PA/VSD/MAPCAs

What a surgeon needs to know

Patrick McConnell, MD
Nationwide Children's Hospital

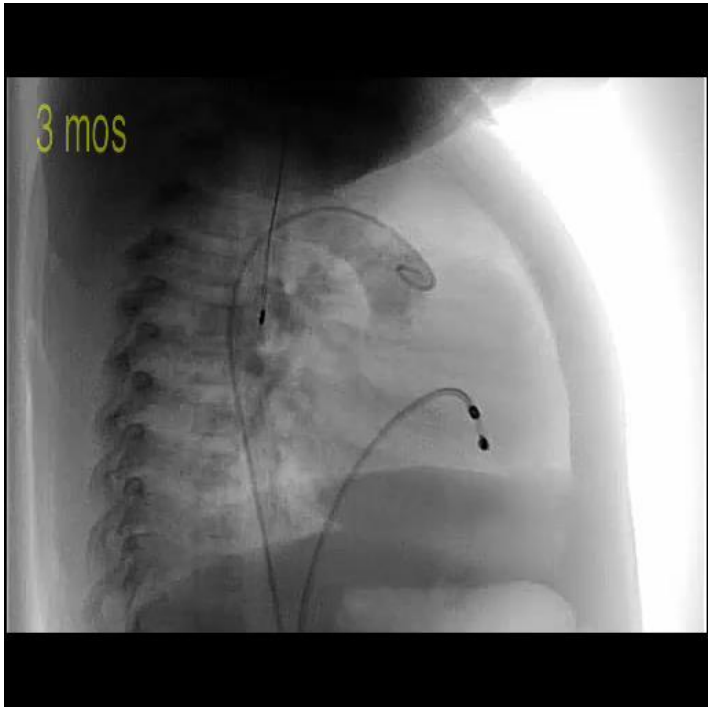
Absolute Surgical Requirements

- Name
- MRN
- Procedure
- Allergies

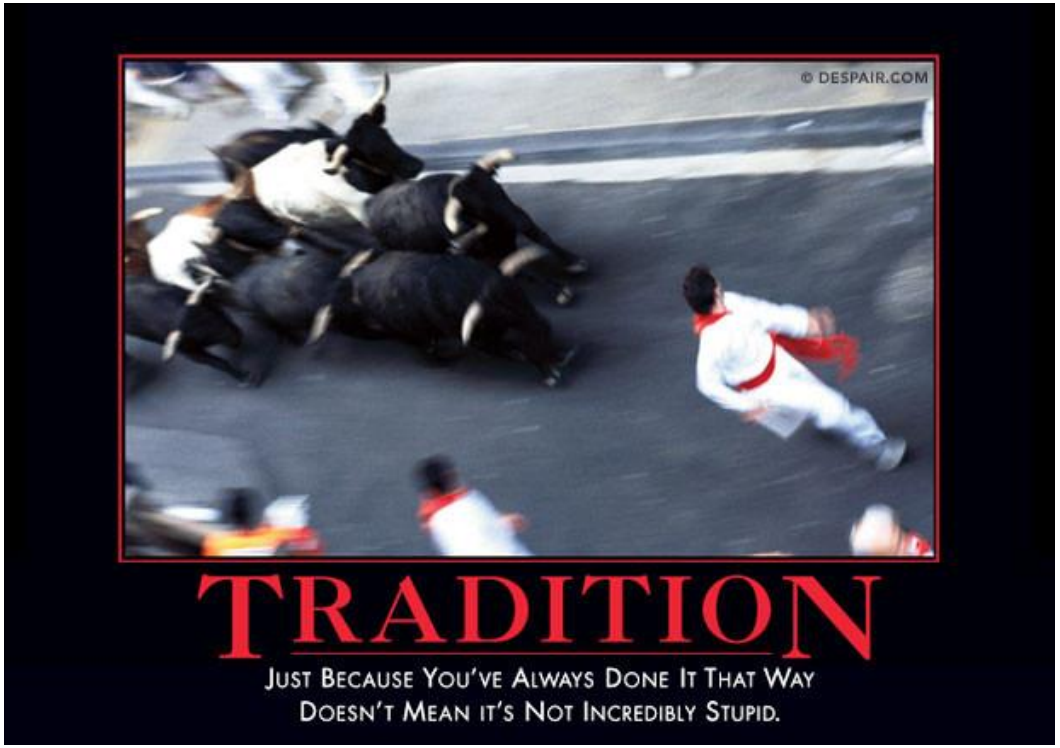
Things needed to complete a ‘Time out’

Overview

1. MAPCAs paradigm?
   - Single stage unifocalization vs. central PA growth
   - Clinical status/intra-cardiac anatomy
     - Age, sats, circulation (over/under), genetics
2. Pulmonary arterial anatomy/hemodynamics
   - Intrapericardial & intrapulmonary characteristics
3. Systemic arterial anatomy/hemodynamics
   - Arch sidedness, MAPCA locations
   - **dual supplied segments
4. Mediastinal/pulmonary anatomy
   - Airway, esophagus, veins, adenopathy, lung status



MAPCAs paradigm . . . When in Rome (Spain?)



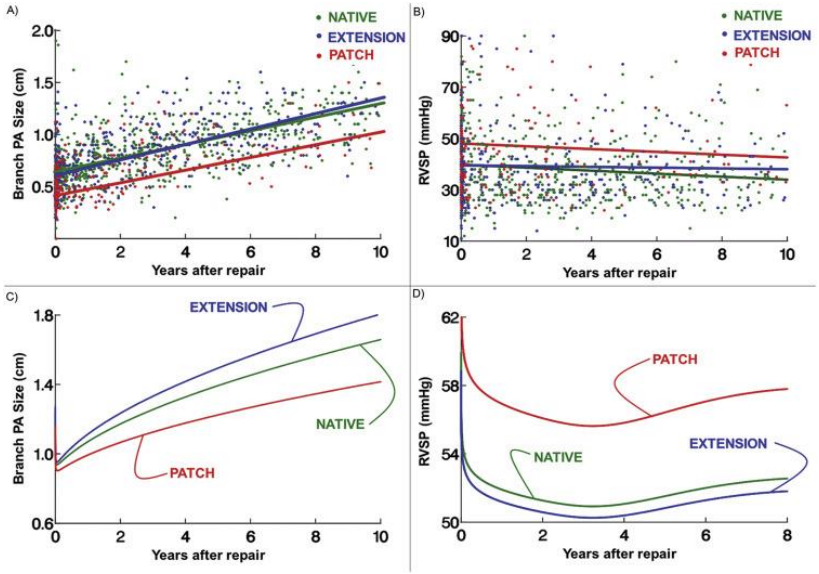
Paradigm

- **Single Stage Unifocalization**
  - Incorporate as many segments into a central PA compartment.
  - Unifocalization to the heart/conduit
  - Intra-cardiac repair
    - VSD closure +/- fenestration
  - 3-9 months of age
    - Average is 7-9 months for most series

