KYACC Meeting
September 10, 2022

Congenital Heart Surgery in Kentucky
A 30+ Year Retrospective

Erle H. Austin III, M.D.
Norton Children's Hospital
University of Louisville
Louisville, Kentucky

Congenital Heart Defects

Most common birth defect

8 per 1000 Live Births

40,000 infants per year in United States

3X the incidence of childhood malignancy

Congenital Heart Disease

1.3 million people in United States

Highest average hospital charges, length of stay, mortality of all birth defects

100,000 years of life lost each year

$6,000,000,000 in acute care costs per year

The Centers

Louisville

Kosair (now Norton) Children's Hospital

University of Louisville

Lexington

Kentucky Children's Hospital

Albert B Chandler Hospital

University of Kentucky

The Team

Success in managing patients with congenital heart disease requires a team

- Cardiologists
- Neonatologists
- Intensivists
- Surgeons
- Anesthesiologists
- Radiologists
- Nurses/ Nurse Practitioners
- Other Caregivers (perfusionists, echo techs, cath lab techs)

The Surgeons - pre-1980s to Present

Louisville

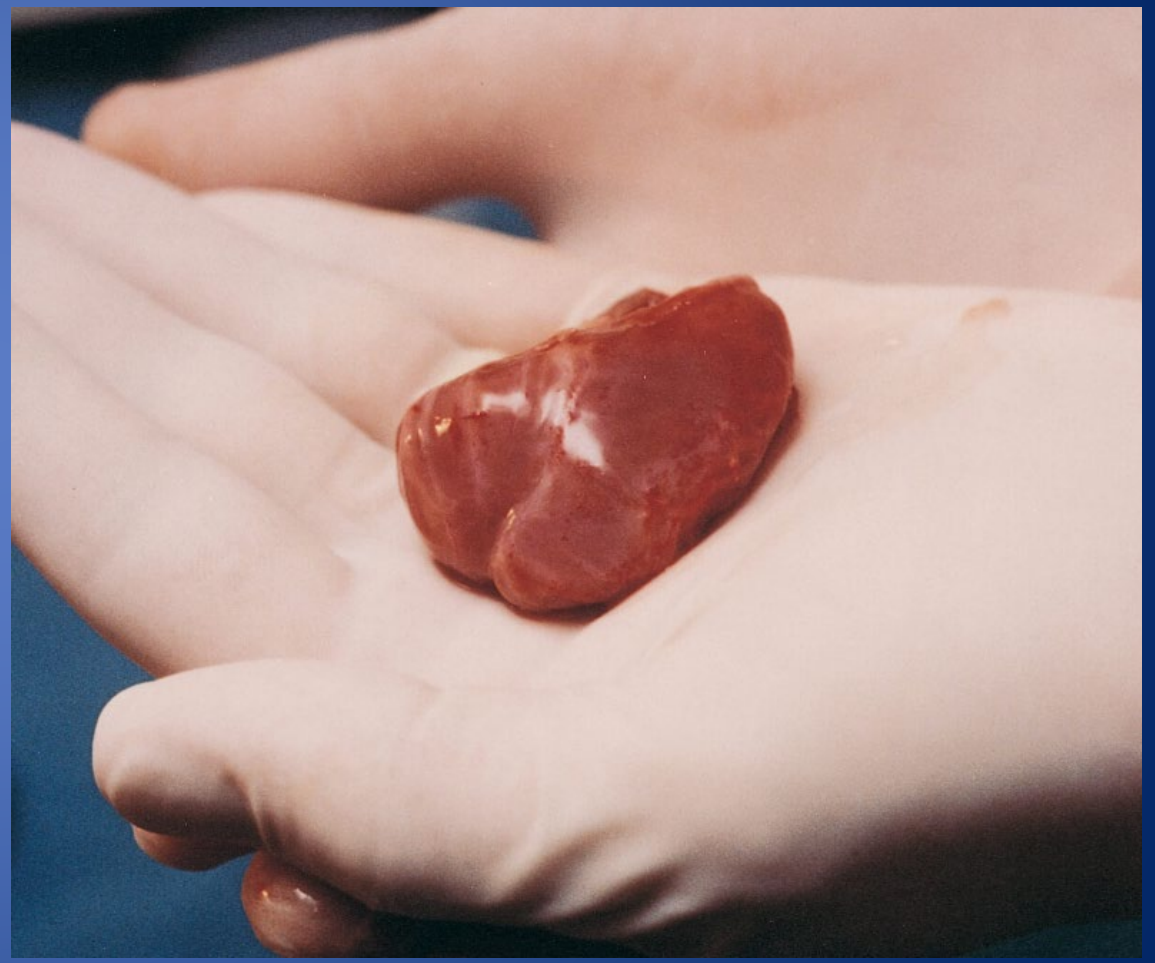
- Allen Lansing
- Constantine Mavroudis
- Erle Austin
- Thomas Yeh, Jr.
- Michael Mitchell
- Christopher Mascio
- Deborah Kozik
- Frank Pigula
- Bahaaldin Alsoufi

Lexington

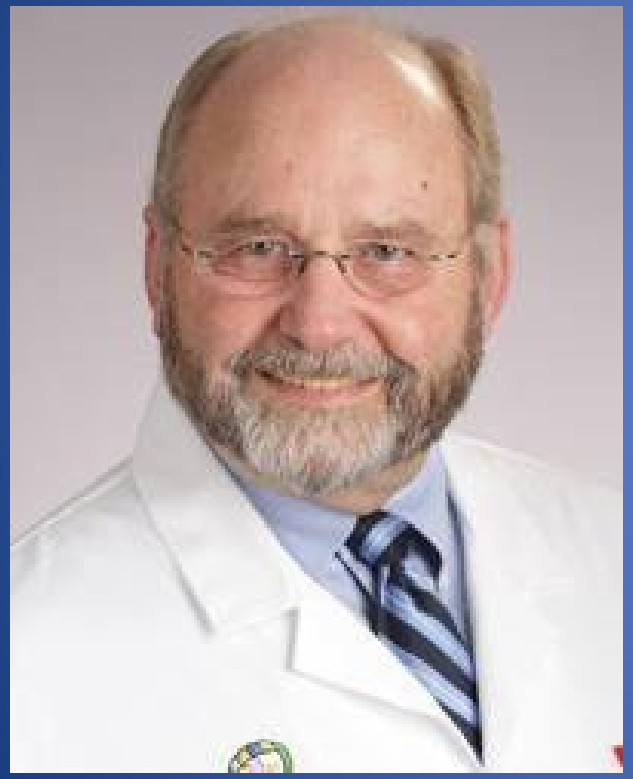
- Frank Spencer
- Gordon Danielson
- Edward Todd
- Robert Salley
- Simon Abraham
- William Douglas
- Mark Plunkett
- Deborah Kozik
- James Quintessenza
- Carl Backer

Second Neonatal Heart Transplant in the World

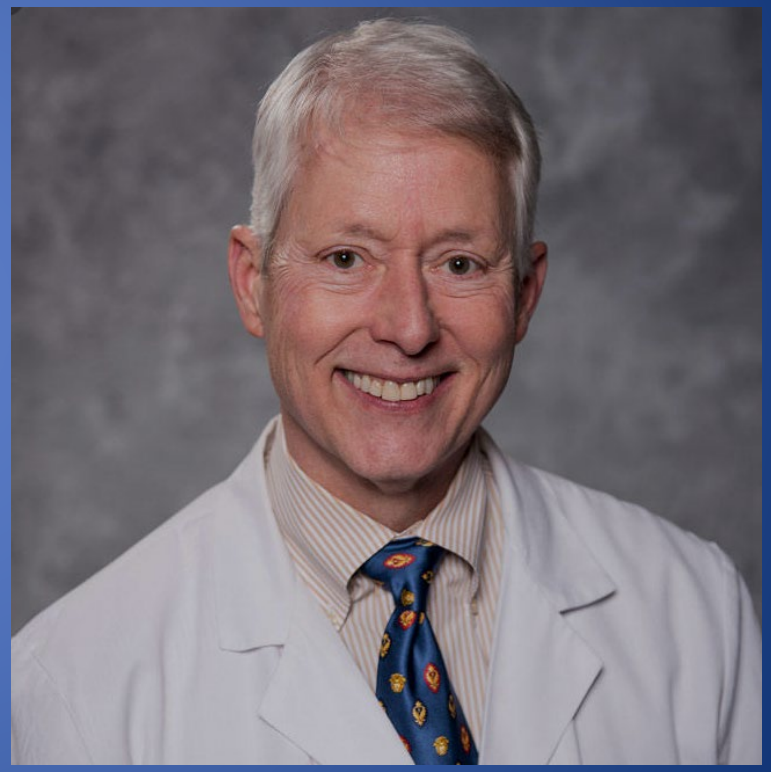
- 23 day old male with Hypoplastic Left Heart Syndrome
- Kosair Children's Hospital
- June 14, 1986
- Surgeon: Constantine Mavroudis, MD



Surgeons Performing Congenital Heart Surgery in Kentucky in 1989



Erle Austin



Robert Salley

My Retrospective View of Significant Advances in Congenital Heart Surgery Since Coming to Kentucky in 1989

Congenital Heart Surgery in Kentucky Has Kept Pace with Congenital Heart Surgery in the Rest of the World

The 1980s Perspective

ASDs and PDAs were closed by surgeons.

Transposition was repaired at the atrial level.

The 1980s Perspective

Single ventricles other than Tricuspid atresia?

What is Hypoplastic Left Heart Syndrome?

The 1980s Perspective

How are we doing compared to other centers?

When the heart stopped, the patient died.

The 1980s Perspective

Is the brain at risk during heart surgery?

Would an adult need surgery for a heart defect?

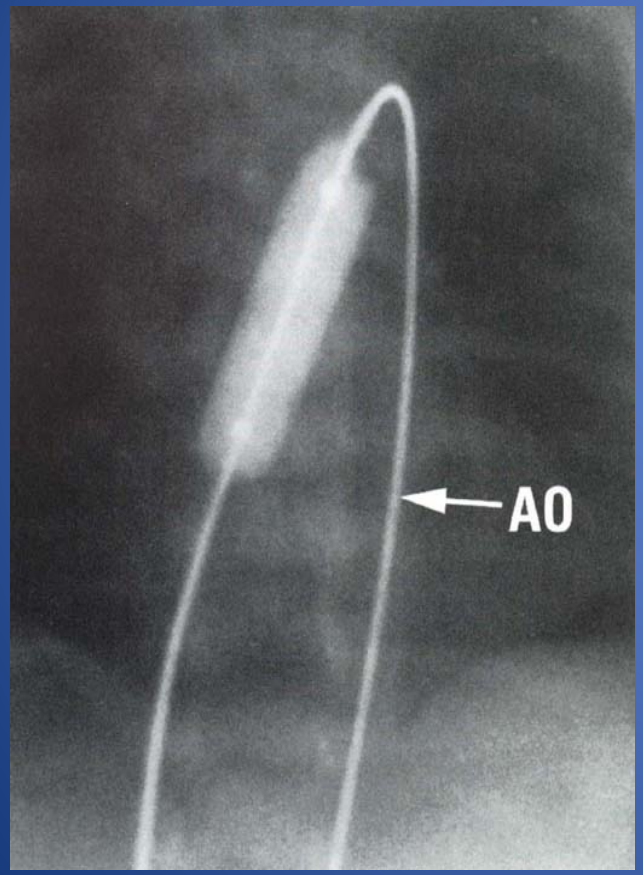
Some Important Advances in Last 30+ Years

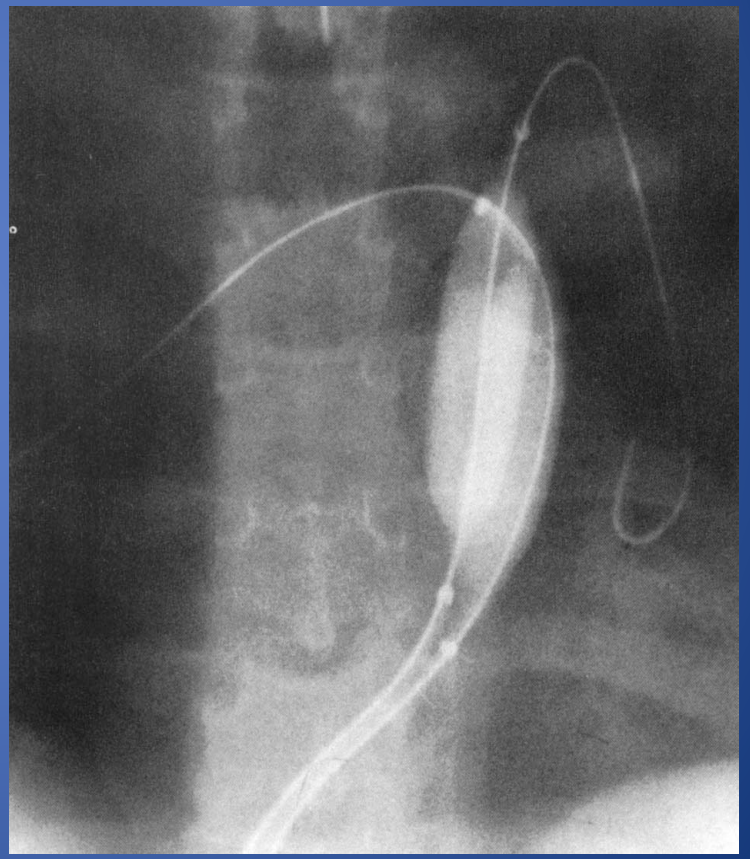
- Interventional Cardiology
- The Arterial Switch for Transposition
- Staged Reconstruction for Hearts with Only One Ventricle
- Intraoperative Neuromonitoring
- Self-evaluation and Databases
- Mechanical support for failing heart
- The Adult with Congenital Heart Disease

Percutaneous Interventions for Congenital Heart Disease

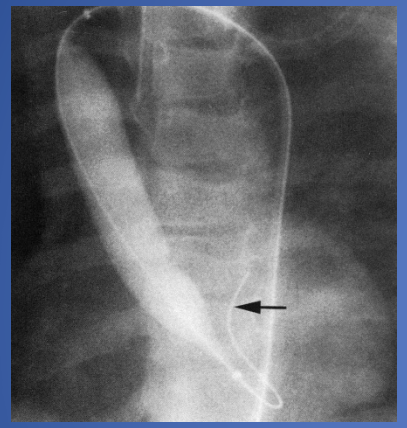
What happened to the easy cases?

