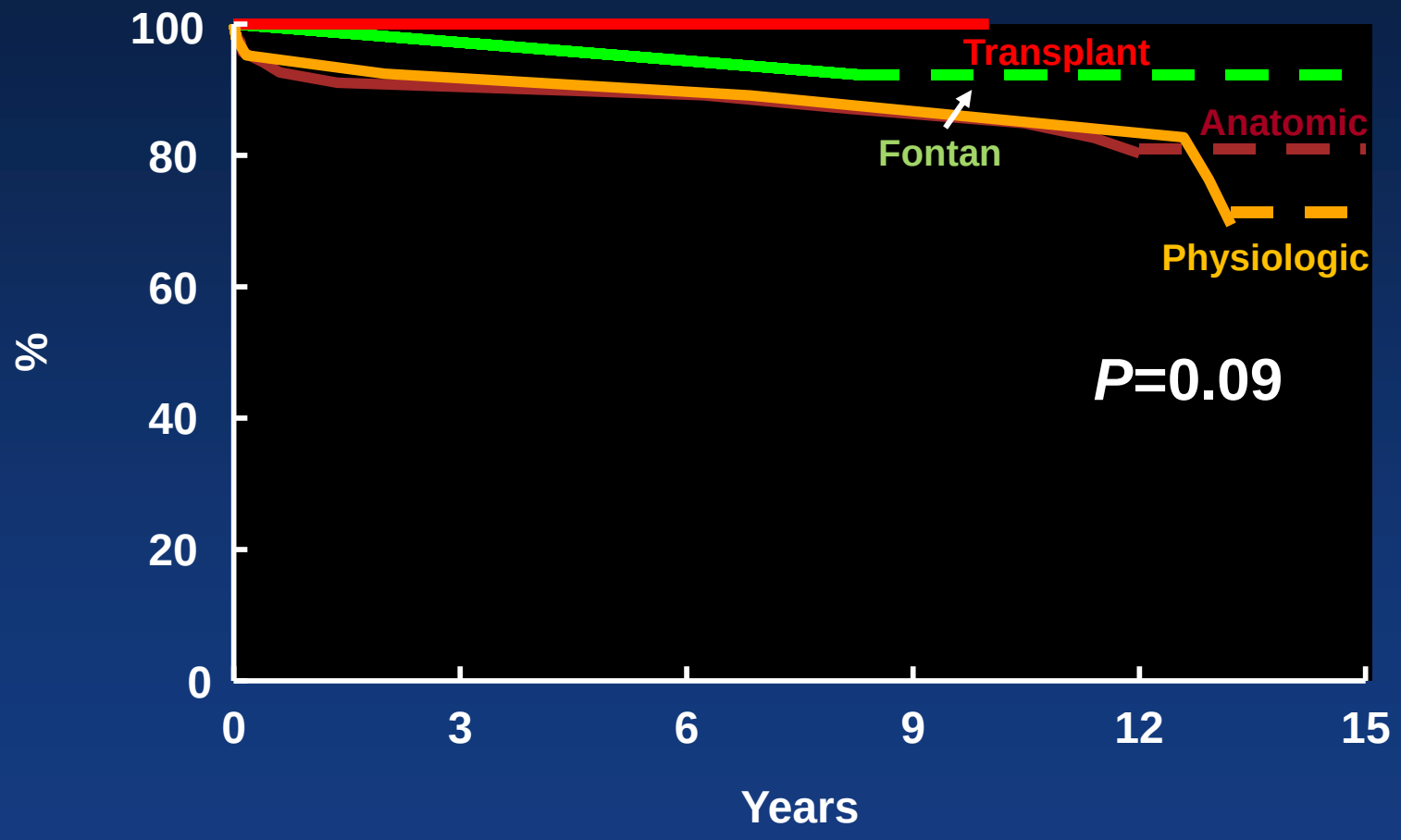
https://doi.org/10.1016/j.jtcvs.2020.11.164