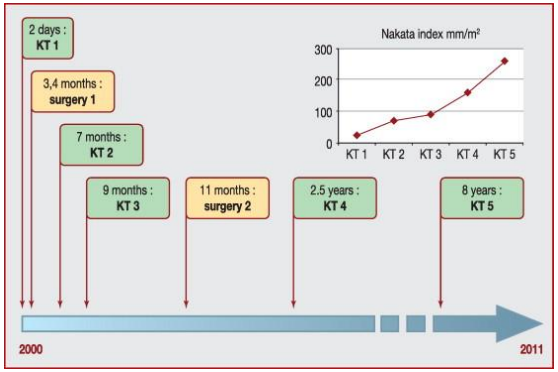
- **Multi-stage or central PA growth focused**
  - Early cath (< 1mos) to delineate central PAs and MAPCAs
  - Surgery (< 3 mos): achieve central PA blood flow
    - RV-PA conduit
    - Central shunt
    - No intracardiac repair vs. fenestrated VSD
  - Later catheterizations to control dual supply MAPCAs and PA growth
  - Surgery (< 2 yo): unifocalize & intra-cardiac repair

Surgery & Central PA growth

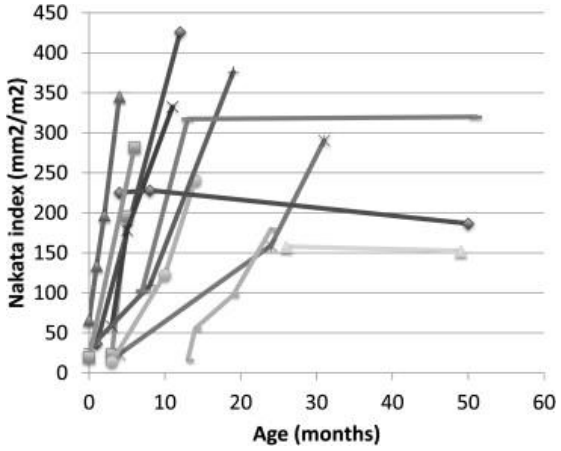
- MAPCAs and surgically manipulated central PAs fail to grow



The Annals of Thoracic Surgery, Volume 101, Issue 3, 2016, 996–1004



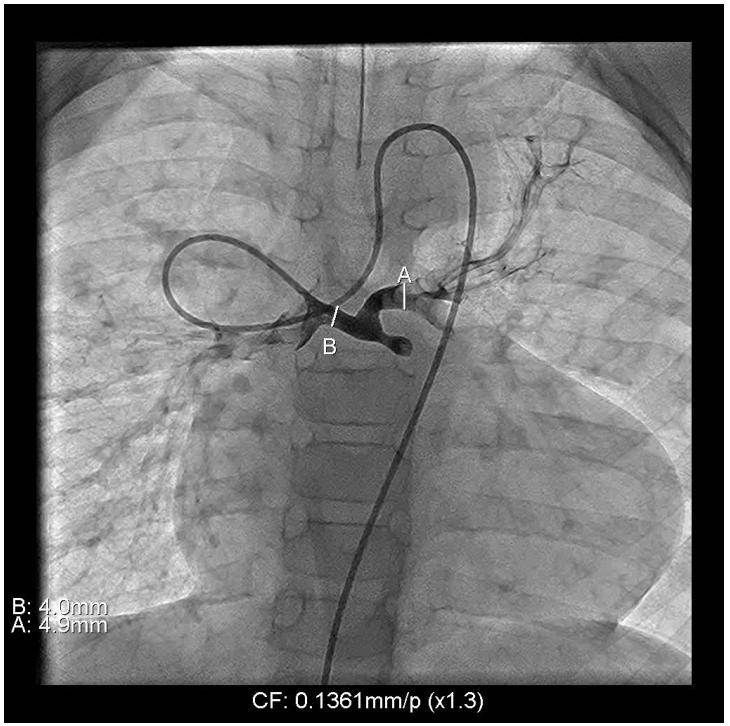
Archives of Cardiovascular Diseases, Volume 105, Issue 12, 2012, 666–675



