

OCTOBER 9TH, 2026

Born Into a Plan

Coordinating Delivery and Immediate
Resuscitation for Critical Fetal Heart Disease

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[UCH/CFCC MFM High Risk Ultrasound Conference](#)



Disclosures

I have nothing to disclose.

Off-label and investigational therapies discussed

- Transplacental antiarrhythmics (digoxin, flecainide, sotalol) and CHB therapies (steroids, IVIG, β -agonists, hydroxychloroquine)
- Transplacental NSAIDs for Ebstein anomaly with circular shunt
- Maternal hyperoxygenation (acute diagnostic testing; chronic therapy is not recommended)
- Fetal cardiac interventions (aortic valvuloplasty, atrial septoplasty/stent) — performed at select centers

Objectives for today

1

Apply a fetal echo–based level-of-care (LOC) framework to plan where, when, and with whom a baby with critical CHD is delivered.

2

Recognize the fetal findings that predict instability in the first hour of life — and where prediction still fails (d-TGA).

3

Describe lesion-specific fetal therapy and first-hour plans for the lesions most likely to be unstable at birth and recognize the larger group that is not.

How this talk is organized

01

The framework

Why a plan matters, what changes at birth, and the level-of-care system

02

Predicting the first hour

How good fetal echo is, why serial scans matter, and hyperoxygenation testing

03

Lesion playbook

Each lesion on one scaffold:
fetal assessment → fetal therapy → perinatal management

04

What you can do

Concrete actions for MFM, OB, and sonographers

The framework

Diagnosis is not the finish line: planning the transition

01

A fetal diagnosis only helps if the first hour is planned

- Fetal diagnosis improves outcomes for many critical lesions by preventing postnatal instability.
- But for a subset, compromise begins in the delivery room — before transport is possible:
 - HLHS or d-TGA with restrictive / intact atrial septum
 - Tetralogy of Fallot with absent pulmonary valve
 - Severe Ebstein anomaly
 - Arrhythmias with hydrops; complete heart block
- For these babies, specialty care has to start in the delivery room.

6.1%

of 670 fetuses with major CHD were judged high-risk for instability within 2 hours of birth (40 fetuses)

50%

of those high-risk fetuses needed intervention, ECMO, or died within 2 hours (70% by 24 hours)

What changes when the cord is clamped

Placenta removed

The fetus loses its oxygen source and low-resistance circuit. Systemic resistance jumps.

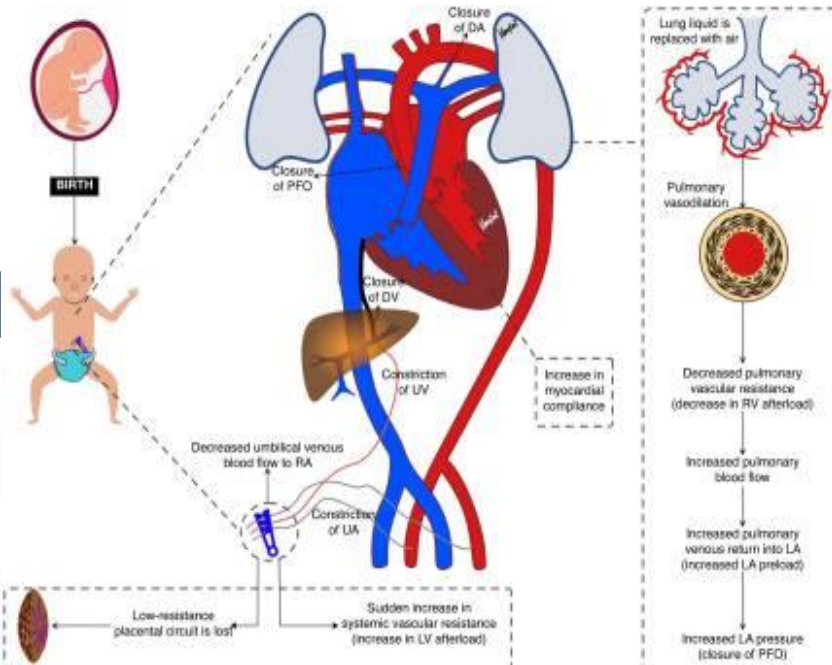
Lungs open

Pulmonary resistance falls; pulmonary blood flow and venous return to the left atrium rise several-fold.

