The Pott's Operation Lives Again!

Pirooz Eghtesady

Cardiology 2024- Scottsdale, Arizona
Feb 15th, 2024

Plenary Session II: What's Old is New Again

Barnes-Jewish Hospital BJC HealthCare | Washington University in St. Louis Physicians | Children's Hospital • St. Louis BJC HealthCare

No Disclosures

- Off label Use:
  - Contegra Conduit
  - Pulmonary Homografts

Manish Aggarwal, MD
Andrea Drussa, CPNP
Mark Grady, MD

Case Scenario- ZC

1 year old White male

- Born Premature at 29 weeks
- Only triplet to survive birth
- Tracheomalacia and Bronchomalacia

No Chromosomal Abnormalities

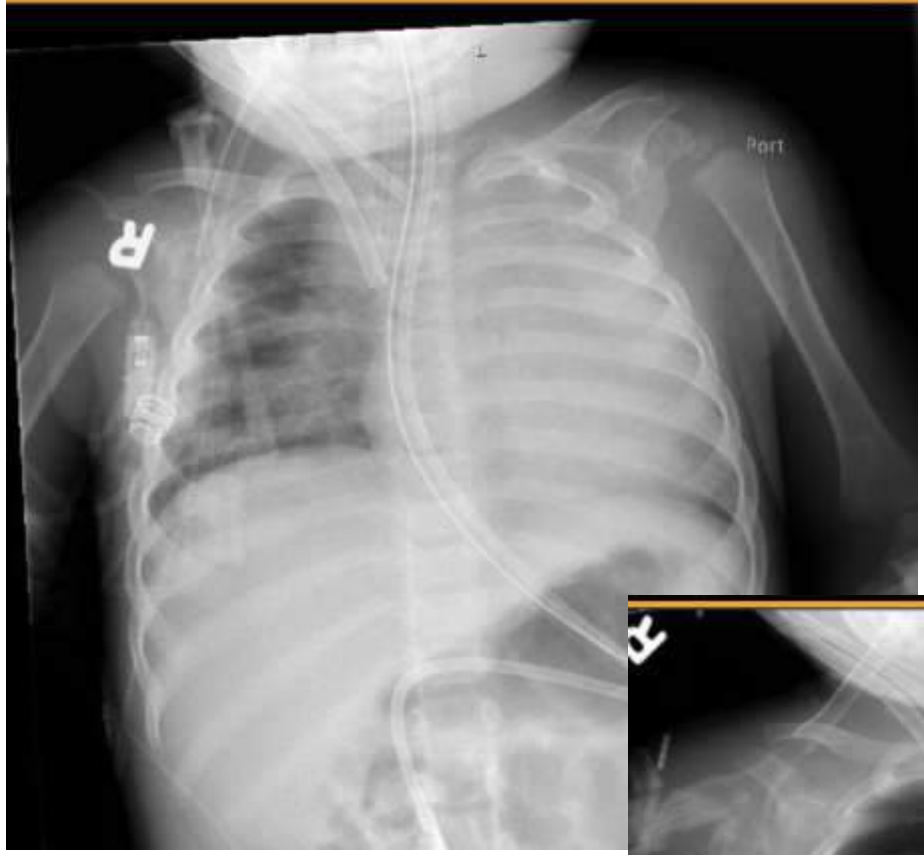
Dx: Pulmonary Hypertension

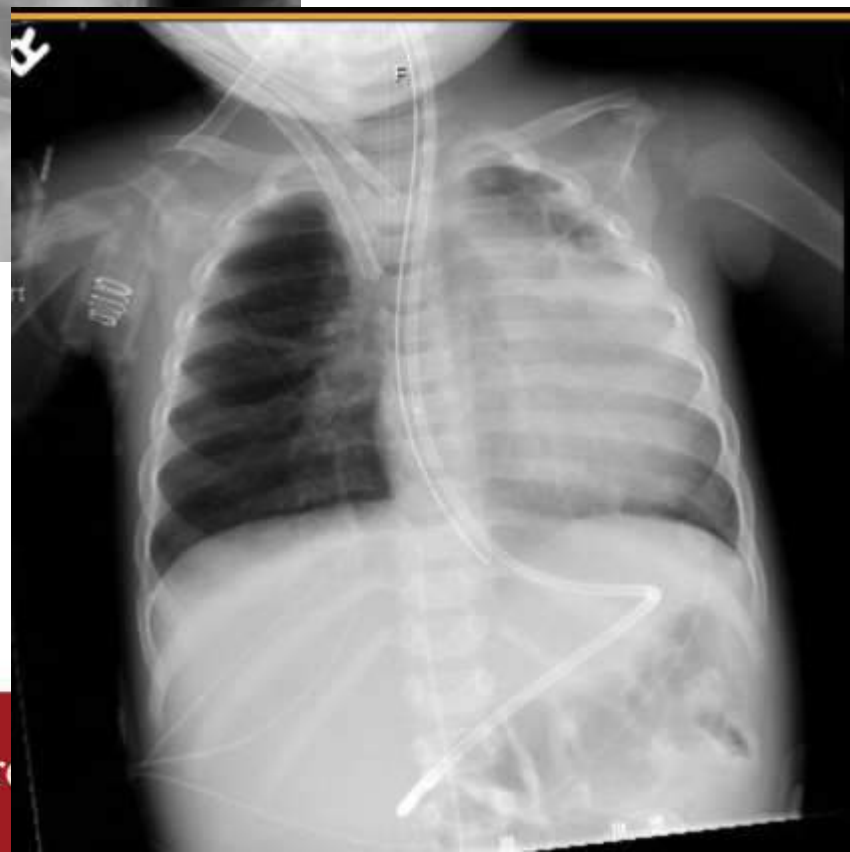
History of seizures

Cath Lab → arrest → ECMO

Head CT: multifocal areas of hypodensity within the right occipital & bilateral frontal and frontoparietal cortex concerning for ischemia and infarct

Admitted on ECMO





ZC- Intraop/Post-op

Surgery Date: 2 days from admittance

Weight at Surgery: 7.6 Kg

Length of Surgery: 1hr 40 min (clamp 16 min)

