Cardiology 2023: 26th Annual Update on Pediatric and Congenital Cardiovascular Disease

How Best to Augment Pulmonary Blood Flow in the Newborn with Congenital Pulmonary Obstruction?

Andrew C. Glatz, MD MSCE
February 25, 2023

Plenary Session IV:
The Pulmonary Circulation in Pediatric & Congenital Heart Disease

Children's Hospital • St. Louis
BJC HealthCare
Washington University in St. Louis
Physicians

Disclosures

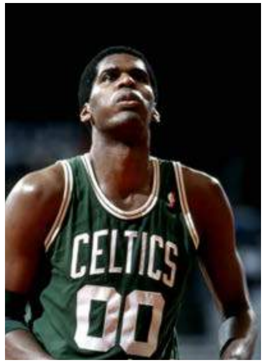
- None

Boston Celtics Starting Five 1985

Criteria for decision-making

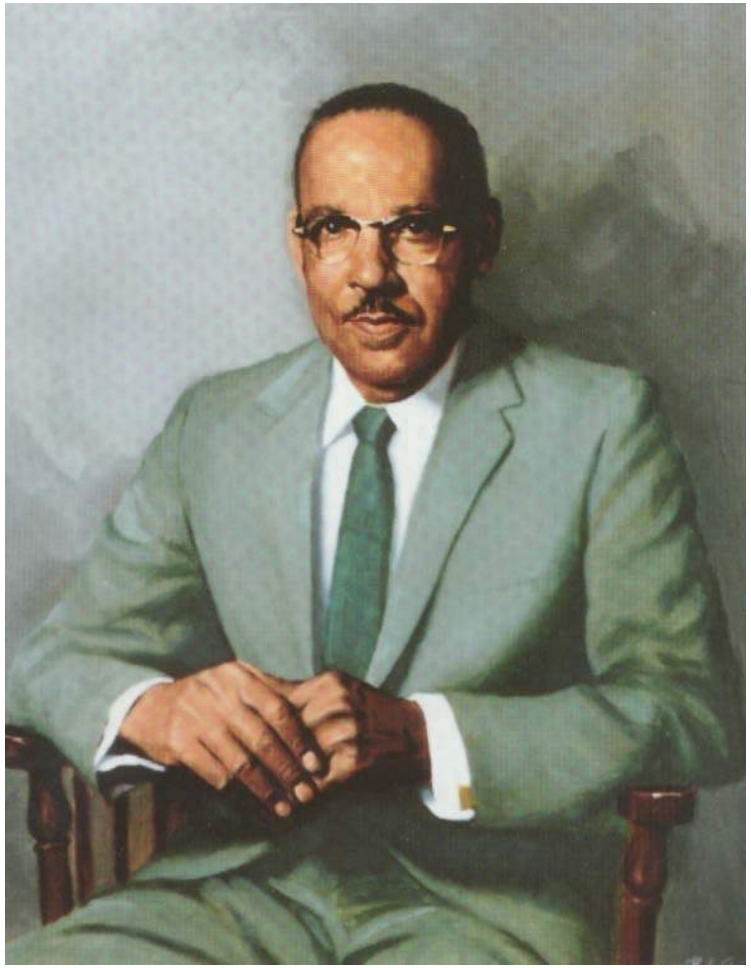
- Minimize morbidity of initial intervention
- Optimize "inter-stage" physiology
- Set up the surgeon for success at next (first) operation
- Maximize longer-term outcomes

November 29, 1944



B-T-T Shunt

Alfred Blalock



Vivien Thomas

Helen Taussig

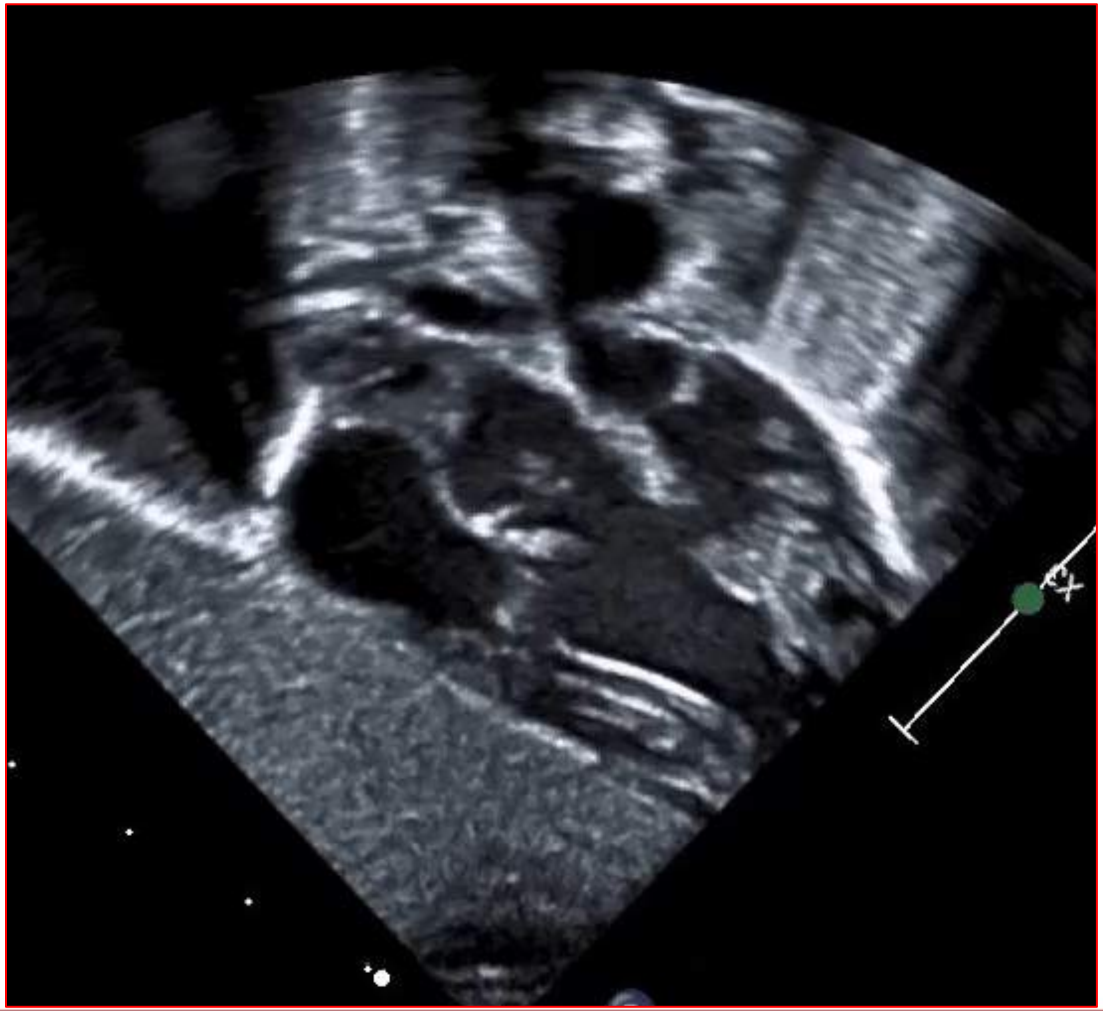
Transcatheter Options

- Overall appeal
  - Reduced exposure to neonatal bypass, anesthesia, ICU morbidities
  - First sternotomy is for complete repair or “stage 2”

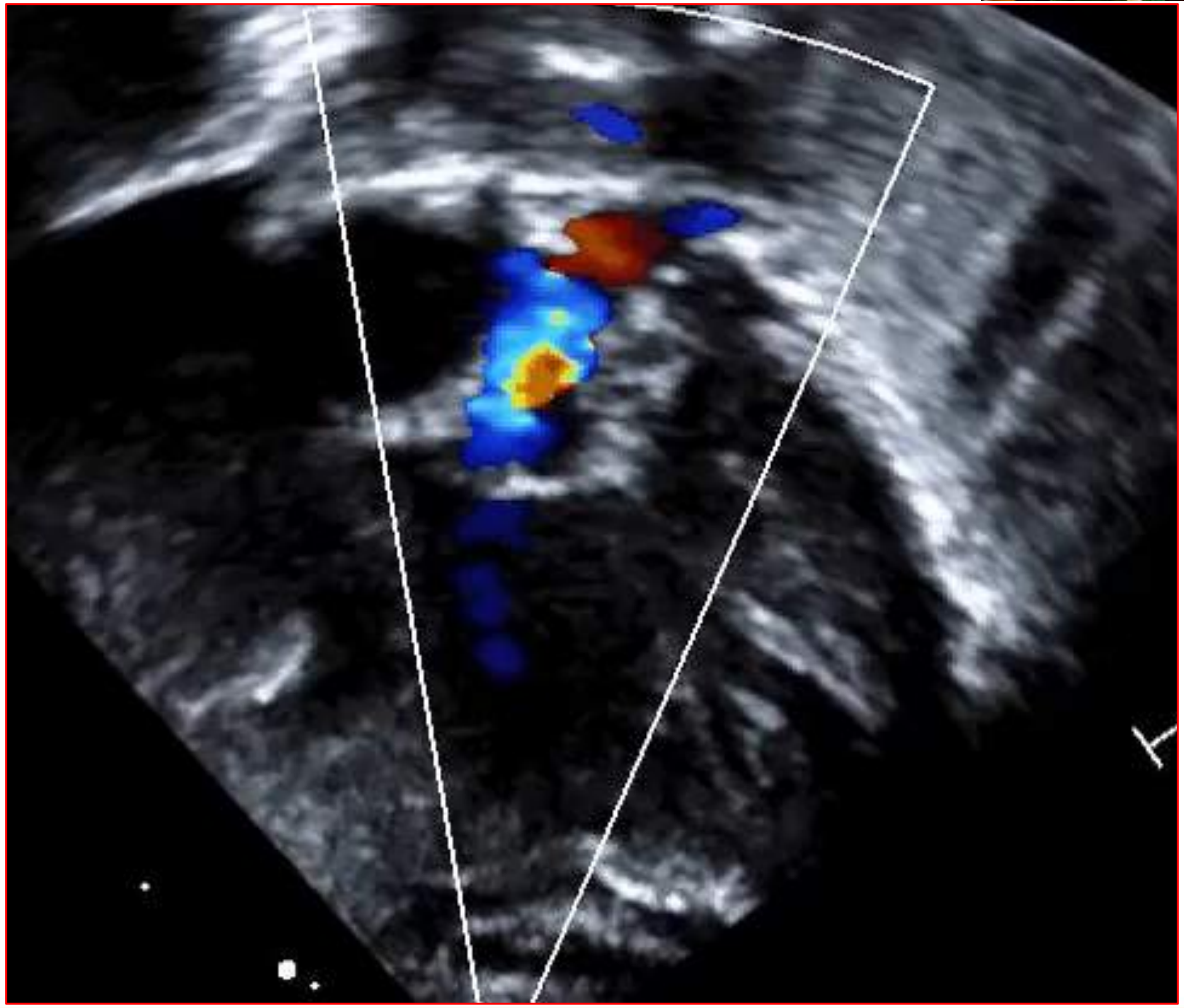
- Pulmonary outflow tract interventions
  - Balloon pulmonary valvuloplasty
  - RVOT stenting