ccTGA Patients

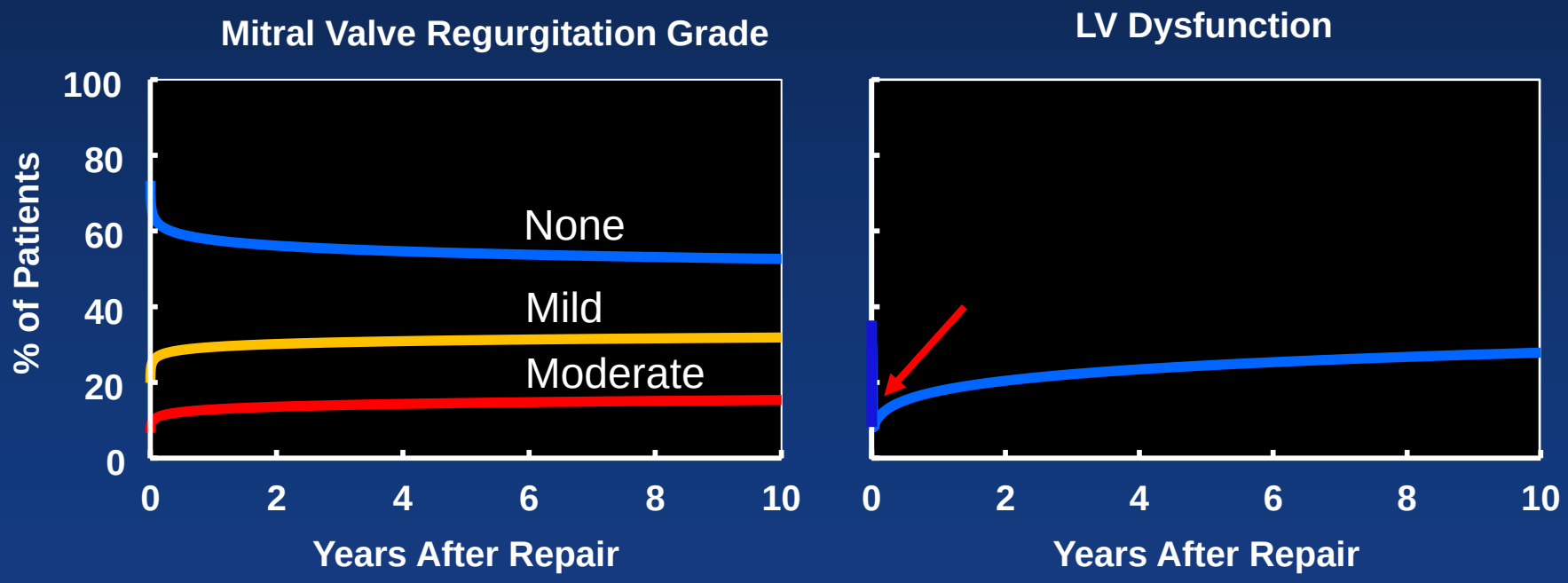


Median age at anatomic repair: 1.9 years

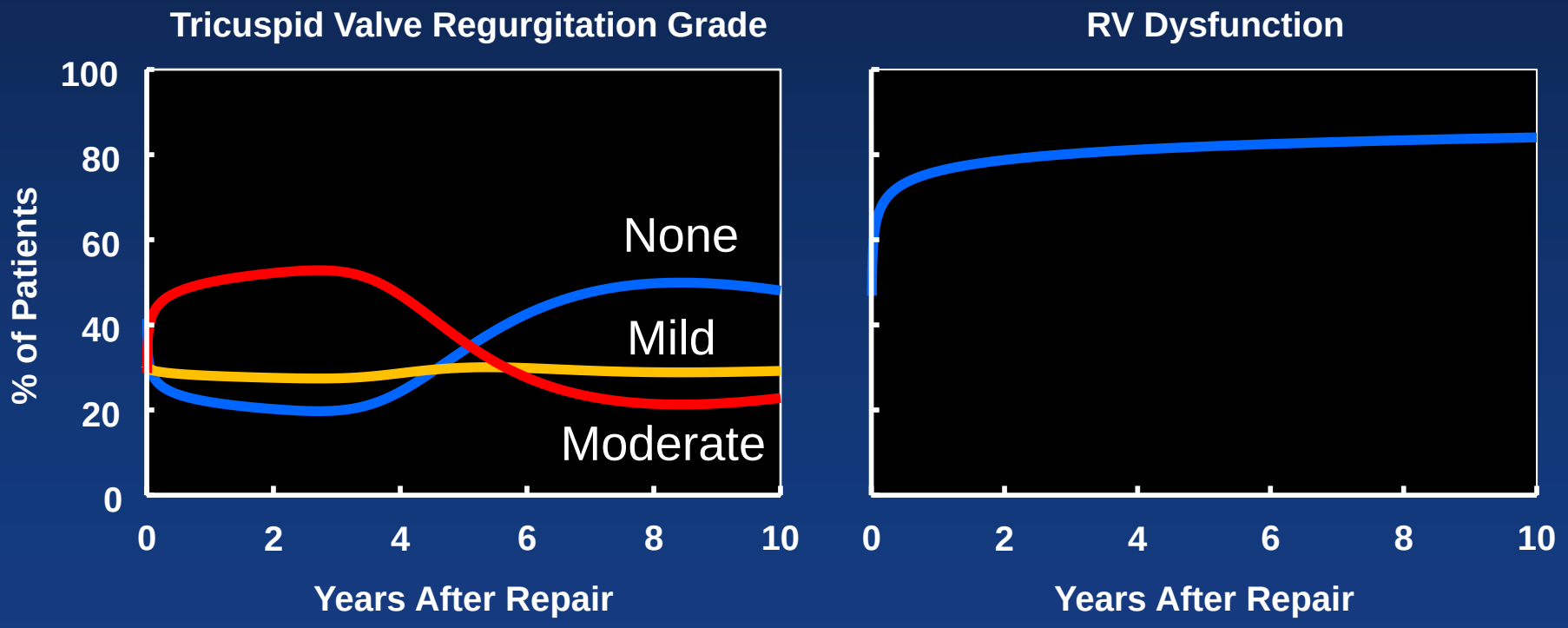
Overall Survival



Systemic AVV and Ventricular Function in Anatomic Repair



Systemic AVV and Ventricular Function in Physiologic Repair



Conclusions

Early anatomic repair may be preferable to physiologic repair

Late attrition after physiologic repair represents failure of expectant management

Patients in the PR group underwent procedures to improve TV function but with failure to rescue the failing RV

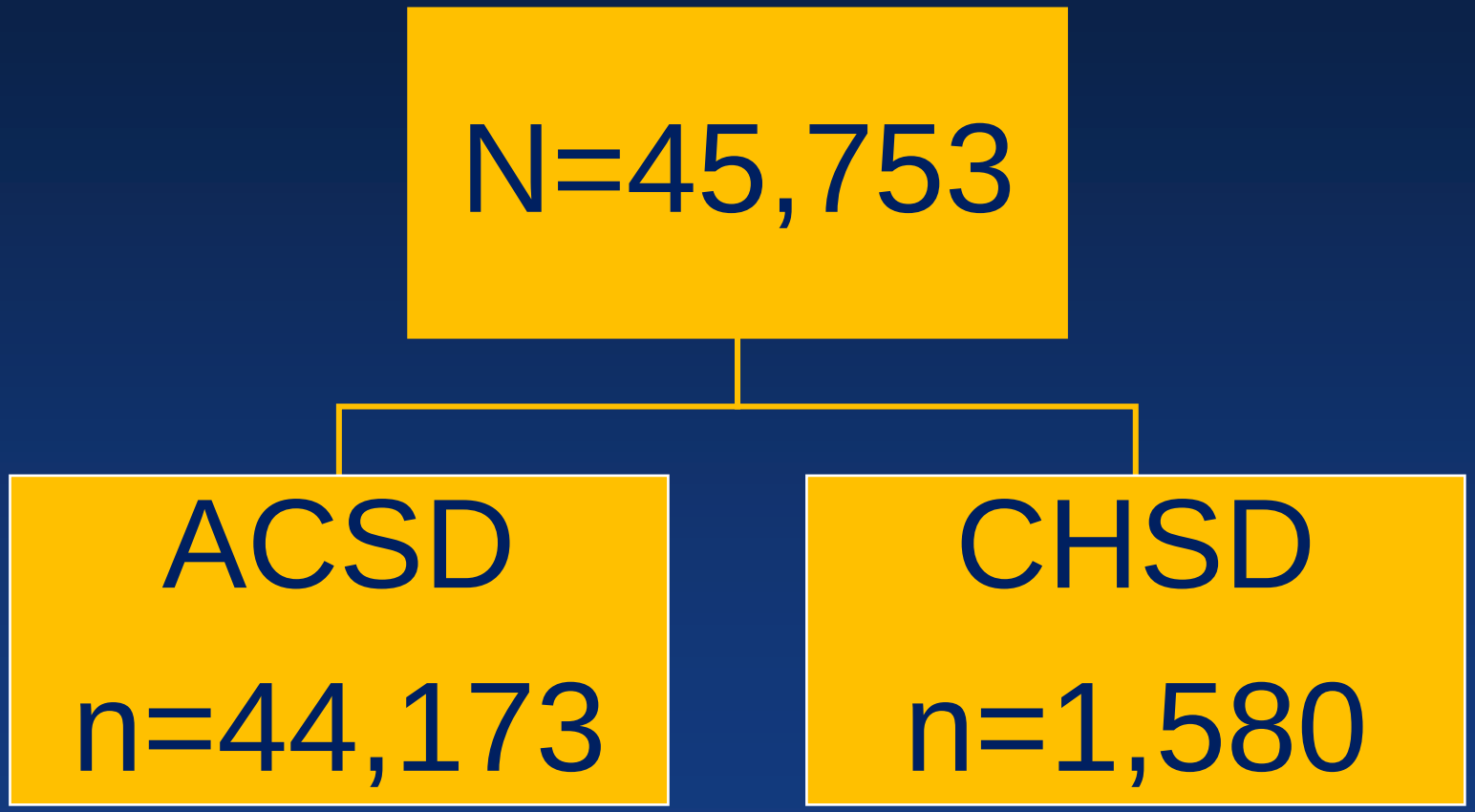
Evolution of Care/Introduction of New Therapies (Era Effect)