The Annals of Thoracic Surgery, Volume 97, Issue 6, 2014, 2129–2133

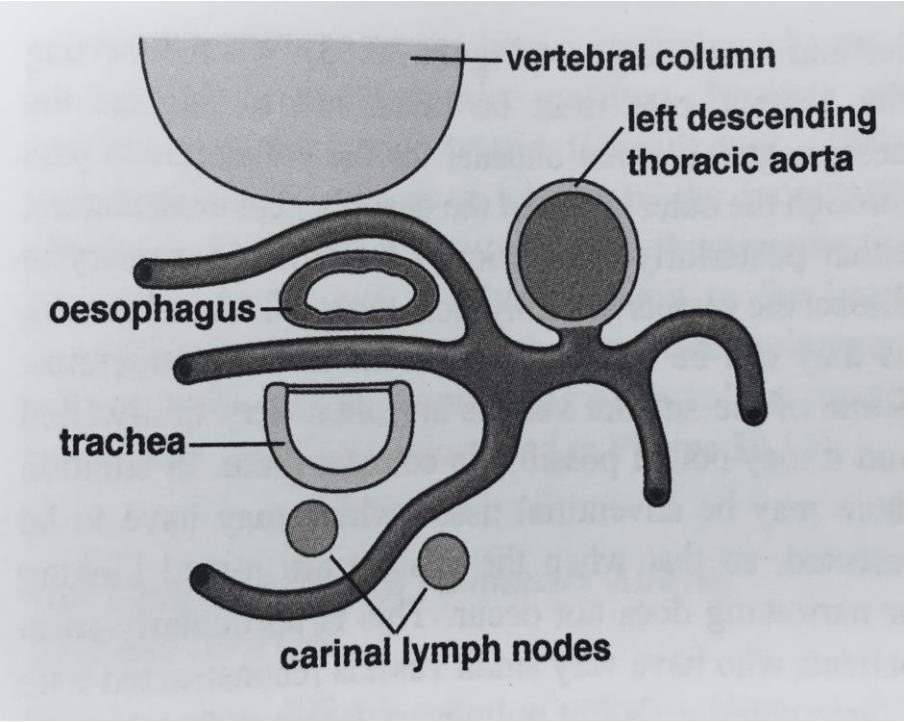
Central PAs

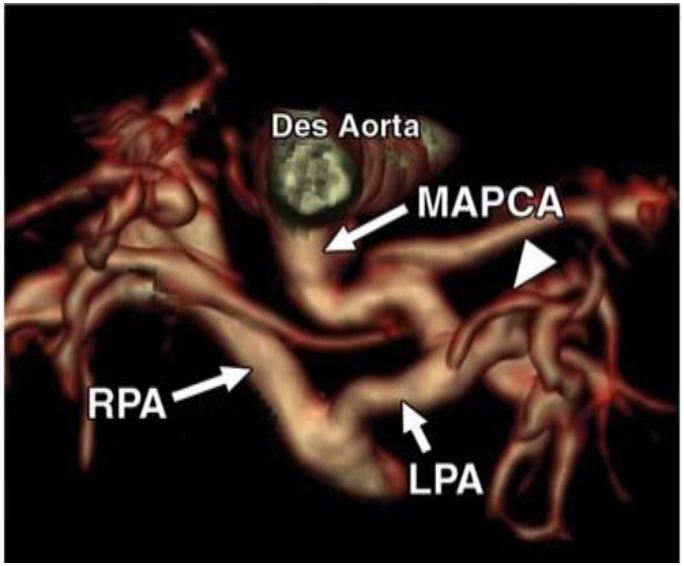
- Central PA Present
  - Branch PA sizes
    - Nakata index
  - Define dual-supply MAPCAs
- Not present
  - MAPCA circulation/pulmonary circulation
    - Confirm by MDCT



5 yo male, unrepaired PA/VSD/MAPCAs. Sats – 72%; Nakata = 155 mm^2/m^2

MAPCAs

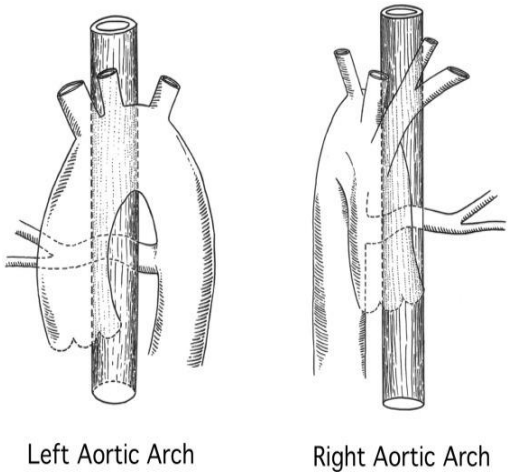


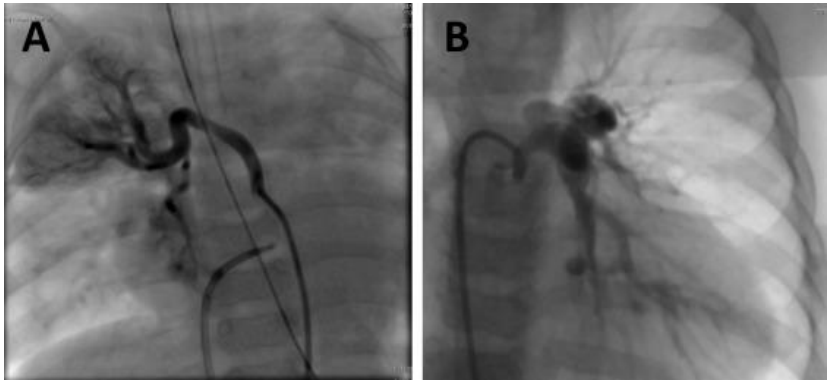


Surgery for Cong Heart Defects. 3rd Edition. Tsang V. Ch 30. Figure 30.11

Retro-Esophageal MAPCAs

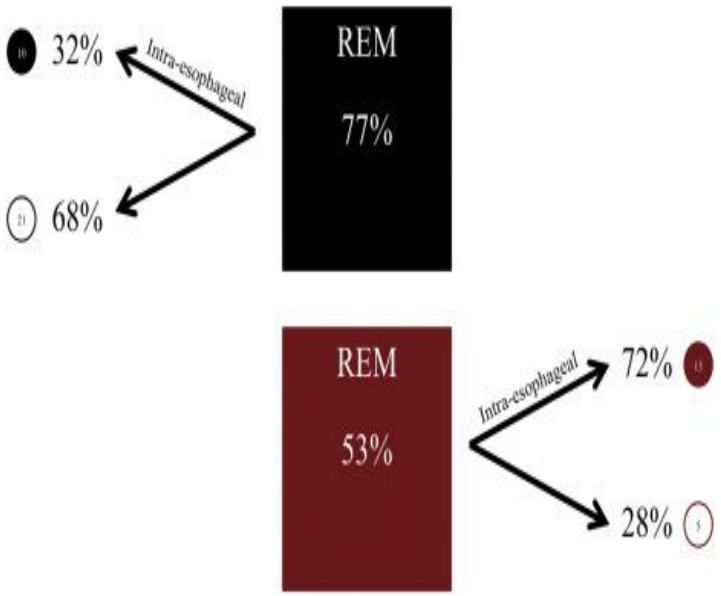
- REMs (Stanford 2011-2013)
  - 68 patients
    - 45 (67%) with REM
  - 84% stenotic
    - 80% Intra and 32% extra
  - LAA
    - 77% REM
      - 32% Intra
  - RAA
    - 53% REM
      - 72% Intra

