Prenatal Care of Ebstein Anomaly: Imaging, Physiology, Treatment, and Counseling

28th Annual Update on Pediatric and Congenital Cardiovascular Disease

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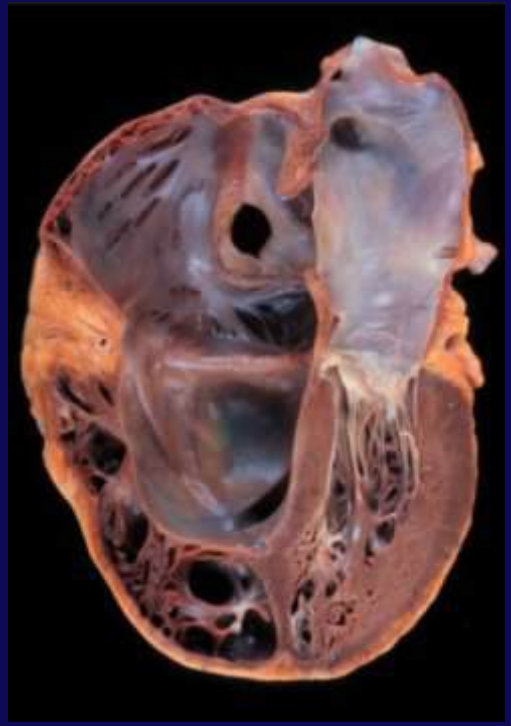
UNIVERSITY OF TORONTO

SickKids | Labatt Family Heart Centre

Introduction

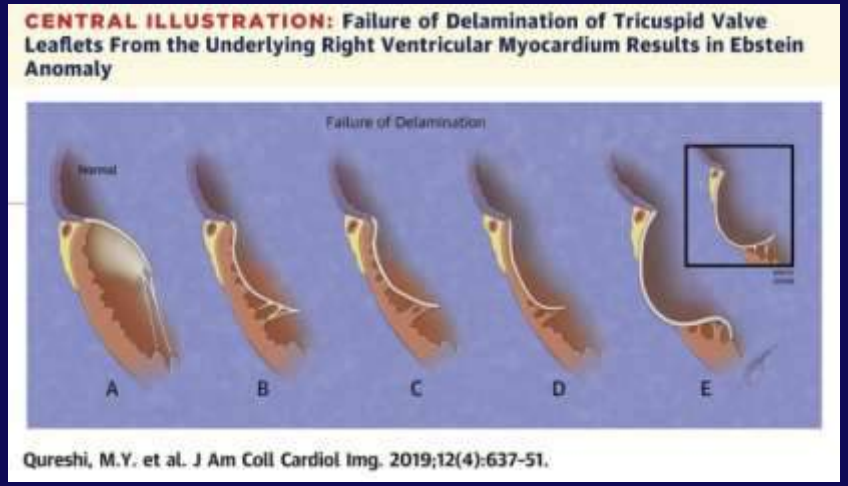
Wilhelm Ebstein (1836-1912)

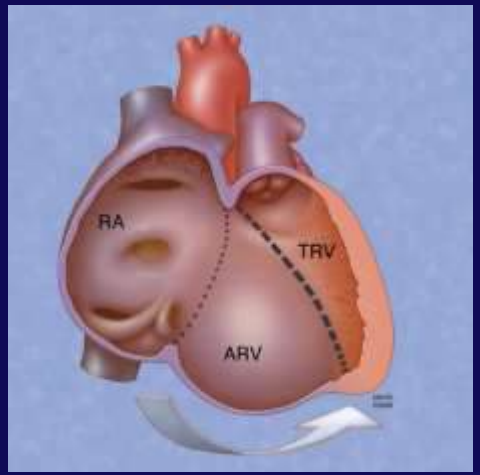


Mann *et al*, *Mayo Clin Proc* 1979

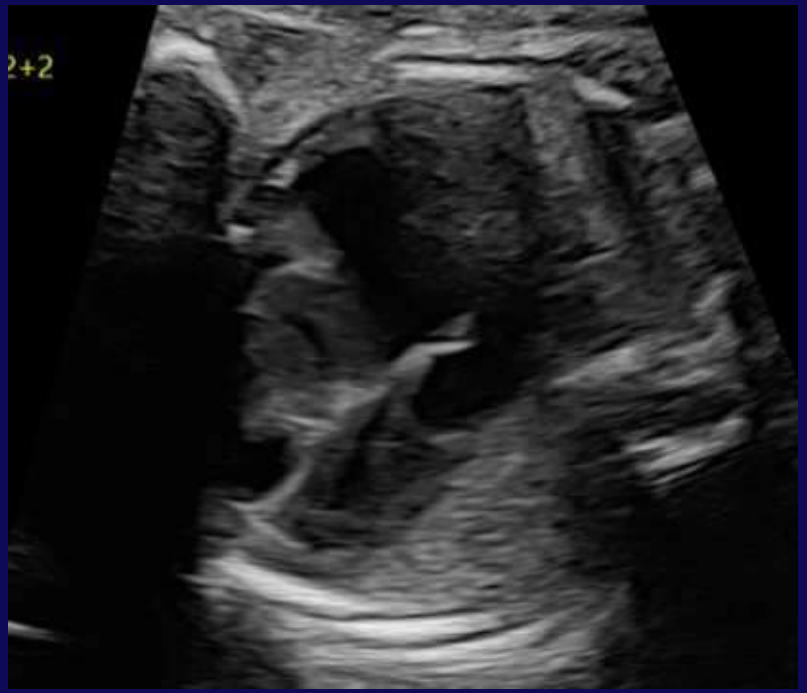
Morphology of Ebstein Anomaly

Wide spectrum of severity with **INFINITE** variations



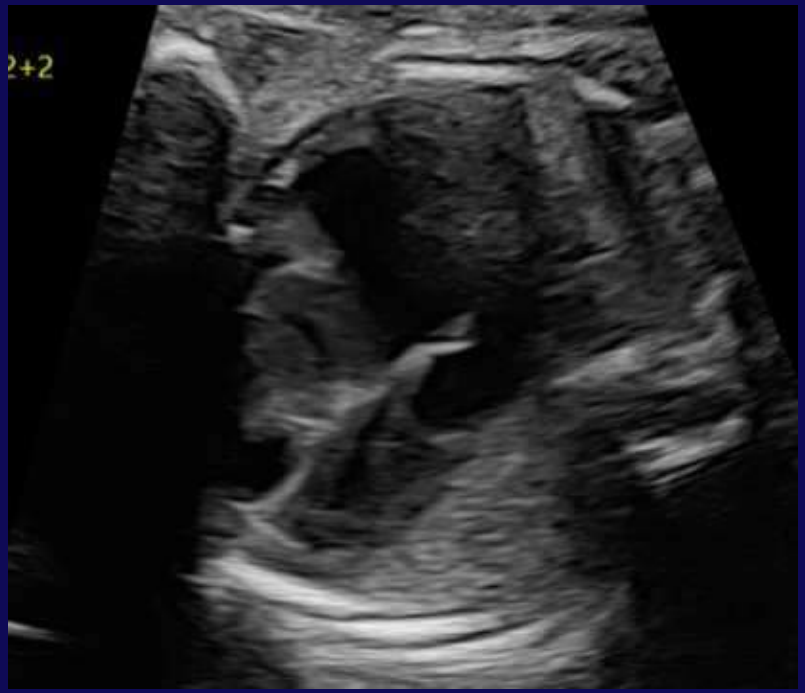


Ebstein Anomaly (EA) / Tricuspid Valve Dysplasia (TVD)



Ebstein Anomaly (EA) / Tricuspid Valve Dysplasia (TVD)

