Case Discussion and Management Challenges

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Disclosures

None

Case

32 1/7 week female, birth weight 1.2kg (z=-3.6)

Prenatal dx HLHS (MA/AA)
Mother 15 years old, G3P1, prior abruption

SpO2 95% on IMV CPAP on arrival to CHONY (0 day of life)



Case

Mild RV dysfunction, mild TR
Posterior deviation of atrial septum, but gradient 5
Normal pulmonary vein anatomy
Ascending aorta 1-2mm, aortic atresia/mitral atresia

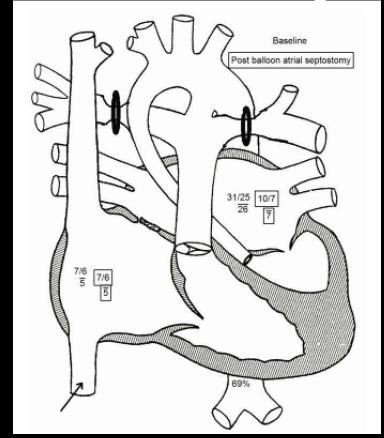
Options

Comfort care

PA bands/Septostomy/Medical Hybrid

Stage 1

Bilateral PA bands and PGE on DOL 8
Increasing hypoxia and transatrial gradient on echo



Remained on CPAP, growing with ND feeds Troponin elevation, concern for retrograde coarctation

CT scan with hypoplastic branch pulmonary arteries





PDA stent vs. Stage 1

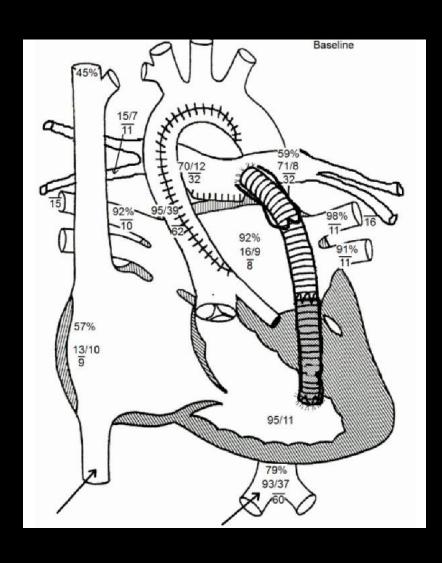
When do you do it?

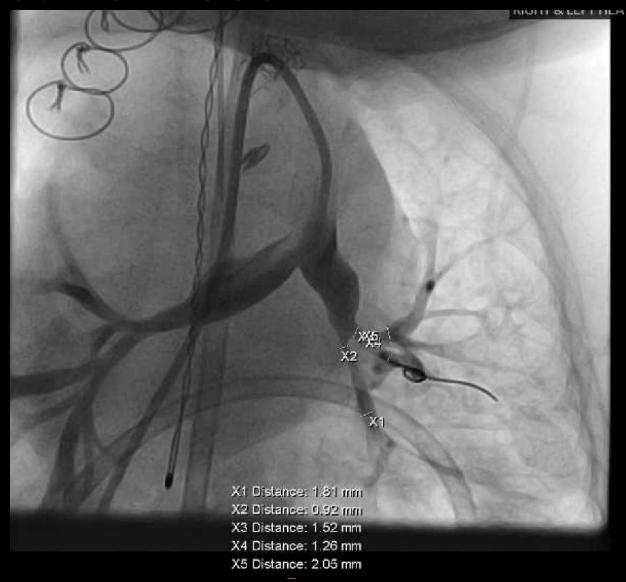
Norwood/Sano at 2.5kg with PA debanding and bilateral PA plasty

Postop ECMO – 3 days, likely I/R lung injury, proximal Sano stent

Recovered, but persistent tachypnea 2 months postop

Cath 2 months post Norwood





Cath vs. surgery?