Aortic Valve Replacement in Young and Middle-Aged Adults: Current and Potential Future Roles for TAVR

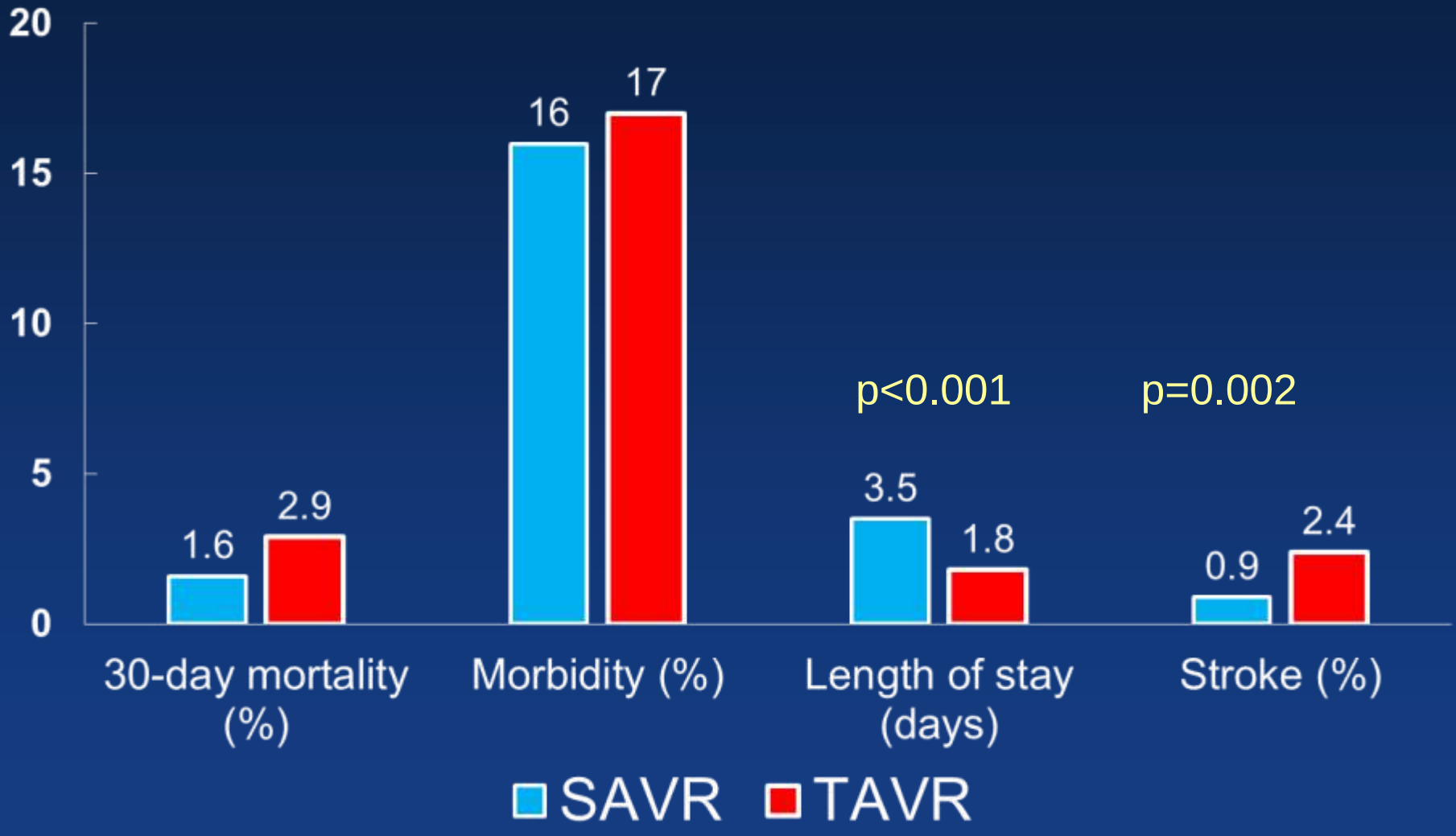
Jennifer S. Nelson, MD, MS; Timothy M. Maul, PhD, CCP; Peter D. Wearden, MD, PhD; Hani K. Najm, MD, MSc; Orkun Baloglu, MD; Douglas R. Johnston, MD; Tara Karamlou, MD, MSc