Severe
TR



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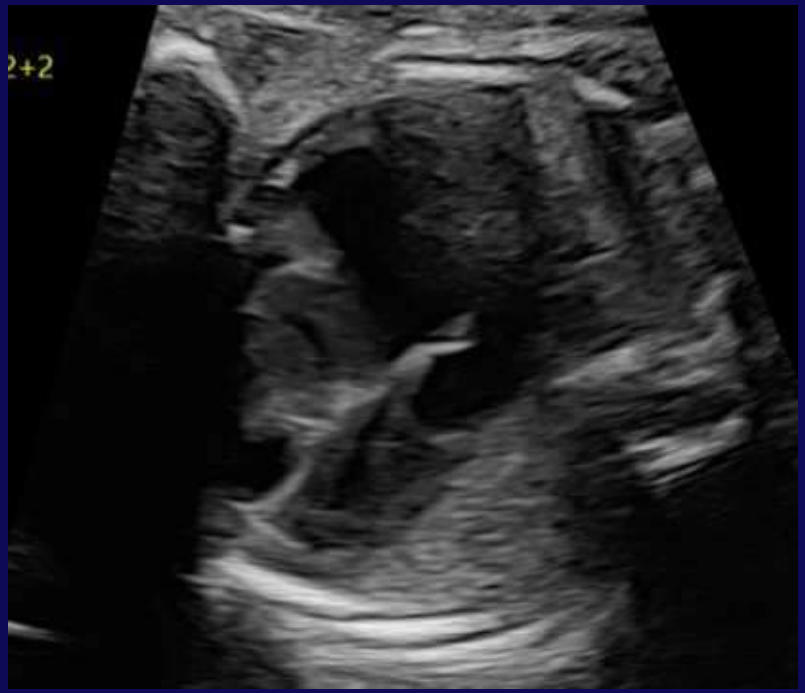
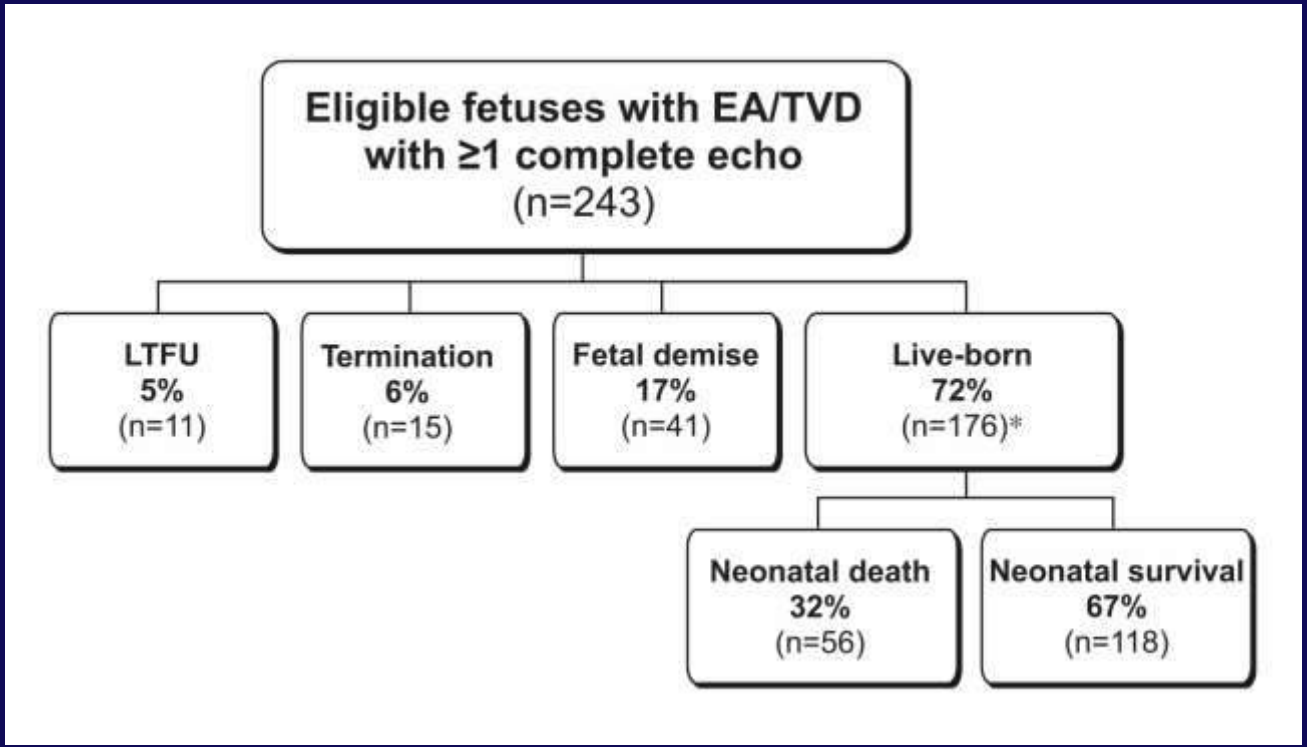