Procedures performed:

- Potts Shunt
- Rigid Bronchoscopy

Passed away on Postop day 30

- Remained Intubated throughout hospital course
- Persistent Left lung collapse despite multiple bronchs
- Hypotension/Desaturation
- Can't come off ECMO
- Anticoagulation difficulties
- Repeat Head CT: hemor. infarct with laminar necrosis

Case Scenario- JB

4 yo- h/o fatigue, cough, dyspnea since age 2 → asthma

⇢ increasing fatigue, new hypoxia (PFO), cardiomegaly on CXR

⇢ pulmonary HTN → Children's Hospital CO (triple therapy and PFO stenting (6mm).

⇢progressive symptoms, near syncope, SOB with short distances, poor weight gain.

Potts Shunt- L thoracotomy; Length of Surgery 43 min (clamp 12 min)

LOS: 9 days

Currently: 10 years out; off prostins/doing great

Case Scenario- PR

14 y.o. male-IPAH diagnosed at 10 months:

- ➢Suboptimal growth and weight gain
- ➢Persistent "wet, wheezing cough”
- ➢CXR= Cardiomegaly
- ➢hypoxemic (unclear etiology)
- ➢cath: "severe PH". Immediately following severe PH crisis/CPR →ICU for 3 weeks …triple therapy