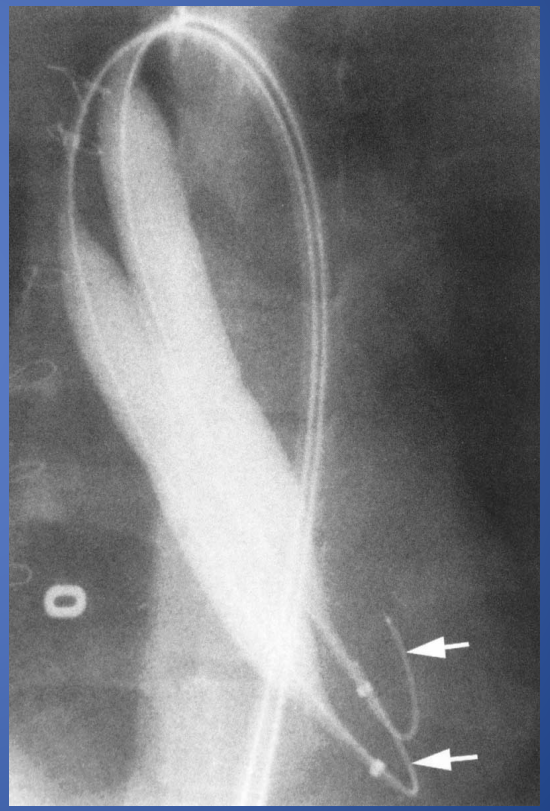
Pulmonary Valvulotomy



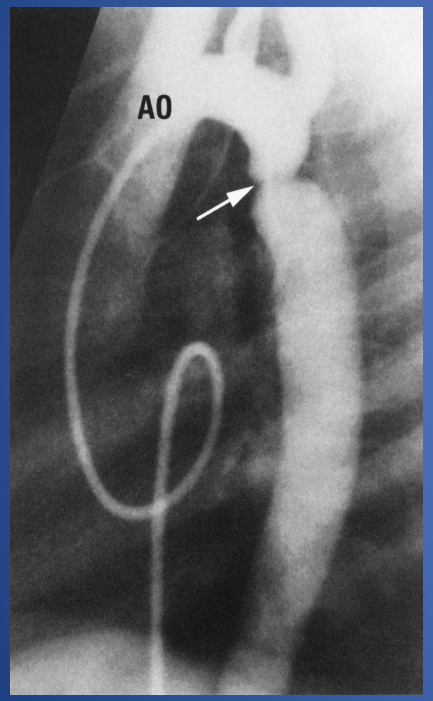


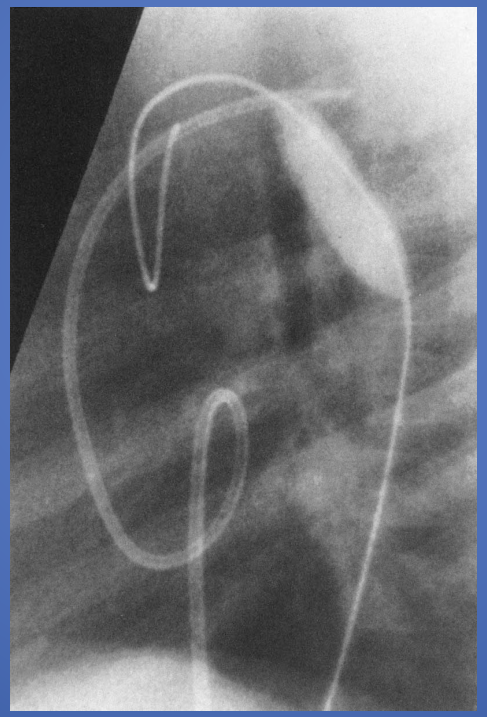
Aortic Valvulotomy

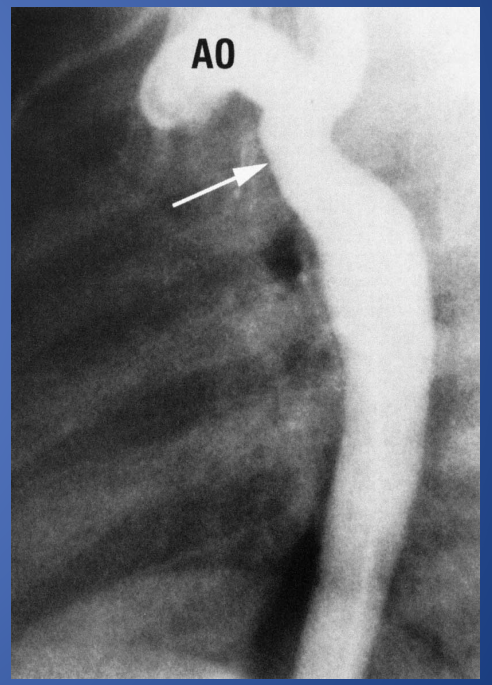




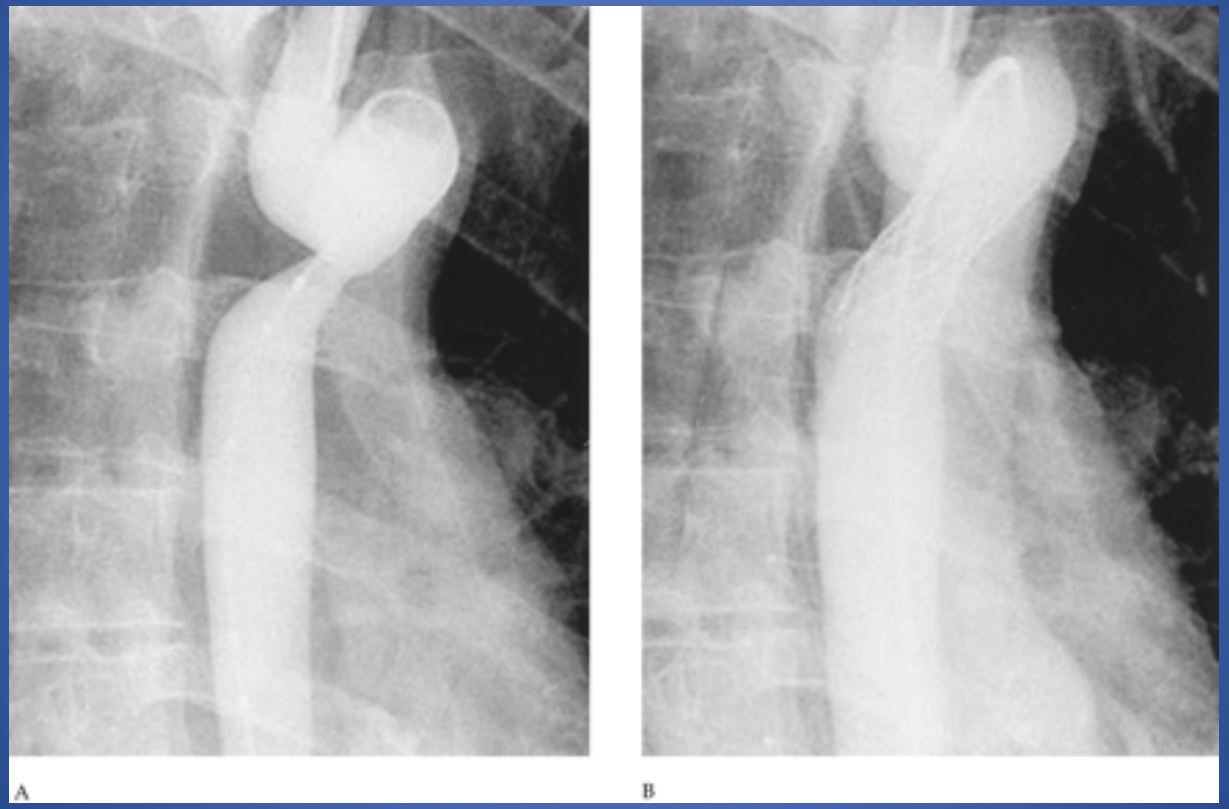
Balloon Dilation of Coarctation



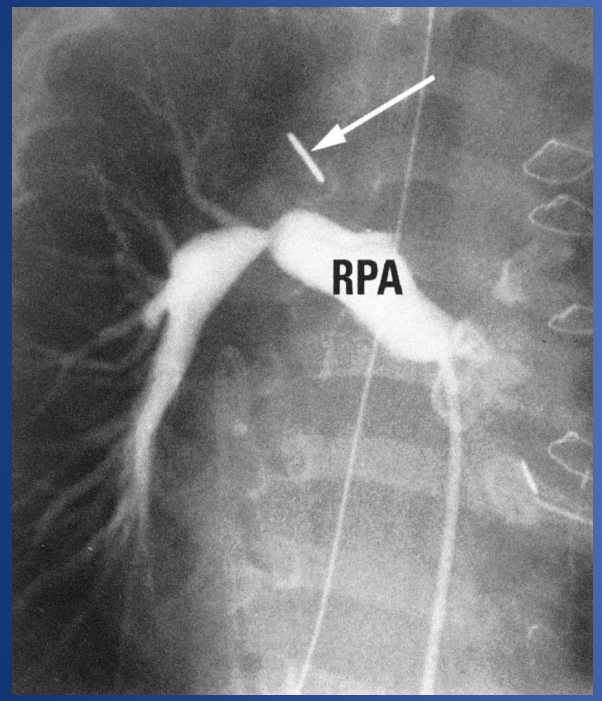


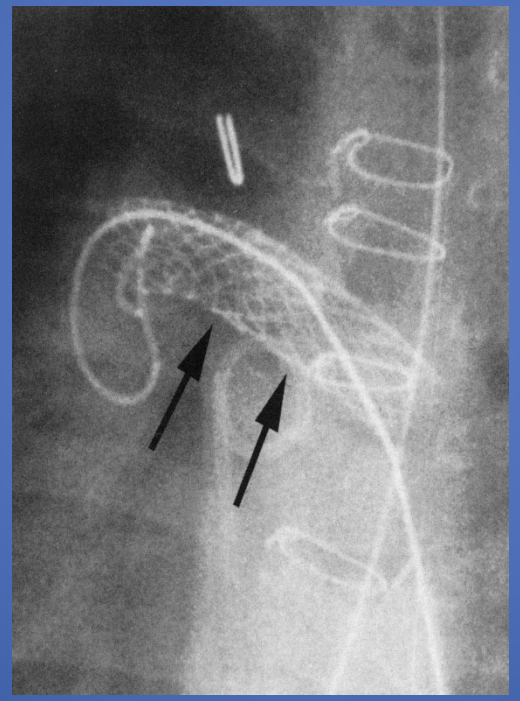


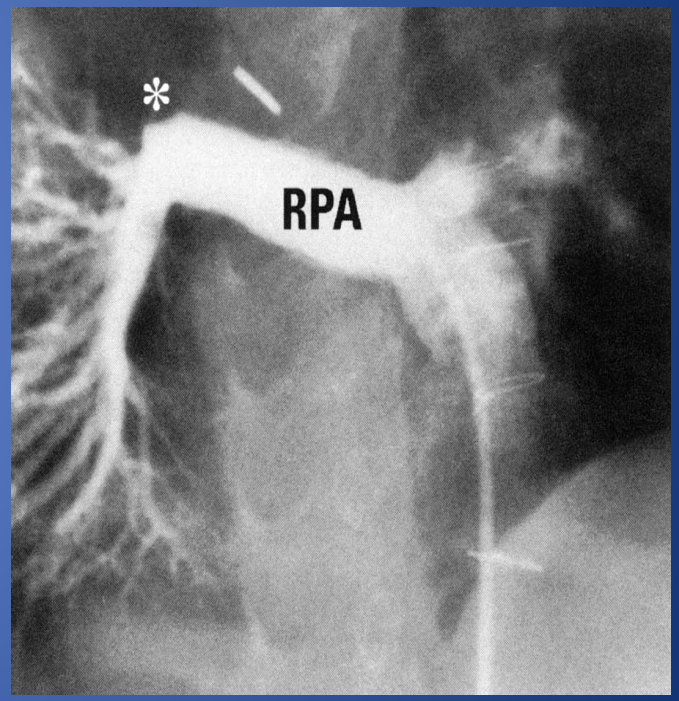
Stenting for Coarctation



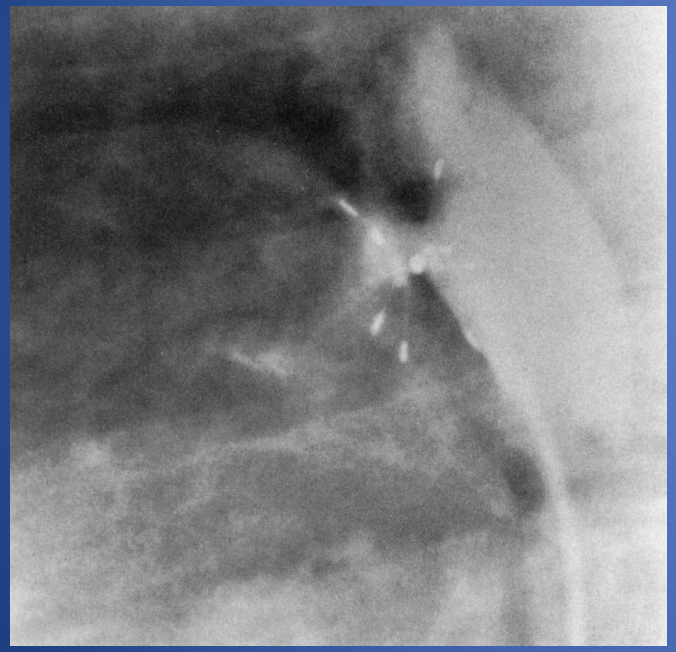
Balloon Dilation and Stenting of Branch Pulmonary Artery Stenosis