Shunts close

The foramen ovale flap often closes as LA pressure rises; the ductus constricts over hours to days.

Lesion	Depends on...	What fails at birth
HLHS with restrictive / intact septum	Left atrial egress across the septum	Pulmonary venous hypertension, hypoxemia
d-TGA/IVS	Mixing at the FO (and ductus)	Profound cyanosis, acidosis
Obstructed TAPVR	Unobstructed vertical vein	Pulmonary edema, low output
TOF / absent pulmonary valve	An airway not compressed by huge PAS	Respiratory failure
Ebstein anomaly	Falling PVR to allow RV forward flow	Cyanosis, low output, circular shunt
CHB / tachyarrhythmia	Adequate rate and function	Low output as afterload rises



The placenta is the fetus's lung, kidney, and safety net — and it is removed in seconds.

Donofrio level-of-care (LOC) assignment

Table 1

Definition of congenital heart disease level of care assignment and coordinating action plan

LOC	Expected Physiology	Example CHD	Delivery Recommendations	Cardiac DR Recommendations
1	No instability expected in first weeks of life	ASD, VSD, AVSD, mild valve disease	Delivery at local hospital	Arrange outpatient follow-up
2	Stability in DR expected but requiring postnatal catheterization or surgery	Ductal-dependent lesions including HLHS, PA/IVS, severe TOF	Planned vaginal induction at about 39 weeks at local hospital; transport to CNMC	Neonatologist in DR; PGE if indicated
3	Instability requiring immediate specialty care in DR before catheterization or surgery	HLHS or TGA with RFO, CHD or arrhythmia with decreased heart function	Planned vaginal induction at WHC at about 38–39 weeks with “bailout” to C/S if necessary	CNMC specialists in DR; medications predetermined by care plan
4	Instability requiring immediate catheterization or surgery in DR	HLHS or TGA with severe RFO or IAS, CHD or arrhythmia with hydrops	Planned C/S at CNMC usually at 38 weeks	Multidisciplinary specialized care team in DR; medications and equipment predetermined by care plan, including ECMO if indicated

How to read it: LOC 1–2 stable in the delivery room · LOC 3 needs a specialist in the room · LOC 4 needs cath or surgery immediately. Refined on the final pre-delivery echo.

Table 1 reproduced from Donofrio MT, et al. Specialized delivery room planning for fetuses with critical congenital heart disease. Am J Cardiol. 2013;111:737–747.

Fetal echo predictors of LOC 3 vs LOC 4

Table 2
Fetal echocardiographic predictors of level of care 3 or 4

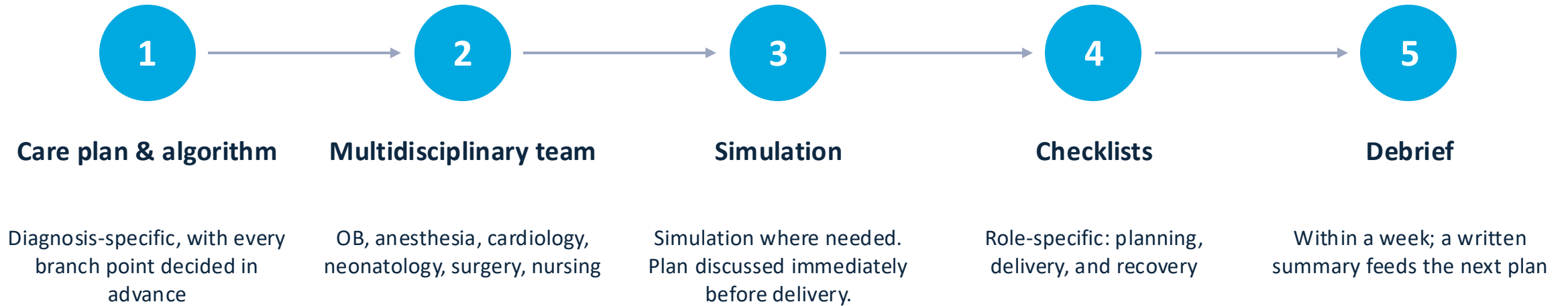
Diagnosis	LOC 3	LOC 4	Care Plan
HLHS with RFO or IAS	PV f/r <5 and >3 (5)	PV f/r <3 (6)	LOC 3: PGE, catheterization pending confirmation of RFO in DR LOC 4: PGE, immediate catheterization
TAPVR (obstructed)	N/A	Accelerated flow in connecting vein	Consider ECMO
TGA with RFO or IAS	Hypermobility (7) or tethered atrial septum or angle of septum primum <30° (8,9) *	Atrial septal bowing >50% into LA, IAS, or RFO with abnormal ductus (z score <2 and/or abnormal flow) (8,9)	LOC 3: PGE, plan for BAS in cardiac ICU LOC 4: PGE, plan for BAS in DR
Severe TOF/APV	Dilated heart with decreased function, significant arrhythmias	Hydrops, CLE on fetal MRI (24)	PHTN therapy if abnormal ductus Specialized ventilation, consider ECMO
Severe Ebstein's anomaly	Dilated heart with decreased function, significant arrhythmias	Hydrops	Therapy to decrease PVR and improve function With hydrops, consider early delivery
Tachyarrhythmias	Uncontrolled arrhythmia with decreased heart function	Hydrops	Cardioversion or medical therapy in DR If hydrops, consider early delivery
Complete heart block	Decreased CVP score to <7 (20), ventricular rate <55 with decreased function	Hydrops	Pacing in DR vs medical chronotrope If hydrops, consider early delivery