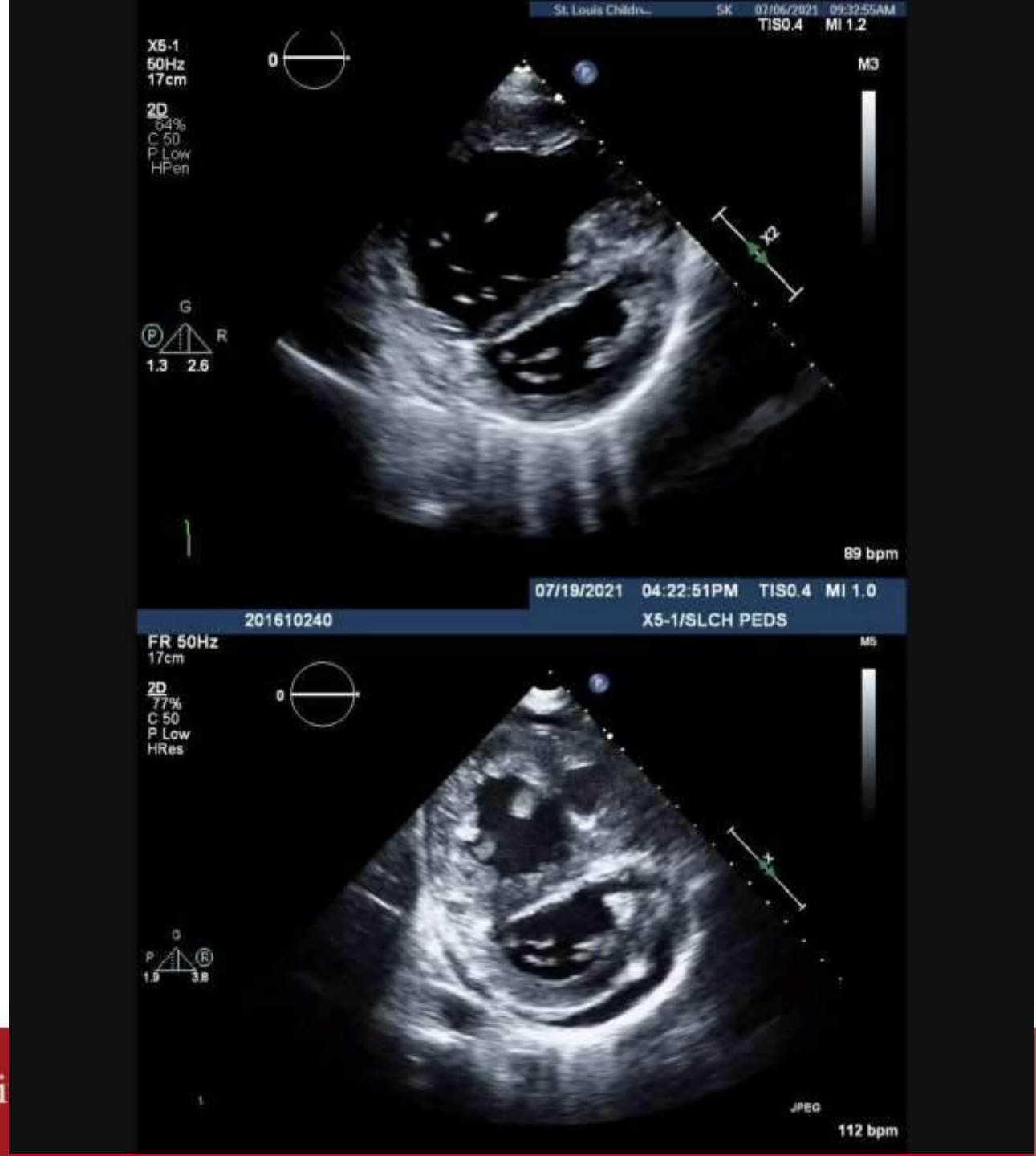
Evidence of worsening disease:

Suprasystemic PH

BNPs increasing.

Diminished activity tolerance (frequent breaks with activity and increased DOE)

- ➢ Placed an 18mm Composite (Contegra inside GoreTex PTFE tube)- 45 min pump run
- ➢ Has done great, off SubQ pump which he had been on for 15 years.



Twenty-Year Experience and Outcomes in a National Pediatric Pulmonary Hypertension Service

Andrew Constantine[1,2,3], Konstantinos Dimopoulos[2,3], Sheila G. Haworth[1,4†], Vivek Muthurangu[4], and Shahin Moledina[1,4]

[1]National Pediatric Pulmonary Hypertension Service UK, Great Ormond Street Hospital for Children National Health Service Foundation Trust, London, United Kingdom; [2]Adult Congenital Heart Centre and Centre for Pulmonary Hypertension, Royal Brompton and Harefield Hospitals, Guy's and St. Thomas' National Health Service Foundation Trust, London, United Kingdom; [3]National Heart and Lung Institute, Faculty of Medicine, Imperial College London, London, United Kingdom; and [4]University College London Institute of Cardiovascular Science, University College London, London, United Kingdom

ORCID ID: 0000-0003-0262-2340 (S.M.).

Am J Respir Crit Care Med. 2022 Sep 15;206(6):758-766.