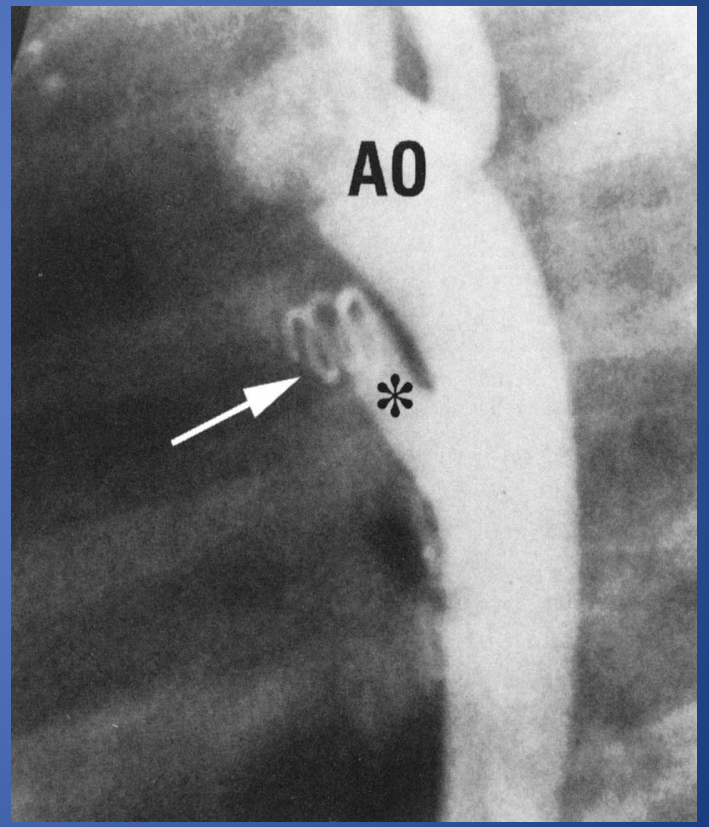




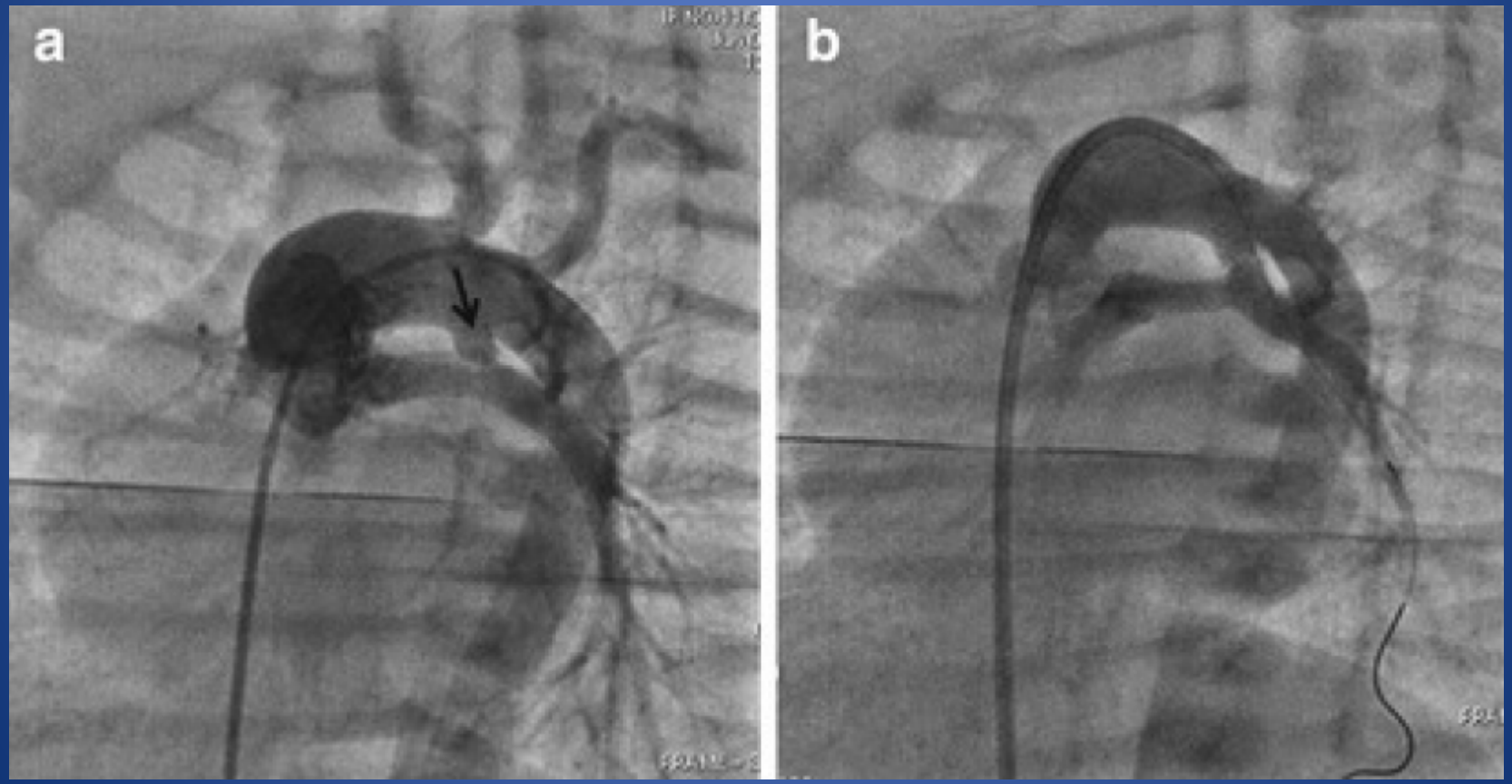


Device Closure of PDA

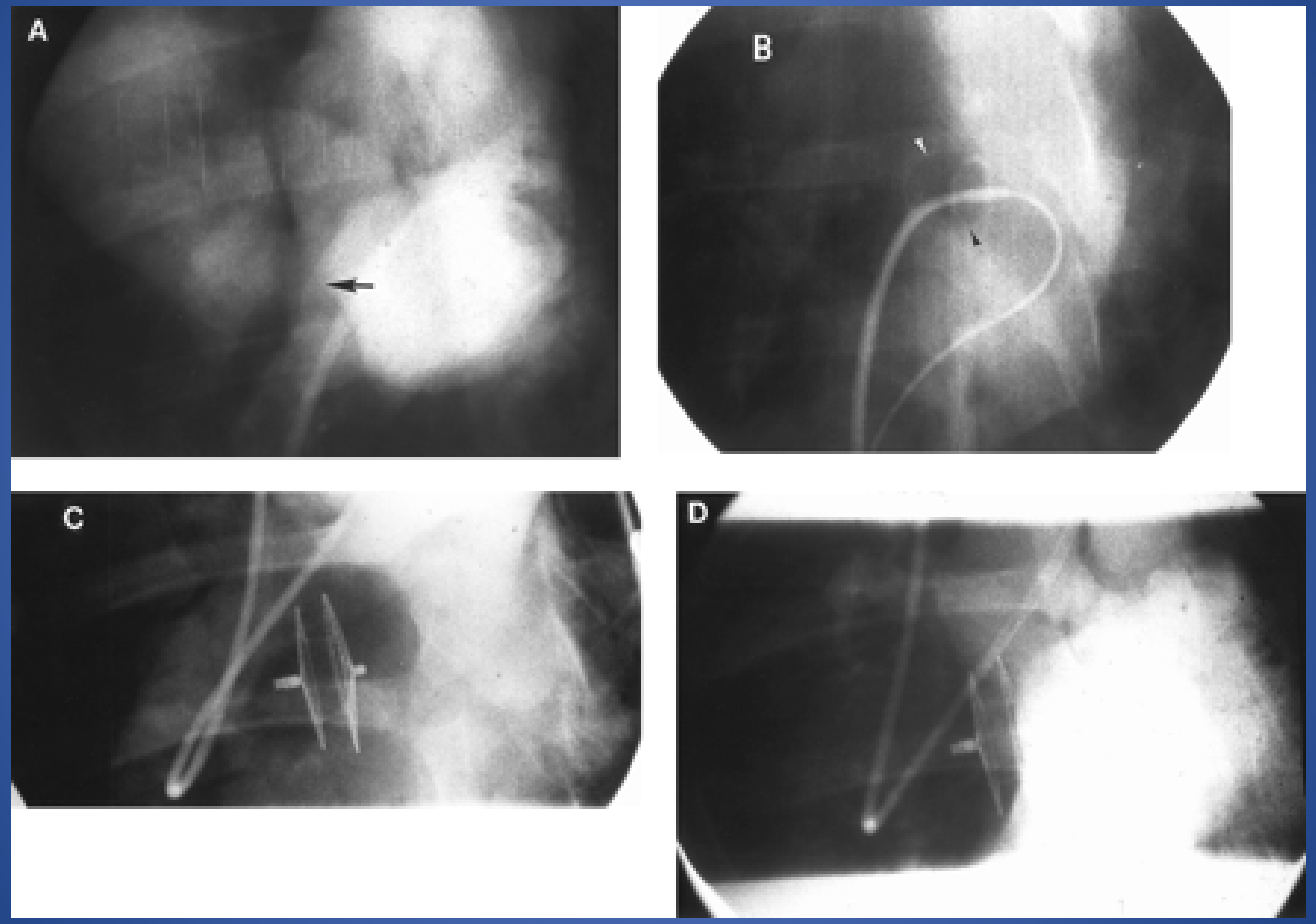




Stenting a PDA



Device Closure of ASD



Transcatheter Pulmonary Valve Replacement

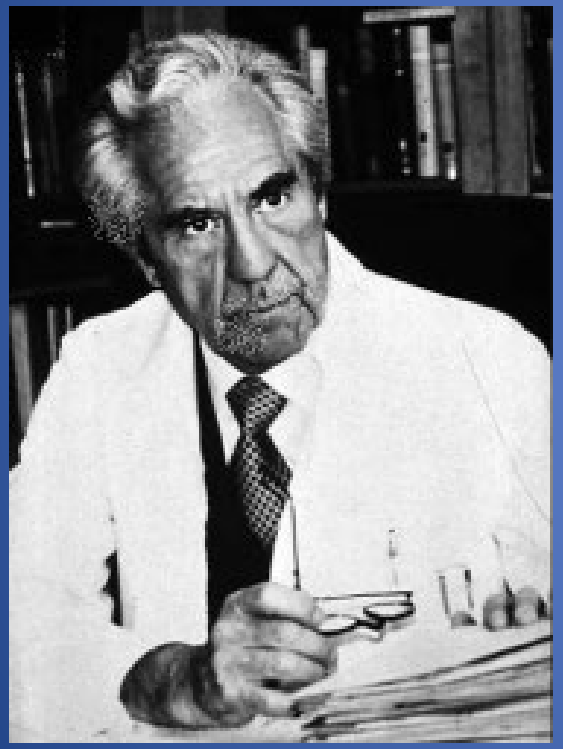
Medtronic Melody
Valve

Surgeon ⟷ Interventional Cardiologist

- Fewer easy cases for the surgeon
- Many difficult surgical cases more easily approached percutaneously
  - recurrent coarctation
  - peripheral PA stenoses
  - multiple collateral vessels
- Combined (hybrid) approaches and spirit of teamwork lead to improved patient outcomes, newer techniques and devices
  - Transcatheter Pulmonary Valve Replacement
  - Hybrid approach to Hypoplastic Left Heart Syndrome

Transposition of the Great Arteries

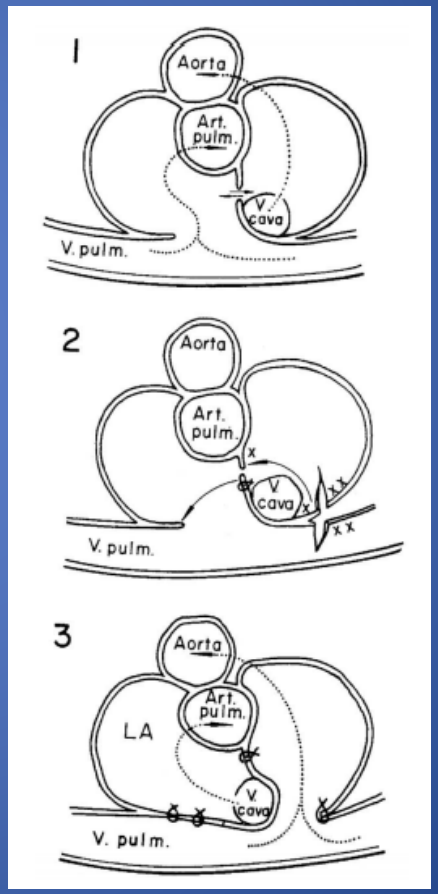
Transposition of the Great Arteries



Ake Senning

1957

The Senning Procedure



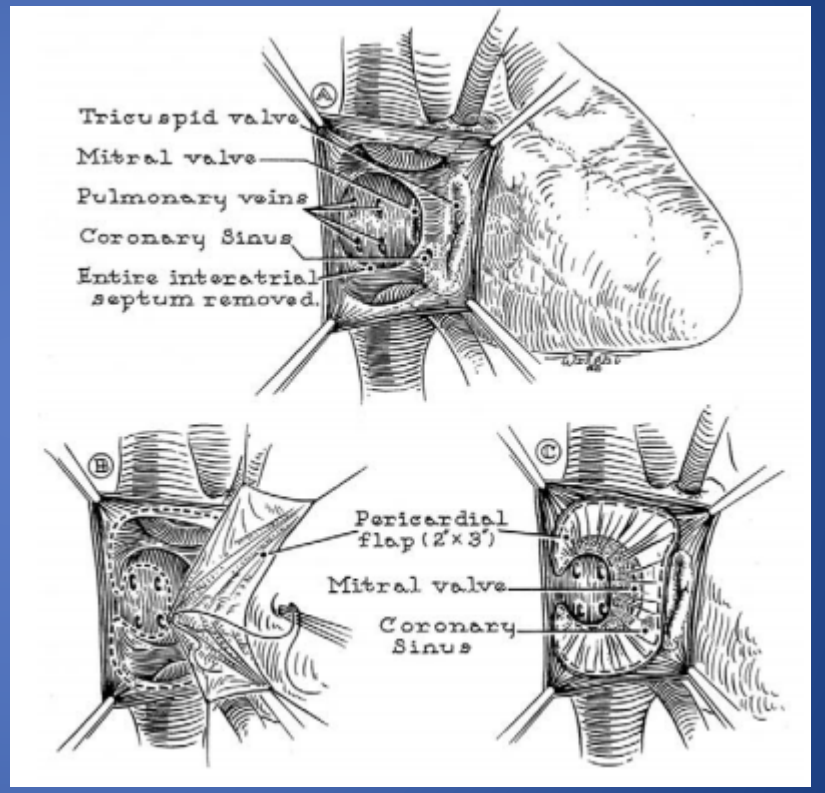
Senning A. Surgery 1959;45:966–80

Transposition of the Great Arteries

William T. Mustard

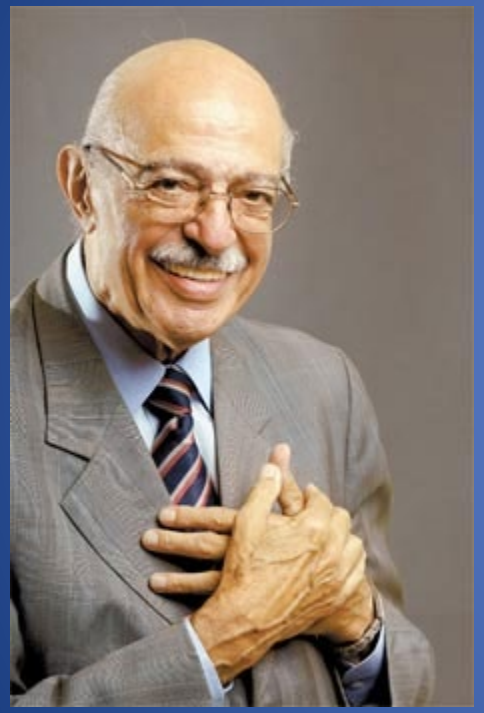
1963

The Mustard Procedure



Mustard WT, et al. JTCVS 1964.

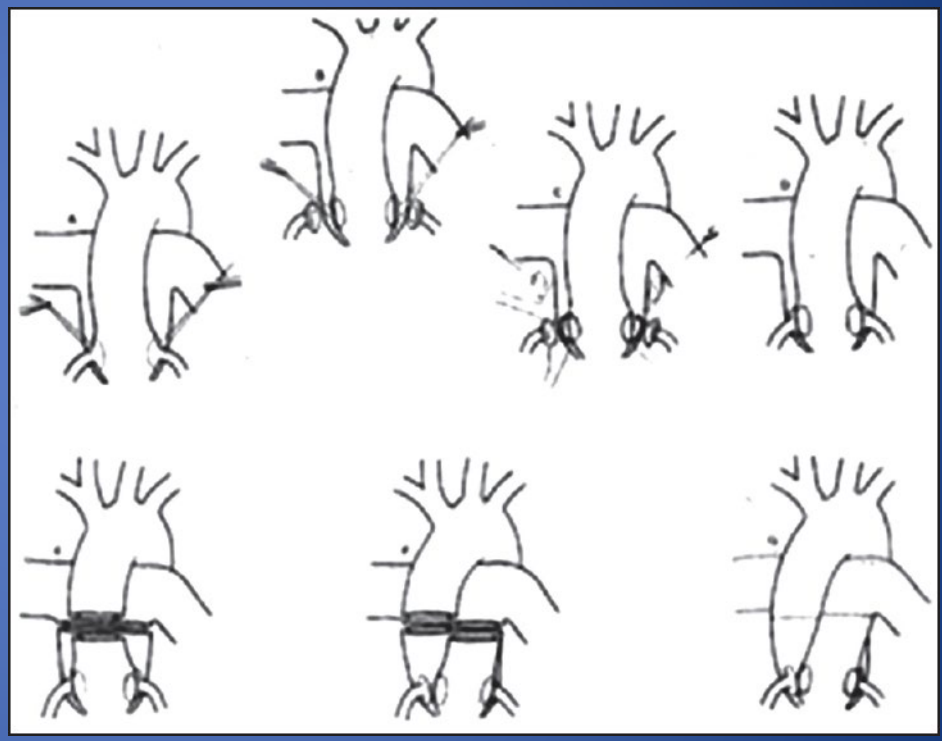
Transposition of the Great Arteries



Adib Jatene

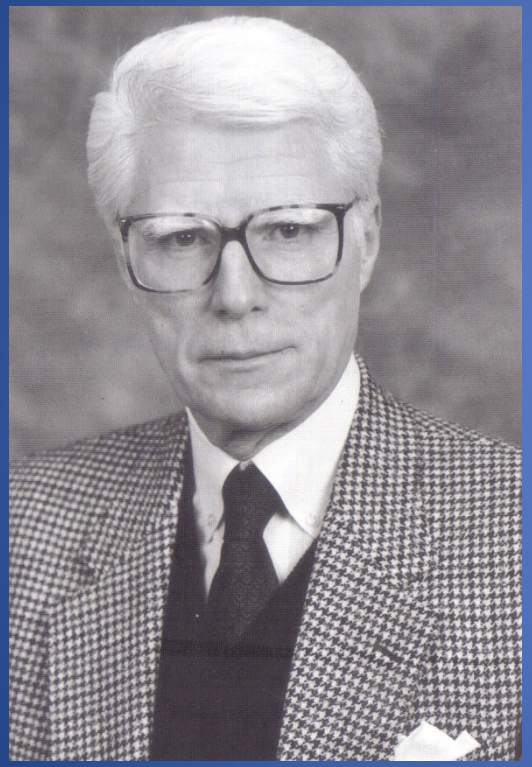
1975

The Jatene Procedure (Arterial Switch)



Jatene et al. Arq Bras Cardiol 1975

Arterial Switch in Newborn with Transposition and Intact Ventricular Septum



Aldo Casteneda

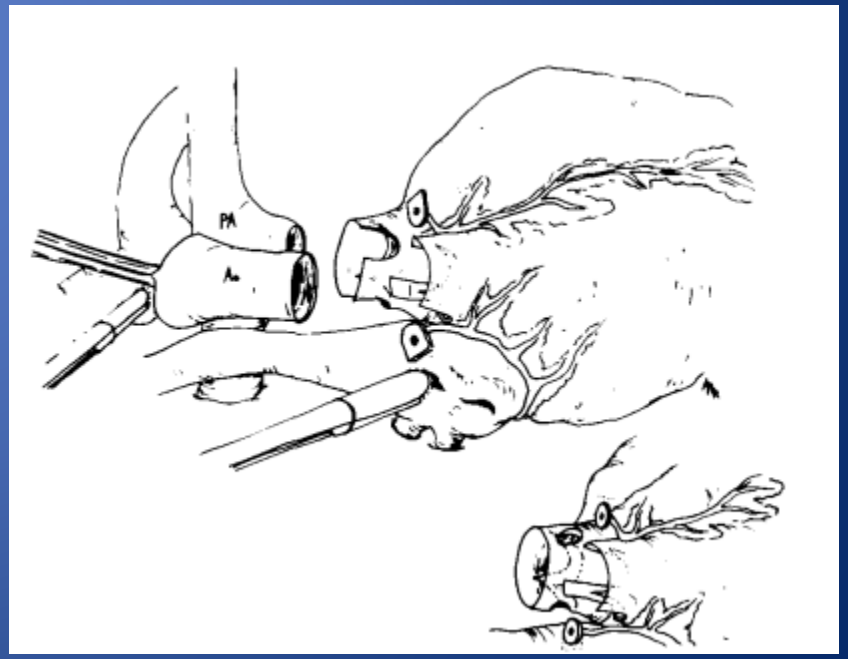
- January 2, 1983
- First arterial switch operation at Boston Children's Hospital
- 11 day old neonate with TGA/IVS

Anatomical Repair of Transposition of the Great Arteries with Intact Ventricular Septum in the Neonate: Guidelines to Avoid Complications

Constantine Mavroudis, M.D.

Ann Thorac Surg 43:495–501, May 1987

Jan, 1985 – Oct, 1986
16 consecutive neonates
2.4 – 4.2kg
1 to 6 days of life
14 of 16 (88%) survival
Kosair Children's/U of L



Current Results of Management in Transposition of the Great Arteries, With Special Emphasis on Patients With Associated Ventricular Septal Defect

GEORGE A. TRUSLER, MD,* ALDO R. CASTANEDA, MD, FACC,†
AMNON ROSENTHAL, MD, FACC,‡ EUGENE H. BLACKSTONE, MD, FACC,§
JOHN W. KIRKLIN, MD, FACC,§ and the CONGENITAL HEART SURGEONS SOCIETY

Toronto, Ontario, Canada, Boston, Massachusetts, Ann Arbor, Michigan and Birmingham, Alabama

Two hundred forty-five patients <15 days of age with transposition of the great arteries with or without a ventricular septal defect or pulmonary stenosis were entered into an ongoing 20 institution treatment study between January 1, 1985 and June 1, 1986. Complete follow-up is available on all patients. The ventricular septal defect narrowed in only 1 of 36 patients with combined transposition of the great arteries and ventricular septal defect; pulmonary stenosis developed or worsened in 3 of these 36 patients and in 3 of the 187 patients with simple transposition. Twelve month overall survival among the 245 patients was 80%. No morphologic feature of transposition was a risk factor for death but major associated cardiac and noncardiac anomalies (more common in patients with combined transposition and ventricular septal defect) and low birth weight were risk factors.

Neither arterial switch repair (n = 86), atrial switch (Mustard) repair (n = 21) nor atrial switch (Senning) repair (n = 39) was a risk factor for death, but results in all surgical groups were better in the last part of the experience. Death before repair was less frequent late in the study. Possibly, in low birth weight infants, survival was better with the arterial than with the atrial switch repair.

These data suggest that survival at 1 year is similar with either the arterial or the atrial switch repair. The early results of repair of combined transposition of the great arteries and ventricular septal defect are as good as those of simple transposition. Special institutional efforts are required to attain good results with the arterial switch repair and to prevent death before repair.