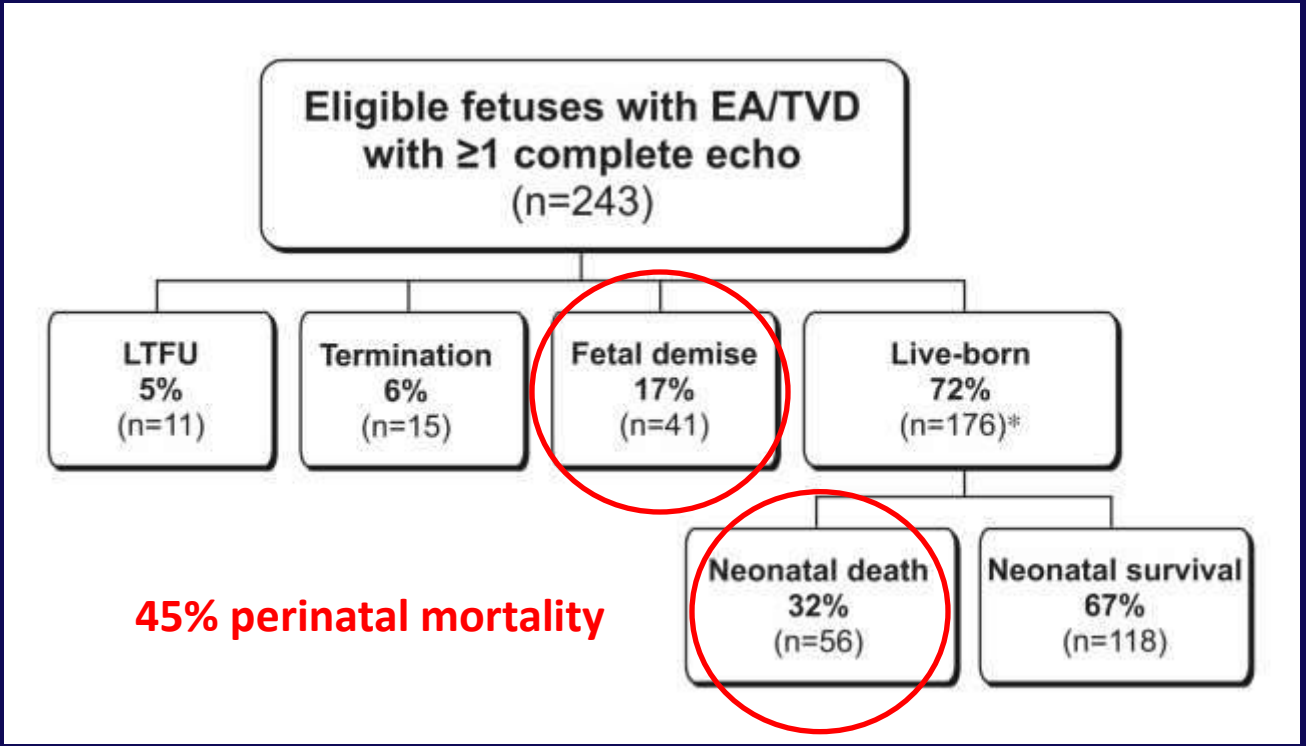
Poorly tolerated in fetal circulation

Perinatal Outcome



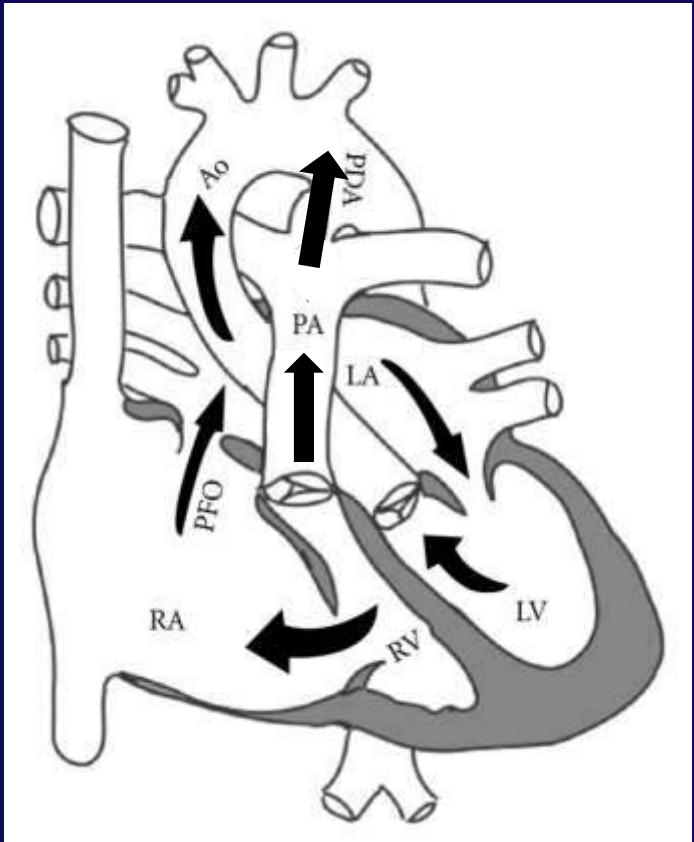
Freud *et al*, *Circ* 2015

Perinatal Outcome



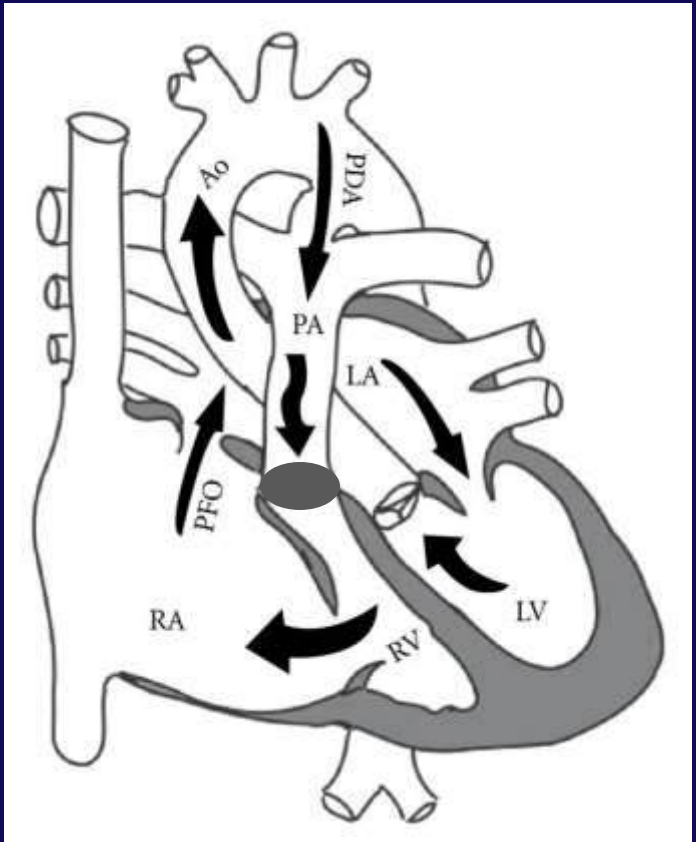
Freud *et al*, *Circ* 2015

Pathophysiology



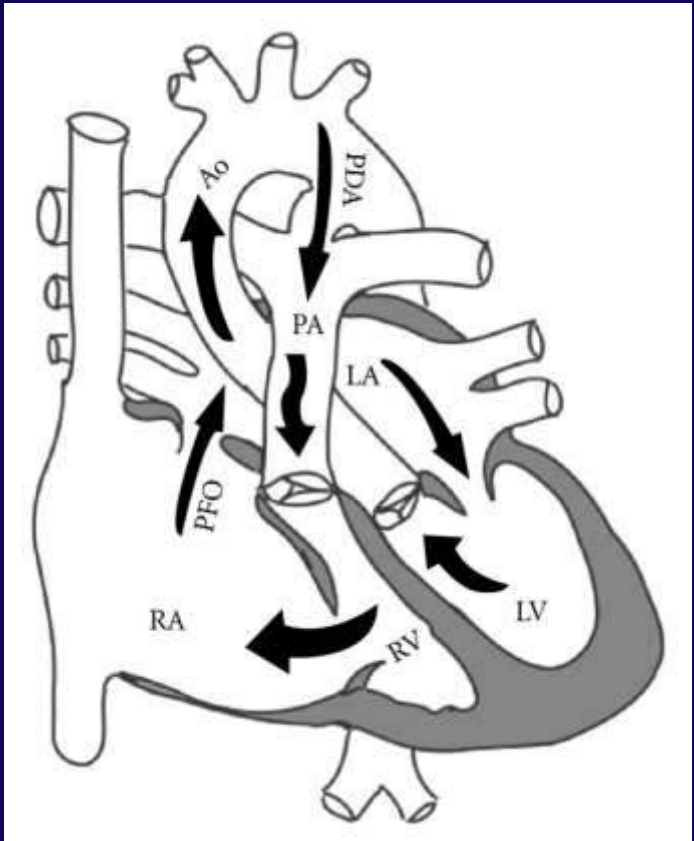
- R → L shunting = GOOD

Pathophysiology



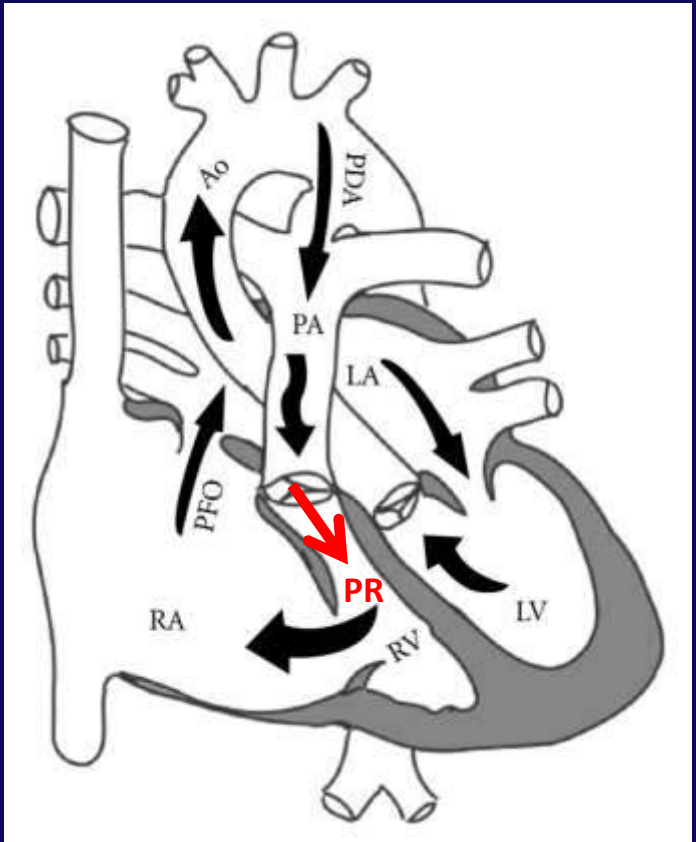
- L → R shunting with fixed anatomic RV outflow tract obstruction = OK

Pathophysiology

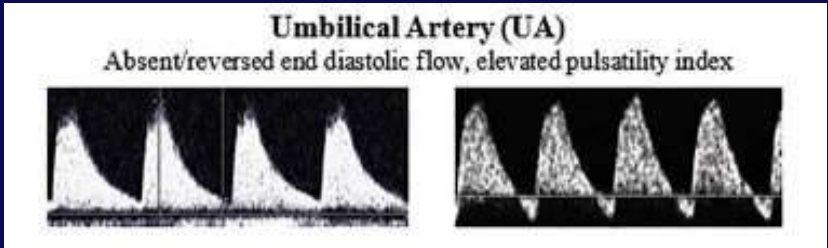


- L → R shunting without fixed obstruction signifies functional pulmonary atresia = BAD

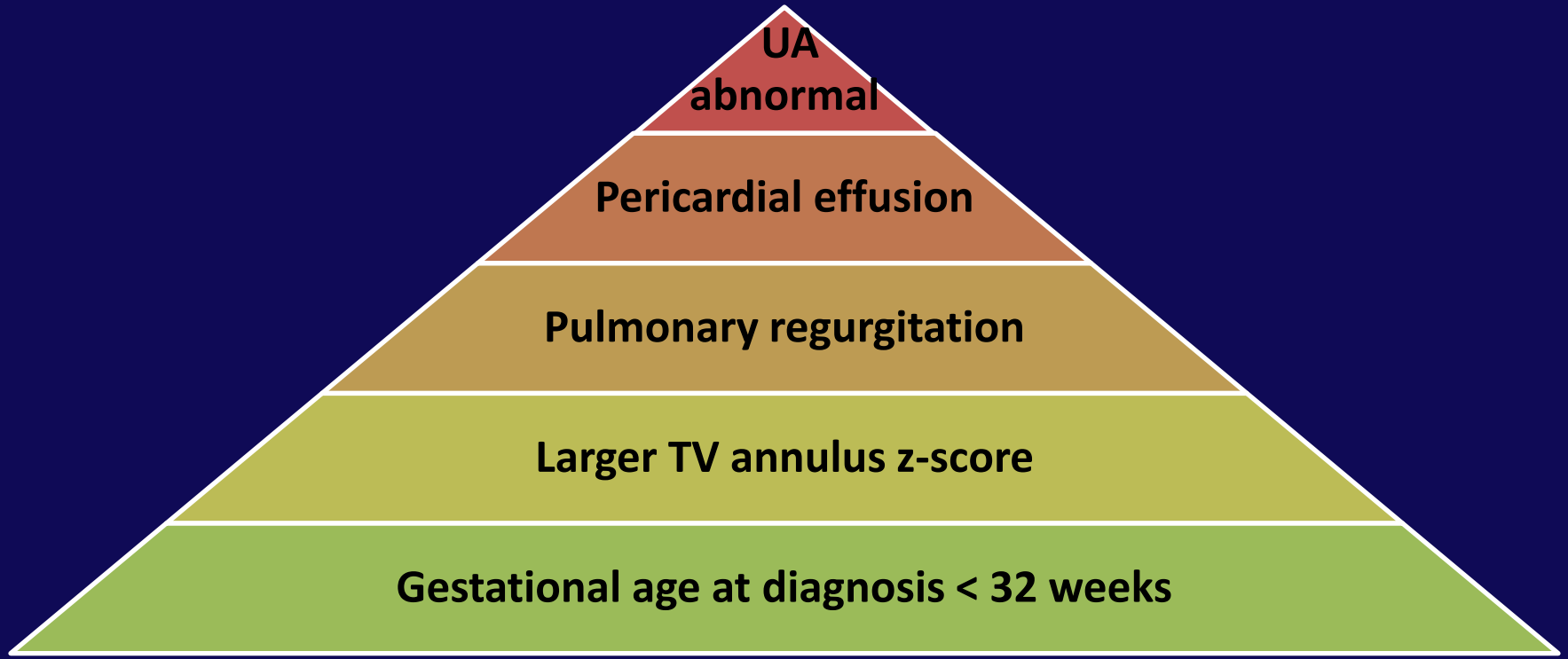
Pathophysiology



- Pulmonary regurgitation completes the “circle of death” = VERY BAD!