ORIGINAL ARTICLE

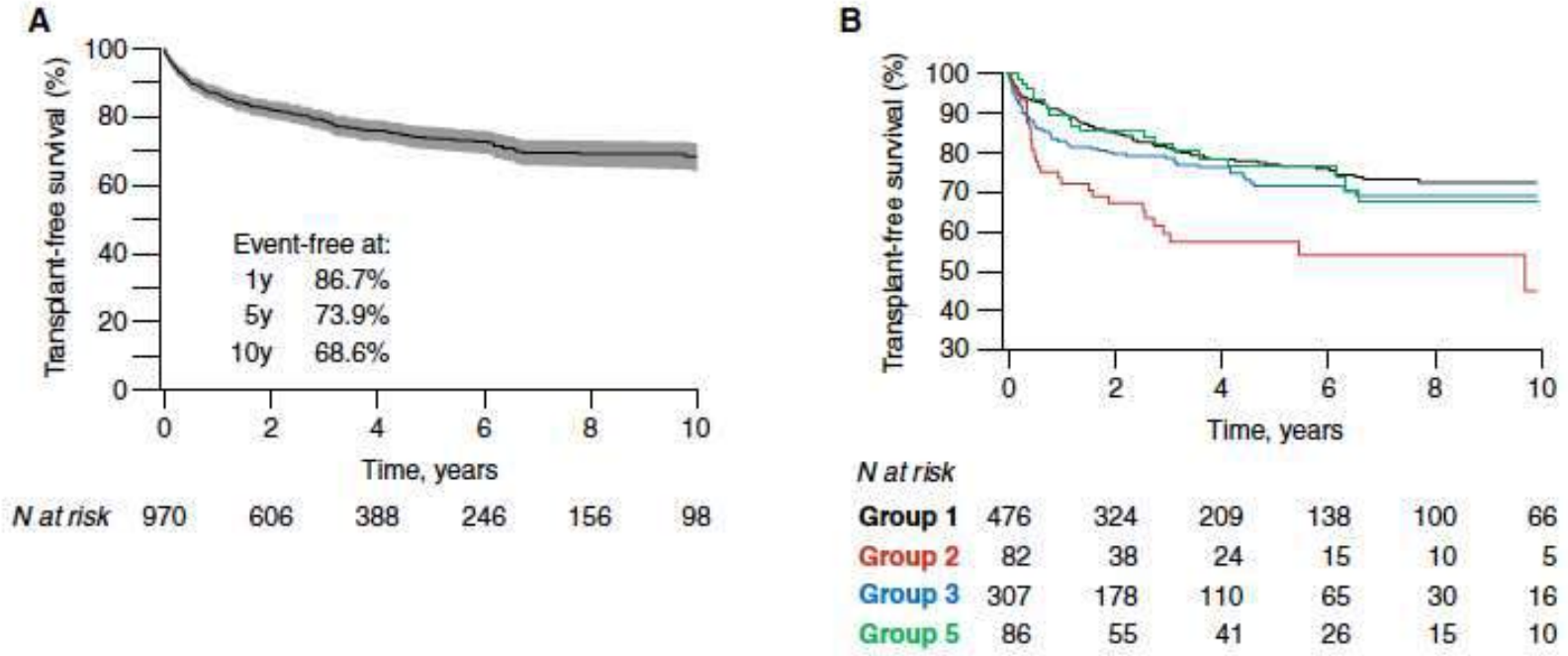
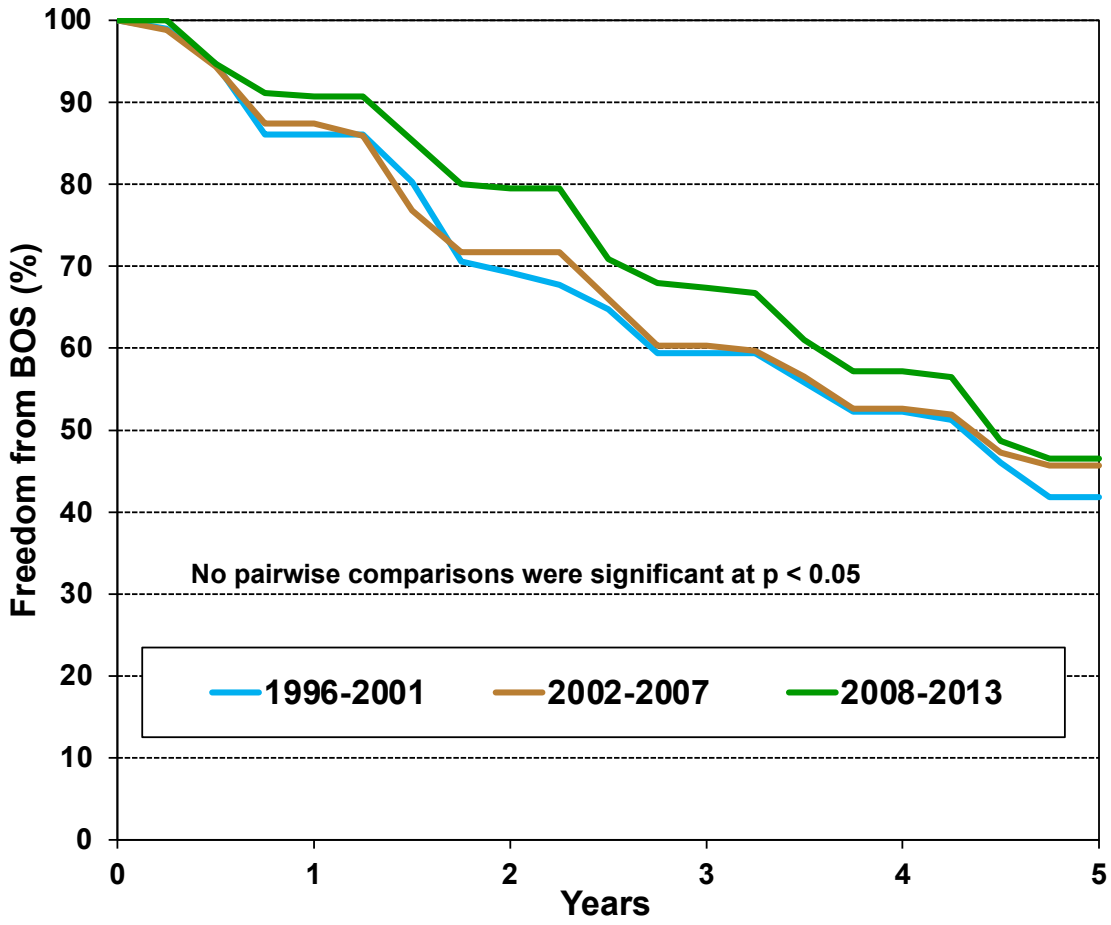