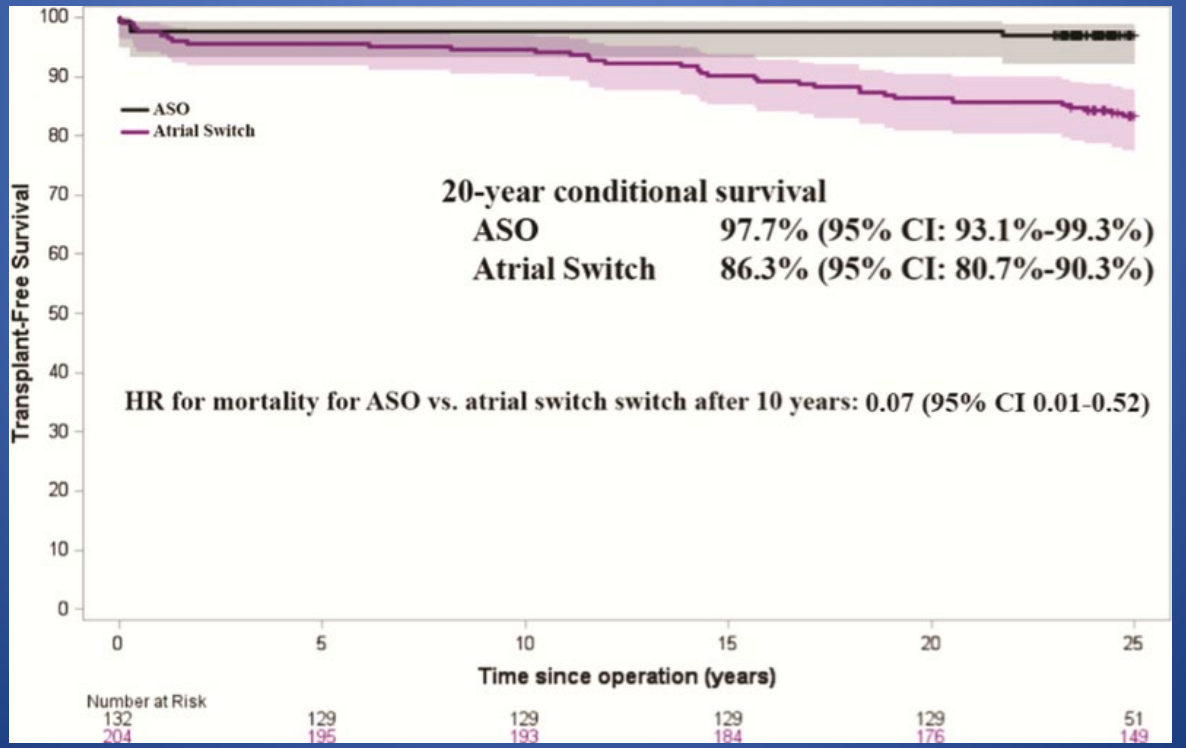
(J Am Coll Cardiol 1987;10:1061–71)

Long-Term Survival After Arterial Versus Atrial Switch in d-Transposition of the Great Arteries

Alexander Kiener, MD, MPH, Michael Kelleman, MSPH, Courtney McCracken, PhD, Lazaros Kochilas, MD, MSCR, James D. St. Louis, MD, and Matthew E. Oster, MD, MPH

Department of Pediatrics, Emory University School of Medicine, Atlanta, Georgia; Emory University Rollins School of Public Health, Atlanta, Georgia; Children's Healthcare of Atlanta, Atlanta, Georgia; and Department of Pediatric Surgery, University of Missouri-Kansas City School of Medicine, Kansas City, Missouri

[Ann Thorac Surg 2018;106:1827–33]



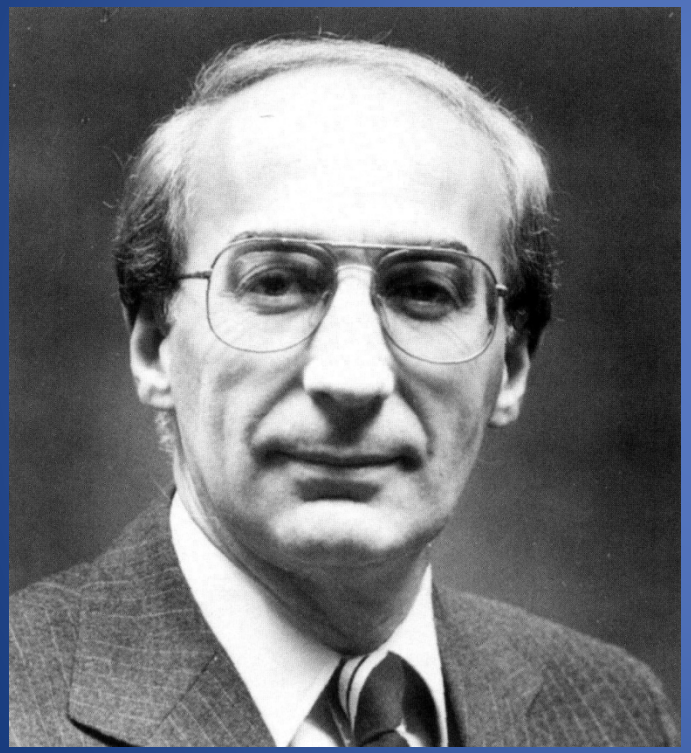
Single Ventricles

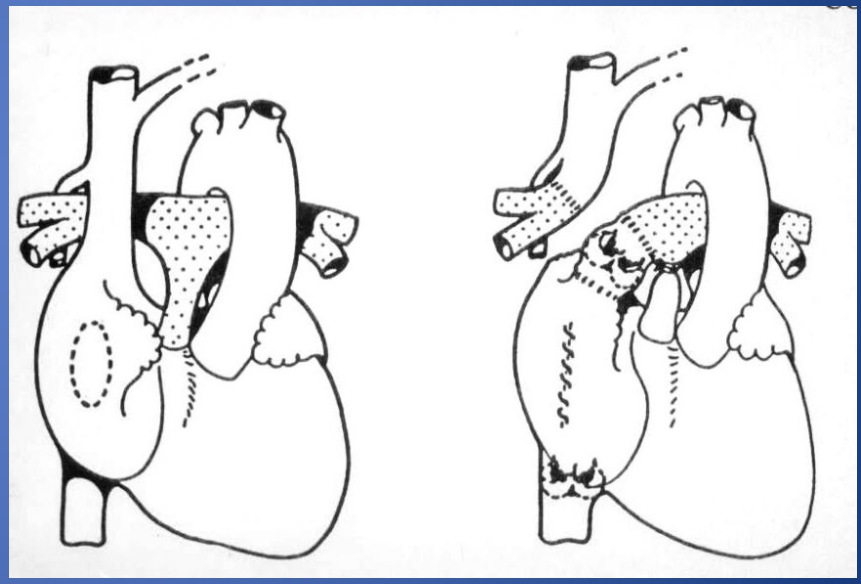
Staged Reconstruction for Univentricular Hearts

- Tricuspid atresia
- Unbalanced AV Canal
- Large VSD with Straddling AV Valves
- Double Inlet Left Ventricle
- Double Inlet Right Ventricle
- Some forms of Double Outlet Right Ventricle
- Pulmonary atresia with intact ventricular septum and inadequate right ventricle
- Mitral Atresia
- Hypoplastic Left Heart Syndrome

Fontan Operation

1968 – Tricuspid Atresia





Fontan and Baudet, Thorax 26:240, 1971

Evolutions in the Fontan Procedure

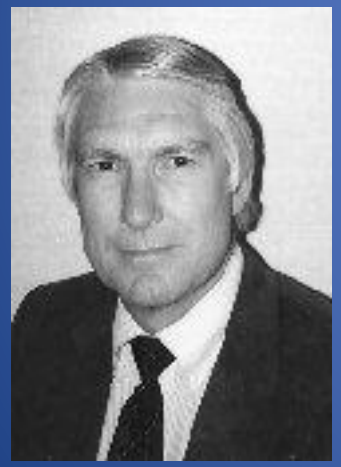
- Applications
  - From Tricuspid Atresia
  - To All forms of Single Ventricle
- Techniques
  - From Unidirectional Cavopulmonary Anastomosis (Classical Glenn procedure)
  - To Extracardiac Conduit Fontan Procedure

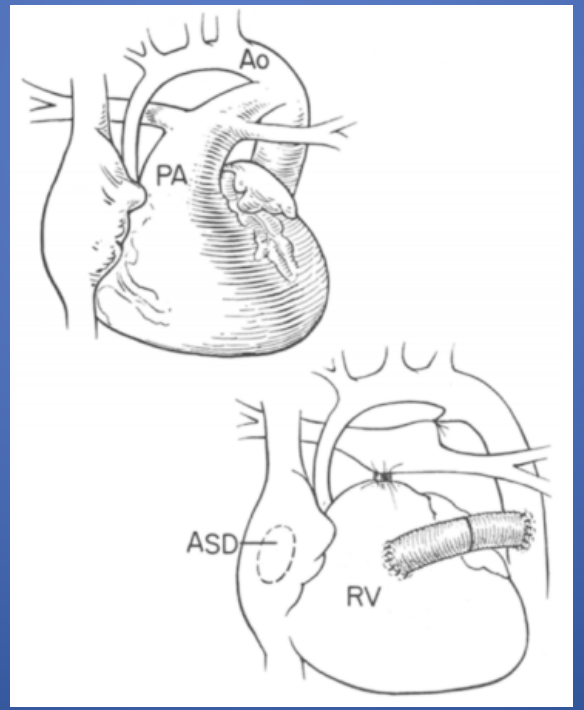
Hypoplastic Left Heart Syndrome: Experience With Palliative Surgery

WILLIAM I. NORWOOD, MD, FACC*
JAMES K. KIRKLIN, MD*
STEPHEN P. SANDERS, MD†

Boston, Massachusetts

Aortic atresia is a form of hypoplastic left heart syndrome always complicated by associated severe hypoplasia of the ascending aorta and various degrees of mitral valve and left ventricular hypoplasia. At present it is a universally fatal lesion in early infancy. This is a report of a new palliative procedure for hypoplastic left heart syndrome that has resulted in early ongoing survival of two infants with aortic atresia. On the basis of experience with a third patient, an operation for future physiologic correction is proposed.





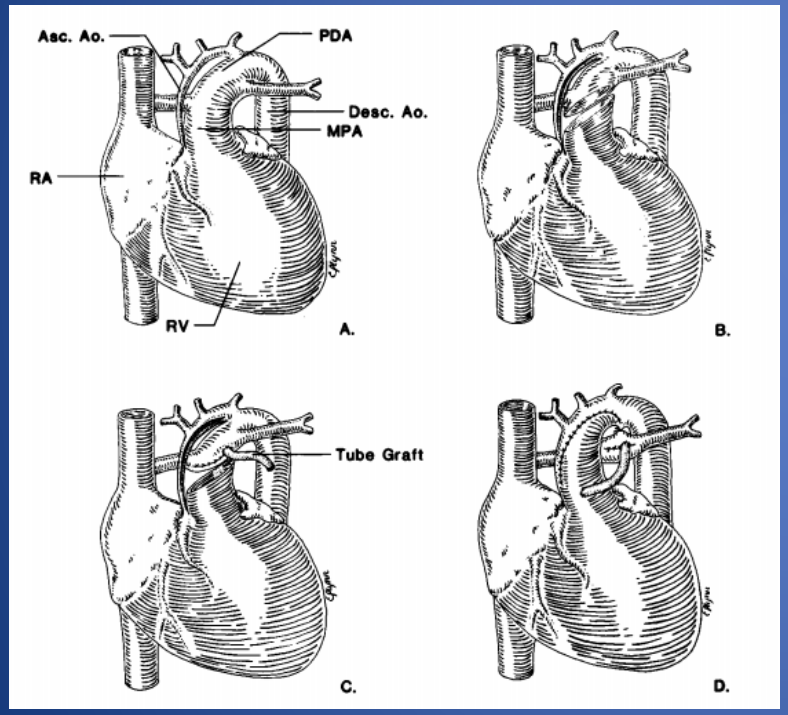
2/6/1979

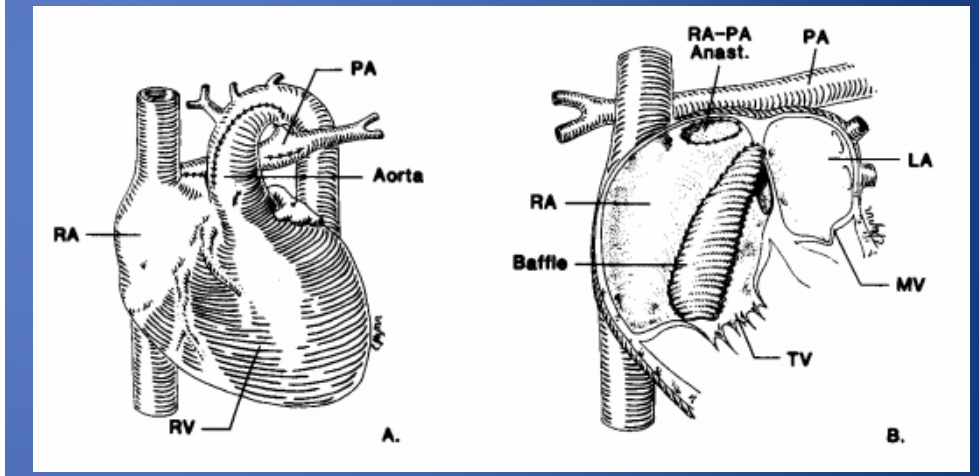
Norwood, Kirklin, Sanders Am J Cardiol 1980

PHYSIOLOGIC REPAIR OF AORTIC ATRESIA–HYPOPLASTIC LEFT HEART SYNDROME

WILLIAM I. NORWOOD, M.D., PETER LANG, M.D., AND DOLLY D. HANSEN, M.D.

N Engl J Med 1983

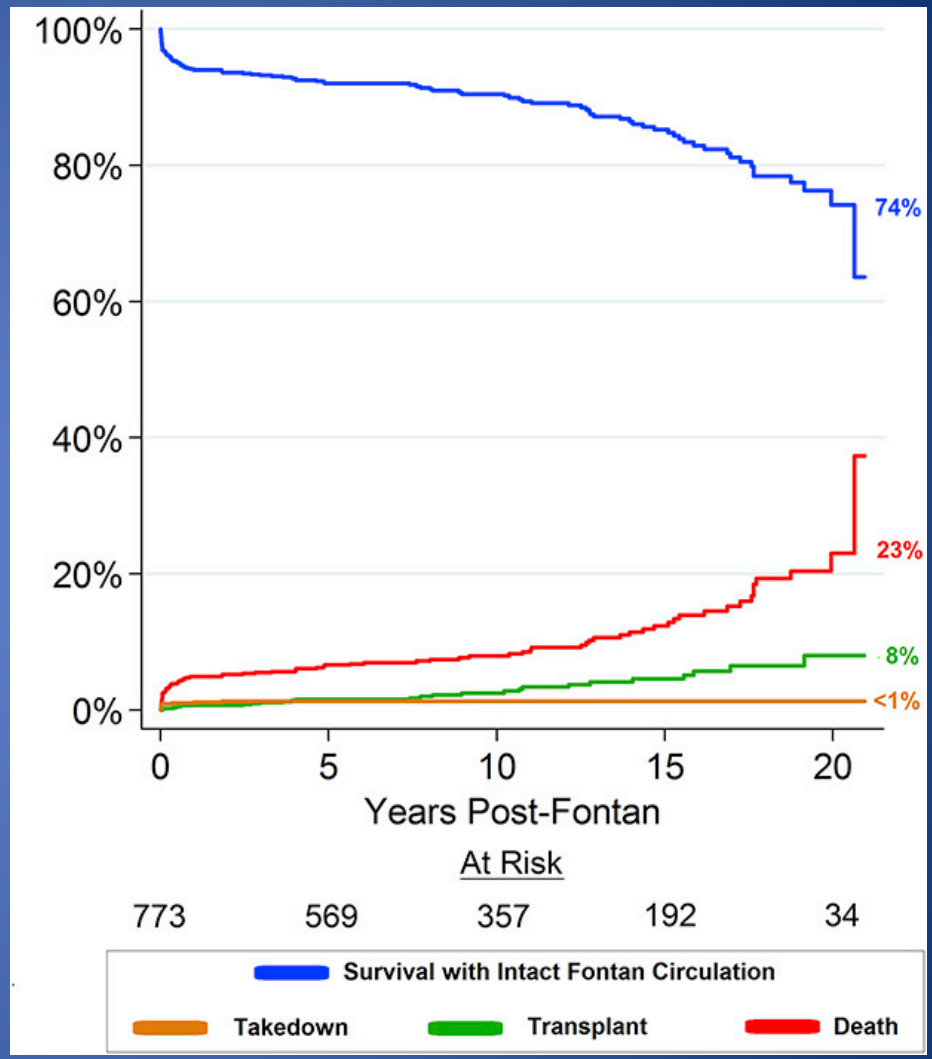




Important Surgical Advances in Single Ventricle Reconstruction

- Bidirectional Glenn 1985
  - –Hopkins/Oldham/Lamberti
- Lateral tunnel
  - –Puga; Jonas; de Leval 1987
- Extracardiac conduit
  - –Marcelletti 1990
- RV-PA conduit for Stage 1
  - –Sano 1998

Two ventricles are still better than one



Downing et al. JTCVS 2017

Intraoperative Neuromonitoring

Intraoperative Neuromonitoring

- Pediatric cardiac surgery

  – Improved survival begs improved quality of life

  Neurologic outcome of primary importance!

  – Neurologic sequelae
    - Up to 25% of patients.