Risk Factors for Perinatal Mortality



Freud *et al*, *Circ* 2015
Wang et al, Prenat Diag 2020

Fetal Echo at 31+6 weeks

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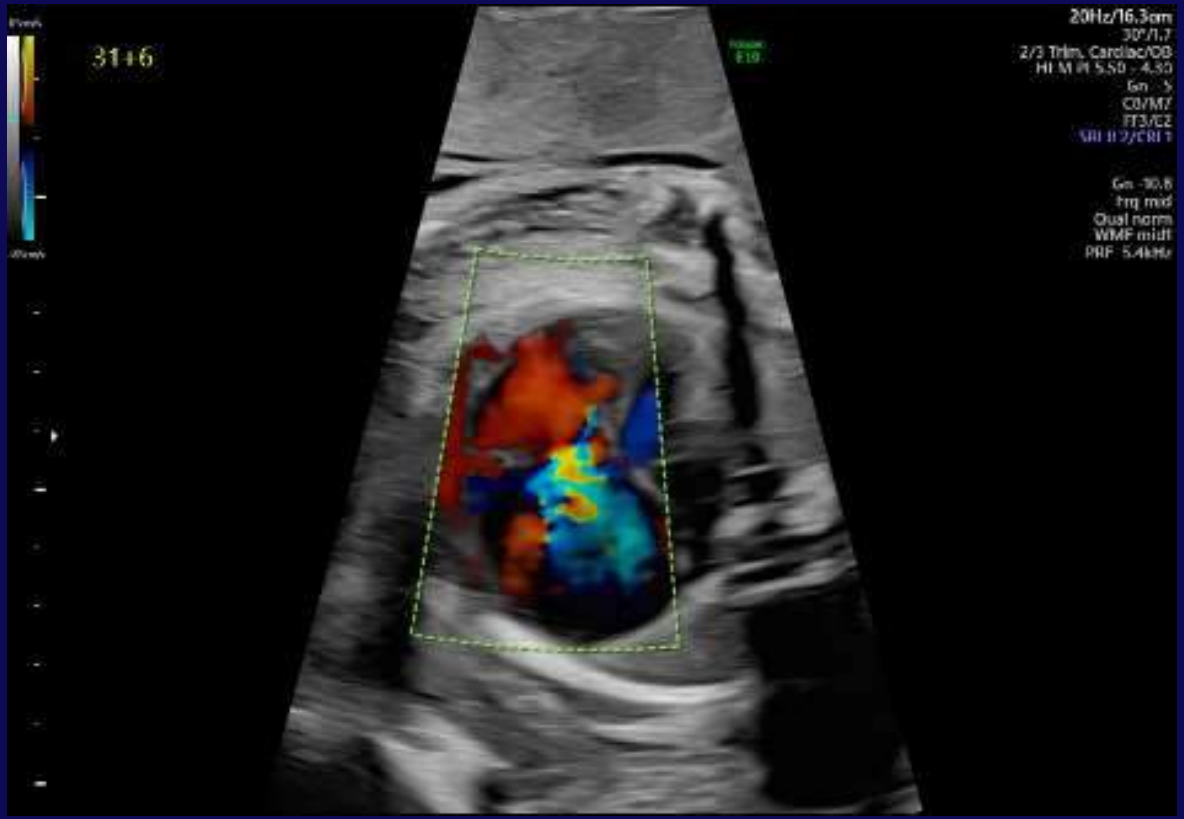
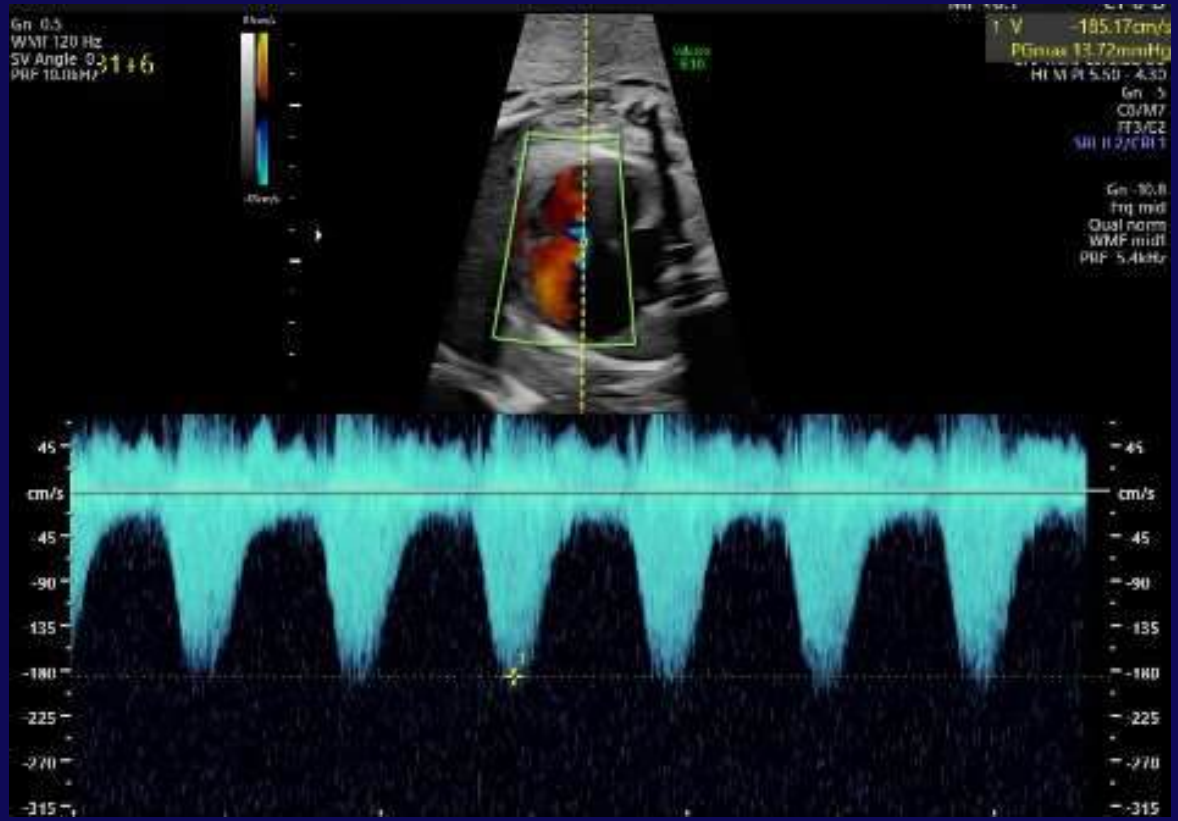
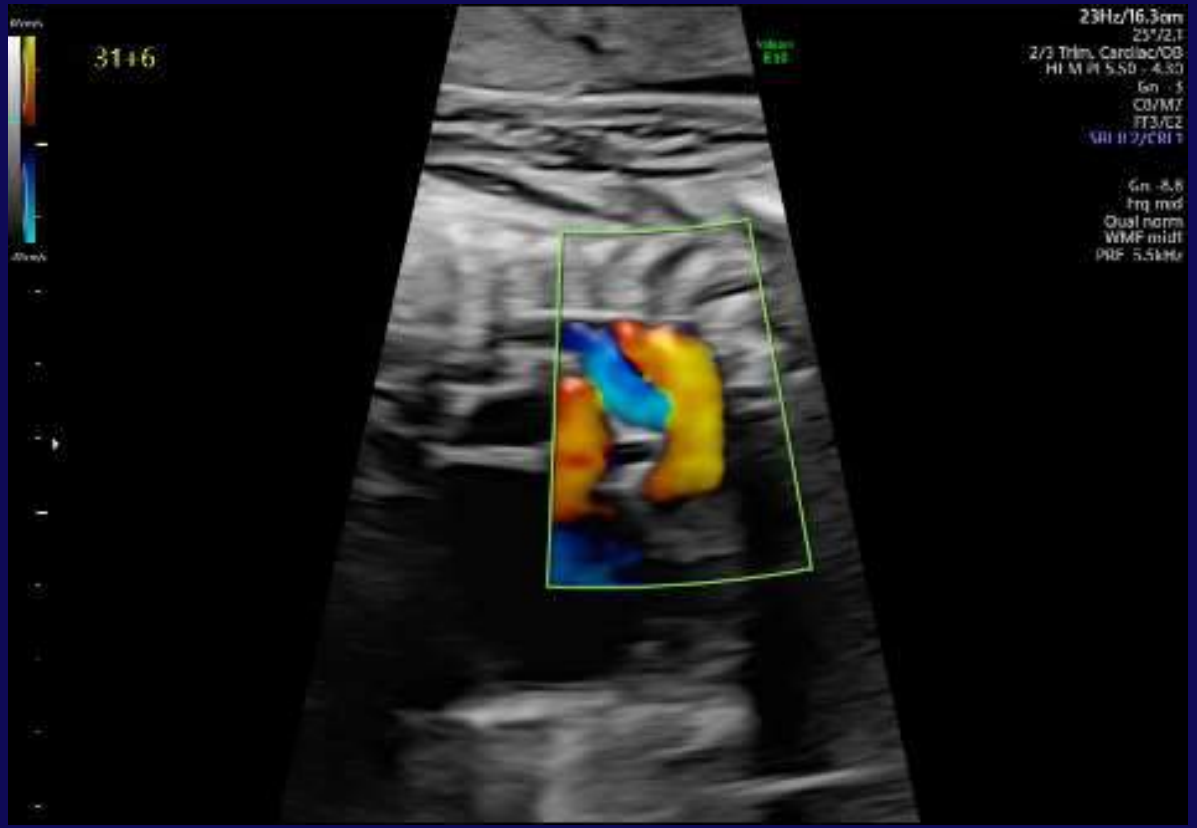