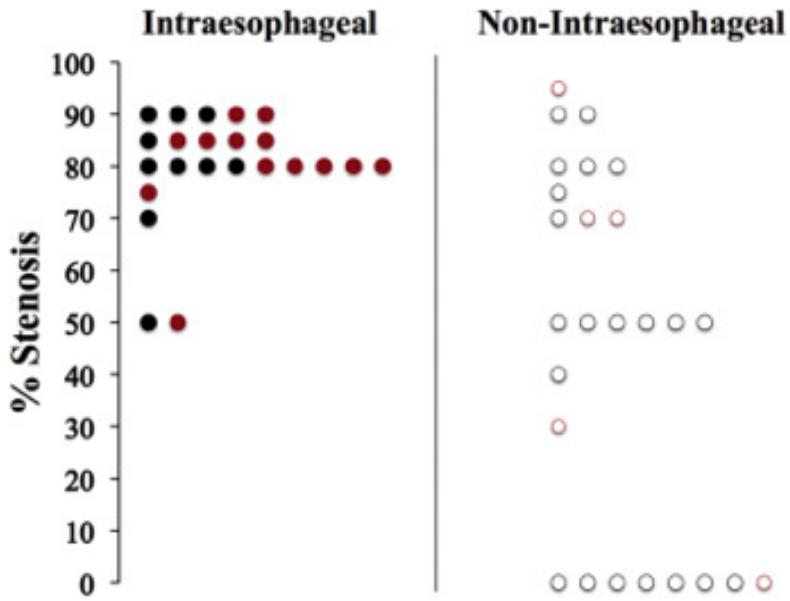




The Annals of Thoracic Surgery, Volume 102, Issue 3, 2016, 877–882

REM and Stenosis

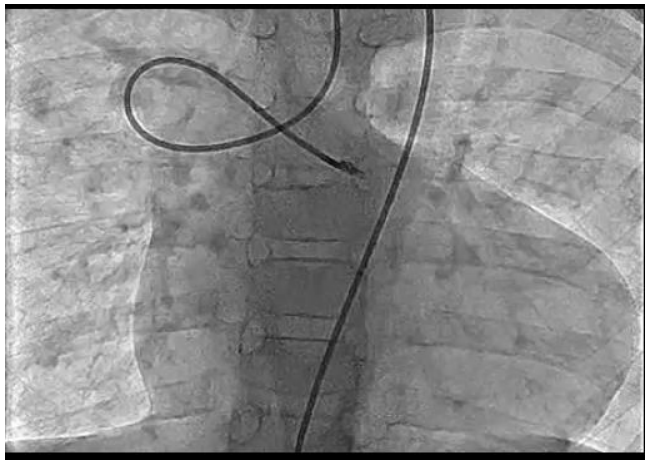


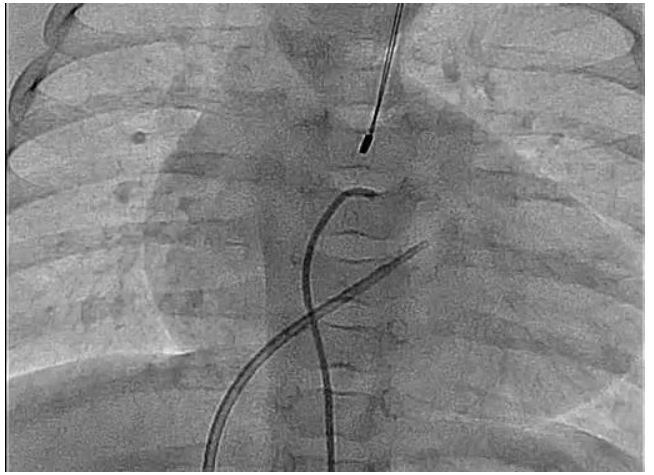


The Annals of Thoracic Surgery, Volume 102, Issue 3, 2016, 877–882

Dual Supply/Over-circulation

- Systemic saturations
  - Lower (< 85%)
    - Plan to keep all non-unifocalized segments
    - Later coil/embolize
  - Higher
    - Ligate non (<50%) stenotic collaterals
- Number and location of single-source segments
- If over-circulated pre-op – very likely there are adequate central PAs and early repair is indicated.





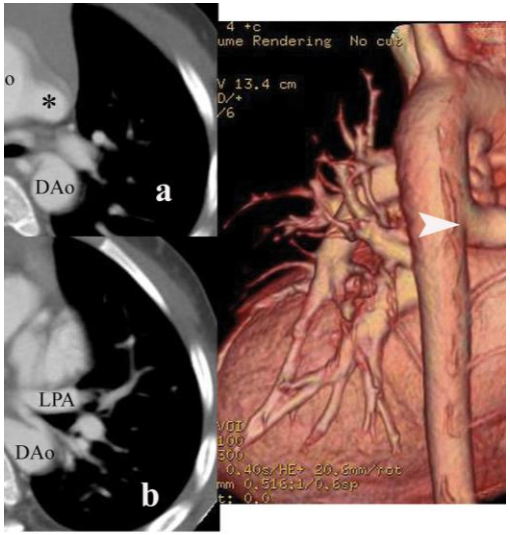
Value of Advanced Imaging?

Complete Preoperative Evaluation of Pulmonary Atresia with Ventricular Septal Defect with Multi-Detector Computed Tomography. PLoS One. 2016; 11(1).

Table 1

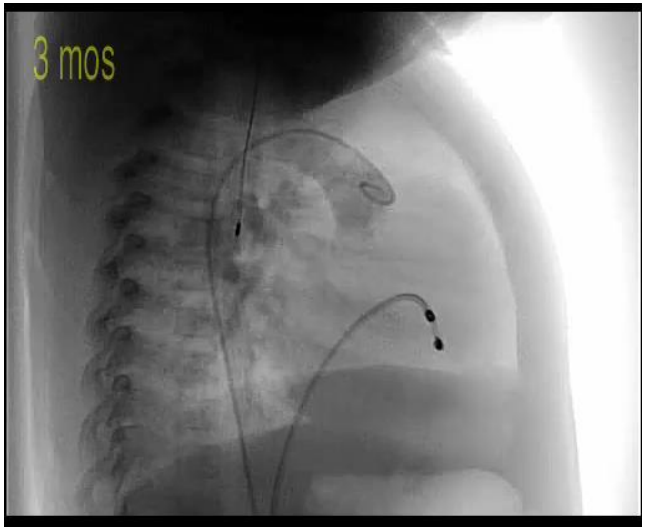
Findings at MD CT, TTE, and Cardiac Catheterization Compared with Findings at surgery.