BAS = balloon septostomy; CLE = congenital lobar emphysema; CVP = cardiovascular profile; ECMO = extracorporeal membrane oxygenation; IAS = intact atrial septum; ICU = intensive care unit; LA = left atrium; MRI = magnetic resonance imaging; PGE = prostaglandin E; PHTN = pulmonary hypertension; PVR = pulmonary vascular resistance; RFO = restrictive foramen ovale; TAPVR = total anomalous pulmonary venous return; TGA = d-transposition of the great arteries; TOF/APV = tetralogy of Fallot with absent pulmonary valve.

* Criteria for LOC 4 planning updated to include these given experience with this group.

Table 2 reproduced from Donofrio MT, et al. Am J Cardiol. 2013;111:737–747.

A plan is more than a location



Result (Donofrio): algorithms anticipated every event in 8 of 12 children's-hospital deliveries; the 4 unanticipated events were all recoverable. Delivery room survival was 100%.

Every person and device has a place

Delivery planning: room setup and team positions

Mother delivers in the delivery suite; the baby moves through the connecting doors to infant stabilization

Infant stabilization suite Resuscitation: NICU, CICU, CT surgery, ECMO, echo

Delivery suite Maternal OR: OB team



OB / maternal team

- 1 OB anesthesia
- 2 Obstetrician (x2)
- 3 OB scrub
- 4 OB circulator

Cardiac surgery / ECMO

- 5 Cardiac anesthesia
- 6 CT surgeon
- 7 CT scrub
- 8 CT circulator
- 9 ECMO specialist

Neonatal / cardiac ICU

- 10 NICU attending
- 11 CICU attending

Echo

- 12 Echo attending
- 13 Sonographer

Respiratory / airway

- 14 Respiratory therapist
- 15 Airway

Key

-  Baby transfer
-  Equipment
-  IV pole
-  Fixed structure
-  Glass doors

Why map the room?

- Multiple roles in one OR
- The echo machine, ECMO cart, and infant warmer all compete for space
- Mapped for every phase of care
- Rehearsed prior to delivery with roles and actions discussed explicitly

Two published models, one lesson

Washington, DC (Donofrio 2013, n=34)

- LOC 3: delivery at the adjacent adult hospital with children's-hospital specialists in the room
- LOC 4: planned cesarean in the children's hospital OR
- Children's-hospital delivery: time to care 1 vs 7 min; time to intervention 64 vs 117 min
- Survival to discharge similar (83% vs 86%)

Edmonton (Moray 2024, n=40)

- LOC 4: planned cesarean in the pediatric cardiac OR, remote from the OB hospital
- No mortality at delivery; 82% survival to discharge
- High-risk HLHS: 80% survival with cardiac OR delivery vs 0% without (4/5 vs 0/2)
- No major maternal complications

The lesson

Detailed planning with a specialist in the room and a rapid path to intervention mattered more than the building the baby was born in.

Consistency and experience create an ideal model for success.