Fetal Echo at 31+6 weeks



Low TR jet velocity → Failing RV

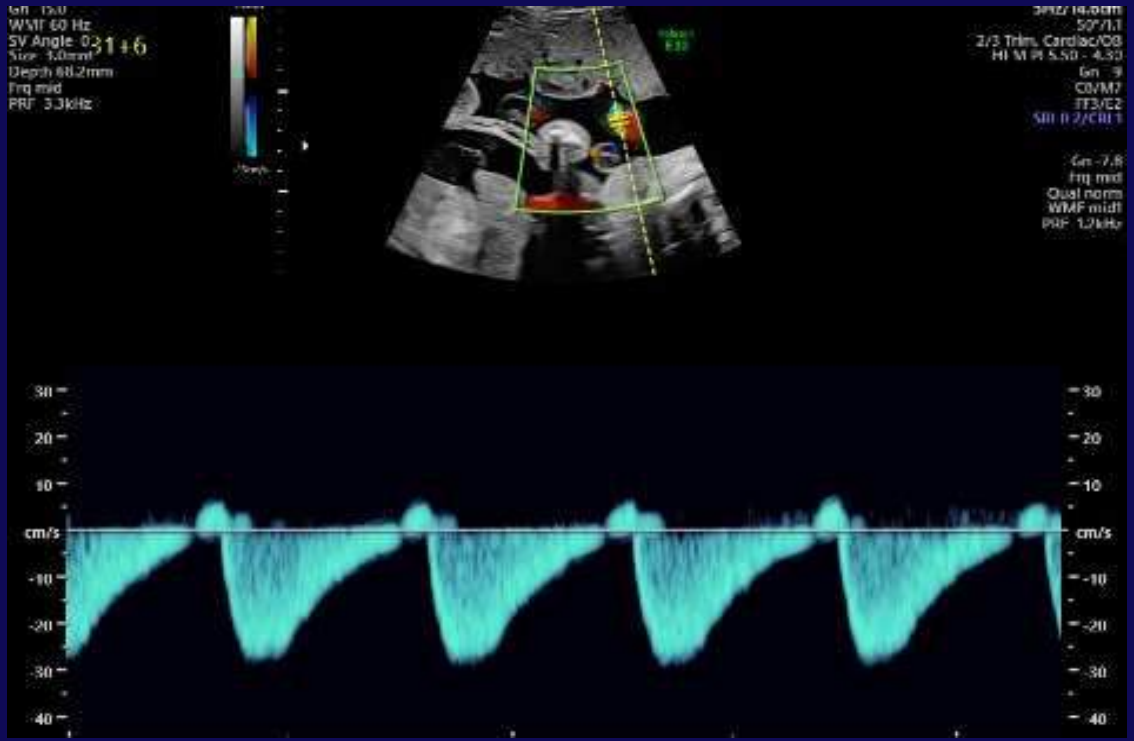
Fetal Echo at 31+6 weeks



Fetal Echo at 31+6 weeks

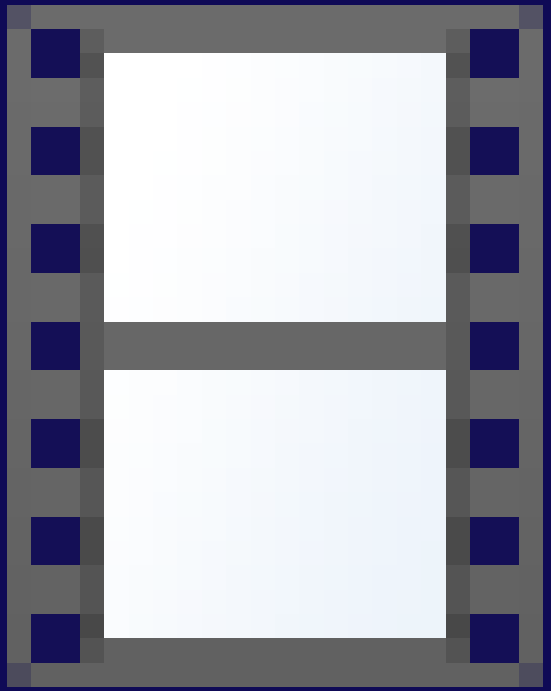
Severe pulmonary regurgitation = Significant circular shunt

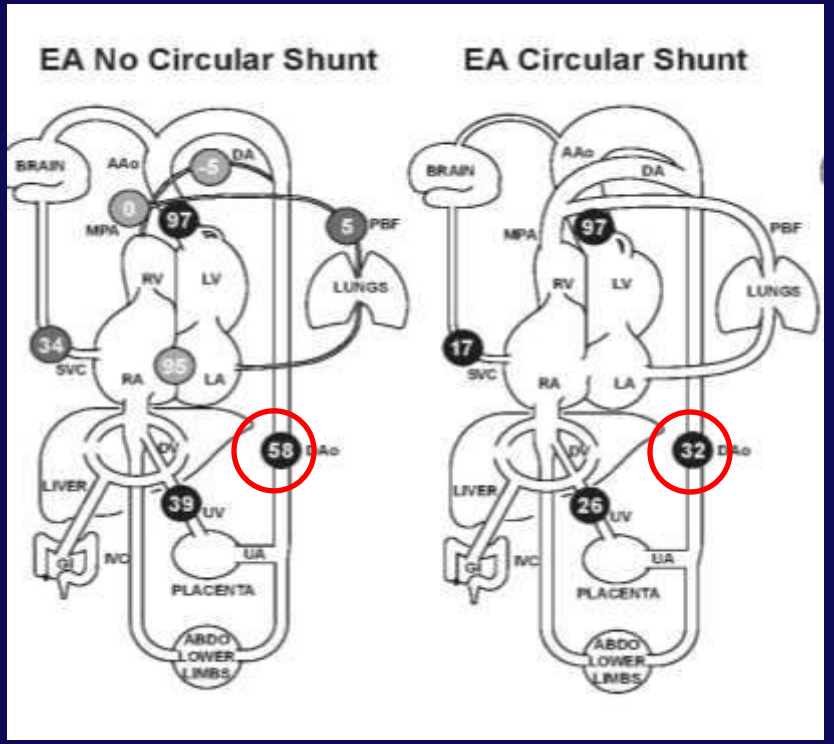
Fetal Echo at 31+6 weeks



Umbilical artery: diastolic flow reversal (systemic steal)