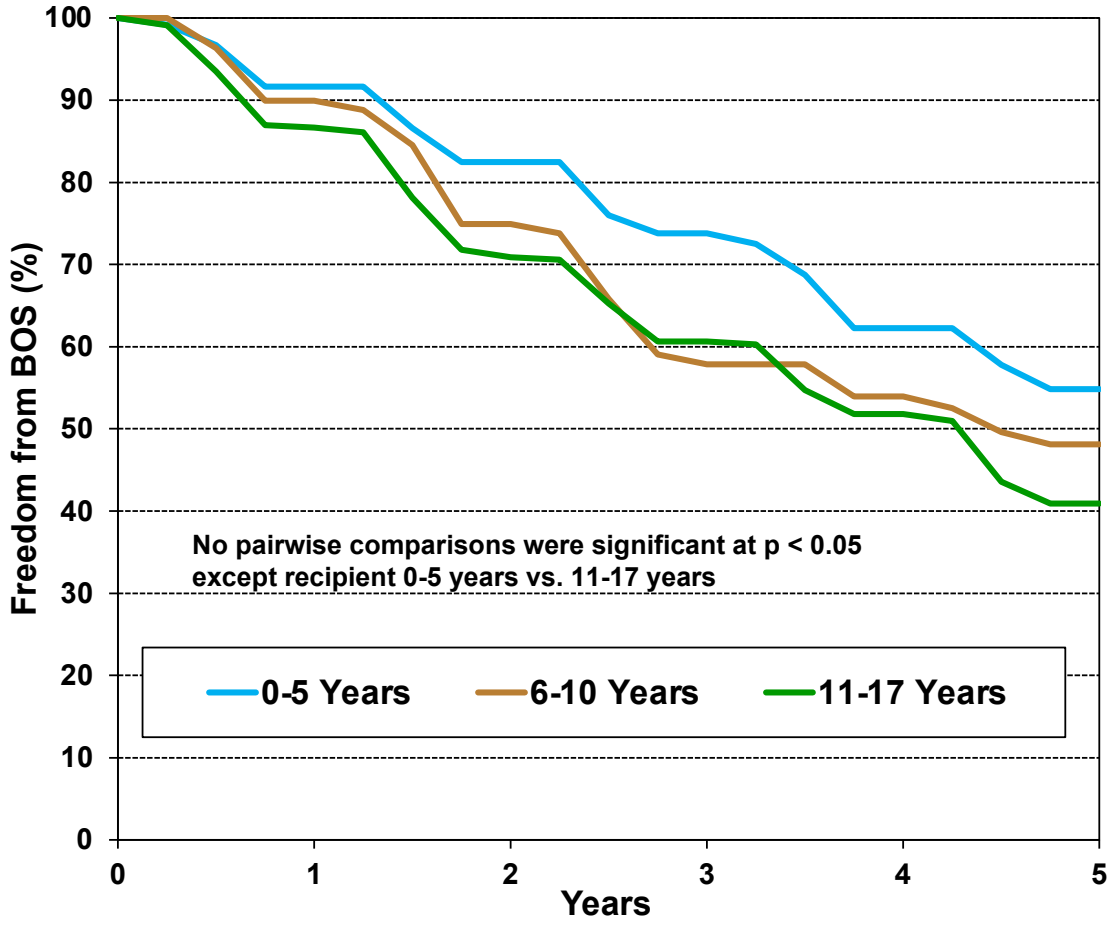
Figure 3. Kaplan-Meier survival curves in children with pulmonary hypertension, (*A*) in the overall cohort and (*B*) for the different pulmonary hypertension groups. The numbers below the horizontal axis represent the number of patients at risk.