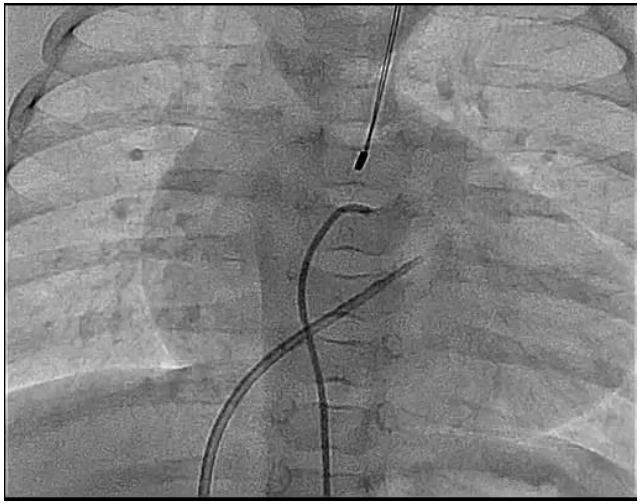
Parameter		No. of Surgical Findings				Test Parameters				
	Technique	True-Positive	False-Negative	True-Negative	False-Positive	Sensitivity	Specificity	PPV	NPV	Accuracy
Main PA	MDCT	33	0	83	0	100%	100%	100%	100%	100%
	Catheterization	30	3	82	1	90.9%	98.8%	96.8%	96.5%	96.6%
	TTE	26	7	80	3	78.8%	96.4%	89.7%	92.0%	91.4%
Right/ Left PA	MDCT	158	0	74	0	100%	100%	100%	100%	100%
	Catheterization	146	12	74	0	92.4%	100%	100%	86.0%	94.8%
	TTE	112	46	70	4	70.9%	94.6%	96.6%	60.3%	78.4%
PA confluence	MDCT	68	0	48	0	100%	100%	100%	100%	100%
	Catheterization	64	4	48	0	94.1%	100%	100%	92.3%	96.6%
	TTE	53	15	43	5	77.9%	89.6%	91.4%	74.1%	82.8%
PA stenosis	MDCT	18	0	97	1	100%	99.0%	94.7%	100%	99.1%
	Catheterization	15	3	97	1	83.3%	99.0%	100%	97.0%	96.6%
	TTE	11	7	94	4	61.1%	95.9%	73.3%	84.7%	90.5%
Overall native PA	MDCT	277	0	302	1	100%	99.7%	99.6%	100%	99.8%
	Catheterization	255	22	301	2	92.1%	99.3%	99.2%	93.2%	95.9%
	TTE	202	75	287	16	72.9%	94.7%	92.7%	79.3%	85.9%
MAPCA stenosis	MDCT	96	2	225	3	98.0%	98.7%	97.0%	99.1%	98.5%
	Catheterization	85	13	222	6	86.7%	97.4%	93.4%	94.4%	94.2%
	TTE	NA	NA	NA	NA	NA	NA	NA	NA	NA
PDA	MDCT	47	0	69	0	100%	100%	100%	100%	100%
	Catheterization	47	0	69	0	100%	100%	100%	100%	100%
	TTE	47	0	69	0	100%	100%	100%	100%	100%

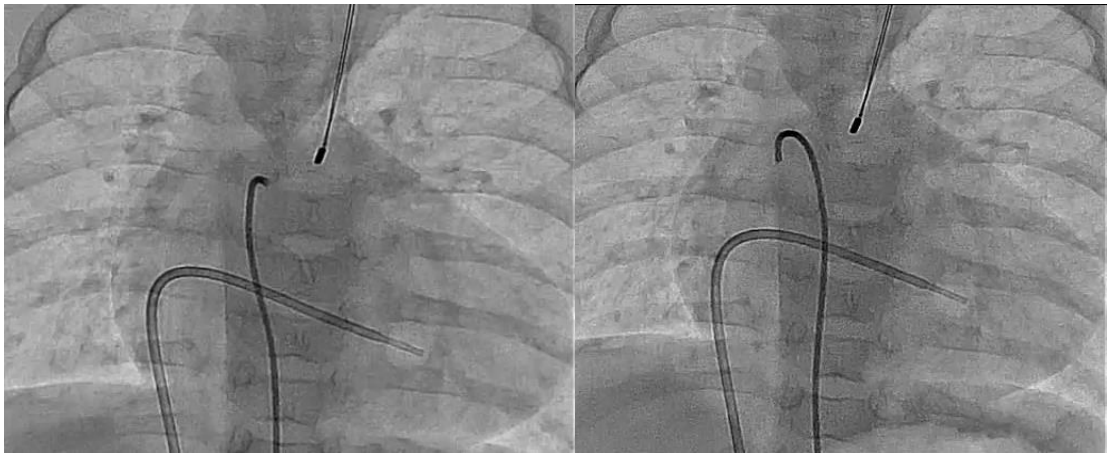


NATIONWIDE CHILDREN'S
When your child needs a hospital, everything matters.℠

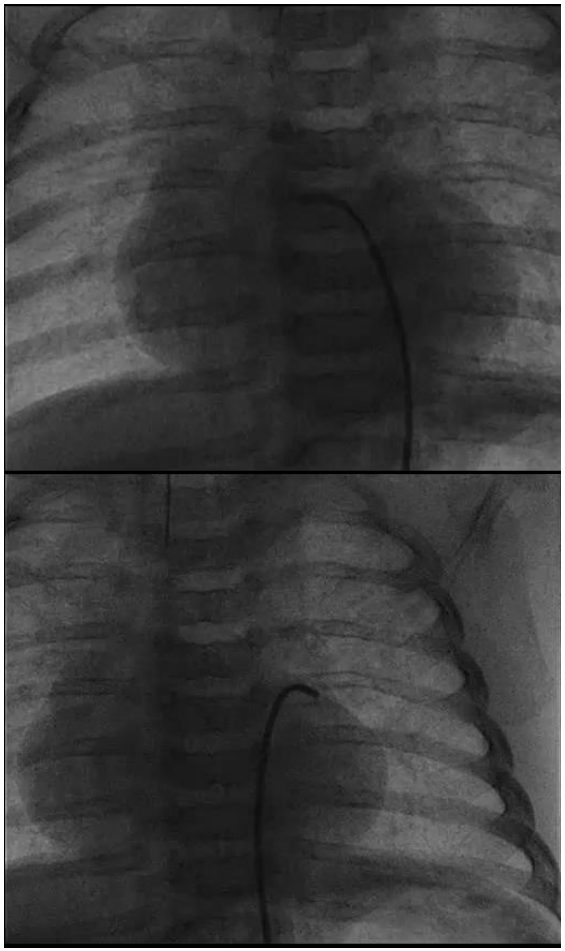
Imaging: What is Helpful?