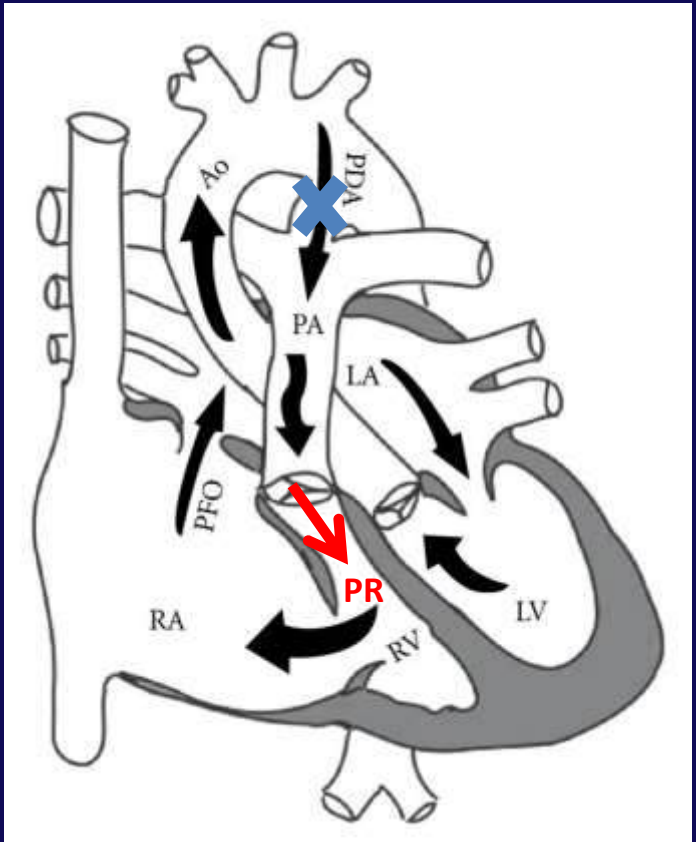
Fetal Cardiac MRI





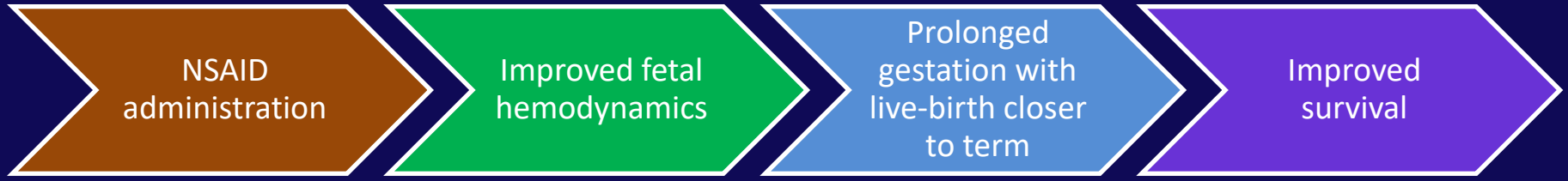
Courtesy of Liqun Sun, Davide Marini, Mike Seed

Fetal Therapy?

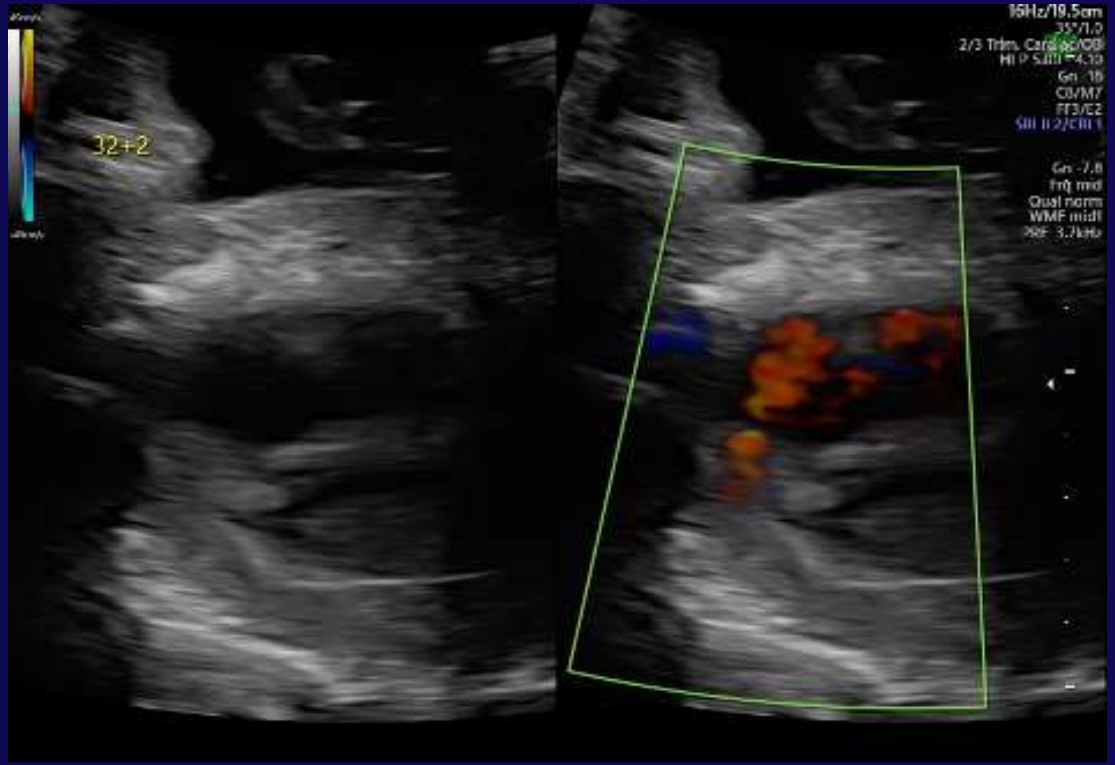


Can constriction of the duct with NSAIDs help interrupt the “circle of death”?