Donofrio MT, et al. Am J Cardiol. 2013;111:737–747 (Table 3). Moray A, et al. J Am Heart Assoc. 2024;13:e031184.

Predicting the first hour

How accurate is fetal echo — and how do we make it better?

02

How well does fetal echo predict the first hour?

84%

of Donofrio's LOC 4 fetuses needed immediate intervention

50%

Moray PPV for intervention / ECMO / death at ≤2 h (all high-risk lesions)

70%

Moray PPV for the same outcome within 24 h

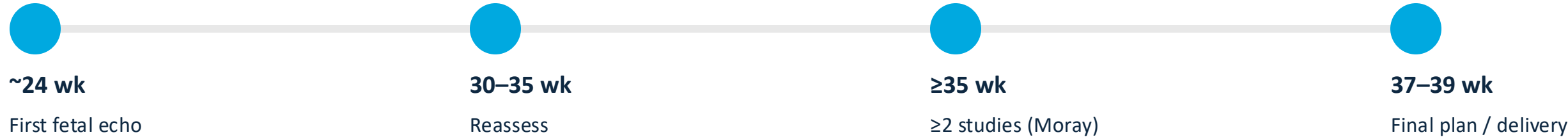
0 / 45

low-risk HLHS needed intervention ≤2 h (Moray)

	Donofrio 2013	Moray 2024
HLHS	PV f/r <3: sensitivity 80%, specificity 100%	Sensitivity 100%, specificity 95%, PPV 71%, NPV 100%
d-TGA/IVS	LOC 4 assignment: sensitivity 40%, specificity 0%	Sensitivity 67%, specificity 93%, PPV 80%, NPV 87%
Advanced LOC (3–4) vs LOC 2	HLHS 100% / 98%; d-TGA 100% / 78% (sens / spec)	6 of 46 “low-risk” d-TGA needed BAS ≤2 h

Donofrio MT, et al. Am J Cardiol. 2013;111:737–747. Moray A, et al. J Am Heart Assoc. 2024;13:e031184.

The 24-week scan does not set the delivery plan



HLHS (n=11)

- Pulmonary vein f/r ratio worsened in every fetus: mean 3.8 at ~24 wk → 1.8 near term (p=0.005)
- Final ratio <3 in all 11
- 7 moved from LOC 3 to LOC 4

d-TGA (n=12)

- Atrial septal restriction increased in 10 of 12
- Ductus abnormal on the final study in 9
- 6 moved from LOC 3 to LOC 4
- Across all lesions: no fetus's findings improved

Donofrio MT, et al. Am J Cardiol. 2013;111:737–747. Moray A, et al. J Am Heart Assoc. 2024;13:e031184.

Previewing the first breath: hyperoxygenation (MHO)

- 100% O₂ to the mother by non-rebreather for ~10 minutes during the fetal echo
- Compare branch pulmonary artery Doppler pulsatility index (PI) before and during
- Reactive vessels: PI falls → more pulmonary blood flow and more venous return to the left atrium
- The ductus constricts and FO flow falls; combined output is unchanged

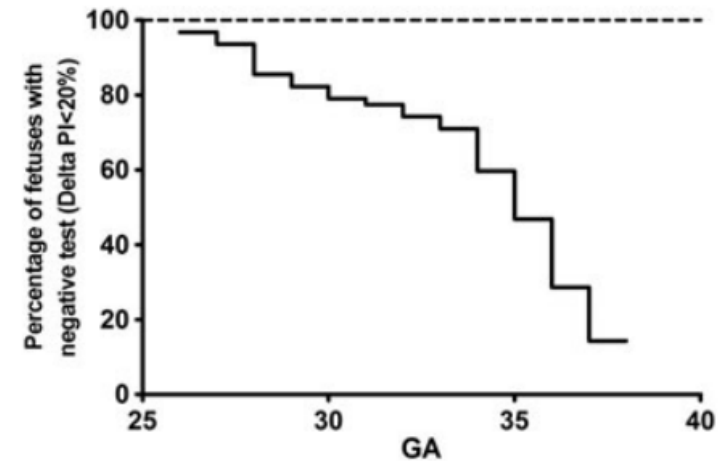


Figure 3 Kaplan-Meier curve expressing the percentage of fetuses with a negative test throughout the pregnancy. GA, gestational age

Normal fetuses with a non-reactive test fall with gestation: ~80% at 30 wk, ~40% at 35 wk.