Pediatric Lung Transplants
Freedom from BOS Conditional on Survival to Discharge

By Era
(Transplants: Jan 1996 - Jun 2013)



By Recipient Age
(Transplants: Jan 1996 - Jun 2013)



ISHLT • INTERNATIONAL SOCIETY FOR HEART AND LUNG TRANSPLANTATION
2021
JHLT. 2021 Oct; 40(10): 1023-1072

Potts Shunt in Patients with Pulmonary Hypertension

TO THE EDITOR: Pulmonary hypertension is a rare complication of transposition of the great arteries.[1] As in other forms of primary pulmonary hypertension, the prognosis is poor, particularly in children who have symptoms.[2] Converting normal cardiac physiological features to features typical of Eisenmenger's syndrome by means of blade atrial septostomy has already been proposed for patients with this condition.[3] But patients with severe right heart failure and markedly elevated pulmonary vascular resistance do not tolerate atrial septostomy, because massive right-to-left shunting may result in insufficient pulmonary blood flow and severe hypoxemia.

Therefore, in the cases of two boys who had suprasystemic pulmonary hypertension and right ventricular failure 4 years and 14 years after an arterial switch performed for transposition of the great arteries, we decided to create an anastomosis between the descending aorta and the left pulmonary artery (Potts shunt) (Fig. 1). The ratio of pulmonary blood flow to systemic blood flow was 1, as there was no intracardiac shunt in either boy. The aim of this intervention was to decrease right ventricular afterload so as to improve right ventricular function and potentially prevent syncope and sudden death. The procedures were uneventful.

The patients' condition improved rapidly, and both are now in New York Heart Association functional class II. Right ventricular function was restored. Right-to-left shunting through the Potts shunt was responsible for cyanosis of the lower limbs, and consequently, polycythemia developed in both patients (the hematocrit values were 51 percent and 55 percent at the last follow-up). The patients did not have any further episodes of syncope after a follow-up of 18 and 6 months, respectively. Hemodynamic evaluation confirmed that the pulmonary-artery pressure and aortic pressure were equal. Left-heart output was preserved.

The advantages of the Potts procedure are the immediate decrease in the right ventricular afterload and the absence of cyanosis of the upper part of the body in the absence of intracardiac shunting. The chief concern remains the operative risk in patients with pulmonary hypertension. Without treatment, the mean survival of children is less than one year after diagnosis.[2] For adults with pulmonary hypertension who are treated with prostacyclin analogues, the survival rate is estimated to be about 55 percent at five years,[4] but comparable studies in children have not been conducted. Median survival in adults with Eisenmenger's syndrome is 53 years.[5] Therefore, the Potts procedure could be an alternative for the treatment of children with severe pulmonary hypertension and right ventricular failure.

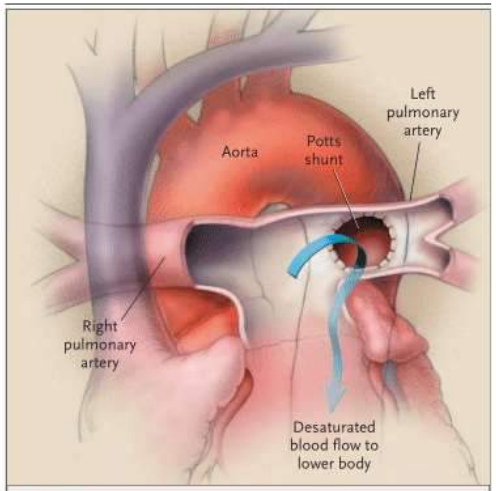


Figure 1. The Potts Shunt Procedure.
The left pulmonary artery is anastomosed to the descending aorta, allowing the desaturated blood to go from the left pulmonary artery to the lower part of the body (arrow). The right pulmonary artery passes in front of the ascending aorta because an arterial-switch procedure has been performed.

Julie Blanc, M.D.
Pascal Vouhé, M.D.
Damien Bonnet, M.D., Ph.D.
Hôpital Necker–Enfants Malades
75015 Paris, France
damien.bonnet@nck.ap-hop-paris.fr

1. Kumar A, Taylor GP, Sandor GG, Patterson MW. Pulmonary vascular disease in neonates with transposition of the great arteries and intact ventricular septum. Br Heart J 1993;69:442-5.
2. Thilenius OG, Nadas AS, Jockin H. Primary pulmonary vascular obstruction in children. Pediatrics 1965;36:75-87.
3. Kerstein D, Levy PS, Hsu DT, Hordof AJ, Gersony WM, Barst RJ. Blade balloon atrial septostomy in patients with severe primary pulmonary hypertension. Circulation 1995;91:2028-35.
4. Sitbon O, Humbert M, Simonneau G. Primary pulmonary hypertension: current therapy. Prog Cardiovasc Dis 2002;45:115-28.
5. Cantor WJ, Harrison DA, Moussadji JS, et al. Determinants of survival and length of survival in adults with Eisenmenger syndrome. Am J Cardiol 1999;84:677-81.