Conceptual Model: Fetal Therapy with NSAIDs



Proof of Concept: 3 days later on NSAIDs



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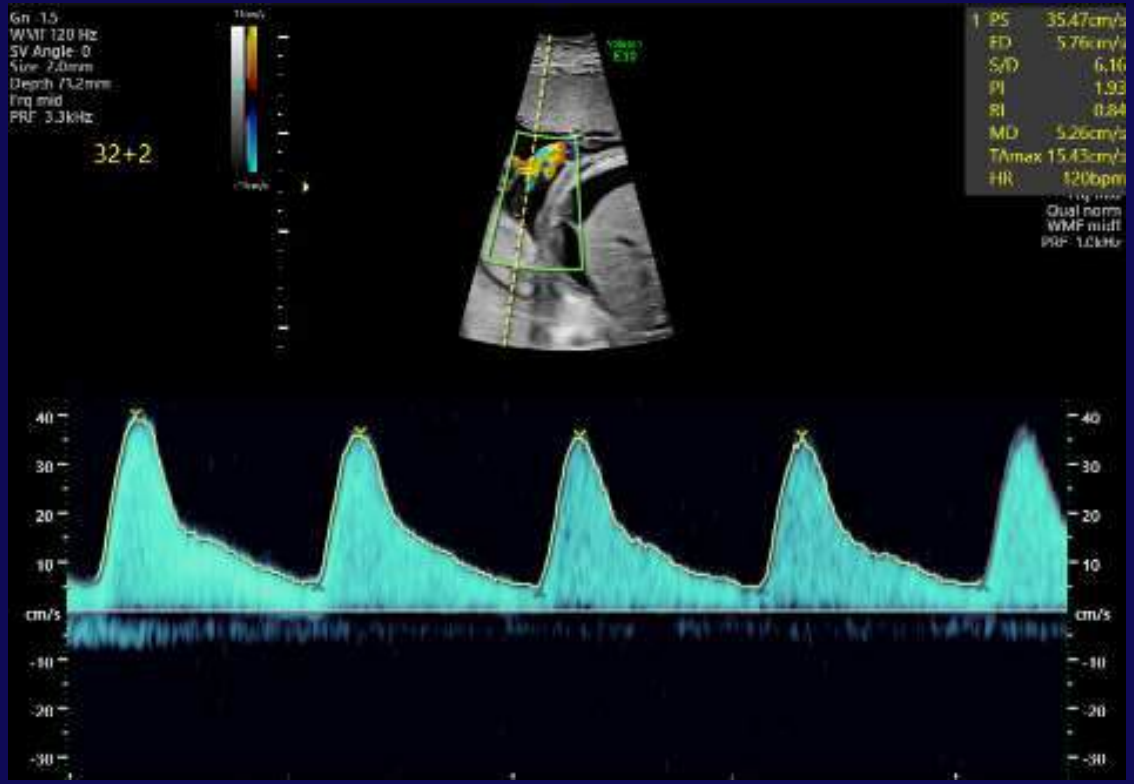
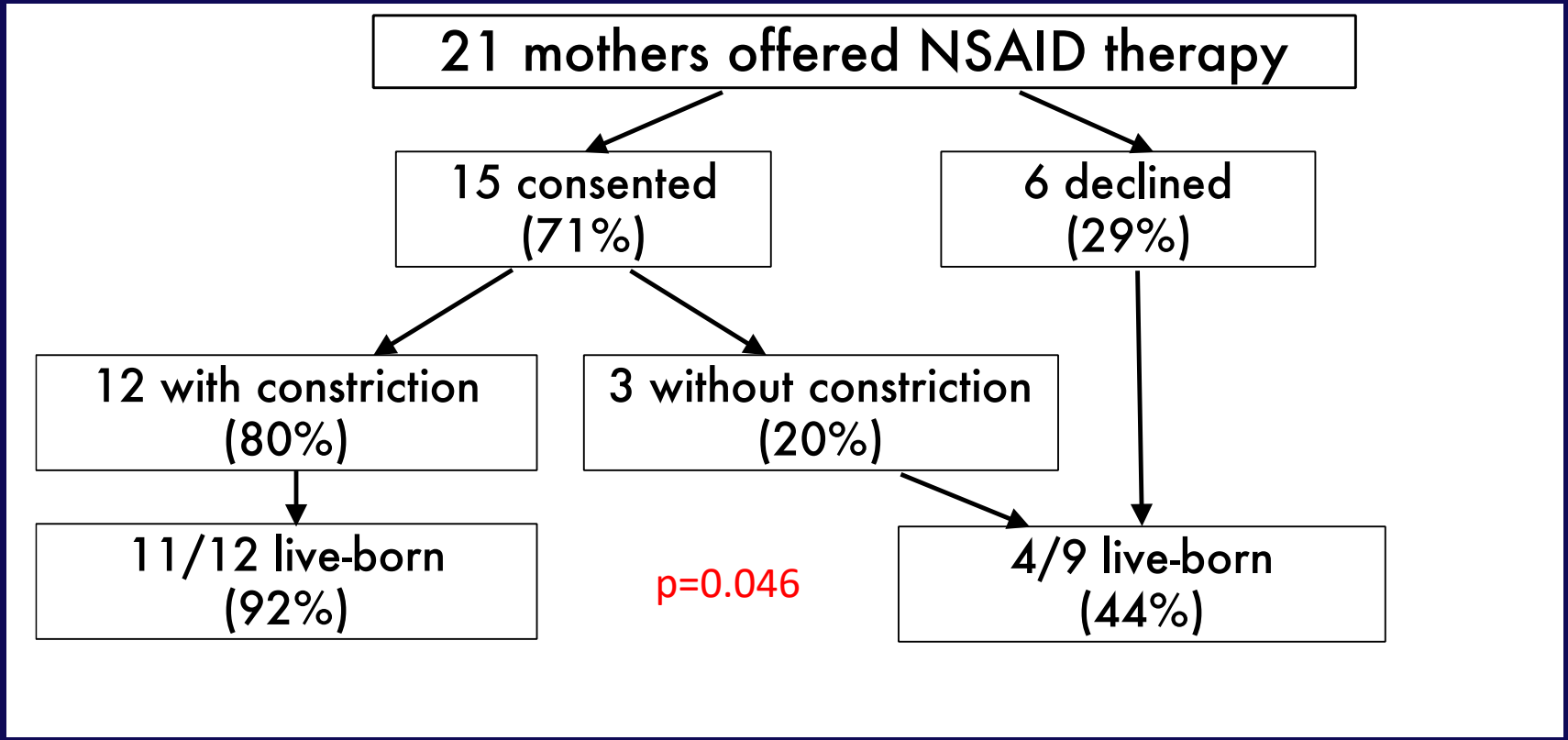
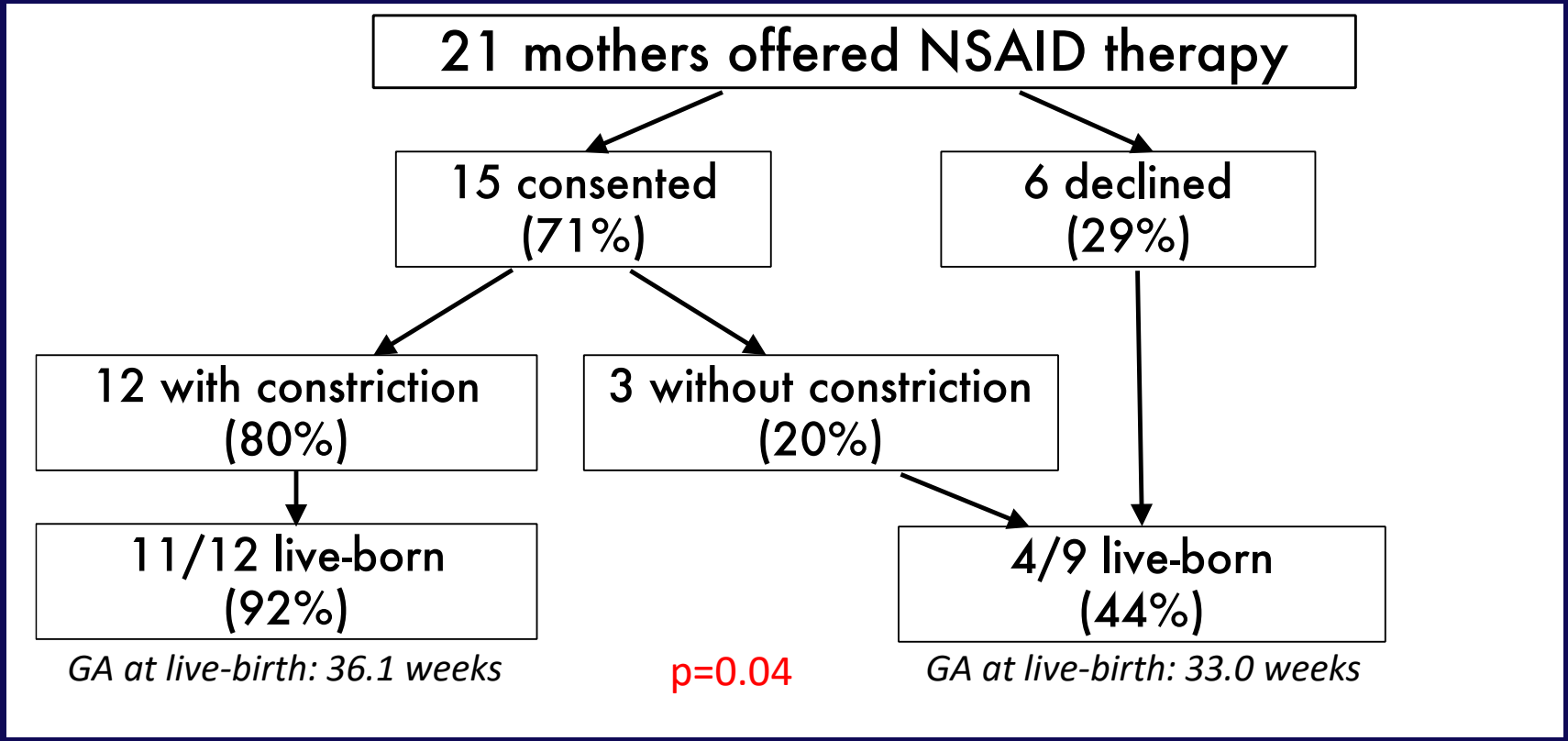


Umbilical artery: restoration of antegrade diastolic flow



Freud *et al*, *AJC* 2021



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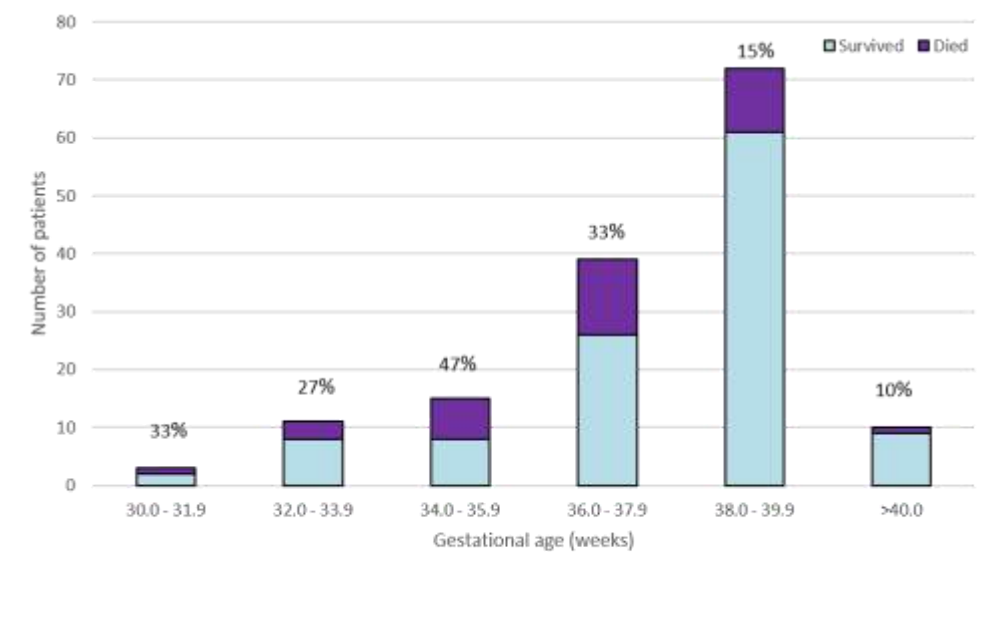
Delivery Planning is Essential for High-Risk Patients with EA/TVD