J. Maxwell Chamberlain Paper
Ann Thorac Surg 2021;112:132-138

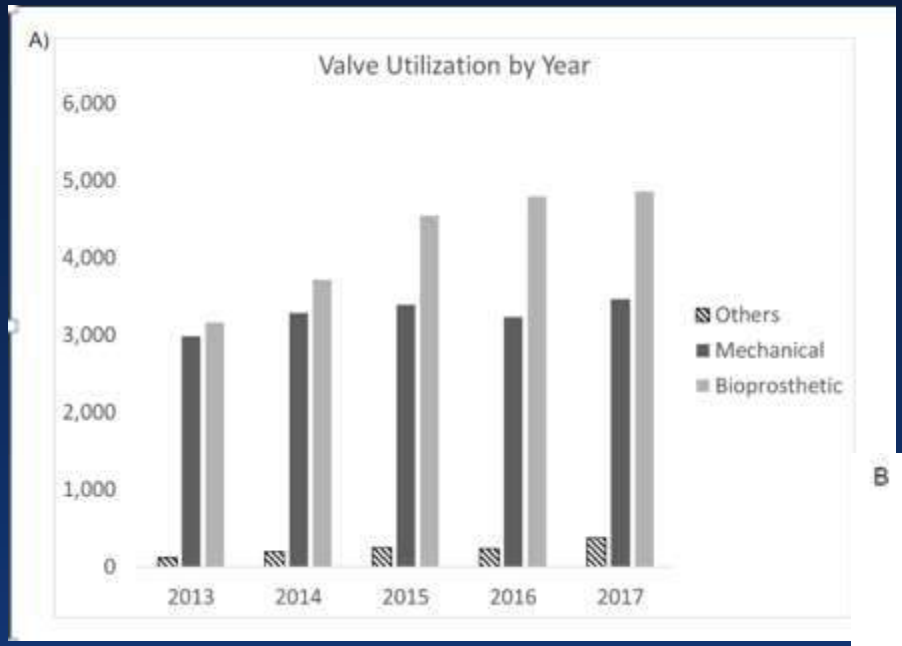
Patients

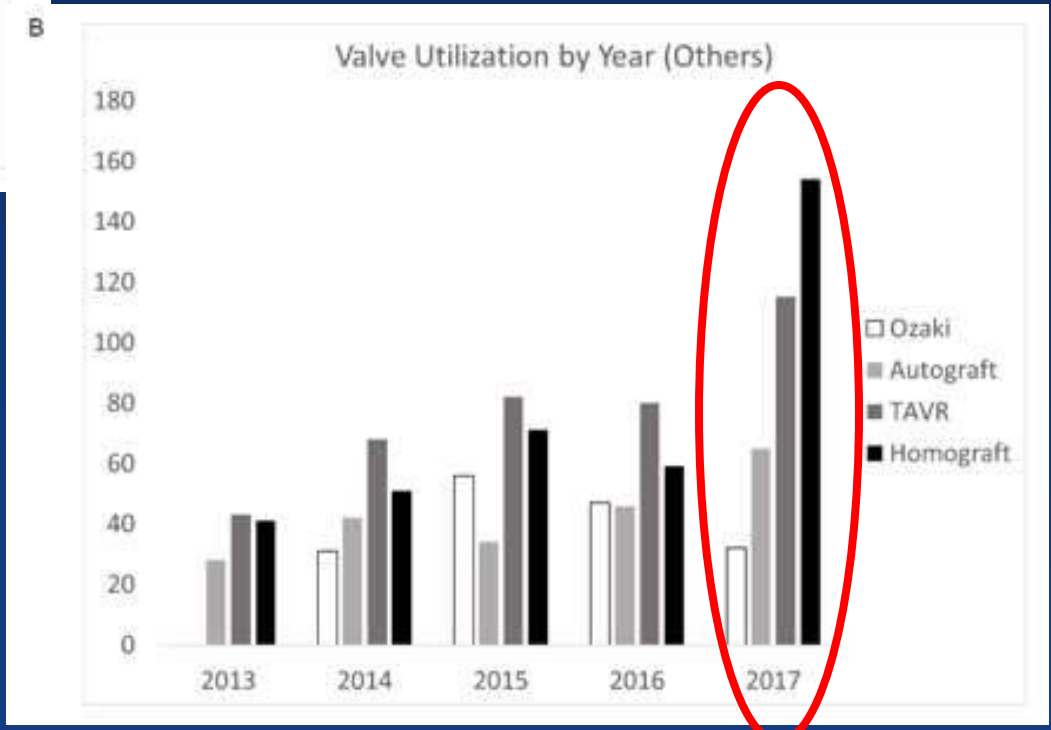

N=45,753
ACSD
n=44,173
CHSD
n=1,580
Jan 2013 – Sept 2018
1,093 Centers
Jan 2013 – June 2018
105 Centers

Isolated SAVR vs TAVR



TAVI USE (Up 163%!)





Imprecise Definitions

John Deanfield 2003

" As a result of the success of paediatric cardiology and cardiac surgery over the last three decades, there will shortly be more adults than children with congenital heart disease. Prior to the advent of surgery, less than 20% of children born with congenital heart malformations survived to adult life. Now, most deaths from congenital heart disease occur in adults. **The 'new population' of patients with congenital heart disease no longer fits within traditional divisions of training and practice, which have separated adult and paediatric cardiology. Adult cardiologists are not equipped to deal with the range and complexity of grown-up patients with congenital heart disease, whereas paediatric cardiologists cannot be expected to manage the many acquired adult diseases in a paediatric medical environment.** Up till now, care has been delivered by a number of enthusiastic centres who have managed the complex medical, surgical and psychosocial needs of the grown-up patients with congenital heart disease. In most countries, however, an organized system is not yet in place. This is needed for continued provision of excellence in clinical care, accumulation of knowledge about the late outcome of management strategies in childhood (with feedback to paediatric practice) as well as for training.

The lack of information regarding numbers, diagnoses and treatment as well as the regular occurrence of avoidable medical problems in this population is testimony to the deficiencies of the current system. The need to reintegrate paediatric and adult cardiac services, and in particular to **provide smooth 'transition' for adolescents is clear."**

Capturing ACHD: What Defines It?

ADULT: ADULT CONGENITAL HEART DISEASE: INVITED EXPERT OPINION

Identifying, capturing, and understanding surgery for adult congenital heart disease: A novel framework

Gösta B. Pettersson, MD, PhD, Tara Karamlou, MD, MSc, and Eugene H. Blackstone, MD

Need - Opportunity

No risk model has been specifically customized for the expanding ACHD population who undergo cardiac operations

STS ACHD Risk Model

Capturing Adult Congenital Heart Disease: Framework for Development of an Adult Congenital Heart Disease Mortality Risk Model

Jennifer S. Nelson, MD, MS • Stephanie Fuller, MD, MS • Yuli Y. Kim, MD • ... S. Adil Husain, MD • Carl L. Backer, MD • Tara Karamlou, MD, MSc • Show all authors

Published: November 03, 2021 • DOI: https://doi.org/10.1016/j.athoracsur.2021.09.063 • Check for updates