- PDA stenting

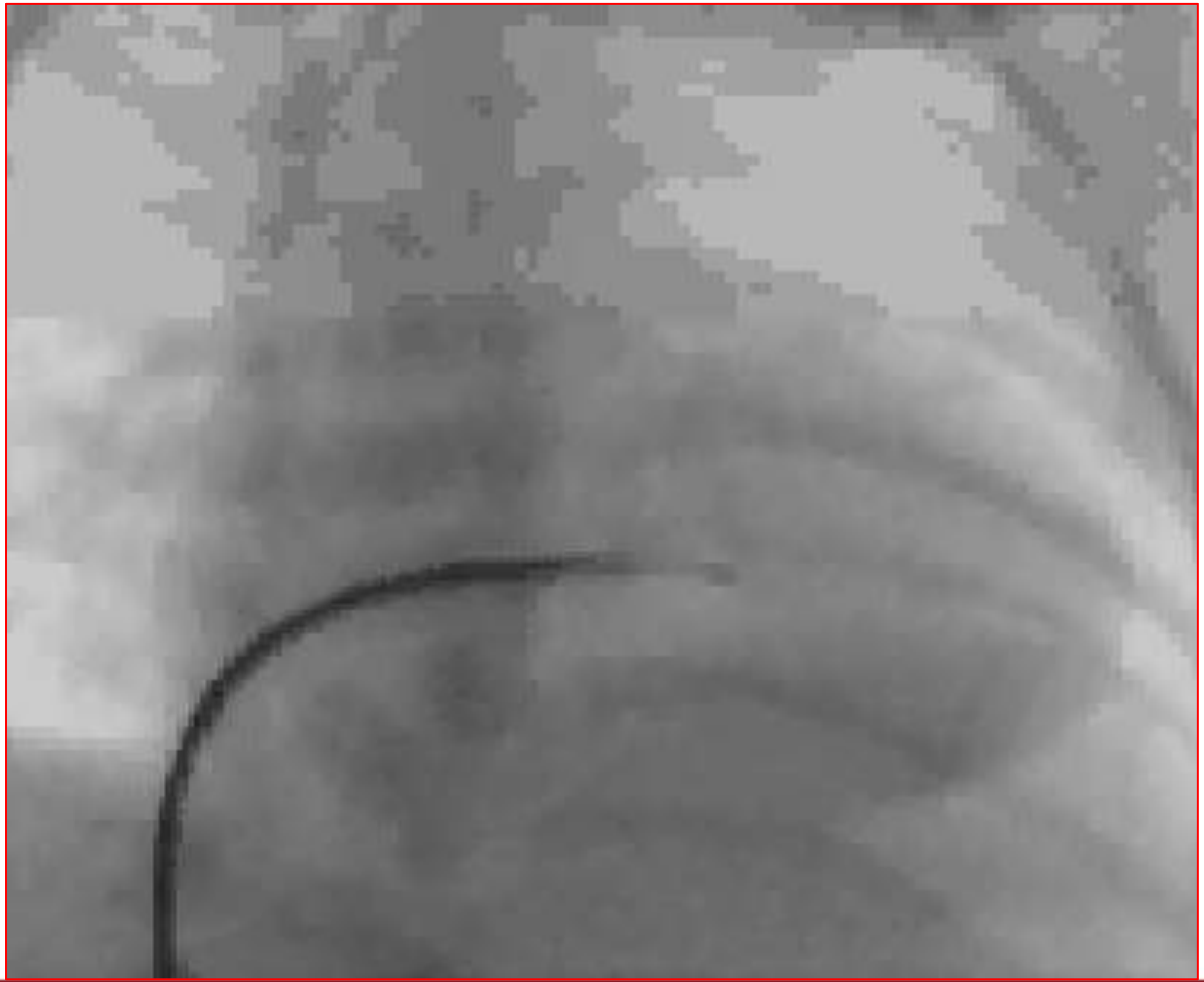
Balloon Pulmonary Valvuloplasty



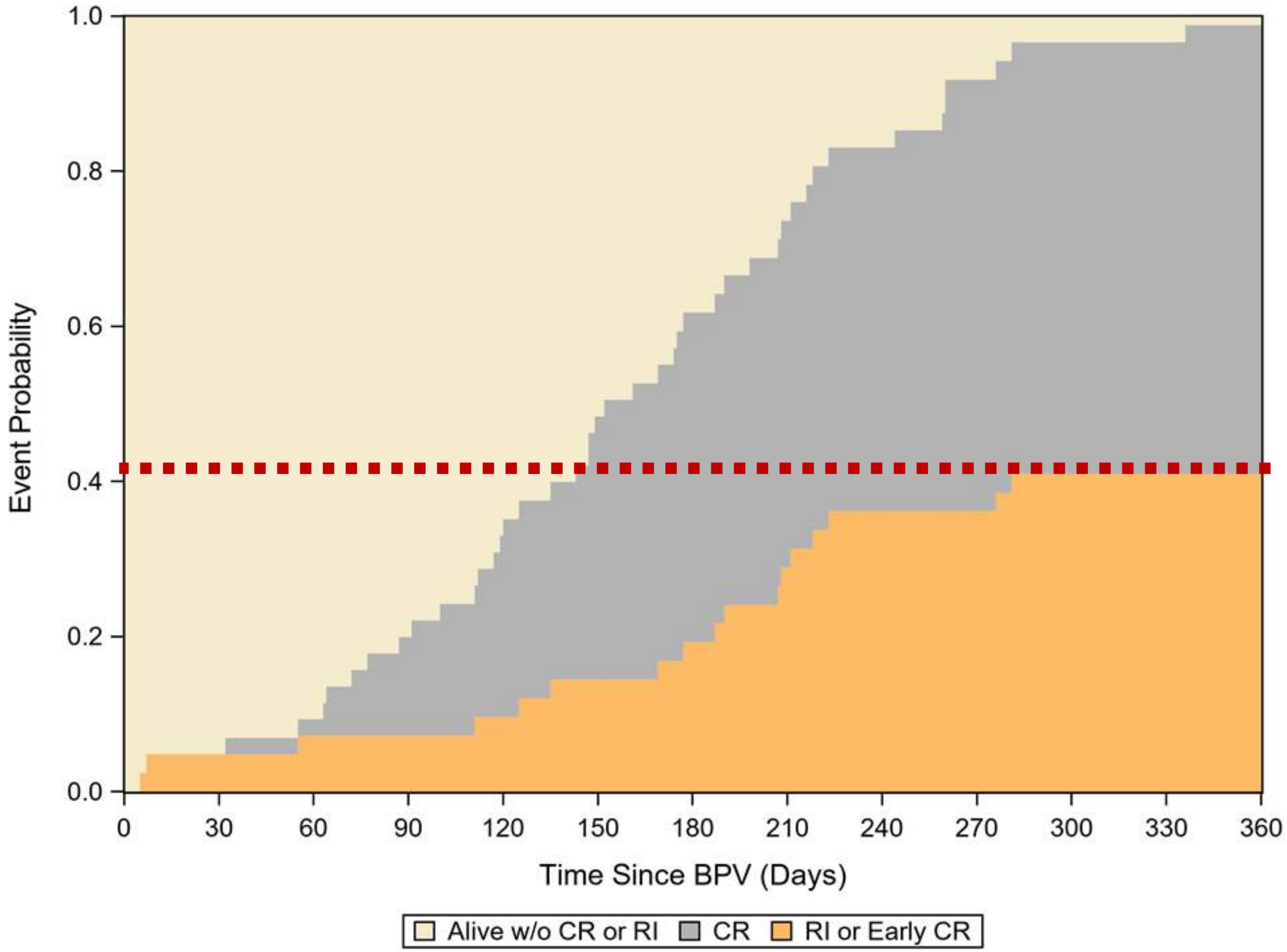
Balloon Pulmonary Valvuloplasty



Balloon Pulmonary Valvuloplasty



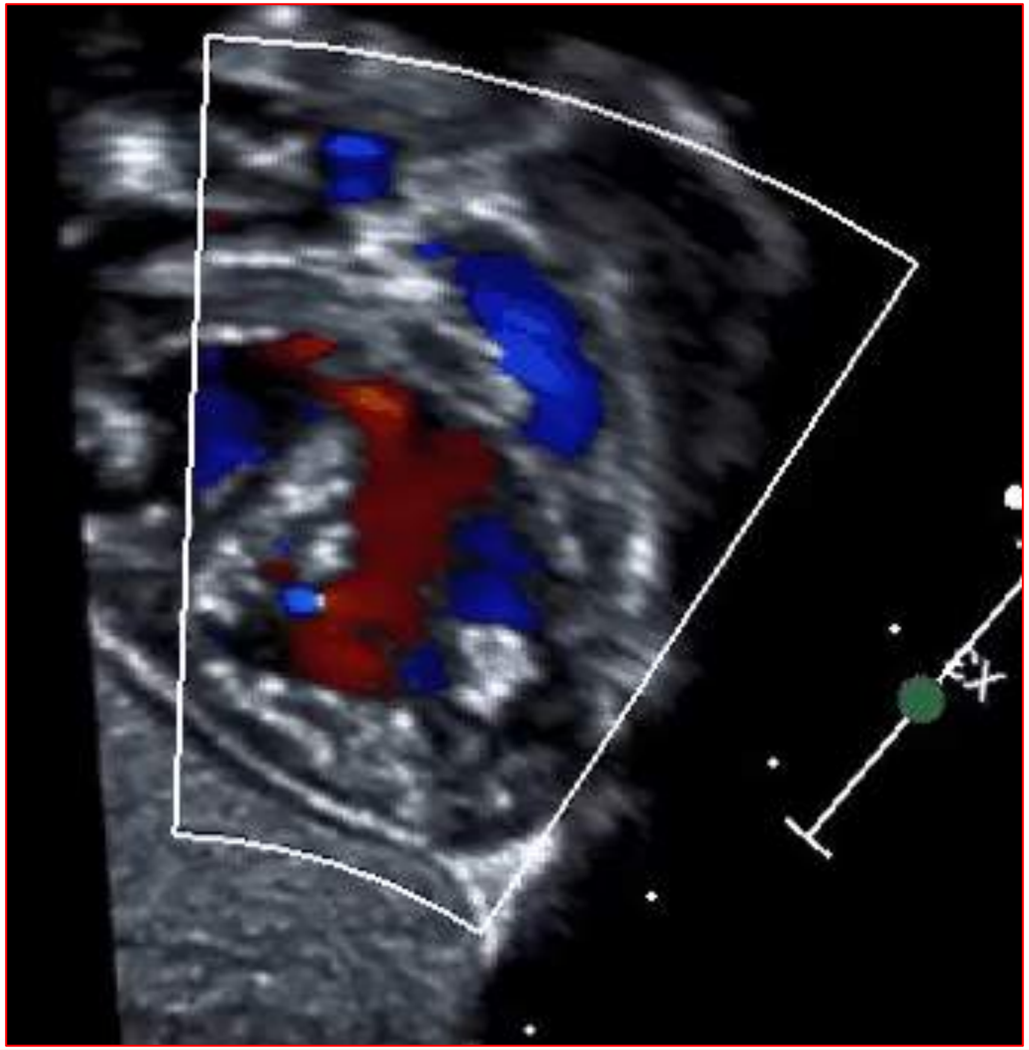
Balloon Pulmonary Valvuloplasty



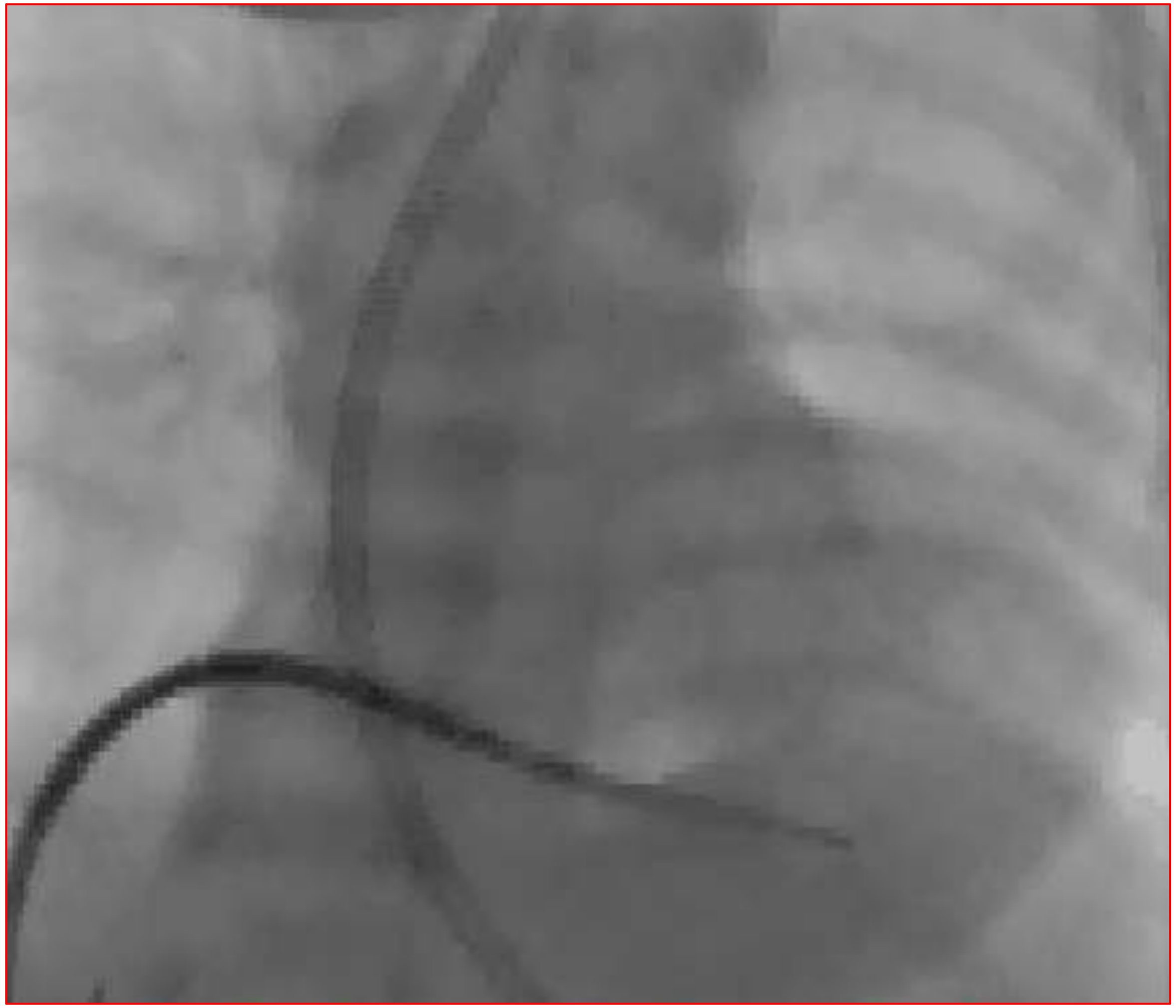
Shahanavaz et al., *Under review*

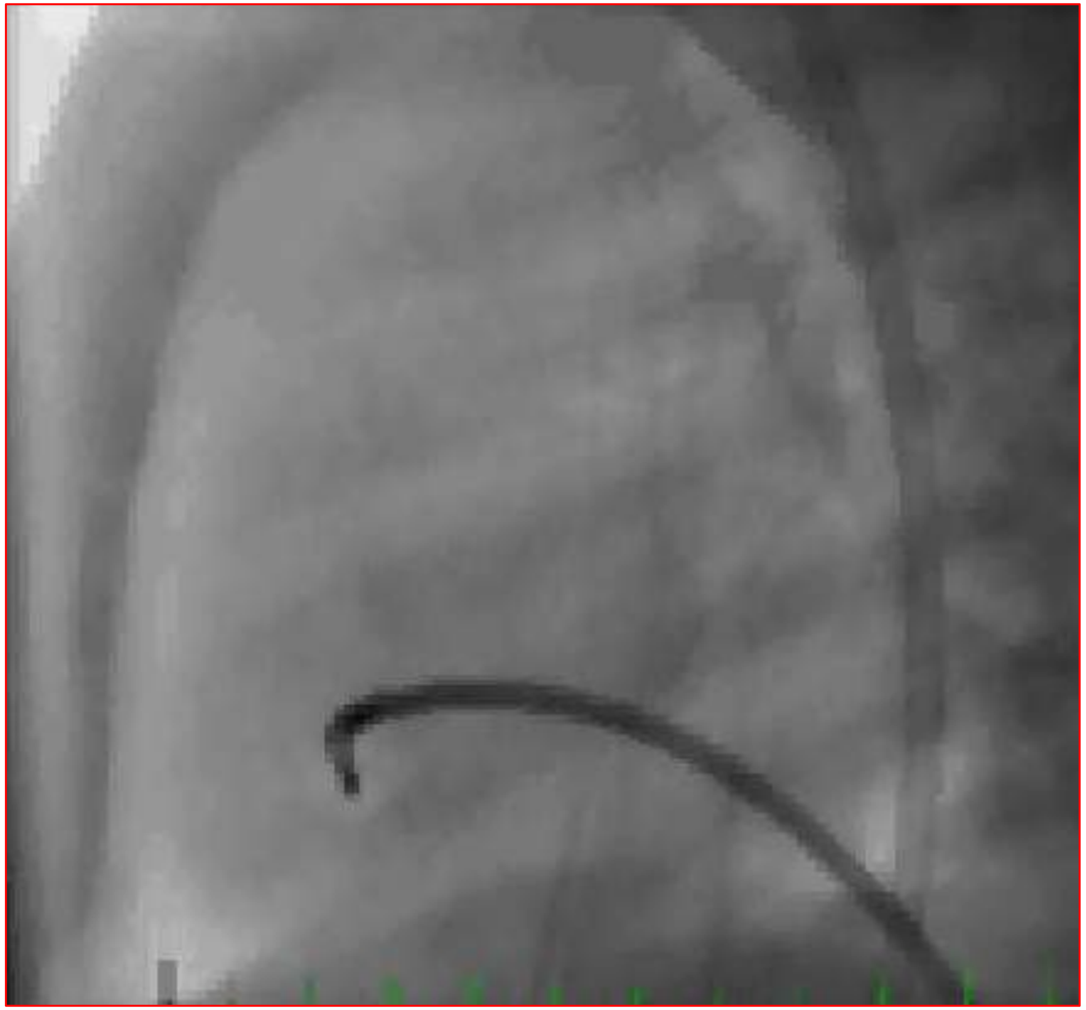
CONGENITAL CARDIAC RESEARCH COLLABORATIVE

RVOT Stenting

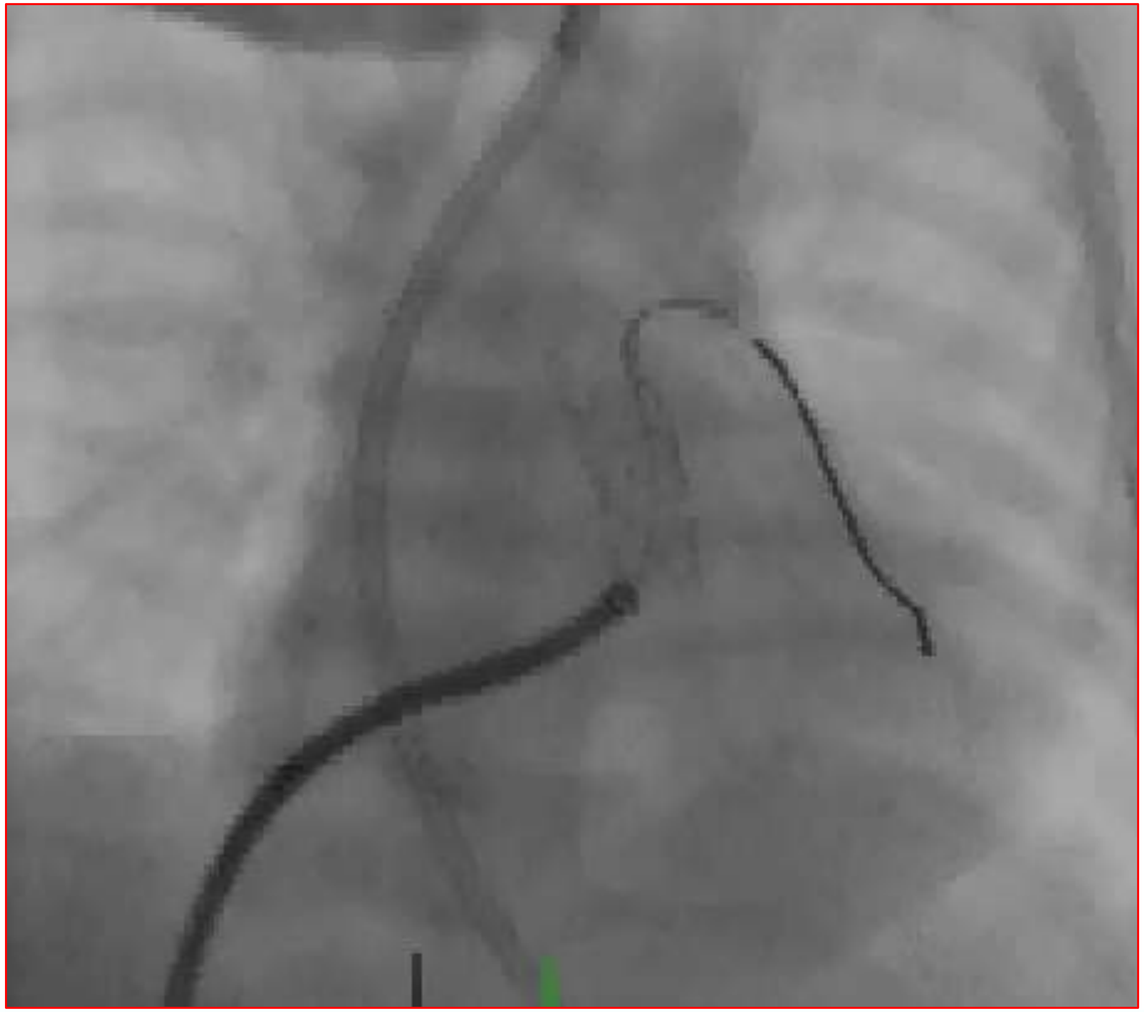


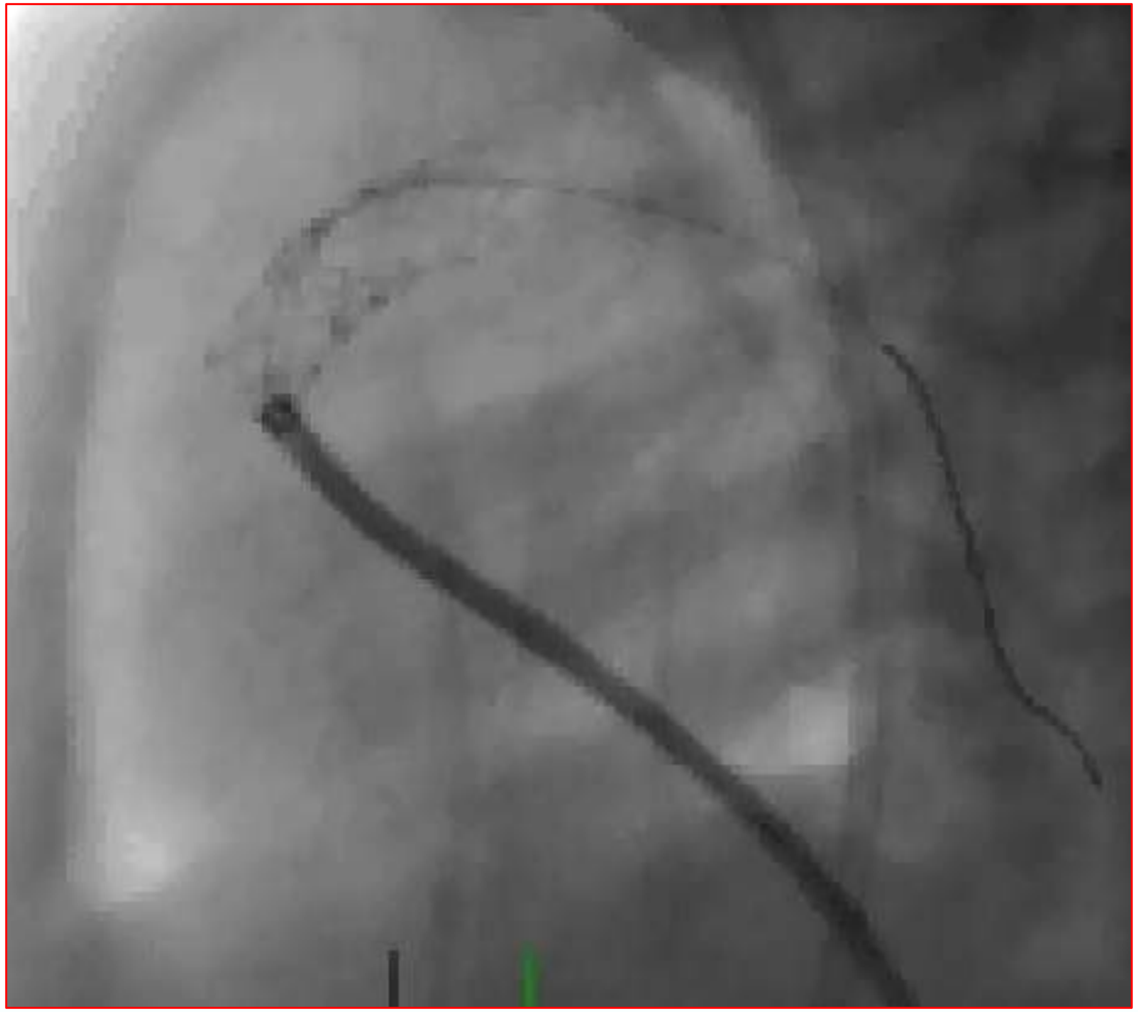
RVOT Stenting





RVOT Stenting





RVOT Stenting

- 42 infants with RVOT stents compared to:
  - Early (<3 months) primary repair
  - Elective primary repair
- RVOT stents
  - Patients with risk factors for primary repair
  - Durable
  - "Catch up" PA growth
  - Similar clinical outcomes at time of complete repair
    - 95% removed completely

Congenital Heart Disease

Right Ventricular Outflow Tract Stenting in Tetralogy of Fallot Infants With Risk Factors for Early Primary Repair

Juan Pablo Sandoval, MD*; Rajiv R. Chaturvedi, MB BChir, MD, PhD*; Lee Benson, MD; Gareth Morgan, MD; Glen Van Arsdell, MD; Osami Honjo, MD, PhD; Christopher Caldarone, MD; Kyong-Jin Lee, MD

Background—Tetralogy of Fallot with cyanosis requiring surgical repair in early infancy reflects poor anatomy and is associated with more clinical instability and longer hospitalization than those who can be electively repaired later. We bridged symptomatic infants with risk factors for early primary repair by right ventricular outflow tract stenting (stent).