Aim for delivery >37 weeks
- 4-fold improvement in survival

Multi-disciplinary planning should occur in late gestation

Freud *et al, JAHA* 2020

Risk Factors for Neonatal Mortality

- All of the previously discussed risk factors

PLUS

- Lower birth weight

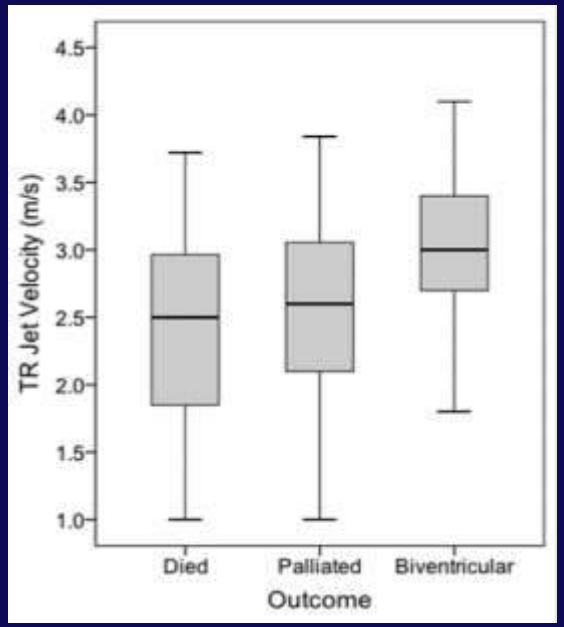
Freud *et al, Circ* 2015
Freud *et al*, *JAHA* 2020

Risk Factors for Neonatal Mortality

- All of the previously discussed risk factors

PLUS

- Lower birth weight
- Lower RV pressure by TR jet



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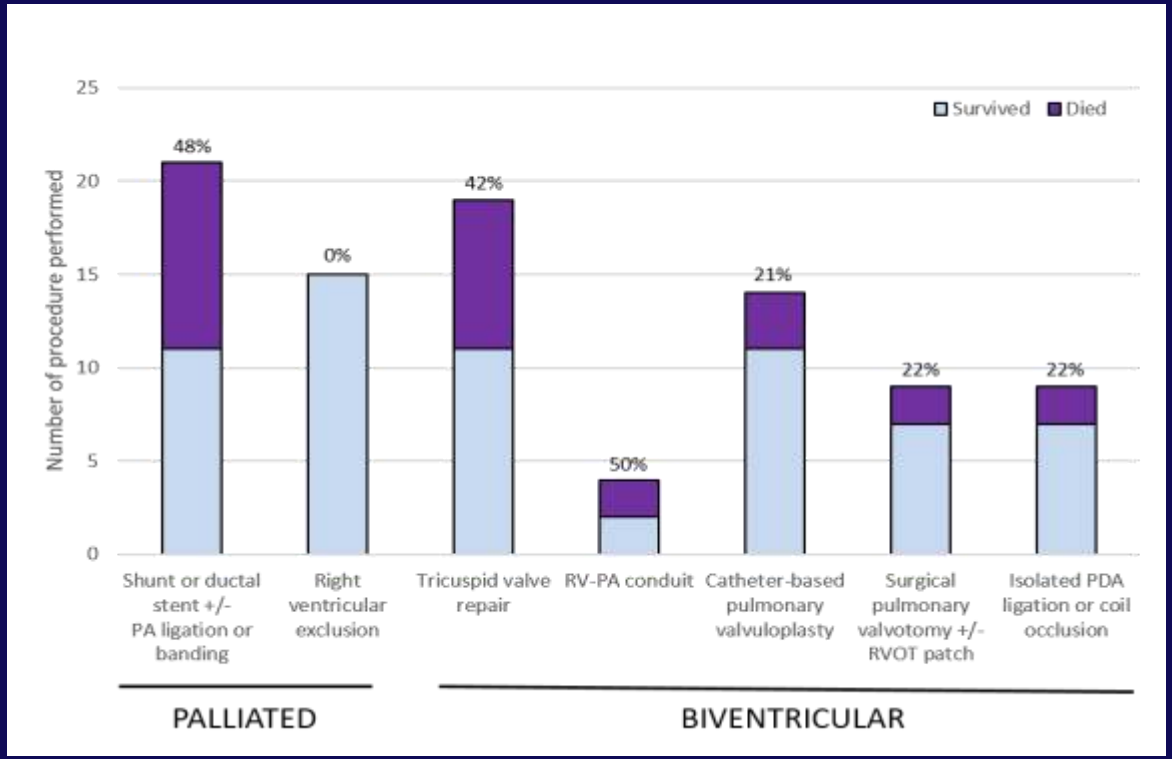
Freud *et al*, *JAHA* 2020

Neonatal Intervention and Mortality

Management

- Approx ½ with catheter or surgical management (n=71)



Freud *et al*, *JAHA* 2020

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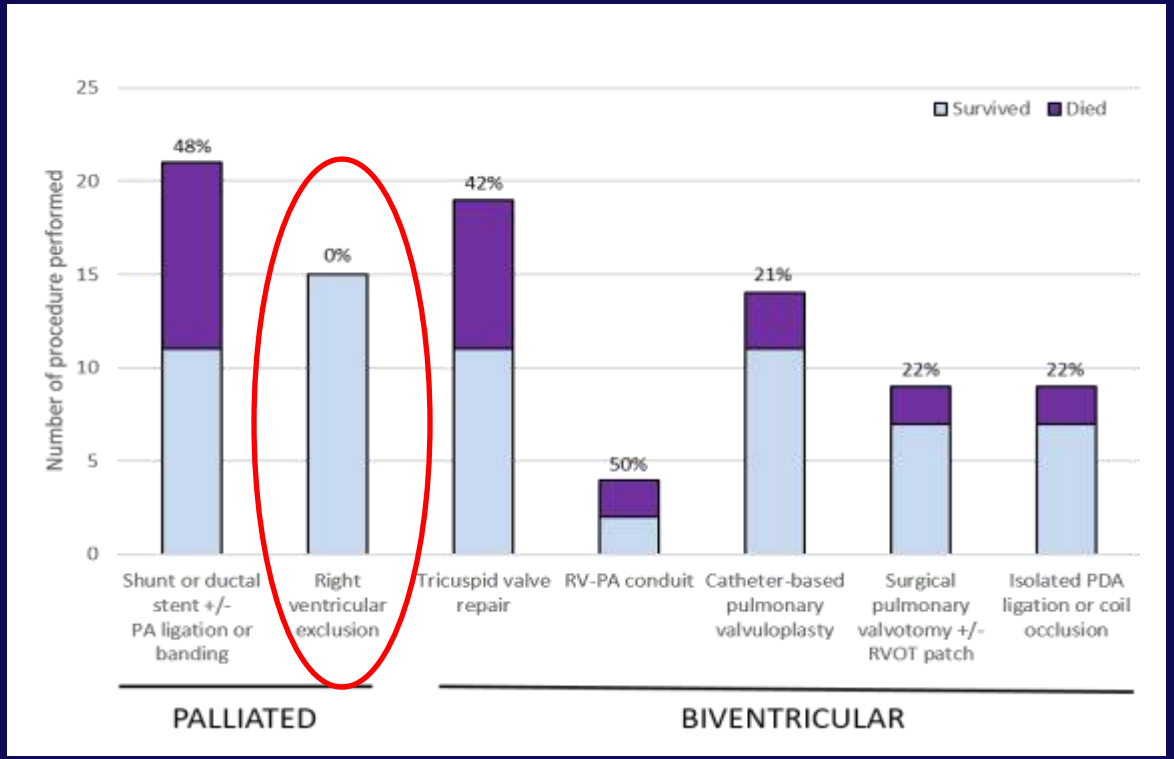

Freud *et al*, *JAHA* 2020

The American Association for Thoracic Surgery (AATS) 2024 expert consensus document: Management of neonates and infants with Ebstein anomaly

Neonates with EA and circular shunt with hemodynamic instability and low RVSP (<20-25 mm Hg) should undergo ligation/occlusion of the main PA and Starnes procedure (class I, level C-EO). Similarly,

Konstantinov *et al, JTCVS* 2024

Single Ventricle Forever? Not Necessarily . . .

- Evolving cases of Starnes takedown once RV has recovered

The completed Da Silva cone repair after a neonatal Starnes procedure.

CENTRAL MESSAGE

Tricuspid valve repair using the cone technique after a neonatal Starnes procedure is feasible, allowing right ventricle rehabilitation and restoring a biventricular physiology.

Da Silva *et al, JTCVS Tech* 2020
Ashraf *et al, JTCVS* 2024

Summary

- Understanding pathophysiology of EA/TVD is essential for
  - Counseling
  - Prenatal management
    - Fetal therapy with NSAIDs
  - Multi-disciplinary delivery planning
  - Neonatal management

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Fetal Ebstein Anomaly and Tricuspid Valve Dysplasia = FEAT Registry

40 sites on 4 continents

Thank you!

Questions? lindsay.freud@sickkids.ca

To Join the FEAT Registry: feat.registry@sickkids.ca

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