JAMES S. TWEDDELL MEMORIAL PAPER FOR CONGENITAL CARDIAC SURGERY

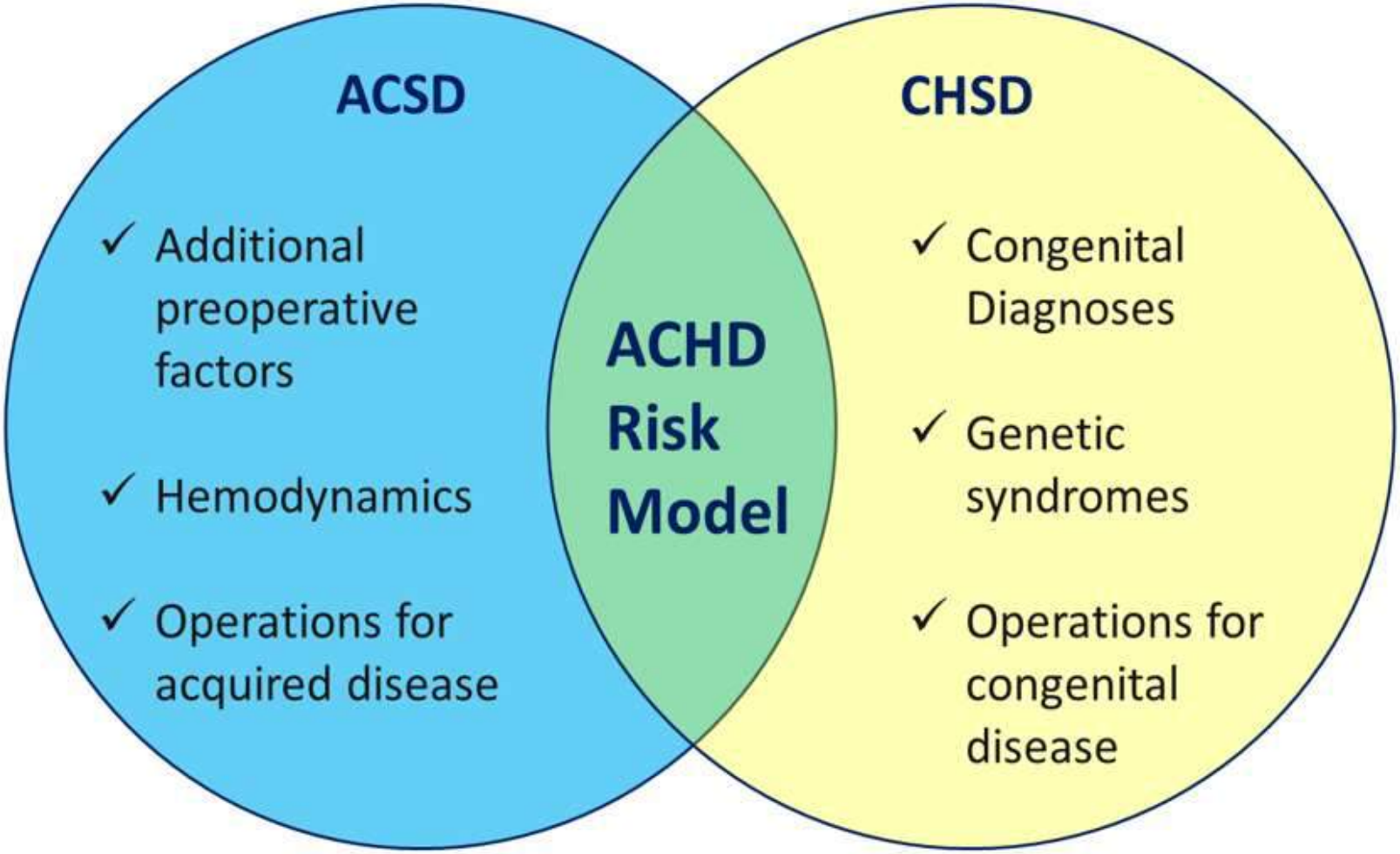
Development of a Novel Society of Thoracic Surgeons Adult Congenital Mortality Risk Model

Jennifer S. Nelson, MD, MS,[1,2] Dylan Thibault, MS,[3] Sean M. O'Brien, PhD,[3] Eric N. Feins, MD,[4] Jeffrey P. Jacobs, MD,[5] John E. Mayer, MD,[4] Hani K. Najm, MD, MSc,[6] David M. Shahian, MD,[7] Kevin D. Hill, MD,[8] Timothy M. Maul, CCP, PhD,[1,2,9] Robert H. Habib, PhD,[10] Jordan P. Bloom, MD, MPH,[7] and Tara Karamlou, MD, MSci[6]