NEJM, 2004

Potts Shunt in Children With Idiopathic Pulmonary Arterial Hypertension: Long-Term Results

Alban-Elouen Baruteau, MD, Alain Serraf, MD, PhD, Maryline Lévy, MD, PhD, Jérôme Petit, MD, Damien Bonnet, MD, PhD, Xavier Jais, MD, Pascal Vouhé, MD, PhD, Gérald Simonneau, MD, Emre Belli, MD, and Marc Humbert, MD, PhD

Centre Chirurgical Marie Lannelongue, Département de Chirurgie cardiaque des cardiopathies congénitales, Le Plessis-Robinson; Centre de Référence Malformations Cardiaques Congénitales Complexes–M3C, Paris; Université Paris-Sud, Le Kremlin-Bicêtre; Hôpital Jacques Cartier, Massy; Université Paris Descartes, Paris; Hôpital Necker-Enfants Malades, APHP, Paris; INSERM U999, Le Plessis-Robinson and Clamart; and Hôpital Antoine Béclère, APHP, Service de Pneumologie et Réanimation Respiratoire, Centre National de Référence de l'Hypertension Pulmonaire Sévère, Clamart, France

Background. Idiopathic pulmonary arterial hypertension (IPAH) remains a progressive fatal disease. Palliative Potts shunt has been proposed in children displaying suprasystemic IPAH.

Methods. A retrospective multicenter study was performed to evaluate Potts shunt in pediatric IPAH.

Results. Between 2003 and 2010, 8 children with suprasystemic IPAH and in World Health Organization functional class IV despite medical pulmonary arterial hypertension therapy underwent Potts shunt. Age at IPAH diagnosis ranged from 4 to 180 months (median age, 64 months). Surgical procedure was performed in a mean delay of 41.9 ± 54.3 months (range, 4 to 167 months; median delay, 20 months) after IPAH diagnosis. Mean size of the Potts shunt was 9.25 ± 3.30 mm. Two patients, whose medical pulmonary arterial hypertension therapy had been interrupted just after surgery, died at postoperative days 11 and 13 of acute pulmonary hypertensive crisis. After a mean follow-up of 63.7 ± 16.1 months, the 6 children who were discharged from the hospital were alive. Functional status improved markedly in the 6 survivors, with a World Health Organization functional class I (n = 4) or II (n = 2) at last follow-up, consistent with significant improvement of 6-minute-walk distance (302 ± 95 m [51% ± 20% of theoretical values] versus 456 ± 91 m [68% ± 10% of theoretical values]; $p = 0.038$) and decrease of brain natriuretic peptide levels (608 ± 109 pg/mL versus 76 ± 45 pg/mL; $p = 0.035$). No Potts shunt was found to be restrictive at last echocardiography.

Conclusions. Palliative Potts shunt constitutes a new alternative to lung transplantation in severely ill children with suprasystemic IPAH, carrying a prolonged survival and persistent improvement in functional capacities.

(Ann Thorac Surg 2012;94:817–24)

Potts Registry

St Louis, USA
Houston, USA
Omaha, USA
Seattle, USA
Boston, USA
Cincinnati, USA
Denver, USA
Milwaukee, USA

Paris, France
London, England
Madrid, Spain
Moscow, Russia
Groningen, Netherlands

JOURNAL OF THE AMERICAN COLLEGE OF CARDIOLOGY VOL. 78, NO. 5, 2021

Pulmonary-to-Systemic Arterial Shunt to Treat Children With Severe Pulmonary Hypertension

R. Mark Grady, MD,[a] Matthew W. Canter, MET,[b] Fei Wan, PhD,[b] Anton A. Shmalts, MD,[c] Ryan D. Coleman, MD,[d] Maurice Beghetti, MD,[e] Rolf M.F. Berger, MD, PhD,[f] Maria J. del Cerro Marin, MD, PhD,[g] Scott E. Fletcher, MD,[h] Russel Hirsch, MD,[i] Tilman Humpl, MD,[j] D. Dunbar Ivy, MD,[k] Edward C. Kirkpatrick, DO,[l] Thomas J. Kulik, MD,[m] Marilyne Levy, MD, PhD,[n] Shahin Moledina, MD,[o] Delphine Yung, MD,[p] Pirooz Eghtesady, MD, PhD,[b] Damien Bonnet, MD, PhD,[n] on behalf of the International Registry Potts Shunt

ABSTRACT

BACKGROUND The placement of a pulmonary-to-systemic arterial shunt in children with severe pulmonary hypertension (PH) has been demonstrated, in relatively small studies, to be an effective palliation for their disease.