Methods and Results—Four groups of tetralogy of Fallot with confluent central pulmonary arteries were studied: stent group (n=42), primary repair (aged <3 months) with pulmonary stenosis (early-PS group; n=44), primary repair (aged <3 months) with pulmonary atresia (early-PA group; n=49), and primary repair between 3 and 11 months of age (surg>3mo group; n=45). Stent patients had the smallest pulmonary arteries with a median (95% credible intervals) Nakata index (mm^2/m^2) of 79 (66–85) compared with the early-PA 139 (129–154), early-PS 136 (121–153), and surg>3mo 167 (153–200) groups. Only stent infants required unifocalization of aortopulmonary collaterals (17%). Stent and early-PA infants had younger age and lower weight than early-PS infants. Stent infants had the most multiple comorbidities. Stenting allowed deferral of complete surgical repair to an age (6 months), weight (6.3 [5.8–7.0] kg), and Nakata index (147 [132–165]) similar to the low-risk surg>3mo group. The 3 early treatment groups had similar intensive care unit/hospital stays and high reintervention rates in the first 12 months after repair, compared with the surg>3mo group.

Conclusions—Right ventricular outflow tract stenting of symptomatic tetralogy of Fallot with poor anatomy (small pulmonary arteries) and adverse factors (multiple comorbidities, low weight) relieves cyanosis and defers surgical repair. This allowed pulmonary arterial and somatic growth with clinical results comparable to early surgical repair in more favorable patients. (*Circ Cardiovasc Interv*. 2016;9:e003979. DOI: 10.1161/CIRCINTERVENTIONS.116.003979.)

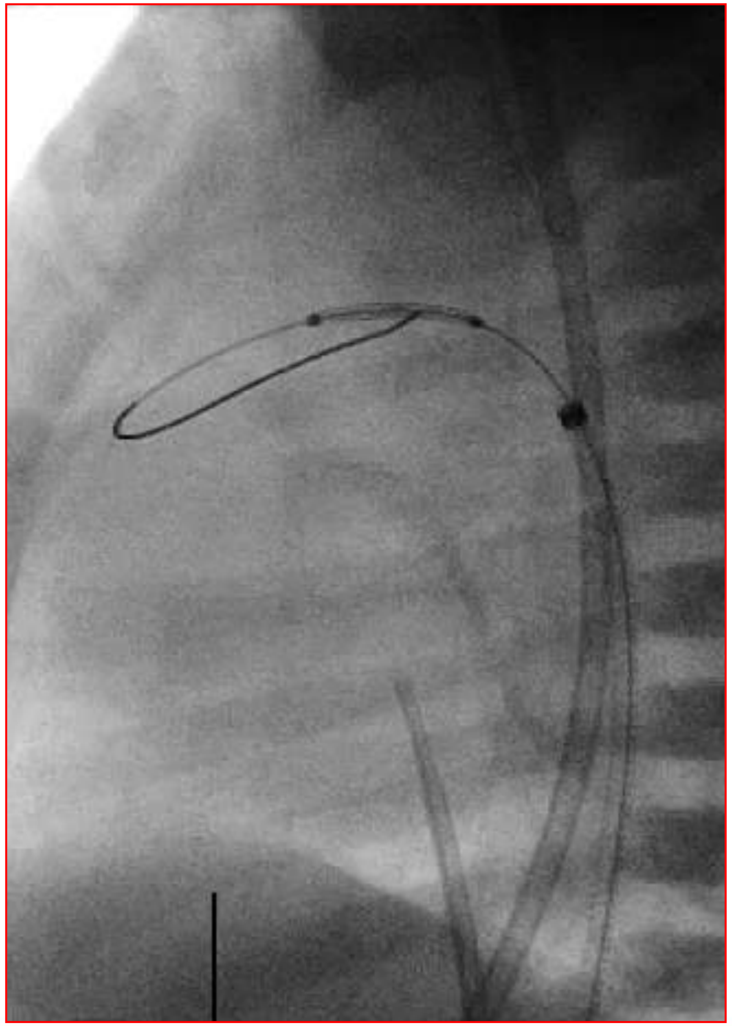
Invited editorial: Glatz, *Circ Cardiovasc Interv*, 2016

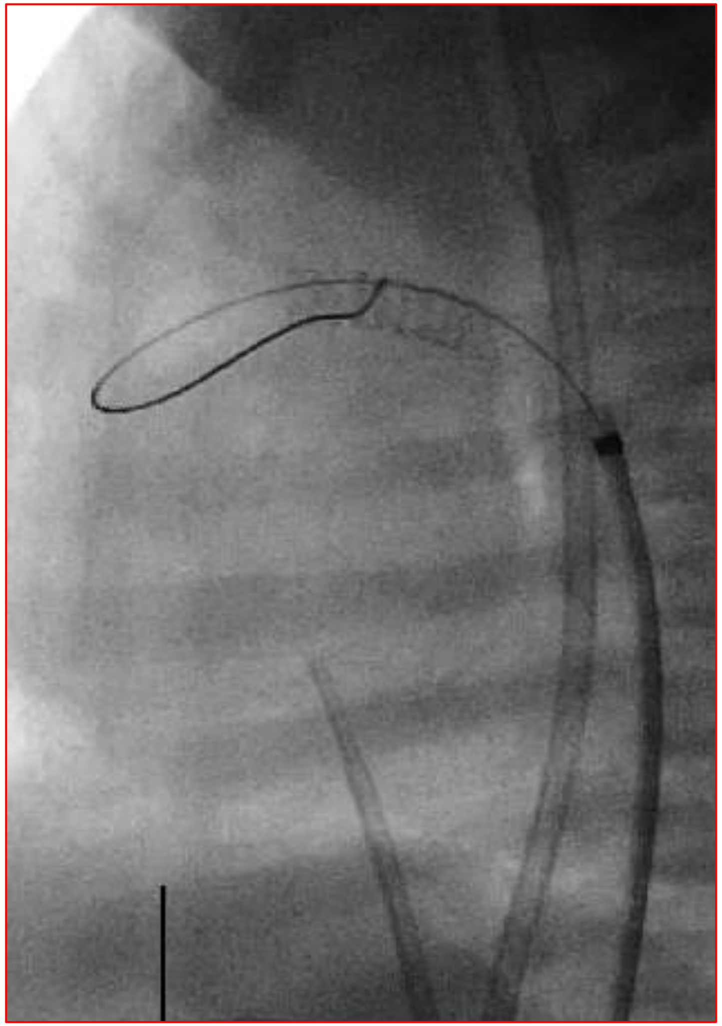
RVOT Interventions


- PROS
  - "Physiologic" pulmonary blood flow
    - Better PA growth (?)
    - No diastolic run-off
  - Straightforward access

- CONS
  - Anatomic restrictions
    - BPV: primarily valvar obstruction
    - RVOT stent: infundibular-MPA continuity
  - Durability (BPV)
  - May complicate subsequent surgery (RVOT stent)
  - Precludes "valve-sparing" repair (RVOT stent)