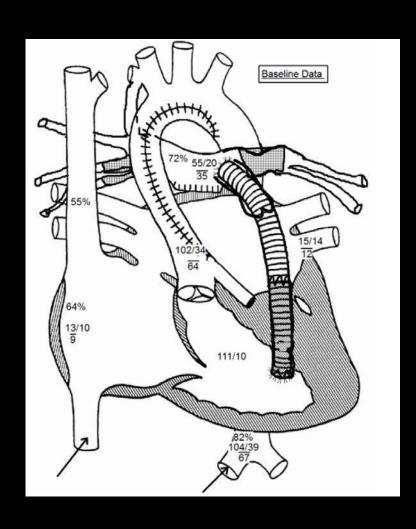
Hospice?

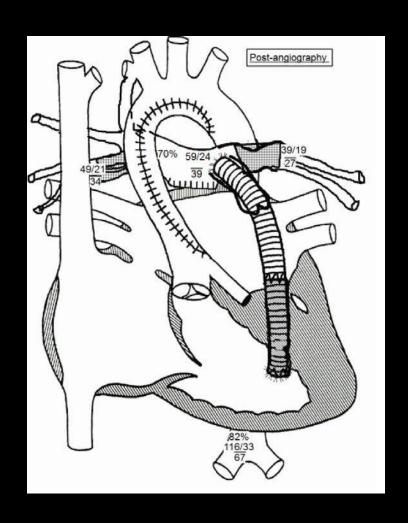
Bilateral PA plasty with addition of mBTT shunt

Persistent tachypnea on 10L HFNC 25% 1 month postop: LPA stent, RML, RLL stents

Repeat cath 2 months after stenting (10 months of age) due to lung perfusion scan 32% to left lung and small LPA on echo

Cath





No further intervention

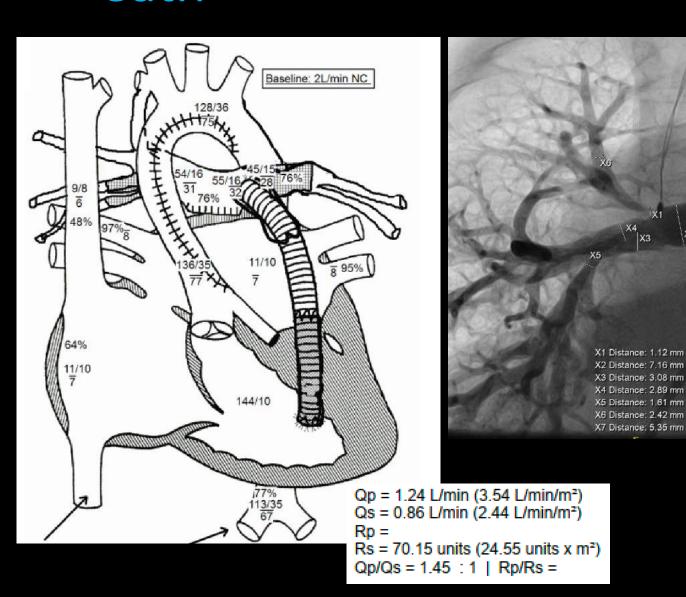
Transcatheter PA rehab

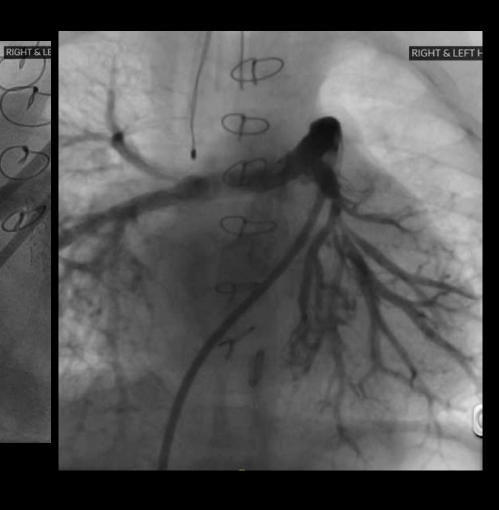
Surgical PA reconstruction Shunt or single-lung Glenn?

Weaned to 0.5LNC 21%, G-tube placed, and discharged to rehab

Readmitted 3 months later (1 yo) and interval cath performed

Cath





No further intervention

Transcatheter PA rehab

Surgical peripheral PA reconstruction Shunt or single-lung Glenn?

Thank You!