Ultimate Neurologic Outcome

- May reflect pre-existing condition
- May reflect other events during hospitalization
- Period of cardiopulmonary bypass (w/wo CA)
  - **Critical period during which the brain is in great jeopardy**
  - **Surgeon's control and responsibility**

Background

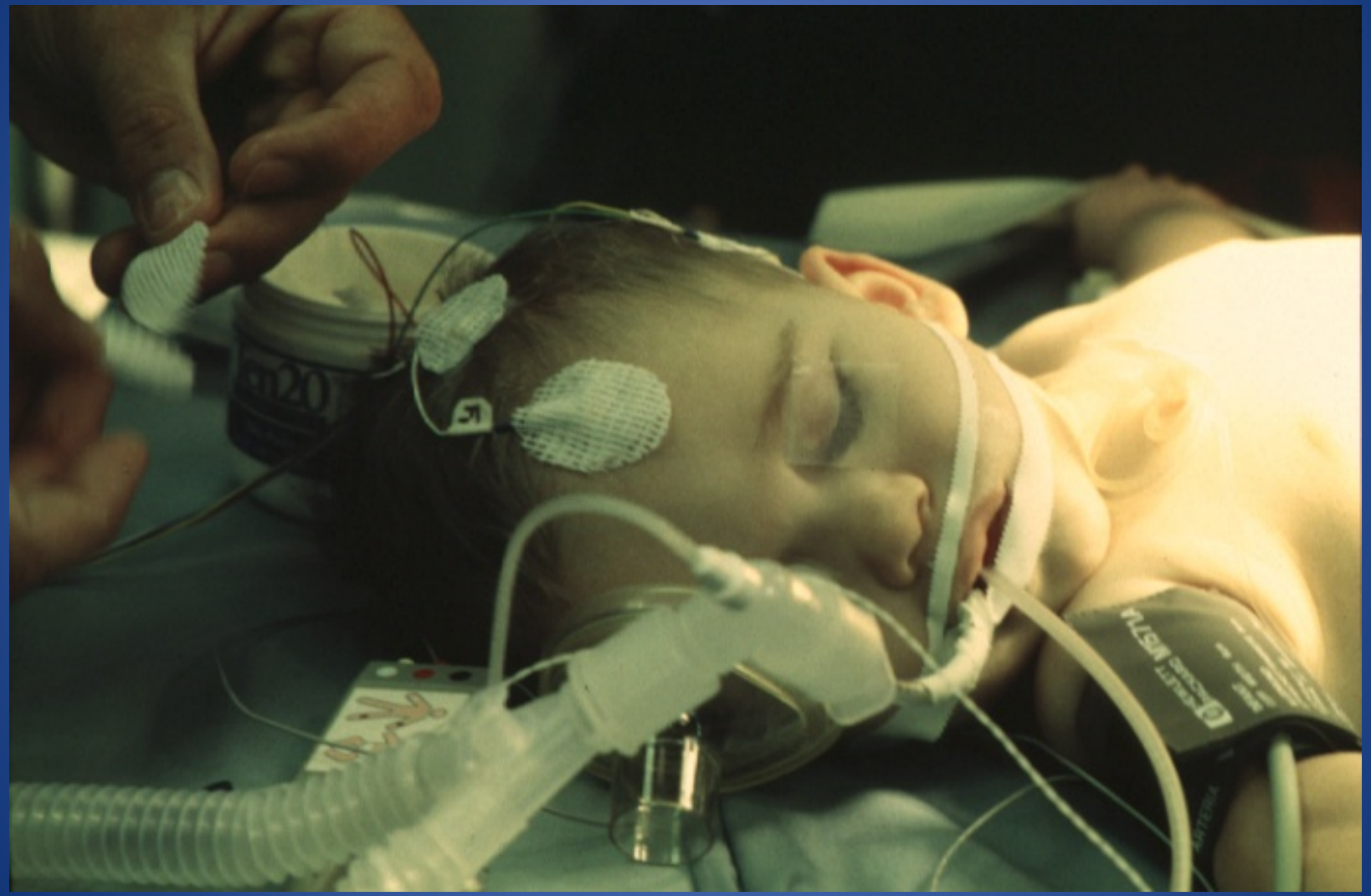
- In 1995 our group began to routinely monitor all patients during pediatric cardiac surgery using all three neuromonitoring modalities
  - EEG (4 channel)
  - Transcranial Doppler
  - Transcranial near-infrared spectroscopy

- Began as an observational study, but in short time began to apply interventions

Multimodality Neuromonitoring

- EEG – demonstrates neural function
  - –Affected by cooling, anesthesia, artifact
- TCD – demonstrates presence of and changes to blood flow to the brain
- NIRS – demonstrates oxygen availability to brain tissue

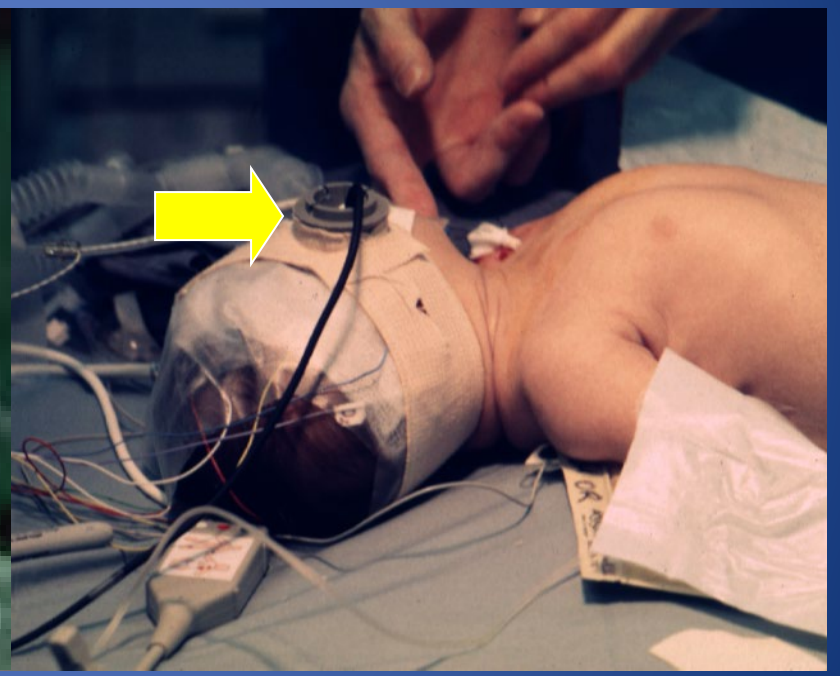
Disk electrodes use conductive paste.
Self-adhesive disks only outside hairline.



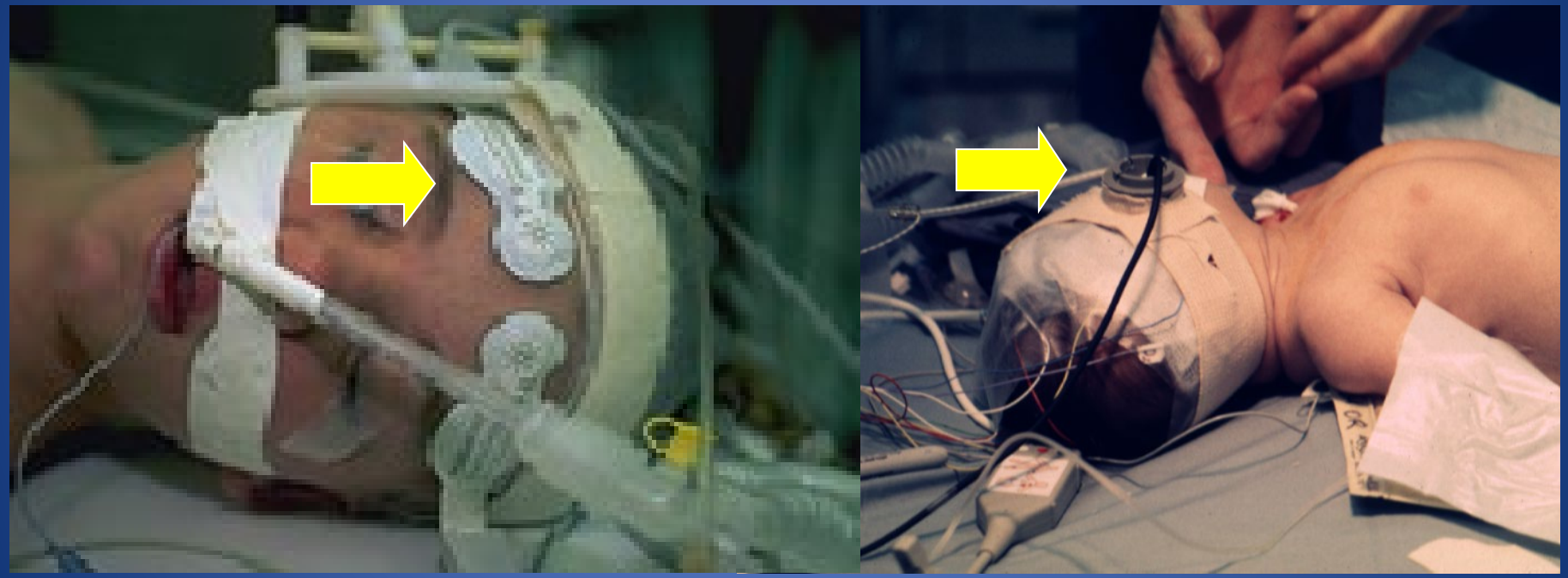
TCD Probe Fixation

Bisonette plastic wheel

Soft flexible plastic socket

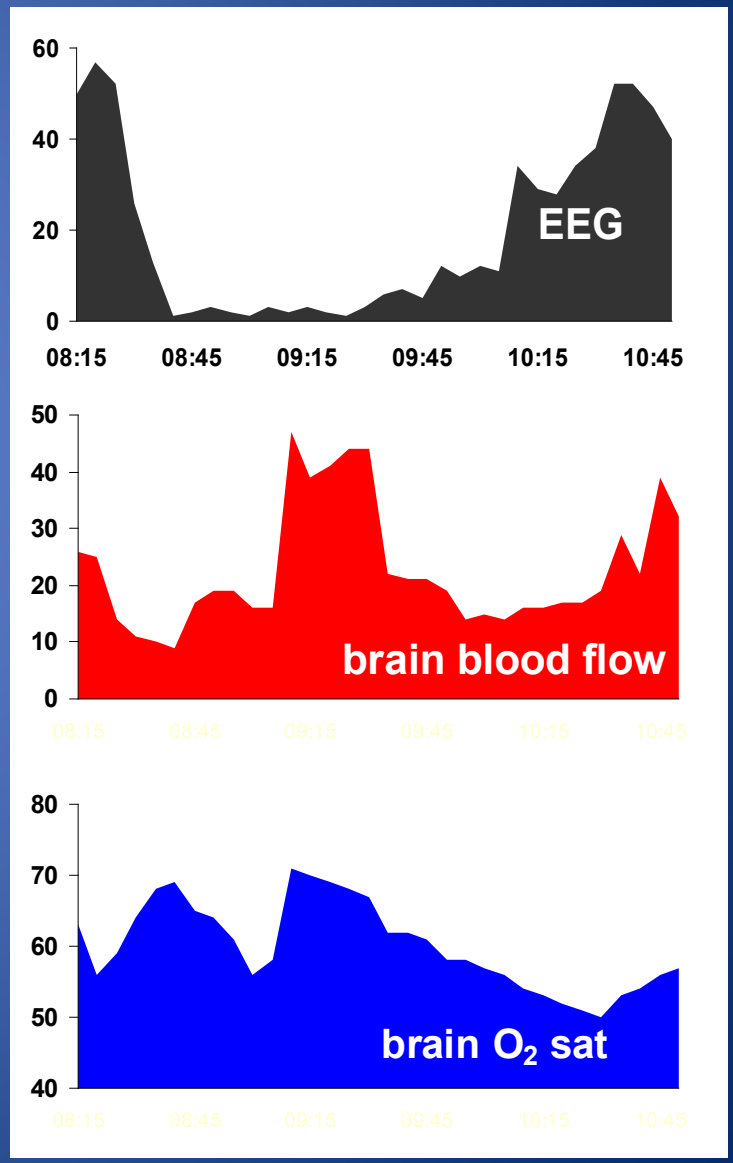


Near-Infrared Spectroscopy Patches



Multi-modality Neuromonitoring

- Synaptic Function
  - EEG
- Large Cerebral Vessel Perfusion
  - transcranial Doppler (TCD)
- Small Cerebral Vessel Oxygenation
  - near-infrared spectroscopy (NIRS)



Multi-modality Neuromonitoring

Retrospective analysis of our first 250 patients

176 patients with significant changes

- Among 42 pts w/o intervention
  - 28% neurologic complications
- Among 134 with intervention
  - 5% neurologic complications

Austin, Edmunds, et al, JTCVS 114:707-15, 1997

Conclusions Reached

- **Cerebral perfusion and oxygenation abnormalities occur commonly during pediatric heart surgery.**
- **Multimodality neuromonitoring allows timely detection of intraoperative events that can threaten neurologic function.**
- **Neuromonitoring-based interventions decrease the incidence of postoperative neurologic sequelae**

As a Result of This Initial Experience

- We have continued to apply multi-modality neuromonitoring on virtually all pediatric cardiovascular surgery cases
- We take all significant changes seriously and intervene as indicated.

Self Evaluation

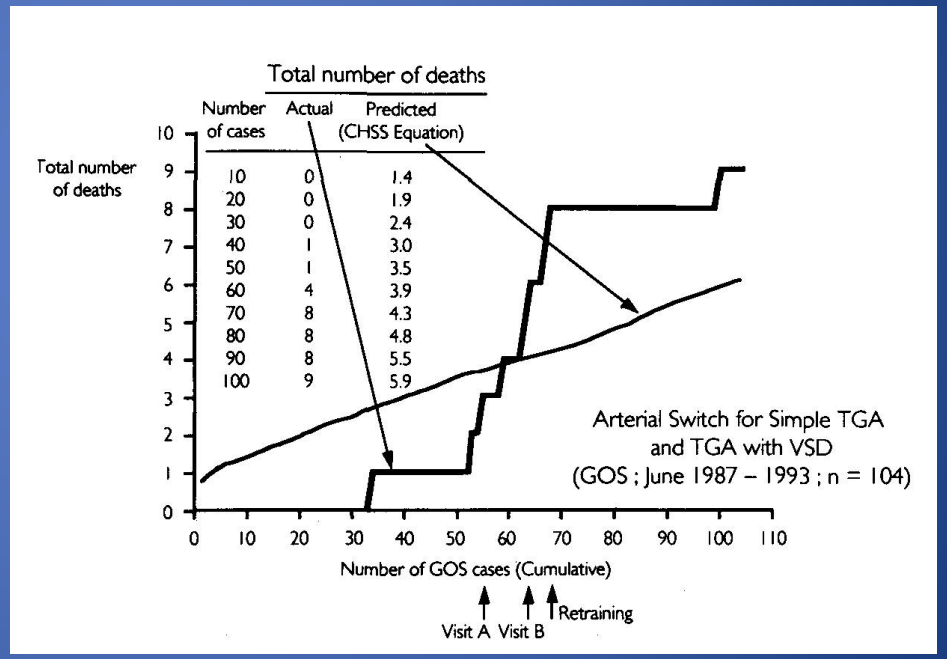
Self Evaluation

- Recognition of failures
  - Importance of keeping track
- Development of databases
  - Permitted tracking at individual centers
  - Identifying “real” outcomes (benchmarks)
  - Variation in outcomes

Analysis of a cluster of surgical failures

Application to a series of neonatal arterial switch operations

JTCVS 107:914, 1994



Marc R. de Leval, MD, FRCS, Katrien François, MD (by invitation), Catherine Bull, MRCP (by invitation), William Brawn, FRCS (by invitation), and David Spiegelhalter, PhD (by invitation), *London, England*

The Report of the Public Inquiry into
children's heart surgery at
the Bristol Royal Infirmary1984–1995

Learning from Bristol

Presented to Parliament by
the Secretary of State for Health
by Command of Her Majesty
July 2001

CM 5207(I)

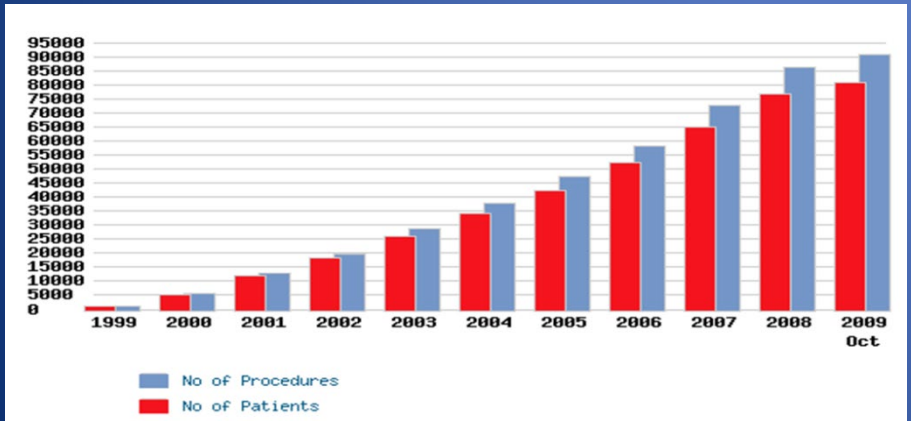
£32 (inc. VAT)

Quality Assessment - Databases

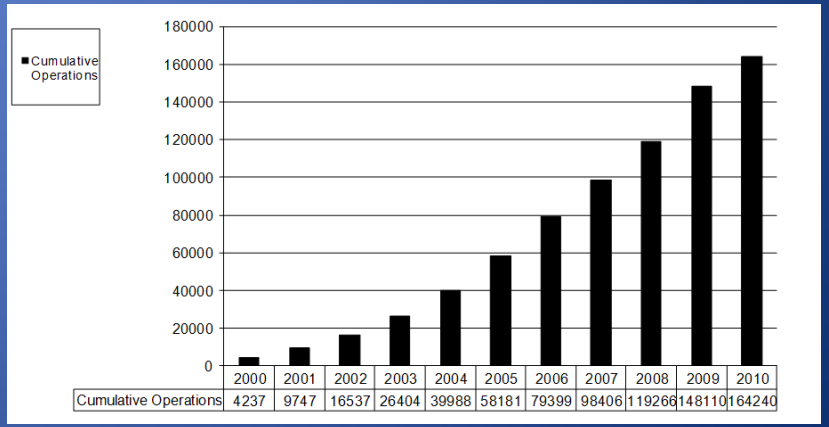
Databases to assess outcomes in Pediatric Heart Surgery

- Pediatric Cardiac Care Consortium - J. Moeller – 1984
- Congenital Heart Surgeons' Society – Datacenter
- EACTS Congenital Database - 1999
- STS Congenital Database - 2000

EACTS Congenital Database



STS Congenital Database



Quality Assessment - Databases

Good news

- Benchmarks derived from outcomes at multiple centers
- Permit individual centers to compare its outcomes to benchmarks