OBJECTIVES The aim of this study was to expand upon these earlier findings using an international registry for children with PH who have undergone

METHODS Retrospective da
from 13 institutions in Europe

RESULTS Seventeen childre
subsequently died or underwe
The overall 1- and 5-year free
those discharged home, respe
functional class ($P < 0.001$),
Postprocedure, 59% of childr
factors for dying in-hospital p
mechanical ventilation (HR: 8.

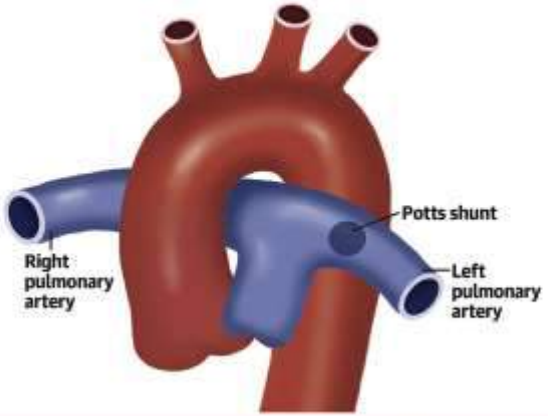
CONCLUSIONS A pulmonar
improvement that is both du
comparable to children under
aggressive intensive care are
© 2021 the American College

Post-procedure, **59%** of children were weaned completely from their prostacyclin infusion ($P < 0.001$)

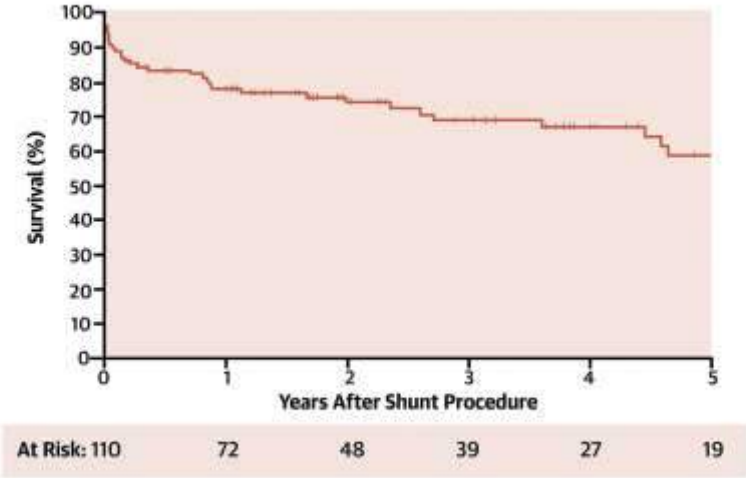
"Potts shunts provide durable, clinical benefit to children with severe (supra-systemic) PH with potentially less medicine"

CENTRAL ILLUSTRATION: Potts Shunt and Pulmonary Hypertension

Potts Shunt and Pulmonary Hypertension



Freedom from Death or Lung Transplant Post-Shunt Procedure



Grady, R.M. et al. J Am Coll Cardiol. 2021;78(5):468-77.

but what about…..

Timing of shunt

Critically ill child with severe PH and a failing RV

Lung transplantation after shunt

Risk Factors for Early Mortality

		Hazard ratio	95% CI	p-value
Pre-procedure data				
Age at time of procedure		1.0	0.9, 1.1	0.7
Gender		1.2	0.5, 3.1	0.7
History of CHD		1.4	0.5, 3.8	0.6
Time from diagnosis to procedure		1.0	0.9,1.0	0.1
WHO PH type:	Type-1	0.4	0.2, 1.2	0.1
	Type-2	0.04	0, 138	0.5
	Type-3	5.4	1.9, 15	0.002
Use of IV/SQ PGI2		0.7	0.3, 1.7	0.4
ICU admission		3.2	1.2, 8.3	0.02
Intravenous inotropes		2.9	1.1, 7.9	0.04
Mechanical ventilation		8.3	3.1, 22	<0.001
ECMO		10.7	3.9, 29	<0.001
Operative data: Shunt type				
Surgical		0.7	0.3, 1.8	0.4
PDA stent		0.7	0.2, 2.3	0.5
Transcatheter		3.6	1.2, 11	0.03

J Am Coll Cardiol. 2021;78(5):468-477

Other Operative considerations

- Age
- Presence of ASD (and size)
- Presence of TR/PR
- Size of Shunt
- Valved or non-valved
- PH due to L side hypoplasia
- Collaterals
- Preop workup
- Post Op Management/Anticoagulation?


CARDIOLOGY
2024
27th Annual Update on
Pediatric and Congenital
Cardiovascular Disease
Actual (Not Artificial)
Intelligence in Pediatric &
Congenital Cardiovascular Disease
What do we know and what

THANK YOU!