PDA stenting





PDA stenting vs. BTTS

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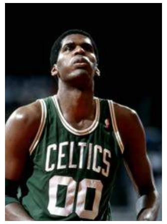
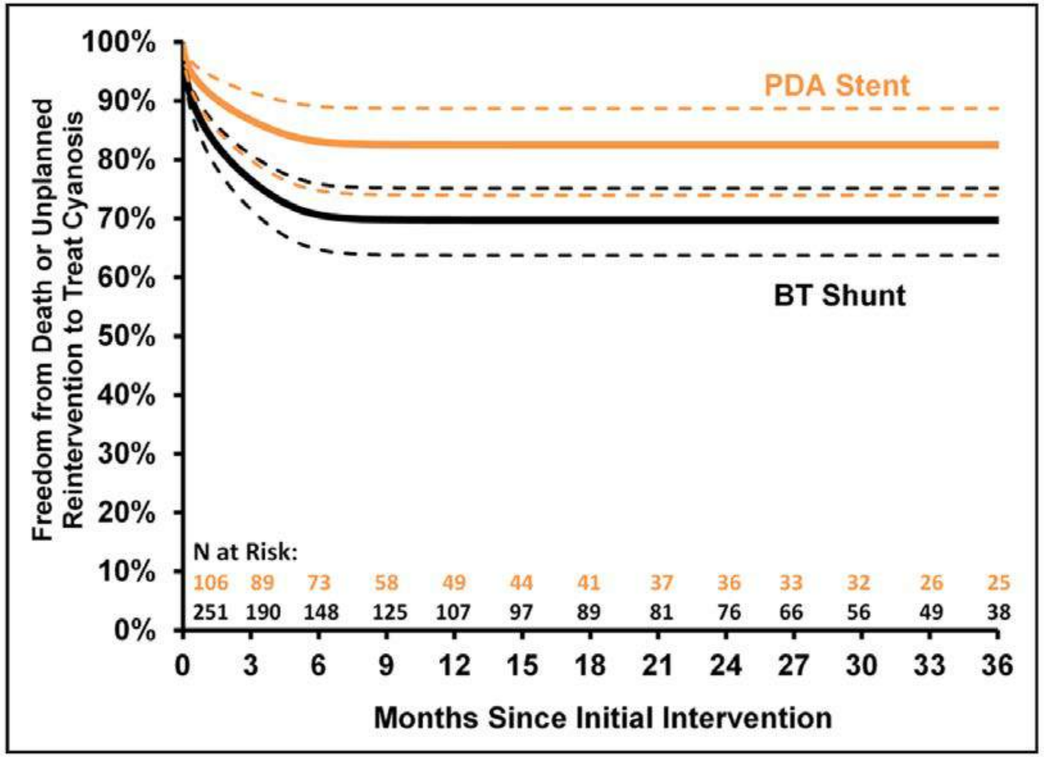
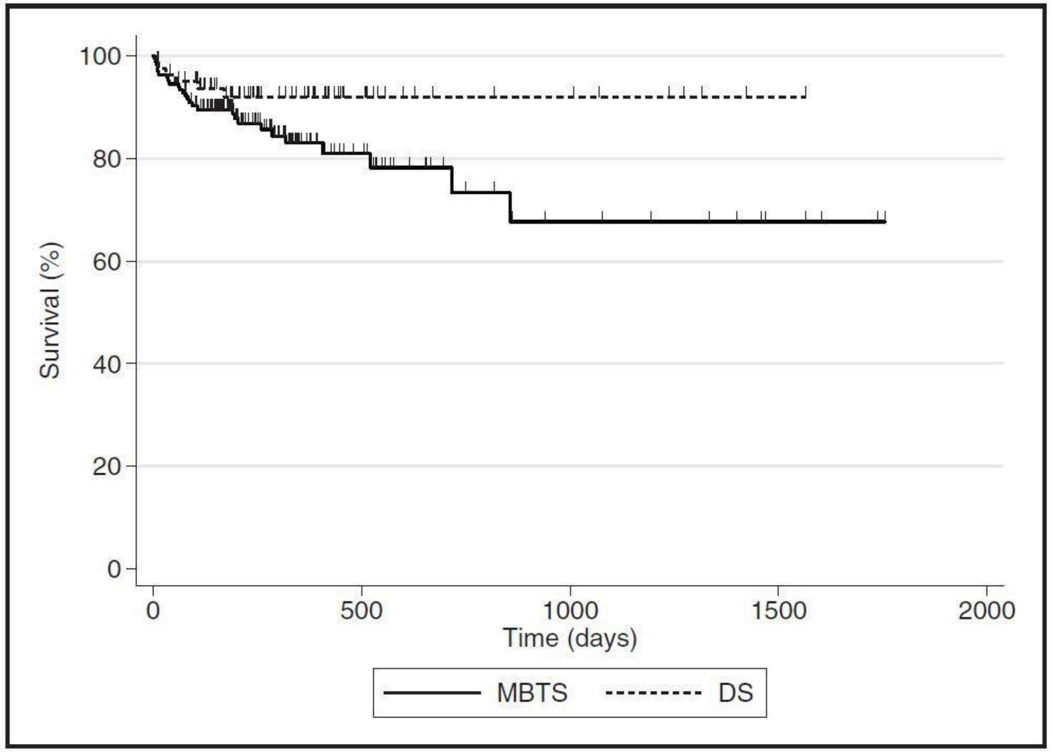




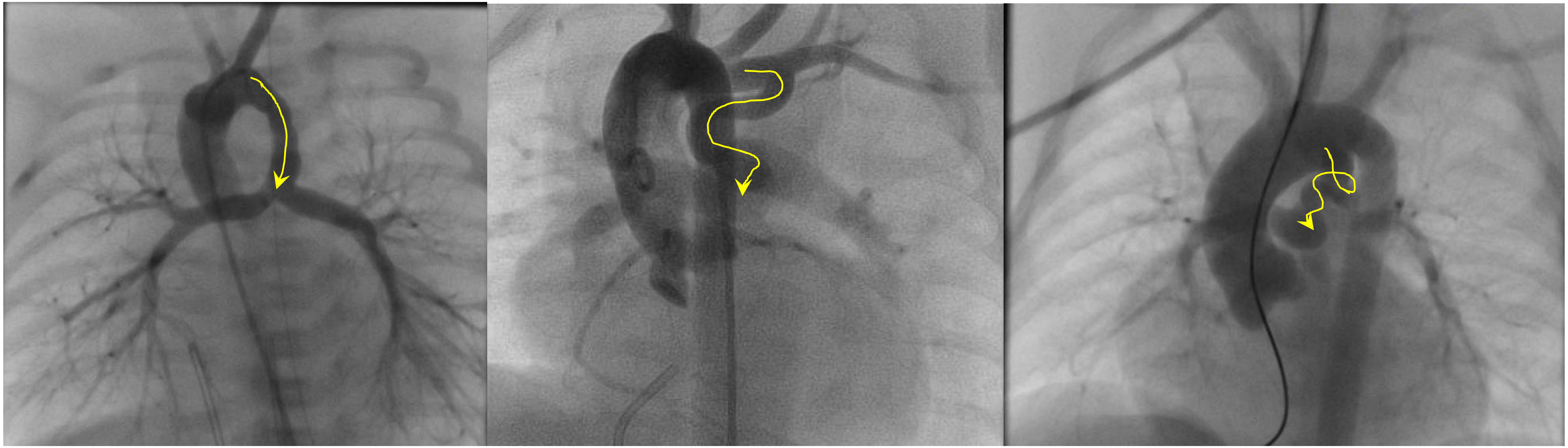
Glatz et al., *Circulation*. 2018



Bentham et al., *Circulation*. 2018

CCRC
CONGENITAL CARDIAC RESEARCH COLLABORATIVE

Can (should) all ducts be stented?



Type I (n=58) | Type II (n=24) | Type III (n=23)