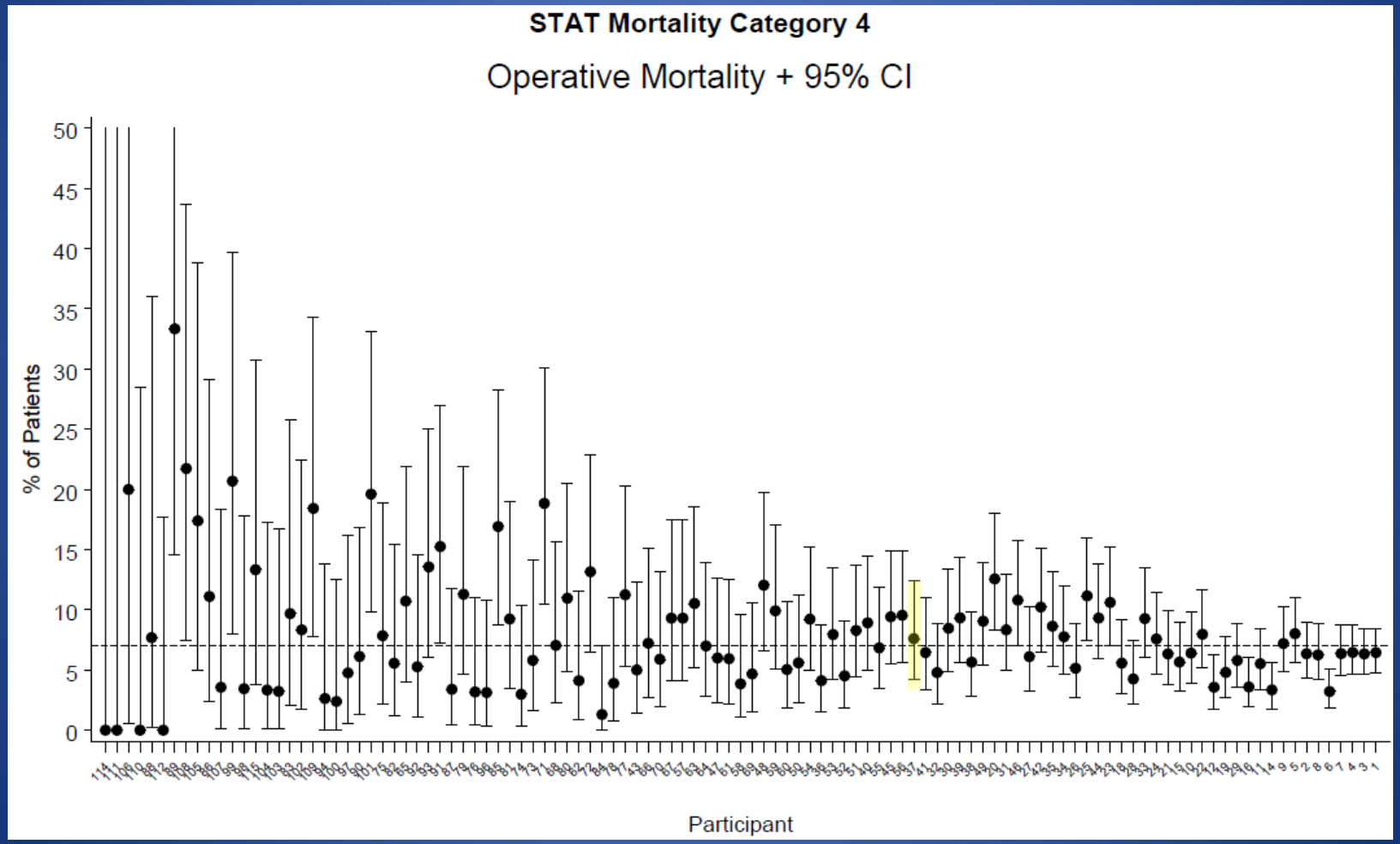
Not so good news

- Push for “transparency”
- Public likes to compare/rate/rank
- Risk adjustment challenging
- Risk aversion becoming a real phenomenon

How We Have Used STS Congenital Database

- Self-assessment with benchmarks for comparison
  - Compare to group means
  - Compare to other (unidentified) centers
- In concert with easily accessible database (CardioAccess)
  - Permits more frequent feedback
  - Dashboards

STS Congenital Database



Dashboard

Feb	Mar	Apr	May	Jun	Jul	Aug	Sep	Oct	Nov	Dec	2015			Jan	Feb	Mar
20	**27**	**32**	**42**	**35**	**35**	**39**	**40**	**23**	**47**	**28**	**388**	**Total Operations in CardioAccess**		**33**	**35**	**30**
9	12	11	21	22	19	16	16	13	12	10	**172**	Cardiopulmonary Bypass (CPB)		16	17	21
6	11	14	7	10	8	15	15	2	16	13	**123**	No CPB Cardiovascular (excluding primary PDA ligations, wt <2500g)		16	10	4
0	0	1	1	1	2	1	3	1	3	4	**17**	Primary PDA Ligations wt <2500g		0	0	0
1/1	2/1	2/4	2/9	2/0	1/2	1/3	2/2	3/0	1/15	0	**20/37**	ECMO, CPS or VAD	Initiation/Other (decan, repositioning, etc)	0	2/5	2/2
3	0	0	0	0	2	1	1	4	0	0	**11**	Thoracic		1	0	1
0	0	0	0	0	0	0	0	0	0	1	**1**	Bronchoscopy		0	0	0
0	1	0	2	0	1	2	1	0	0	0	**7**	Minor Procedure/Other		0	0	0
0	0	0	0	0	0	0	0	0	0	0	**0**	Interventional Cardiology (includes pacemakers done by Cardiology in OR)		0	0	0
												STAT Mortality Categories (Mortalities/Surgeries)	**STS Median and Interquartile Range**			
0/4(0%)	0/3(0%)	0/8(0%)	0/7(0%)	0/11(0%)	0/8(0%)	0/3(0%)	0/6(0%)	0/4(0%)	0/7(0%)	0/10(0%)	**0/75(0%)**	STAT Category 1	0.6% (0.4%-0.8%)	0/10(0%)	0/12(0%)	0/7(0%)
1/1(100%)	0/3(0%)	0/6(0%)	1/9(11%)	0/5(0%)	0/7(0%)	0/10(0%)	0/9(0%)	0/5(0%)	0/2(0%)	1/2(50%)	**3/61(5%)**	STAT Category 2	1.6% (1.3% - 2.0%)	0/6(0%)	0/3(0%)	0/5(0%)
0/5(0%)	0/2(0%)	0/2(0%)	0/3(0%)	0/2(0%)	0/3(0%)	0/1(0%)	0/2(0%)	0/3(0%)	0/3(0%)	0/0(0%)	**0/28(0%)**	STAT Category 3	2.6% (2.0% - 3.3%)	0/6(0%)	0/4(0%)	0/4(0%)
0/3(0%)	0/3(0%)	0/3(0%)	1/6(17%)	0/7(0%)	1/6(17%)	1/4(25%)	0/4(0%)	0/3(0%)	1/6(17%)	0/3	**4/54(7%)**	STAT Category 4	7.4% (6.6% - 8.2%)	1/5(20%)	0/2(0%)	0/8(0%)
1/1(100%)	0/1(0%)	0/0(0%)	0/0(0%	0/0(0%)	0/0(0%)	0/3(0%	0/1(0%)	0/0(0%)	0/0(0%)	0/1(0%)	**1/9(11%)**	STAT Category 5	15.2% (13% - 17.8%)	0/1(0%)	0/1(0%)	0/0(0%)
1/14(7%)	0/12(0%)	0/19(0%)	2/26(8%)	0/25(0%)	1/24(4%)	1/20(5%)	0/22(0%)	0/15(0%)	1/18(6%	1/16	**8/227(4%)**	# of pts in STAT categories		1/28(3%)	0/22(0%)	0/26(0%)
17	**16**	**23**	**30**	**28**	**30**	**22**	**26**	**19**	**25**	**20**	**272**	**Number of Patients in CardioAccess**		**28**	**24**	**24**
3	4	5	4	8	4	6	9	3	6	7	**63**	Neonate (<31 days)		6	4	5
6	6	9	8	3	8	6	4	9	11	7	**80**	Infant (31 days - 1 yr)		10	10	13
7	5	5	13	15	14	9	9	5	6	4	**100**	Child (1 yr - 18 yrs)		7	6	5
1	1	4	5	2	4	1	4	2	2	2	**29**	Adult (>18 yrs)		5	4	1
0	1	0	0	3	1	0	0	0	0	0	**5**	**Transplant Program**		**0**	0	0

Dashboard

Encourages regular assessment of outcomes

Encourages regular comparison to prior experience and benchmarks

Identifies areas requiring quality improvement

Provides continuous monitoring of performance

A Word of Caution in Public Reporting

Thomas L. Spray and J. William Gaynor

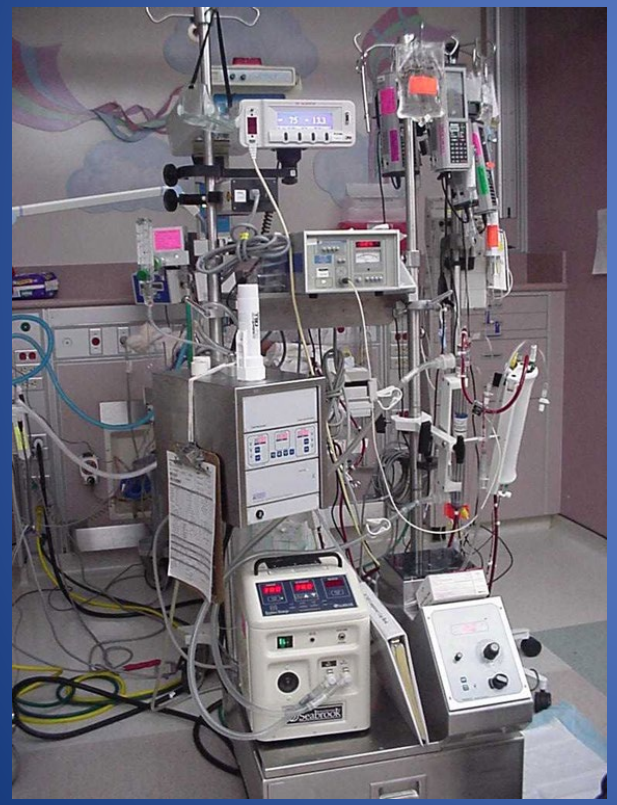
Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2017; 20:49-55.

We must be very careful that we don't create a system where we are afraid to take on a patient with increased risk of mortality because it may affect our overall ranking.

Mechanical Support

Mechanical Support

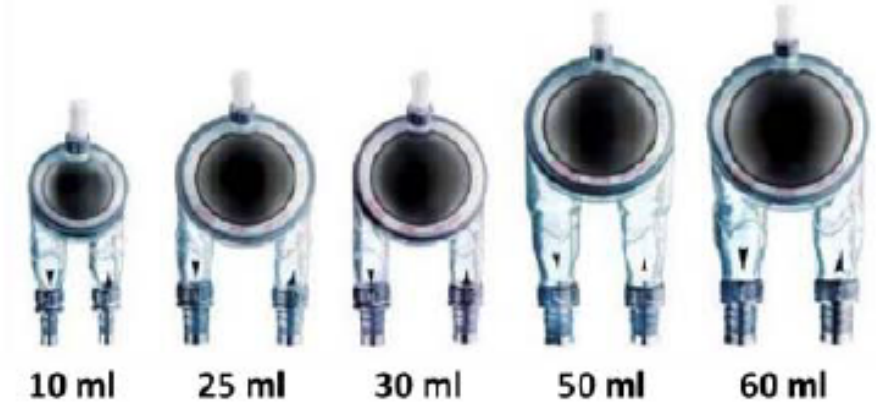
ECMO

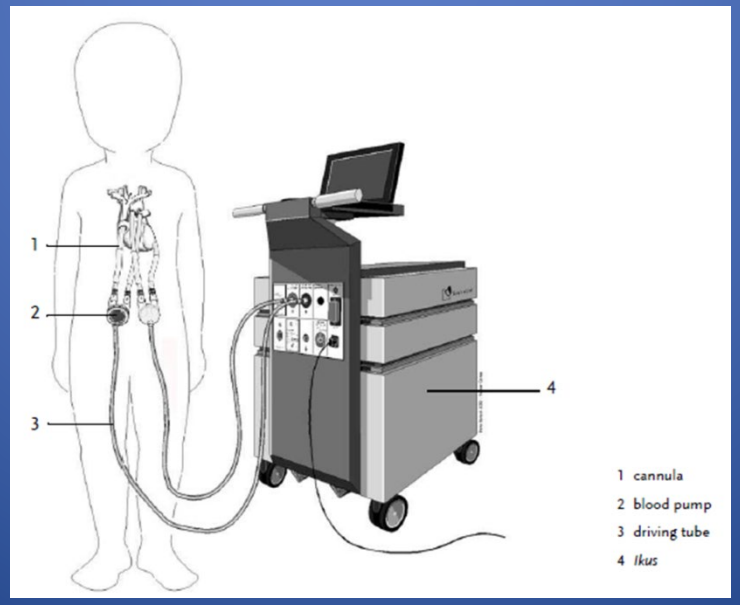


VAD - Berlin

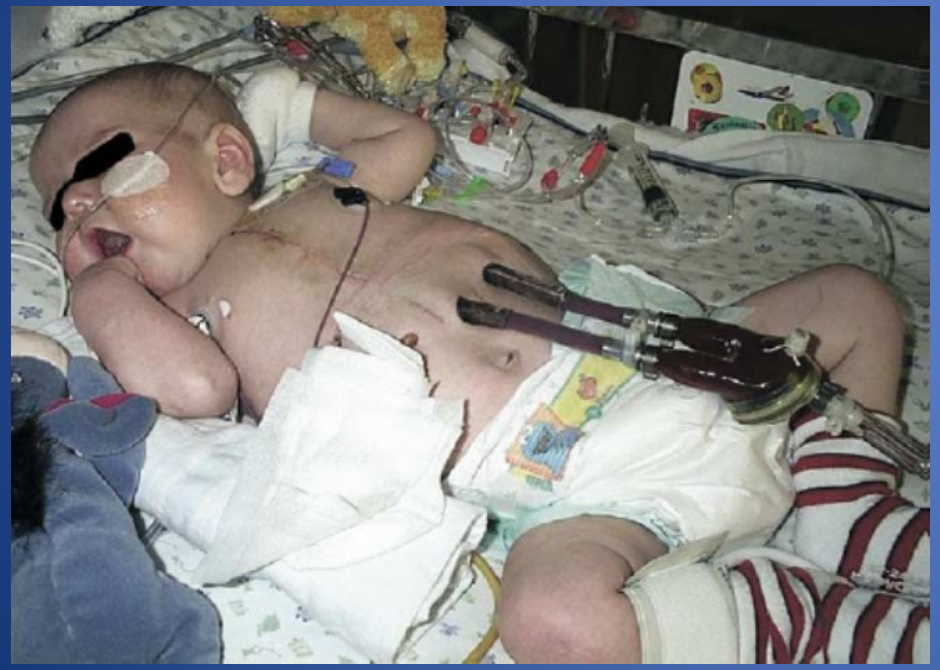
Berlin Heart EXCOR® HDE

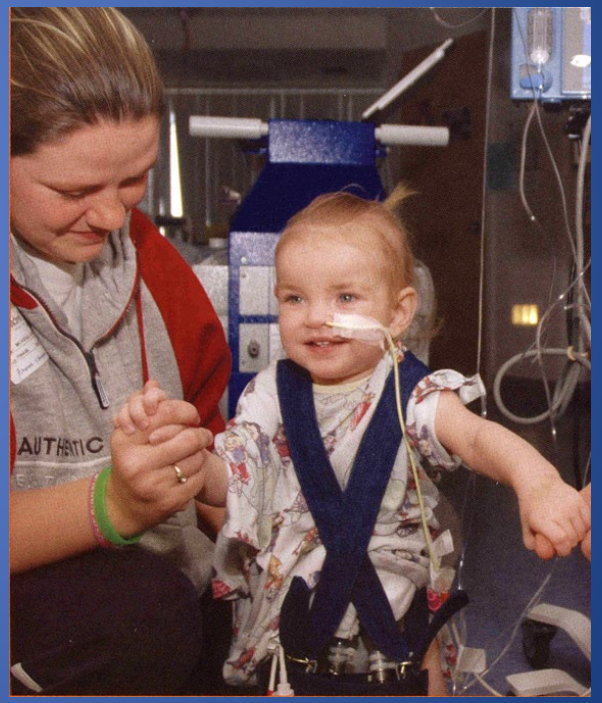
Weight Range	Pump Size
0-8 kg	10 ml
8-25 kg	25 ml
15-30 kg	30 ml
30-55 kg	50 ml
35-60 kg	60 ml





Berlin Heart EXCOR®





Berlin Heart EXCOR®

First Clinical Implant 1990 Germany

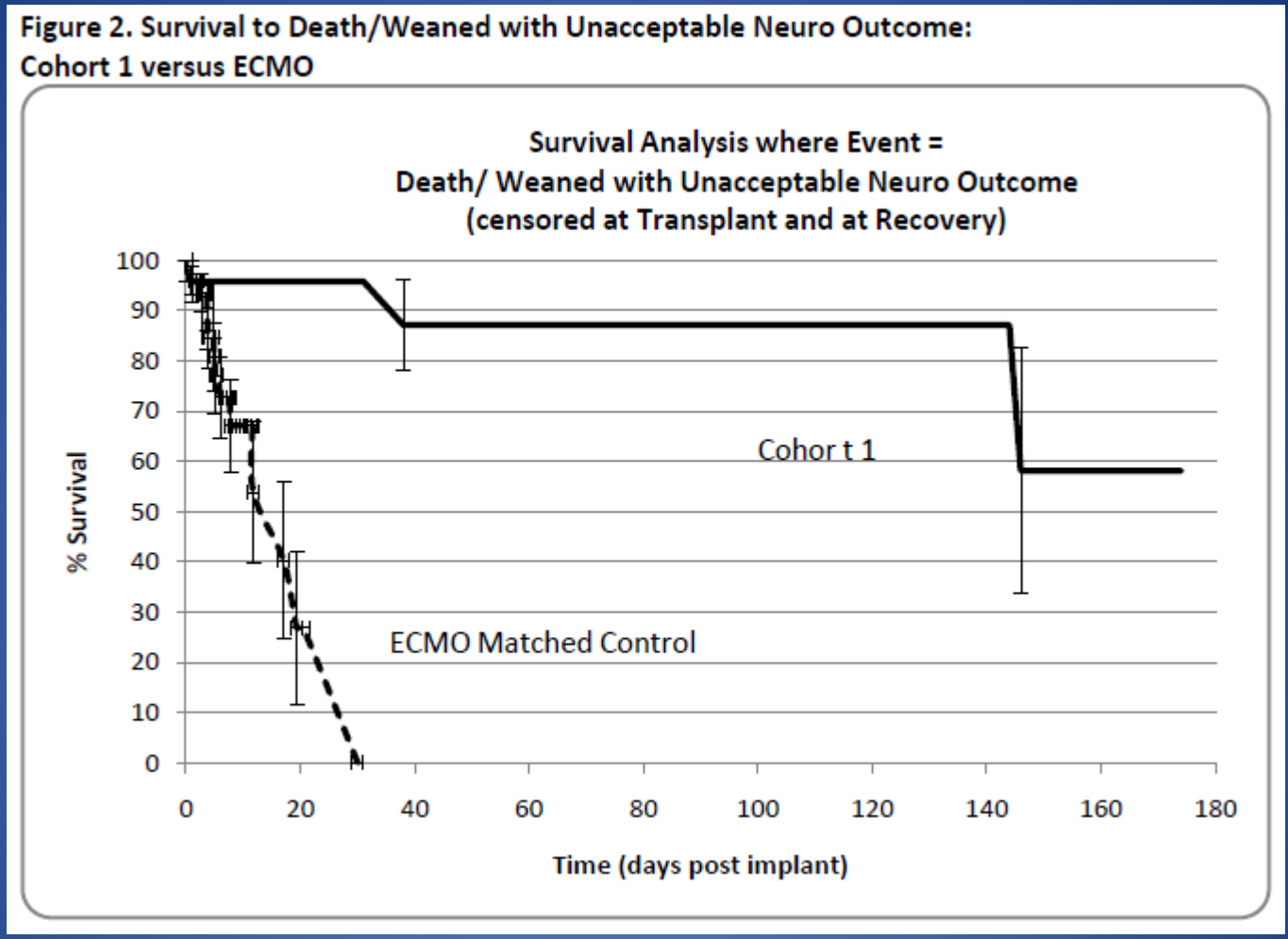
First US Clinical Implant - 2000 - U of Arizona