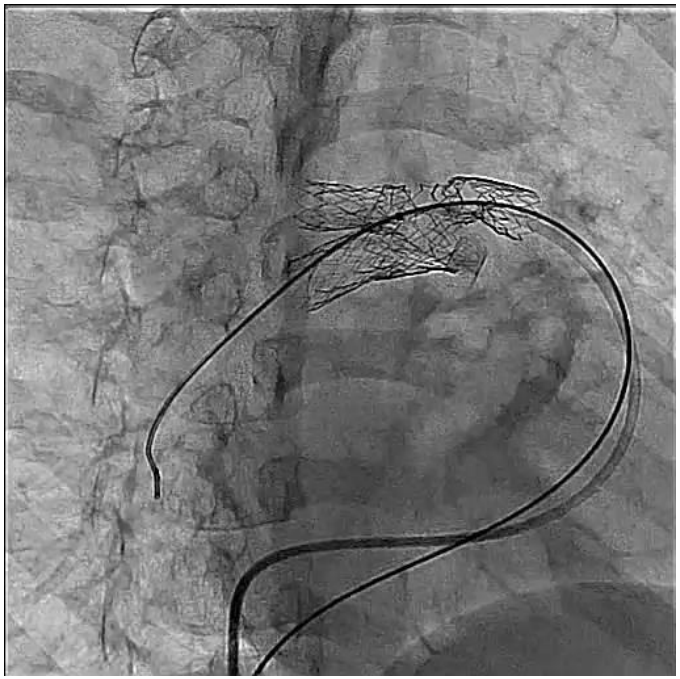


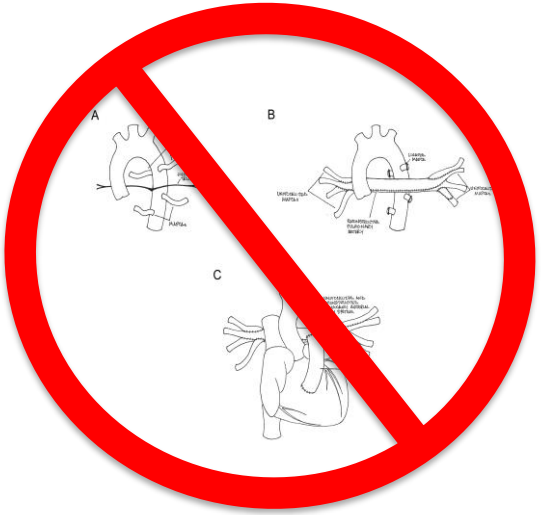




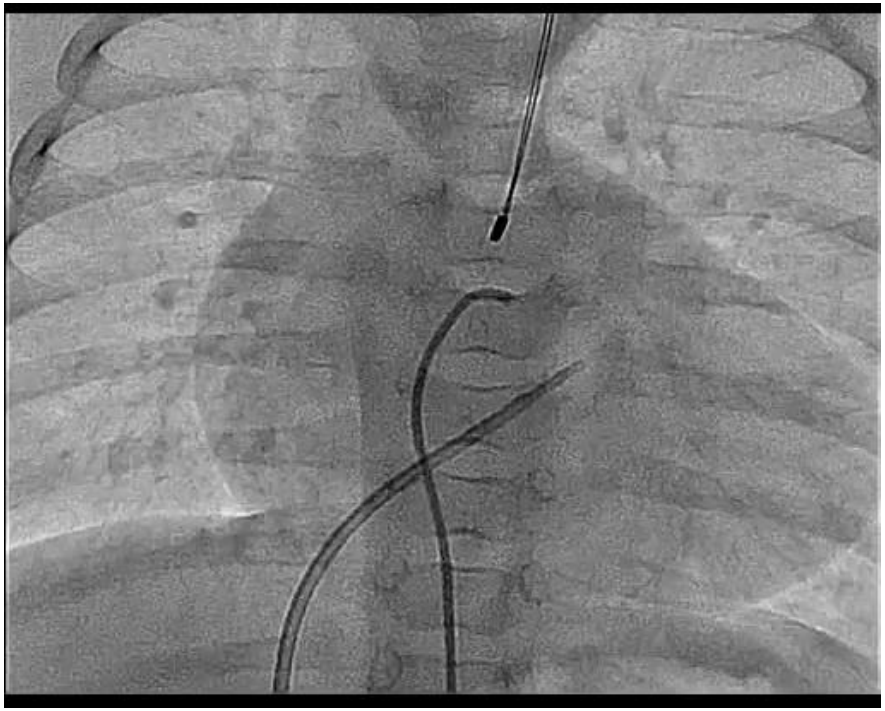
Traditional Outcome



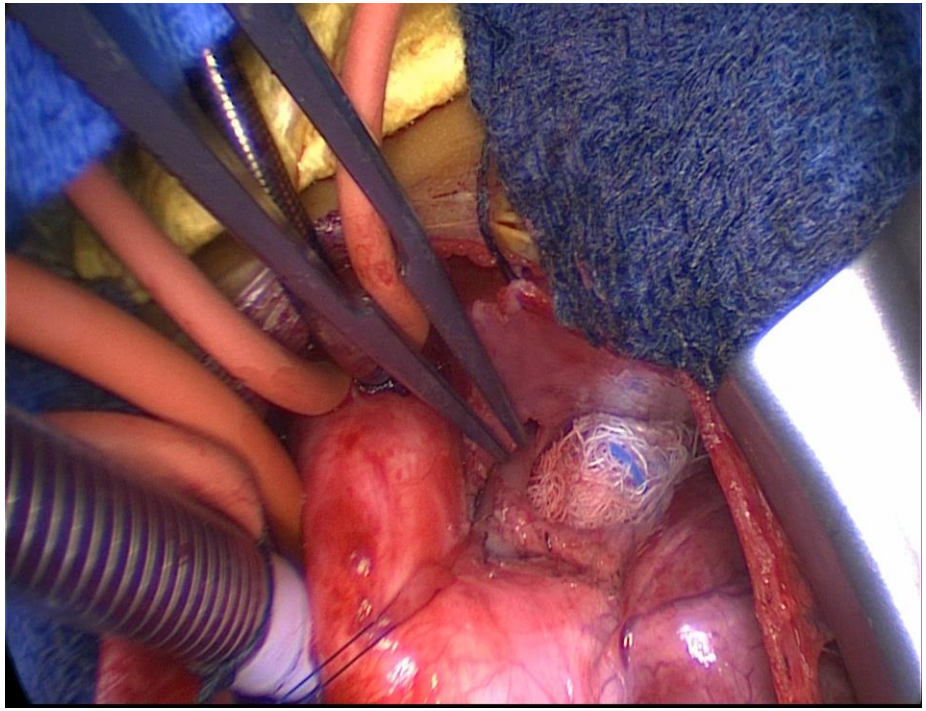




Less > More

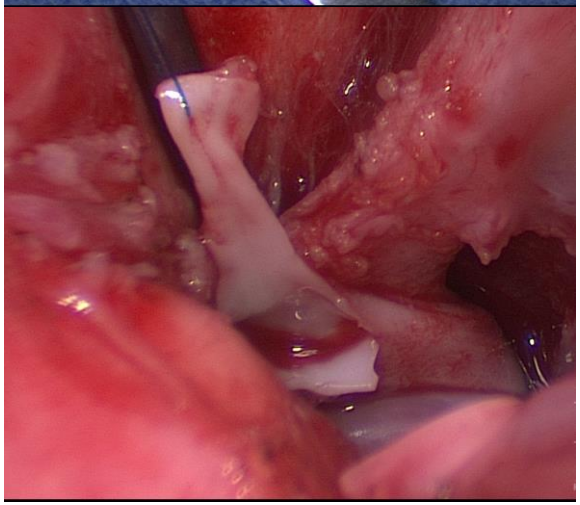


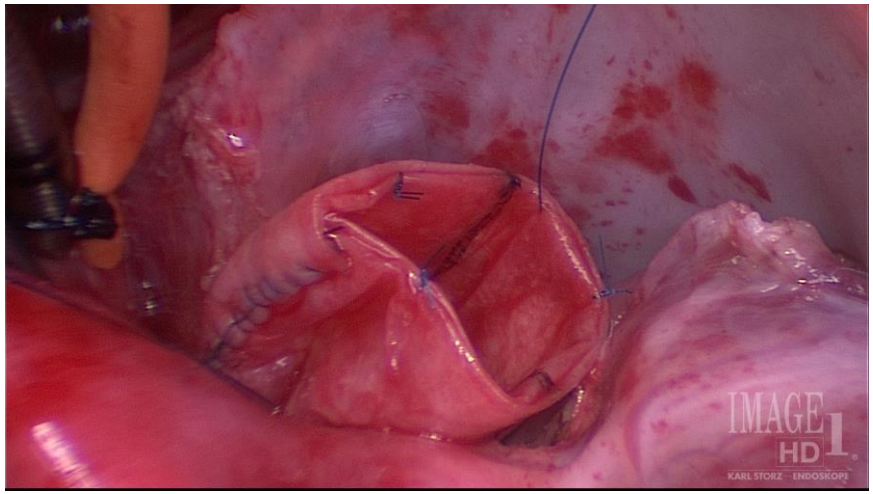
4 mos - preop

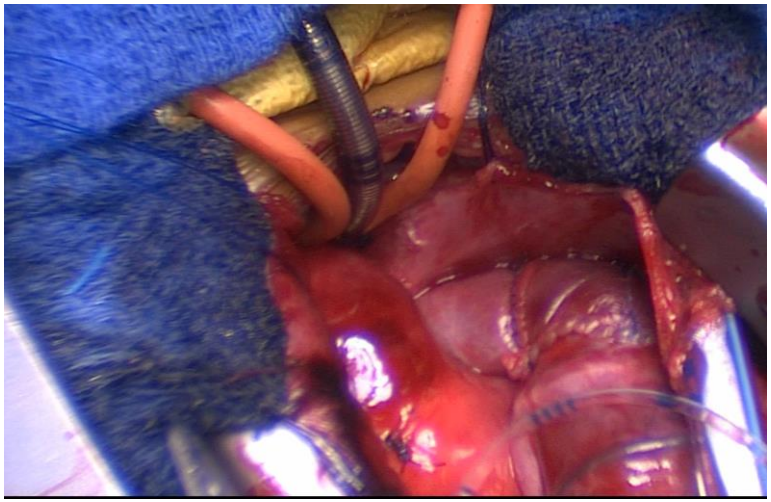


5 mos – intra-op

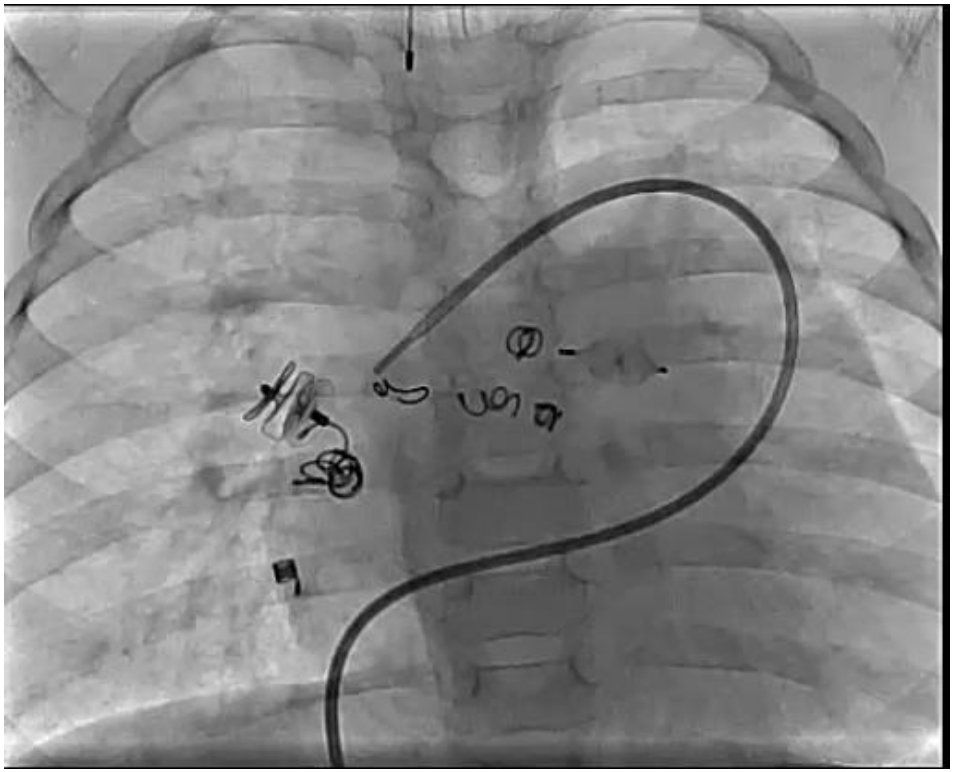
Less > More