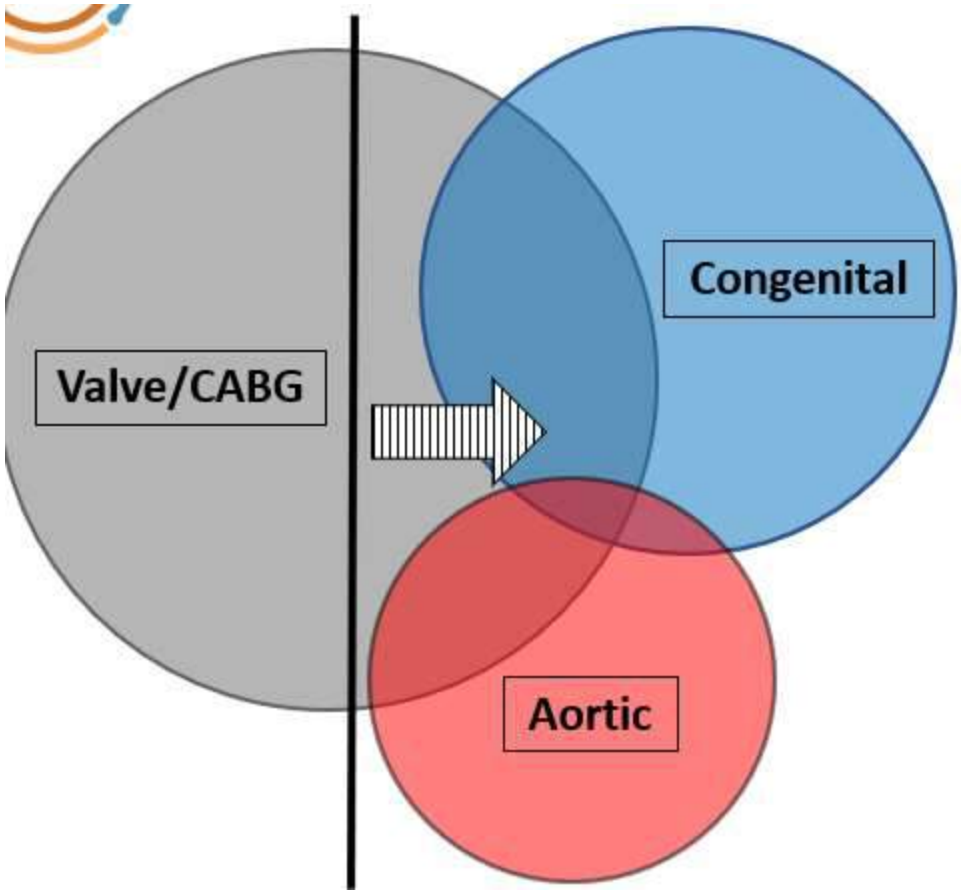
Check for updates

Clevelar

Existing STS Mortality Risk Models



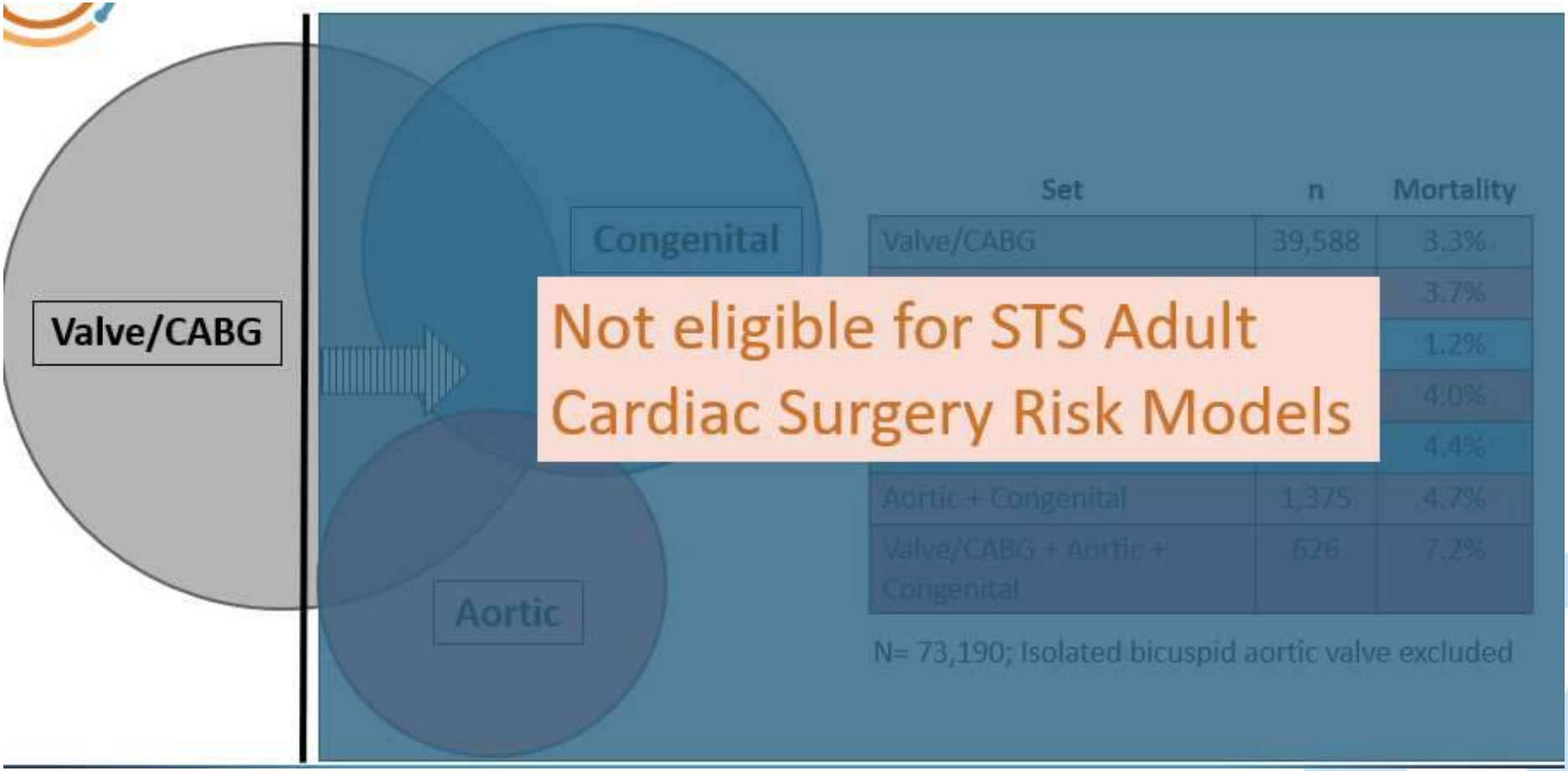
Combination/Complex Procedures



Set	n	Mortality
Valve/CABG	39,588	3.3%
Aortic	15,515	3.7%
Congenital	26,089	1.2%
Valve/CABG + Aortic	7,169	4.0%
Valve/CABG + Congenital	10,977	4.4%
Aortic + Congenital	1,375	4.7%
Valve/CABG + Aortic + Congenital	626	7.2%

N= 73,190; Isolated bicuspid aortic valve excluded

Ineligible Patients!