North American Implants (00-07) – 97

IDE approval – June 2007

FDA Humanitarian Device Exemption – Dec 2011

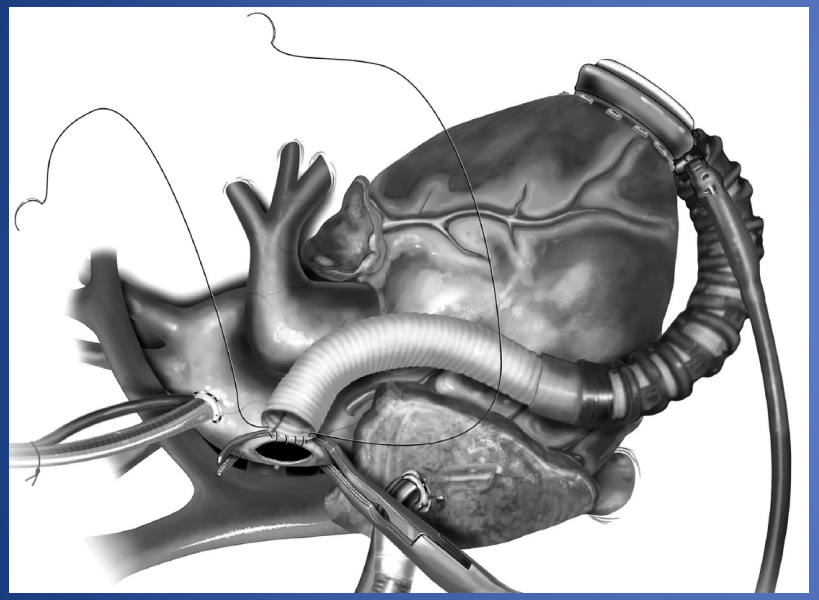
Figure 2. Survival to Death/Weaned with Unacceptable Neuro Outcome: Cohort 1 versus ECMO



Median wait time for heart transplant in children = 119 days (JHLT 2011)

Mechanical Support for Children – Adult Devices

HeartWare LVAD



Honjo & Rao OpTechTCVSurg 2014

Youngest 6 yo

Lowest weight 17 kg

Withdrawn from Market in 2021!

HeartMate 3

O'Connor et al. JHeartLungTransplantation 2020

28 pts <18 yo

Youngest 8 yo

Lowest weight 19.1 kg

Progress in Mechanical Support for Small Children Lags behind what has been achieved for Adults

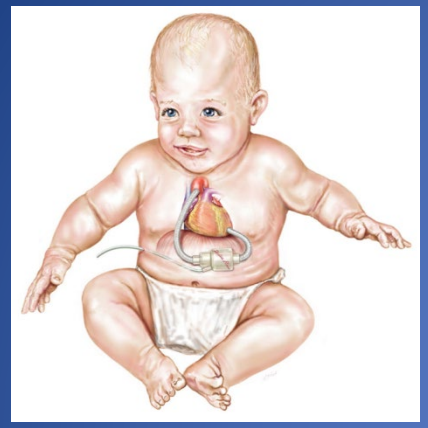
NHLBI Pediatric Circulatory Support Program

November, 2002 – Proposals solicited for novel circulatory support systems for children weighing 2 to 25 kg

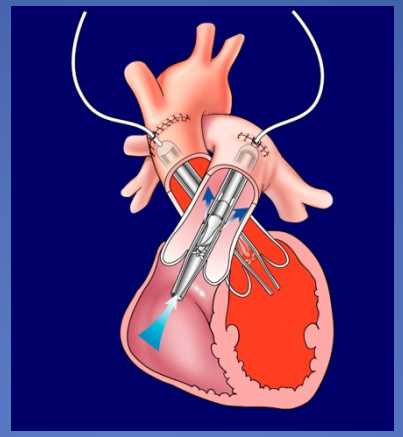
System requirements

- Deployable in less than 1 hour
- Minimize infection, bleeding, hemolysis, thrombosis
- Capable of providing up to 6 months support

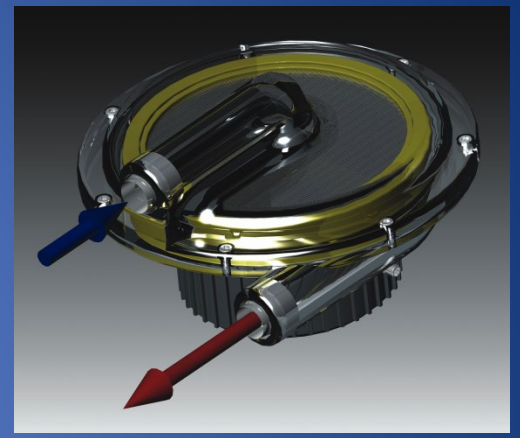
Pediatric Circulatory Support Systems



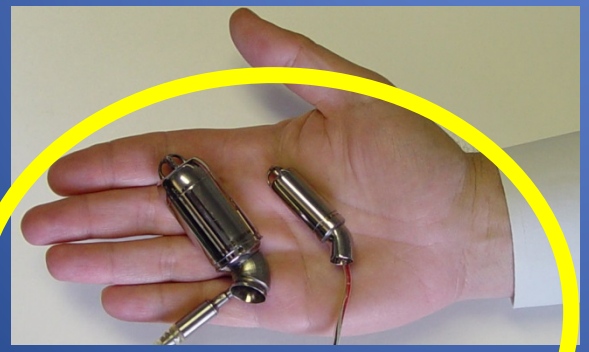
PediaFlow VAD
University of Pittsburgh
Harvey Borovetz, Ph.D.



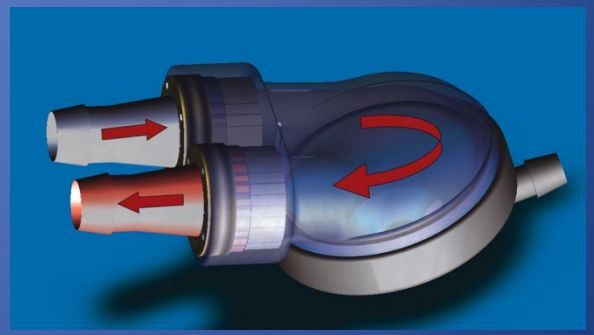
PediPump
Cleveland Clinic
Lerner College of Medicine-CWRU
Brian Duncan, M.D.



pCAS
Ension, Inc., Univ of Louisville
Mark Gartner, M.S.

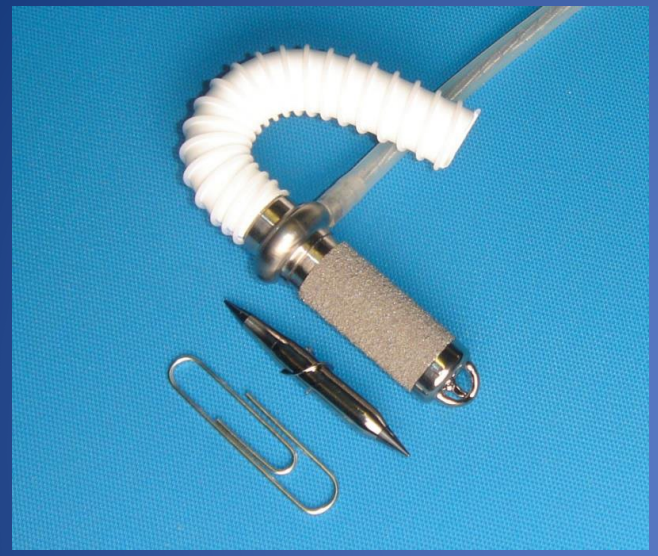


Child-size and Infant-size
Jarvik 2000 LVADs
Jarvik Heart, Inc.
Robert Jarvik, M.D.



Pulsatile Pediatric Ventricular
Assist Devices
Penn State University
Bill Weiss, Ph.D.

Infant Jarvik 2000 VAD



- 11 mm x 52.0 mm
- Miniature power/controller cables
- Modified analog control system
- Ceramic Cone Bearings
- Continuous Flow up to 1.5 l/min
- For 4-15 kg children

Infant Jarvik 2015 vs 2000

Infant Jarvik 2000

Infant Jarvik 2015

Infant Jarvik 2015 VAD

- 15 mm x 55.4 mm
- Miniature power/controller cables
- Modified analog control system
- Ceramic Cone Bearings
- Continuous Flow up to 3.0 l/min
- For 4-25 kg children

Current Status of PumpKIN Trial

Originally designed as prospective two-arm randomized study

Now completing a single-arm Feasibility trial – 8 to 20 kg

7 children at 4 US centers implanted with Jarvik 2015 as bridge to tx

Age range 7 months to 7 years; weight range 8.2 to 19 kg

Median duration of support 143 days (range 5 to 188 days)

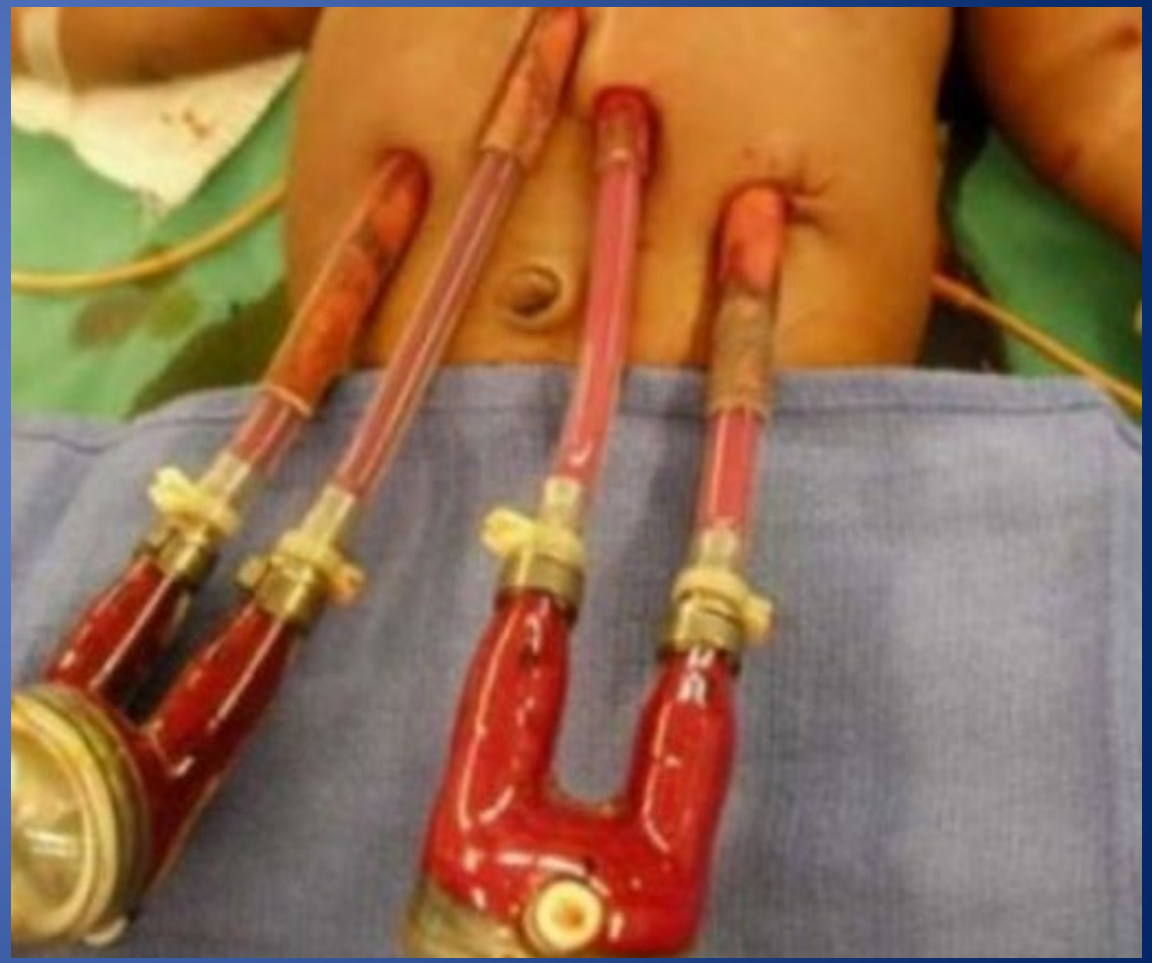
100% survival to transplant

Pivotal trial designed (n=22) – Enrollment to begin soon.

Continuing Status for Small Child or Infant

The Berlin Heart EXCOR® is currently the only FDA-approved durable VAD option for smaller children.

No intracorporeal devices available for patients < 20 kg.



Adult Congenital Heart Disease

Adults with Congenital Heart Disease

- 1 million adults with congenital heart disease
  - Exceeds number of children with CHD
- Consists of
  - Unrepaired congenital defects
  - Heart failure
  - Complications associated with earlier repairs
  - Newly recognized CHDs not found until adulthood
  - Complex arrhythmias
  - Pregnancy issues

Outcomes in adult congenital heart surgery: Analysis of the Society of Thoracic Surgeons Database

Christopher E. Mascio, MD,[a] Sara K. Pasquali, MD,[b] Jeffrey P. Jacobs, MD,[c] Marshall L. Jacobs, MD,[d] and Erle H. Austin III, MD[a]

Objective: Outcomes data for adults undergoing congenital heart surgery are limited. Previous analyses used administrative data or focused on single-center outcomes. We describe the most common operations, patient characteristics, and postoperative outcomes using a multicenter clinical database.

Methods: The study included adults (aged ≥ 18 years) listed in the Society of Thoracic Surgeons Congenital Heart Surgery Database (2000–2009). We describe patient characteristics and morbidity and mortality, and examine congenital procedures in the Society of Thoracic Surgeons Adult Cardiac Surgery Database to permit consideration of the primary dataset within a broader context.

Results: A total of 5265 patients (68 centers) from the Society of Thoracic Surgeons Congenital Heart Surgery Database were included. Patients' median age was 25 years (interquartile range, 20–35). Common preoperative risk factors included noncardiac abnormalities (17%) and arrhythmia (14%). Overall, in-hospital mortality was 2.1%, 27% had 1 or more complication, and median length of stay was 5 days. Common operations included right ventricular outflow tract procedures (21%) and pacemaker/arrhythmia procedures (20%). We further evaluated cardiopulmonary bypass procedures in more than 100 patients. Mortality ranged from 0% (atrial septal defect repair) to 11% (Fontan revision/conversion). Separate evaluation of the Society of Thoracic Surgeons Adult Cardiac Surgery Database revealed 39,872 adults undergoing congenital heart operations.