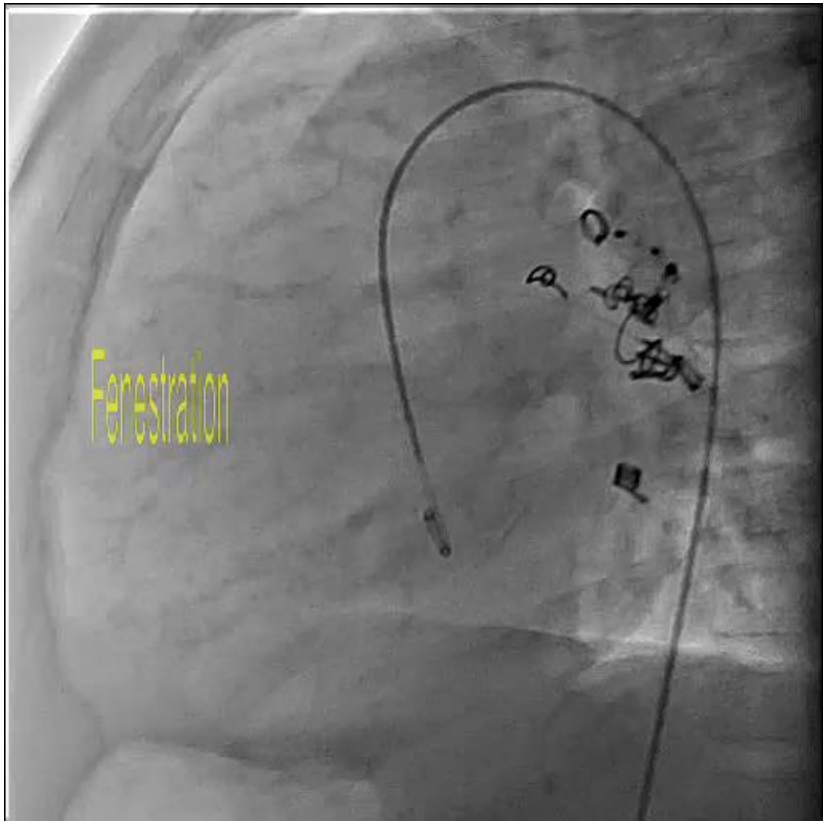


Less > More



1 year post-op

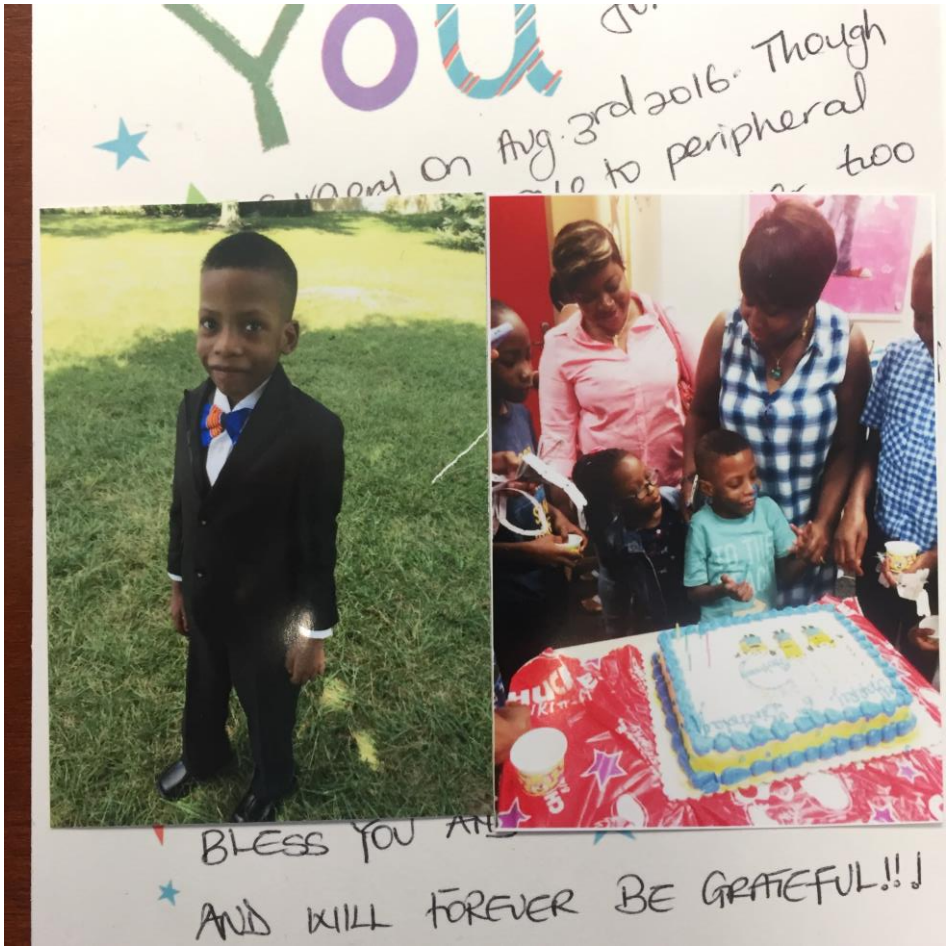
Less > More



Summary

- Clinical status and the status of the central PAs should dictate palliative strategy.
- MAPCAs need to be defined (+/- stenoses)
  - Dual-supply
  - Single source
  - Mediastinal structures involved
- Diminutive central PAs have growth potential
  - Data/experience demonstrates that surgically manipulated PAs and MAPCAs do not grow.
    - Avoid surgical and stent based interventions in early PA rehab
  - Focus interventions controlling dual sources overtime.
- 3-D rotational angiography would seem less informative than selective MAPCA angios to define location, dual-supply and stenoses and advanced imaging may be value added.

Thank you



Pre-op sats = 72%

Post-op sats = 93%

NATIONWIDE CHILDREN'S
When your child needs a hospital, everything matters.℠