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		1.2%
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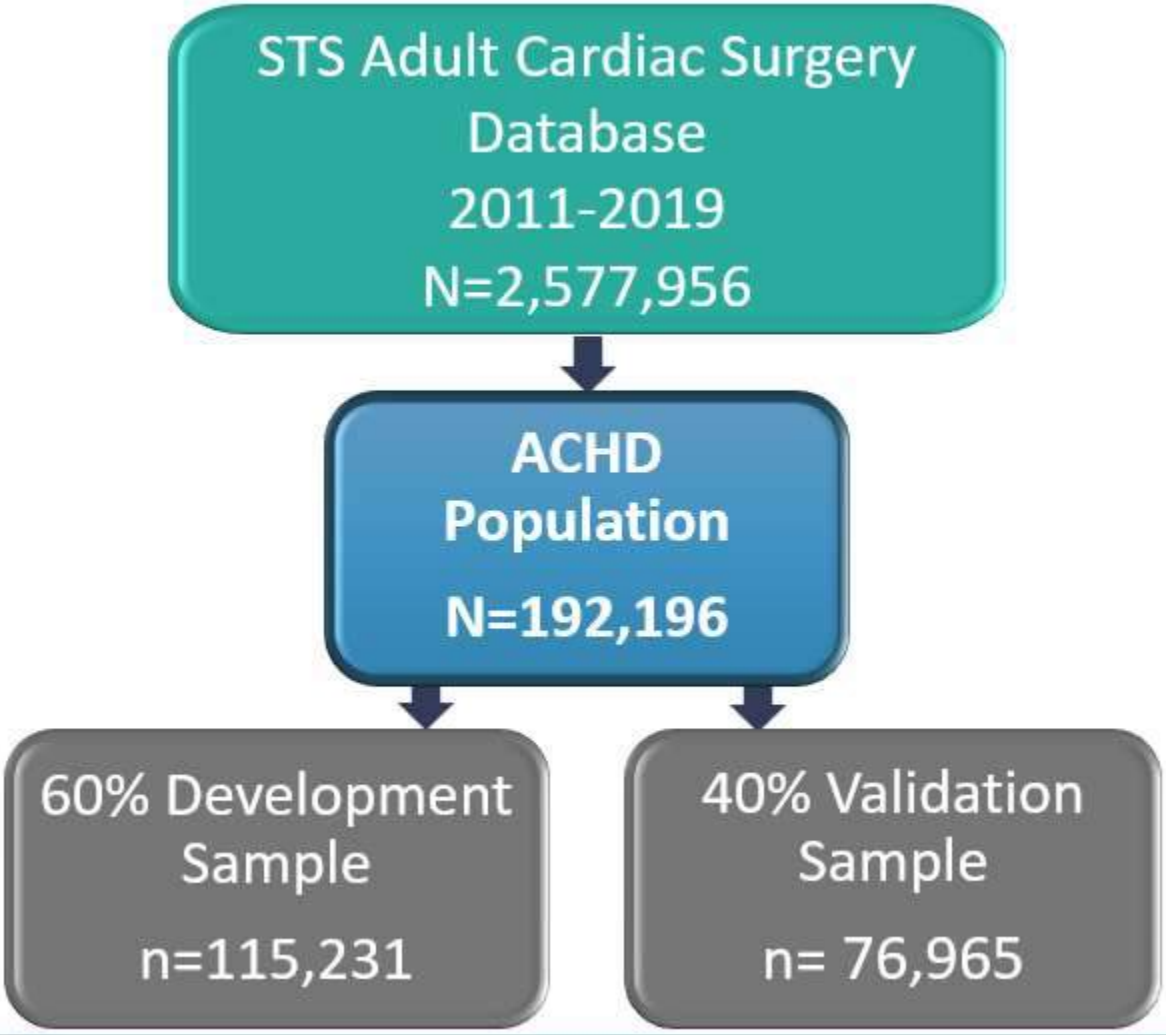
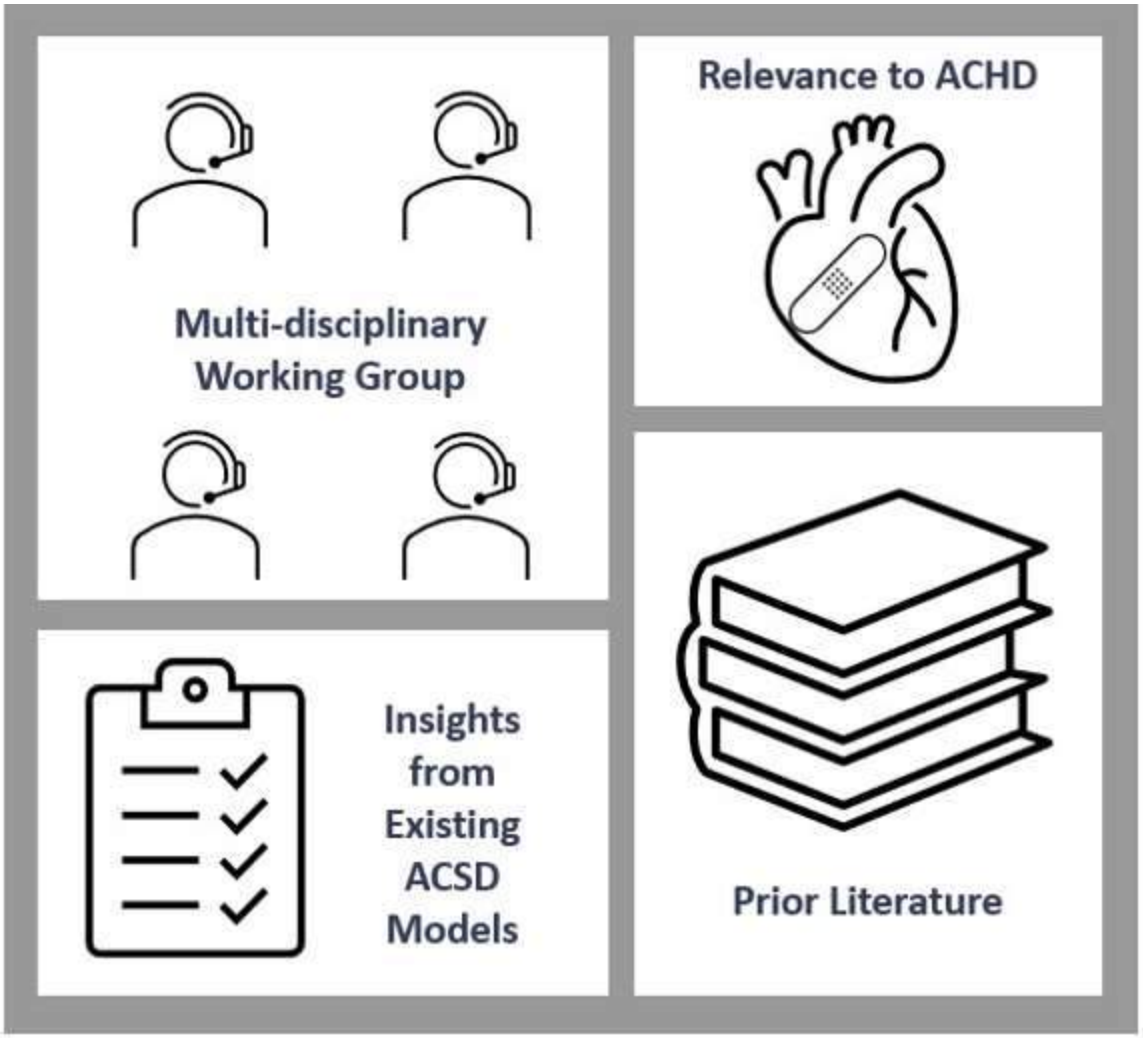

STS Adult Cardiac Surgery Database
2011-2019
N=2,577,956
ACHD Population
N=192,196
60% Development Sample
n=115,231
40% Validation Sample
n= 76,965