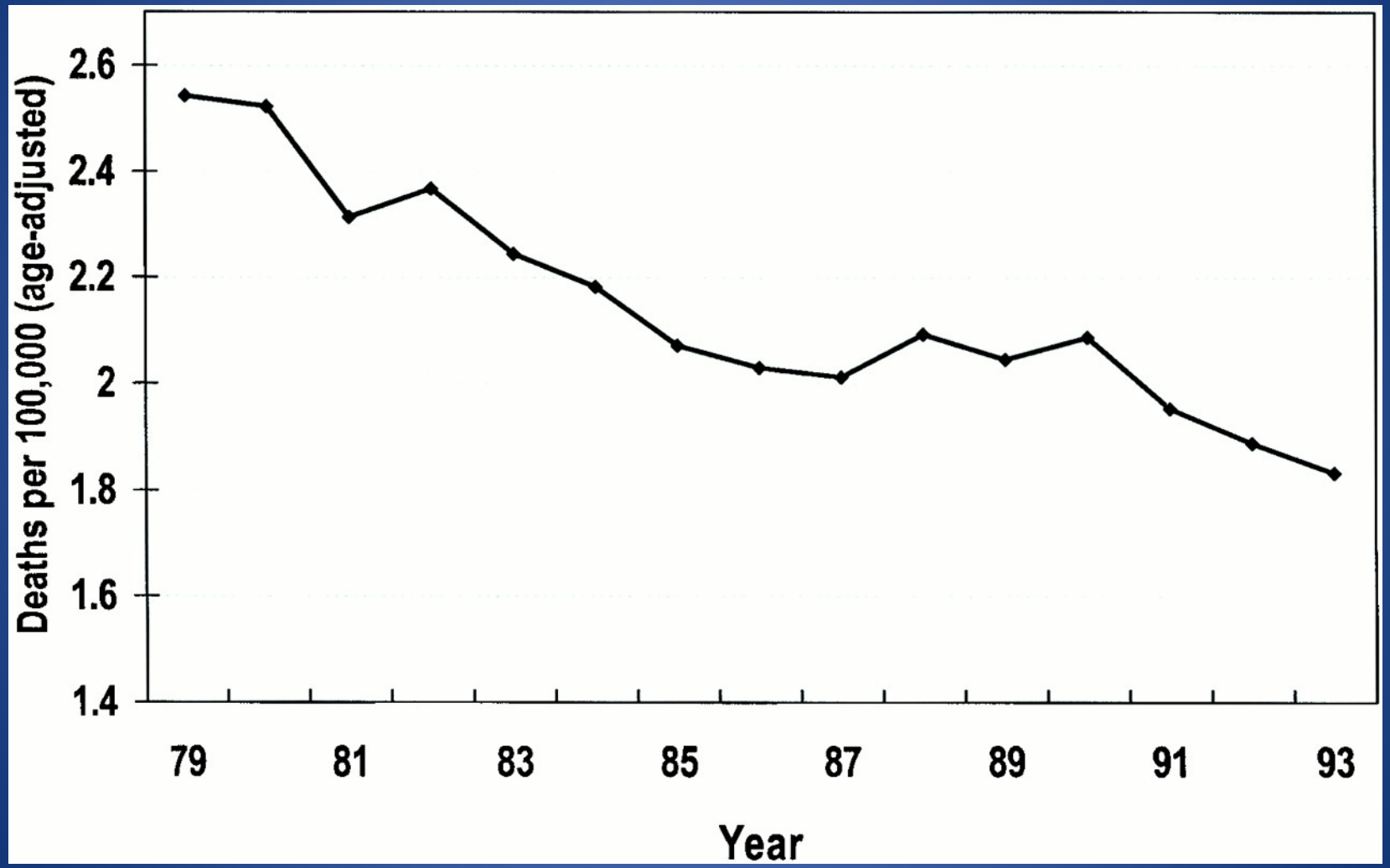
Conclusions: Most adult congenital heart operations listed in the Society of Thoracic Surgeons Congenital Heart Surgery Database are performed in the third to fourth decades of life; approximately half are for right heart pathology or arrhythmia. Many patients have complications, but mortality is low with the exception of those undergoing Fontan revision/conversion. Many more adults undergoing congenital heart surgery are entered into the Society of Thoracic Surgeons Adult Cardiac Surgery Database. (J Thorac Cardiovasc Surg 2011;142:1090-7)

Adults with Congenital Heart Disease

- Adult congenital population is increasing in number (and complexity) thanks to our successes at younger ages
- Cardiologists dedicated to adults with congenital heart disease are increasing in number and knowledge
- Adult Congenital Heart Association established in 1998, began Accreditation Program in 2017
  - (Norton Children's and Cincinnati Children's are both accredited)
- Centers in both Louisville and Lexington now have sections devoted to this growing segment of patients

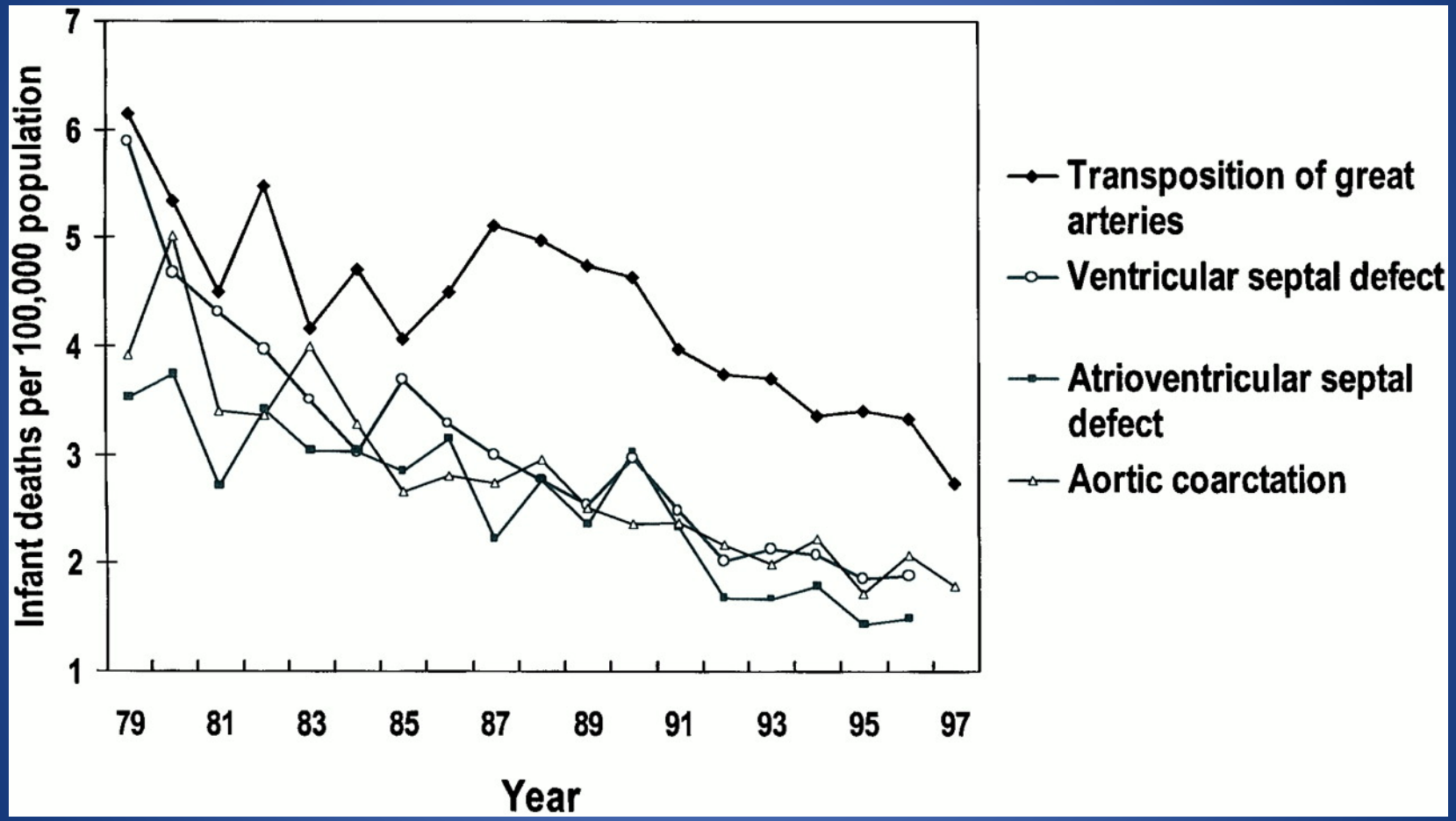
So, how have we done?

Death rate due to congenital heart disease, United States, 1979–1997



Boneva et al. Circulation 2001

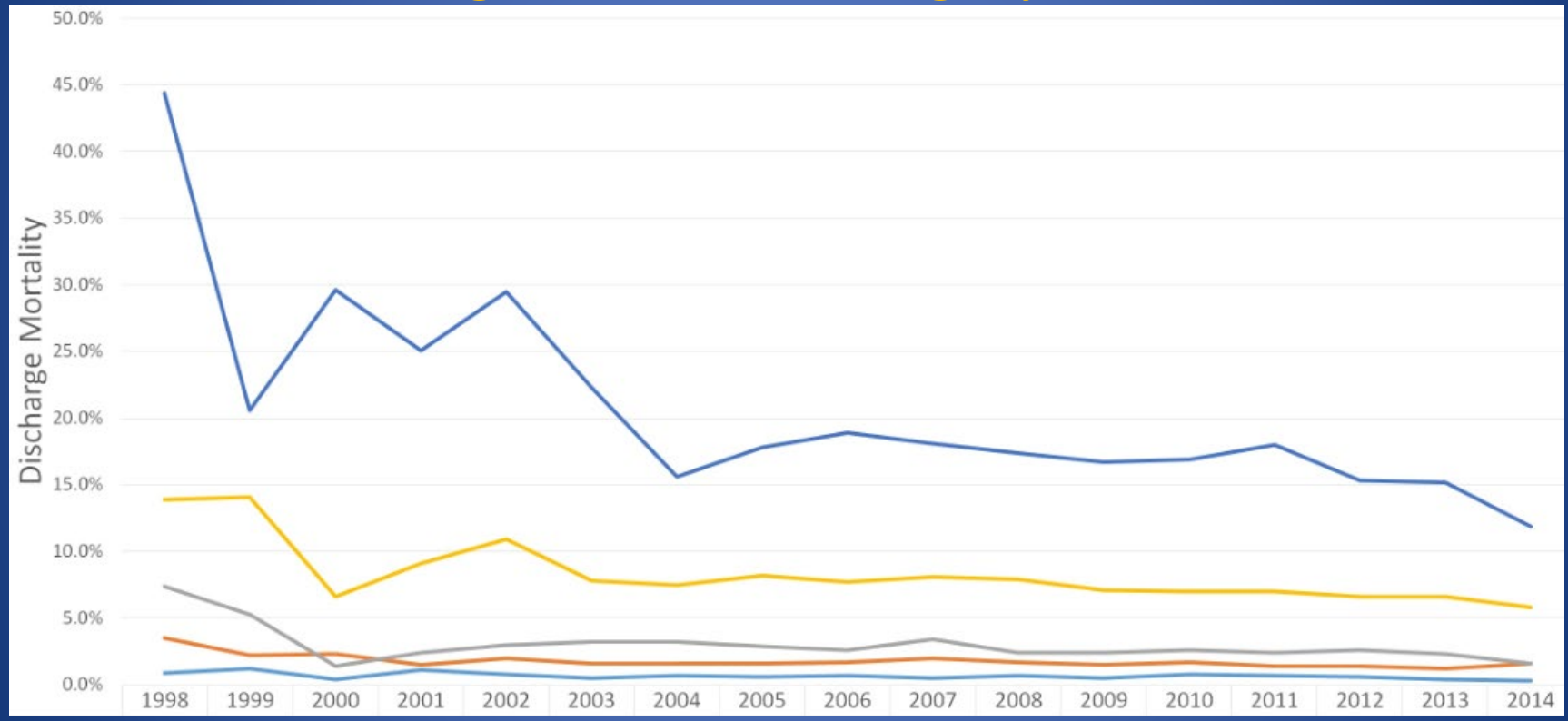
Infant mortality due to selected CHDs, United States, 1979–1997



Boneva et al. Circulation 2001

Discharge Mortality versus Year of Surgery stratified by STAT Category as of 2014

STS Congenital Heart Surgery Database



Jacobs, He, Mayer, Austin, et al. Ann Thor Surg 2016

Looking back 30+ years

Much has been learned and much has been accomplished

Less complex lesions are now managed with excellent success

- Many now managed with catheter intervention alone

Significant improvement achieved for the complex ones

- Scientific advancement, collaboration, team-work and self analysis

And in Kentucky we are doing more than keeping pace, we are forging ahead!

In Lexington at Kentucky Children's Hospital and UK

- Congenital Heart Clinic
- Joint Pediatric Heart Care Program with Cincinnati Children's Hospital
  - Two-sites model
- Carl Backer, MD (past President of Congenital Heart Surgeons' Society)
  - Chief of Pediatric Cardiothoracic Surgery at Kentucky Children's
  - New editor of World Journal of Pediatric and Congenital Heart Surgery
- Full set of cardiology and pediatric specialists in Lexington and Cincinnati

And in Kentucky we are doing more than keeping pace, we are forging ahead!

In Louisville at Norton Children's Hospital and U of L

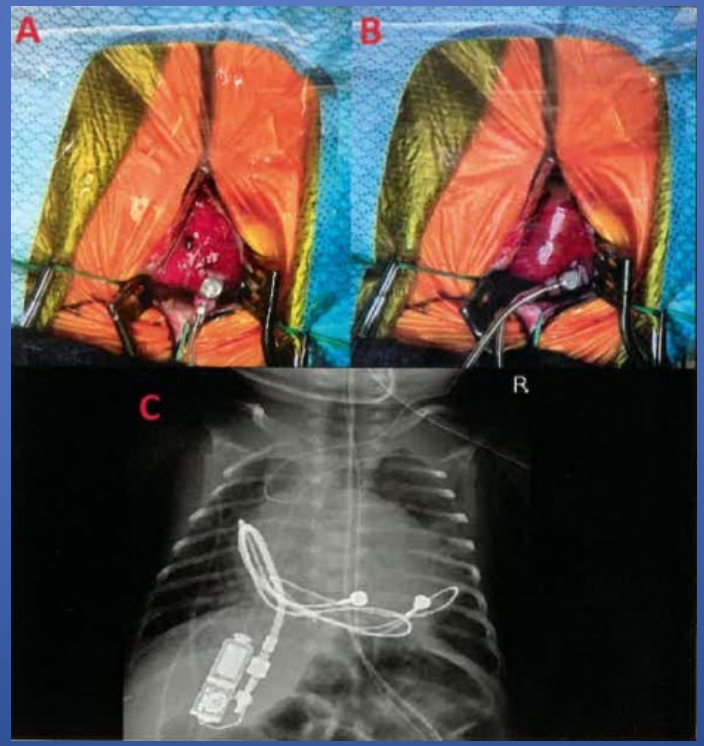
- Norton Children's Heart Institute
  - Collaboration amongst services (Surgery,Cardiology,CICU,NICU,Anesthesia,Nursing)
  - Co-directors: Bahaaldin Alsoufi, MD and Brian Holland, MD
- Surgeons
  - Bahaaldin Alsoufi, MD and Deborah Kozik, DO
- Cardiologists
  - 31 providers (up from 4 in 1989) with dedicated sections
    - Interventional cardiology
    - Electrophysiology
    - Heart failure and transplantation
    - Adult congenital
    - General pediatric cardiology – 16 outreach sites

MEDICAL NEWS

A First for the United States: Norton Children's Heart Institute Physicians Implant Tiny Pacemaker, Saving Infant's Life

Patient Born at 28 Weeks with Slow Heart Rate and Congenital Heart Disease Receives Never Before Used Pacemaker Implant

Congenital Cardiology Today 20:12 Sept 2022



Recent literature contributions from Norton Children's - 1

Heart Transplantation:

- **Increasing donor-recipient weight mismatch is associated with shorter waitlist duration and no increased morbidity or mortality. Alsoufi B. EJCTS 2022.**
- **Associated factors and impact of persistent renal dysfunction in children listed for heart transplantation. Alsoufi B. ATS 2022.**
- **Systemic Venous Reconstructions During Pediatric Heart Transplantation. Altin F, Alsoufi B, et al. WJCPHS 2022.**
- **Interplay between donor and recipient factors impacts outcomes after pediatric heart transplantation: An analysis from the united network for organ sharing database. DAS B, Alsoufi B, et al. ASAIO 2021.**
- **ABO-Incompatible Heart Transplant in Infants. Kozik D. ATS 2021.**
- **Did outcomes of infants listed for heart transplantation change? Alsoufi B. EJCTS 2022.**
- **Trends and outcomes of heart transplantation in adults with congenital heart disease. Alsoufi B . ATS 2022.**

Recent contributions from us at Norton Children's - 2

Mechanical circulatory support:

- ECMO:
  - **Repeat ECMO Is Appropriate in Selected Children With Cardiac Disease: An Extracorporeal Life Support Organization Study. Alsoufi B. WJPCHS 2021.**
  - **Factors Associated With Survival Following Extracorporeal Cardiopulmonary Resuscitation in Children. Alsoufi B. WJPCHS 2020.**

- MCS outcomes:
  - **Recent Era Outcomes of Mechanical Circulatory Support in Children With Congenital Heart Disease as a Bridge to Heart Transplantation. Das B. ASAIO 2022.**
  - **Meeting needs-pushing boundaries: Mechanical circulatory support in children. Deshpande S, Sparks J, Alsoufi B. JTCVS 2021.**

- Special situations:
  - **Single ventricle VAD and Hybrid palliation in neonate with HLHS and dysplastic severe TR. Alsoufi B. 2020**
  - **Total artificial heart in 7 years old to salvage primary graft dysfunction. Alsoufi B. 2022.**

Recent contributions from us at Norton Children's - 3

Complex valve repairs and replacement:

- **Early Outcomes of Patients Undergoing Neoaortic Valve Repair Incorporating Geometric Ring Annuloplasty. Kupferschmid J…Austin E, Alsoufi B, et al. WJPCHS 2022.**
- **Repair of neoaortic valve following the Ross procedure using the Geometric Ring annuloplasty. Austin E. AATS 2022.**
- **Outcomes Following Aortic Valve Replacement in Children With Conotruncal Anomalies. Alsoufi B. WJPCHS 2022.**
- **Are Mechanical Prostheses Valid Alternatives to the Ross Procedure in Young Children Under 6 Years Old? Alsoufi B. ATS 2022.**
- **Multicentre comparative analysis of long-term outcomes after aortic valve replacement in children. Knight J,..Alsoufi,..et al. Heart 2022.**

Recent contributions from us at Norton Children's - 4

Innovative surgeries and hybrid approaches:

- **Norwood Operation With Partial Left Ventriculectomy for Neonatal Dilated Cardiomyopathy. Alsoufi B. ATS 2022.**
- **Unusual Case of Common Arterial Trunk With Atresia of the Right Pulmonary Artery and Aortopulmonary Collaterals. Kozik D. WJPCHS 2020.**
- **Simultaneous Norwood and Aortic Uncrossing in Neonate With Aortic Atresia and Right Circumflex Aorta. Alsoufi B. STCVS 2020.**
- **Modified Microvascular Plug as a Flow Restrictor in Hypoplastic Left Heart Syndrome with Dysplastic Tricuspid and Pulmonary Valves. Kurtz J. Pediatric Cardiology 2021.**
- **Hybrid strategy for management of high risk neonates with complex biventricular anomalies and arch obstruction. Alsoufi B. Pediatric Cardiothoracic Surgery Annual 2023.**

Bottom Line

We’ve done pretty well for patients with congenital heart disease in Kentucky over the past 30 years.

The future is bright!!!


THANK
YOU