- Higher risk of:
  - PA jailing
  - Reintervention
  - PA plasty

Qureshi et al., *Catheter Cardiovasc Interv*. 2019

CONGENITAL CARDIAC RESEARCH COLLABORATIVE

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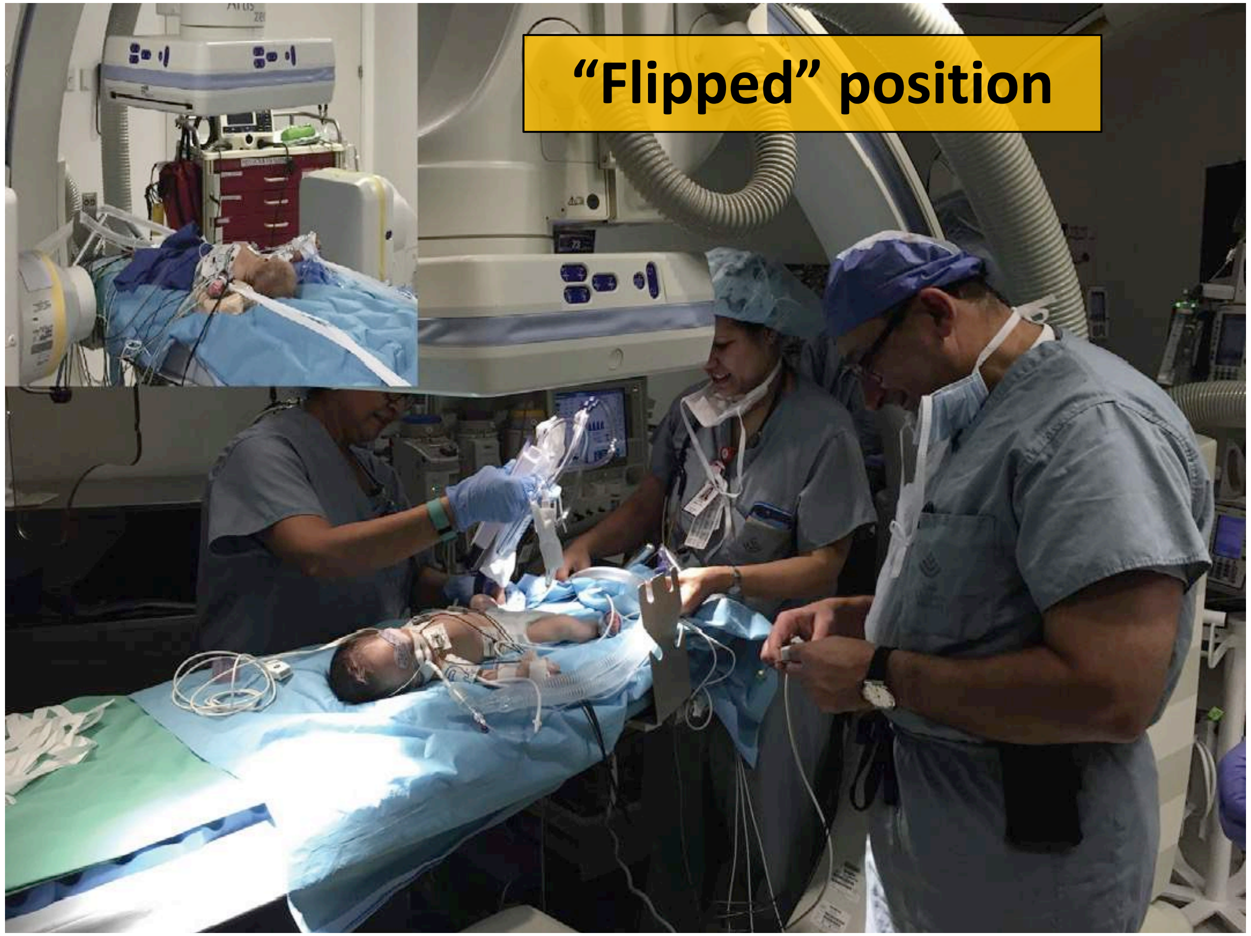
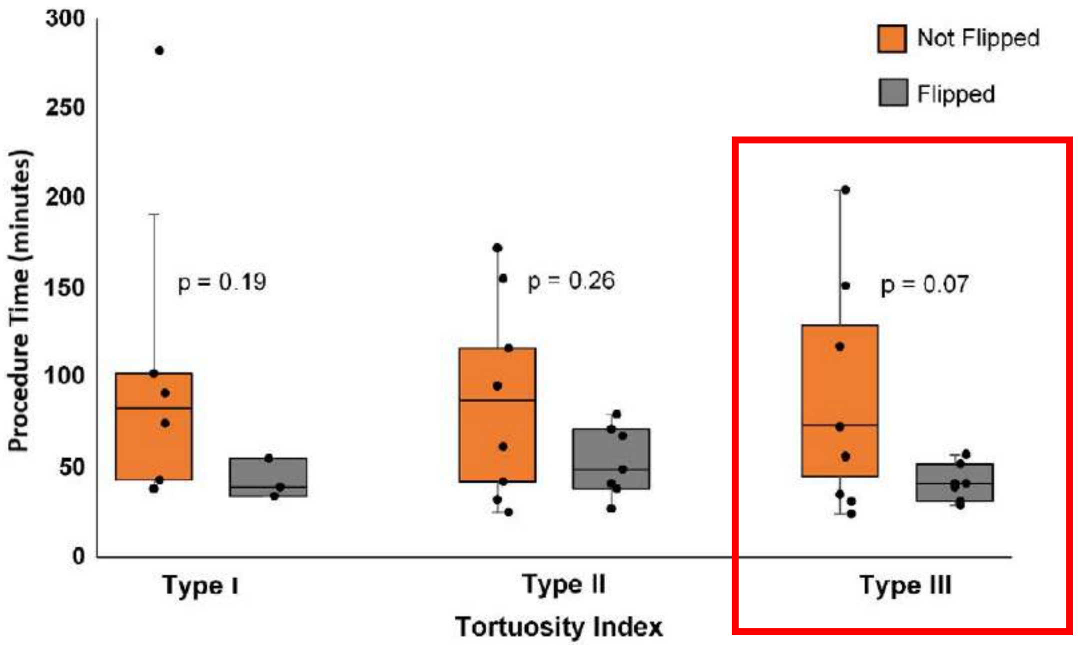


Bauser-Heaton et al., *JACC Cardiovasc Interv*, 2019



Bauser-Heaton et al., *Catheter Cardiovasc Interv*, 2020

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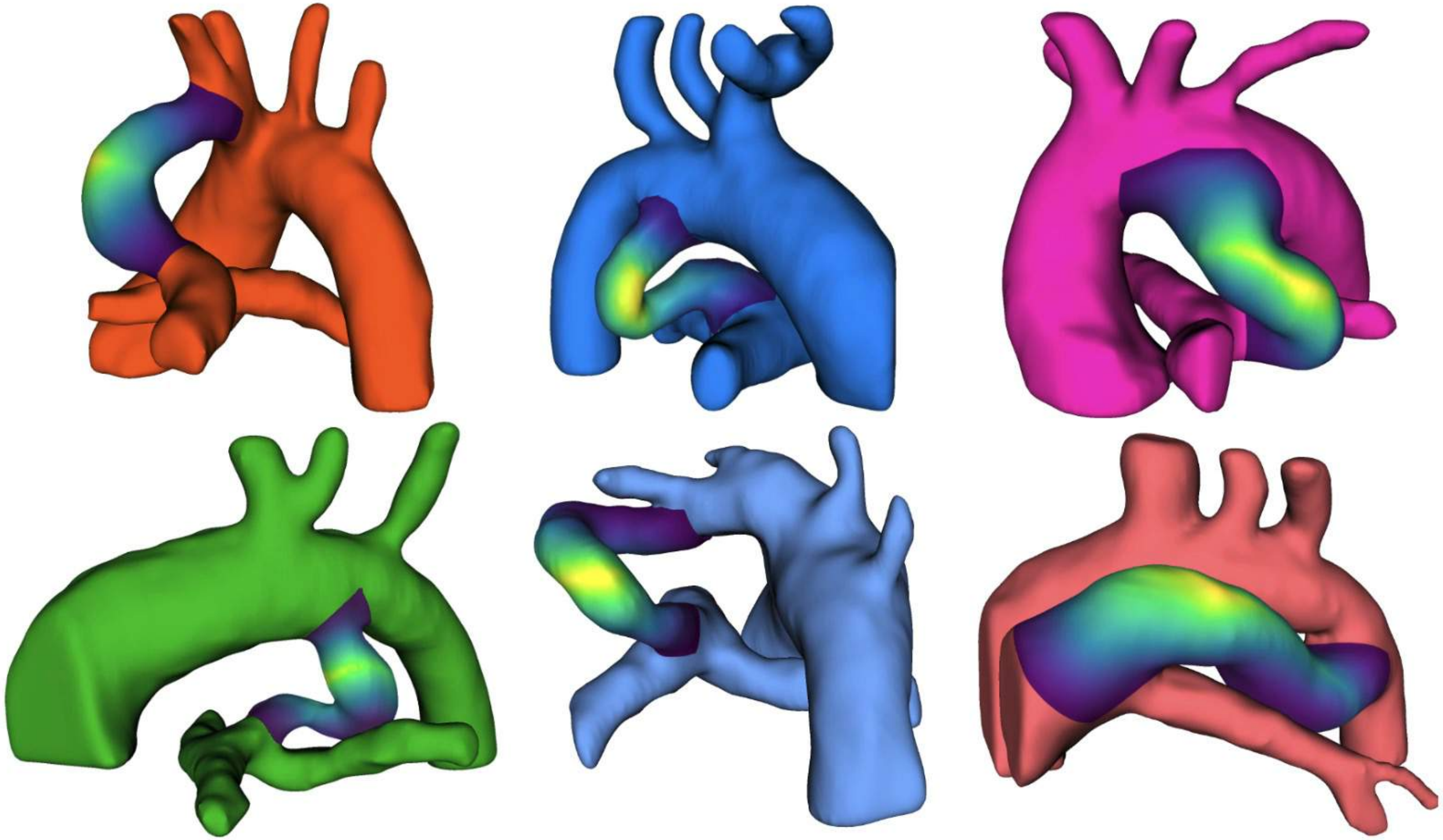
RESEARCH COLLABORATIVE



Video courtesy of Mudit Gupta MD PhD and Matthew Jolley MD

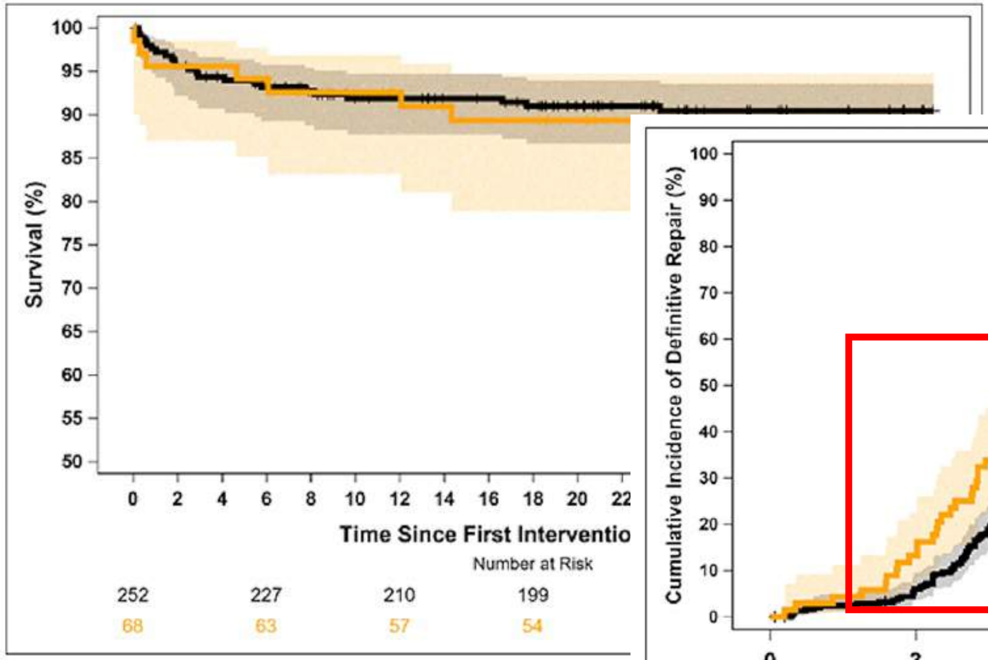
PDA stenting (compared to BTTS)