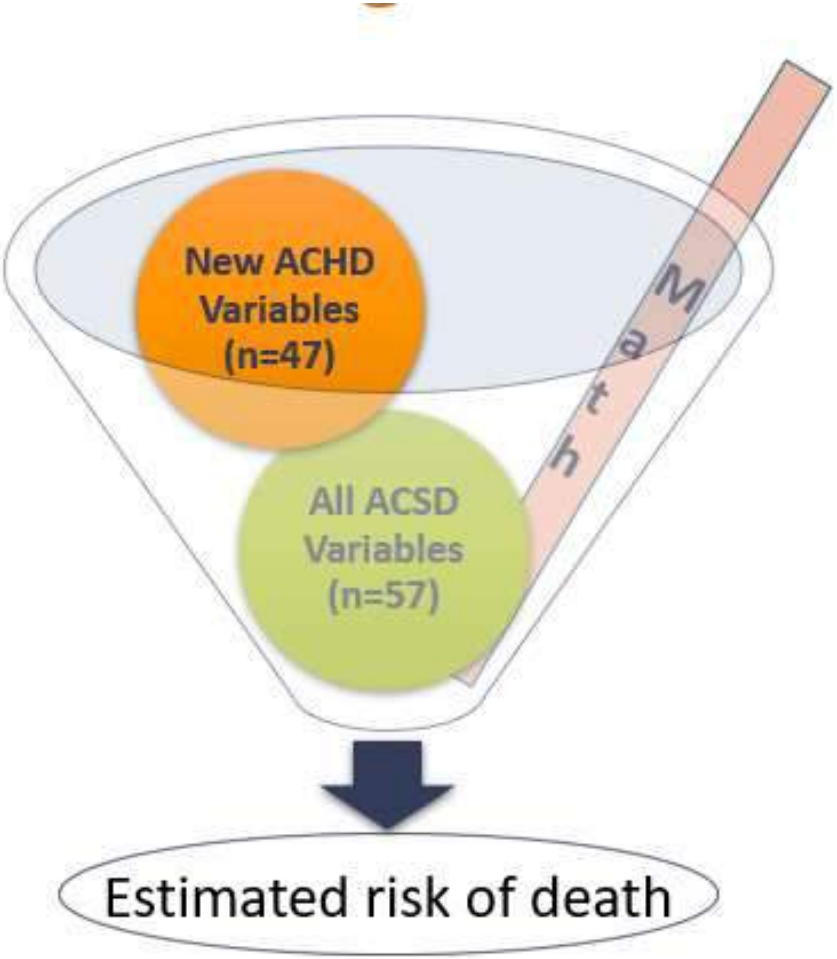
Model Development



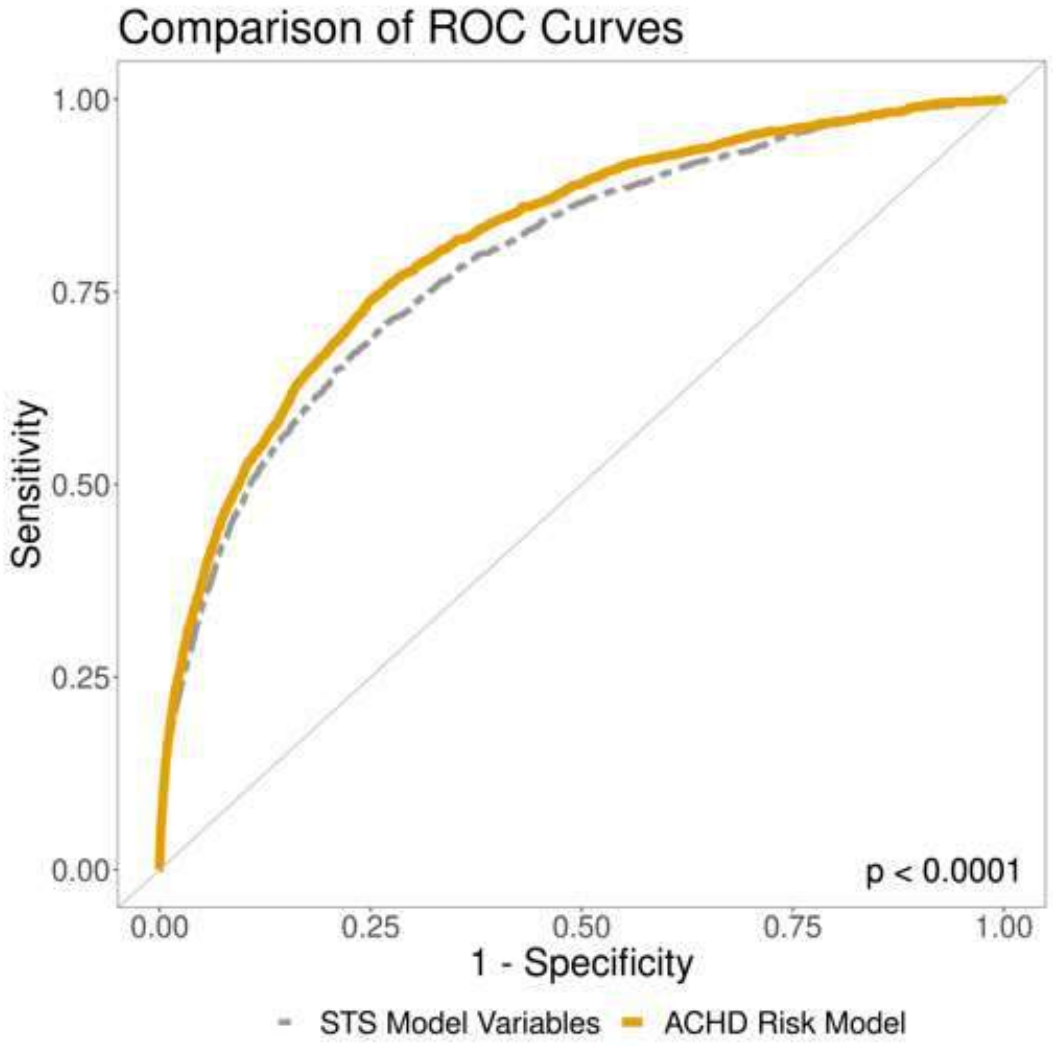
New Congenital Variables Selected

18 Diagnoses + 29 Procedures (n=47)

Final ACHD Risk Model

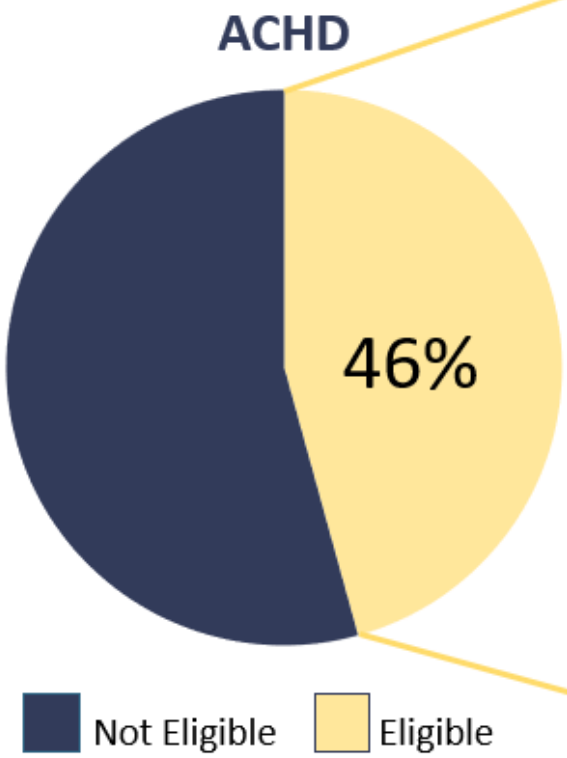


ACHD Model Performance



ACHD Model:
C-Statistic = 0.815
Vs.
STS Variables only:
C-statistic = 0.791
P < 0.0001

Model Comparison



STS Risk Model Procedure Group	ACHD Model AUC	STS Model AUC	P-value
CABG	0.7905	0.7945	0.74
AVR	**0.7616**	**0.7181**	**<0.0001**
MVR	0.7591	0.7487	0.26
AVR + CABG	0.7304	0.7145	0.05
MVR + CABG	0.7153	0.7091	0.78
MV repair	0.7763	0.7806	0.77
MV repair + CABG	0.7272	0.7187	0.79
AVR + MV repair	0.8256	0.7899	0.16
AVR + MVR	0.7776	0.7522	0.09
AVR + MV repair + CABG	0.7696	0.7724	0.93
AVR + MVR + CABG	0.6142	0.6317	0.61

Cleveland Clinic

Every life deserves world class care.