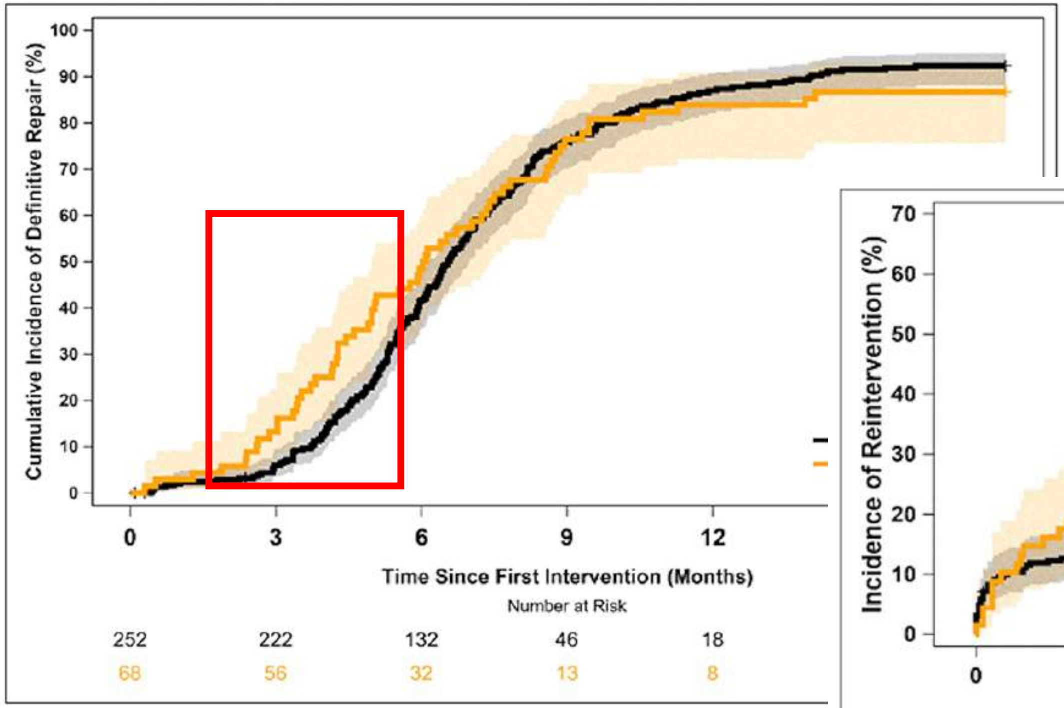
- Pros
  - Minimally invasive
    - Less morbidity and ?mortality
  - Better PA growth
  - Dilatable
- Cons
  - Reintervention
  - Anatomic complexity
  - Technically challenging

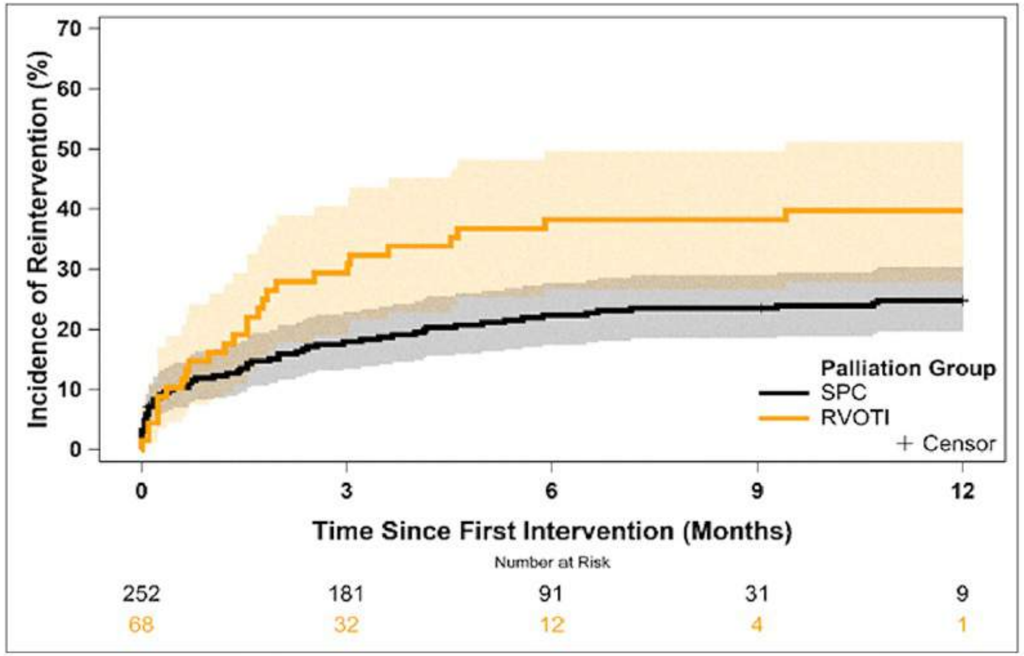
Which physiology is better?

BTTS and DAS

BPV and RVOT stent







Law et al., *Pediatric Cardiology*,

CONGENITAL CARDIAC RESEARCH COLLABORATIVE

Criteria for decision-making

- Minimize morbidity of initial intervention  

- Optimize "inter-stage" physiology **?**
- Set up the surgeon for success at next (first) operation 
- Maximize longer-term outcomes **?**

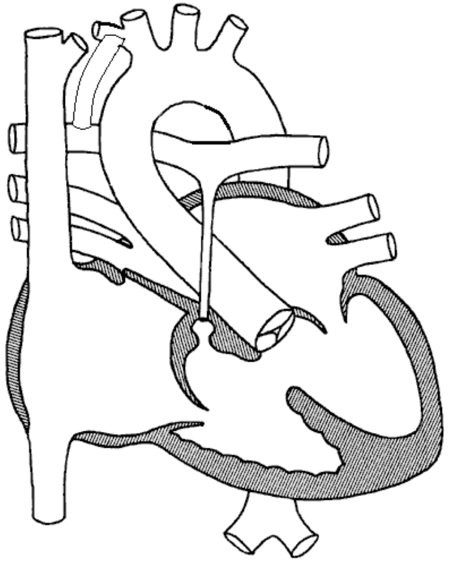
COMPASS Trial

COmparison of **M**ethods for **P**ulmonary Blood Flow **A**ugmentation in Neonates: **S**hunt versus **S**tent

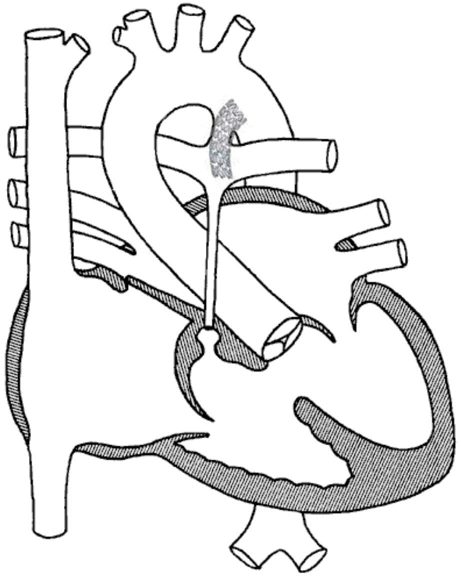
COMPASS

COMPASS Protocol

300 *neonates with DD-PBF randomized to:*



OR



SPS **DAS**

2 years enrollment, 12 months follow-up

COMPASS Sites

COMPASS

Children's Hospital of Philadelphia

Children's Hospital • St. Louis
BJC HealthCare

Morgan Stanley Children's Hospital of NewYork-Presbyterian
Columbia University Medical Center

C.S. Mott Children's Hospital
Michigan Medicine

Texas Children's Hospital

Boston Children's Hospital

Children's Wisconsin

Le Bonheur Children's Hospital

Children's National

Intermountain Primary Children's Hospital

children's health

Joe DiMaggio Children's Hospital

Children's Hospital Los Angeles

Children's Healthcare of Atlanta

MUSC Medical University of South Carolina

Children's Hospital Colorado

Phoenix Children's

Atrium Health Levine Children's

Lucile Packard Children's Hospital Stanford

UCSF Benioff Children's Hospitals

Children's Hospital of Pittsburgh of UPMC

SickKids

Cincinnati Children's

Children's of Alabama

Prospective Longitudinal Registries

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Ductal-dependent pulmonary blood flow

Tetralogy of Fallot


CELTICS
3
CELTICS
CELTICS
44
SPALDING

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The Heart Center

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Washington University in St. Louis Physicians