Think of it as a stress test

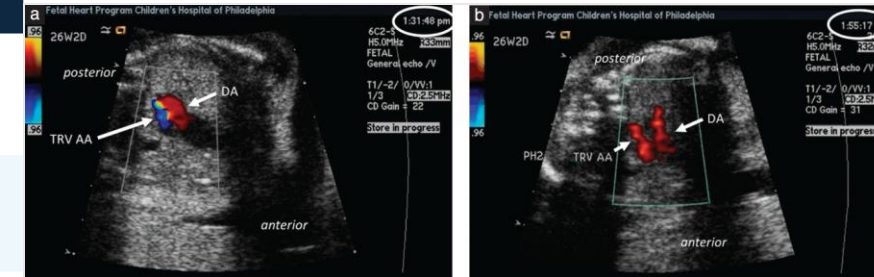
It mimics what the lungs do at birth, so we can see whether the atrial septum, left heart, or a vertical vein can handle the postnatal load.

Altitude caveat

These curves are from sea level and have not been validated at altitude, where neonatal pulmonary hypertension is more common.

How CFCC uses acute MHO

Indication	What changes with MHO	Source
HLHS: is the septum restrictive?	No fall in PA PI; PV f/r stays <7 on MHO	Szwast 2010; Mardy 2021
TAPVC: will the vein obstruct?	Vertical vein gradient rises (2 → 12 mmHg in one case)	Schidlow 2018
Aneurysmal septum vs coarctation	Slender LV fills; reversed arch flow turns antegrade	Channing 2015
Borderline left heart	Mitral/aortic dimensions and LV filling increase	CFCC protocol
d-TGA	Not routine; multicenter FHS study pending	FHS (unpublished)



Aneurysmal septum: retrograde arch flow at baseline becomes antegrade with MHO.

Acute testing only. Chronic MHO to “grow” the left heart slowed fetal head growth and is no longer recommended — families still find those older studies.

Lesion playbook

03



Most critical CHD is stable at birth

Typical lesions

- HLHS with an open atrial septum
- Pulmonary atresia / intact septum; critical pulmonary stenosis
- TOF with pulmonary atresia or severe stenosis
- Tricuspid atresia with pulmonary obstruction
- Coarctation / interrupted arch
- Critical aortic stenosis with preserved LV function

Plan (Donofrio LOC 2)

- Stable in the delivery room; needs cath or surgery later
- Planned induction ~39 weeks; vaginal delivery appropriate
- Neonatologist in the room; PGE if indicated
- Transfer to the cardiac center (Edmonton: within hours)

Reassurance data

- Donofrio: 39 HLHS and 7 d-TGA assigned LOC 2 — none needed emergent intervention
- Moray: 45 low-risk HLHS — none needed intervention ≤ 2 h
- The exception: 6/46 “low-risk” d-TGA needed urgent BAS

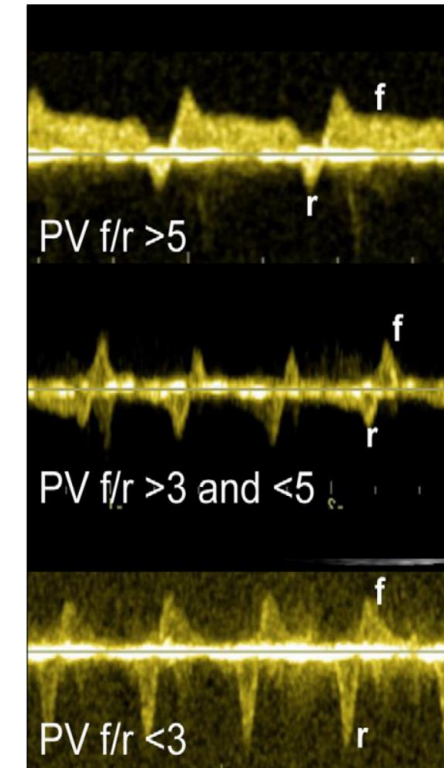
Donofrio MT, et al. Am J Cardiol. 2013;111:737–747 (Table 1). Moray A, et al. J Am Heart Assoc. 2024;13:e031184.

HLHS WITH RESTRICTIVE / INTACT ATRIAL SEPTUM

Fetal assessment

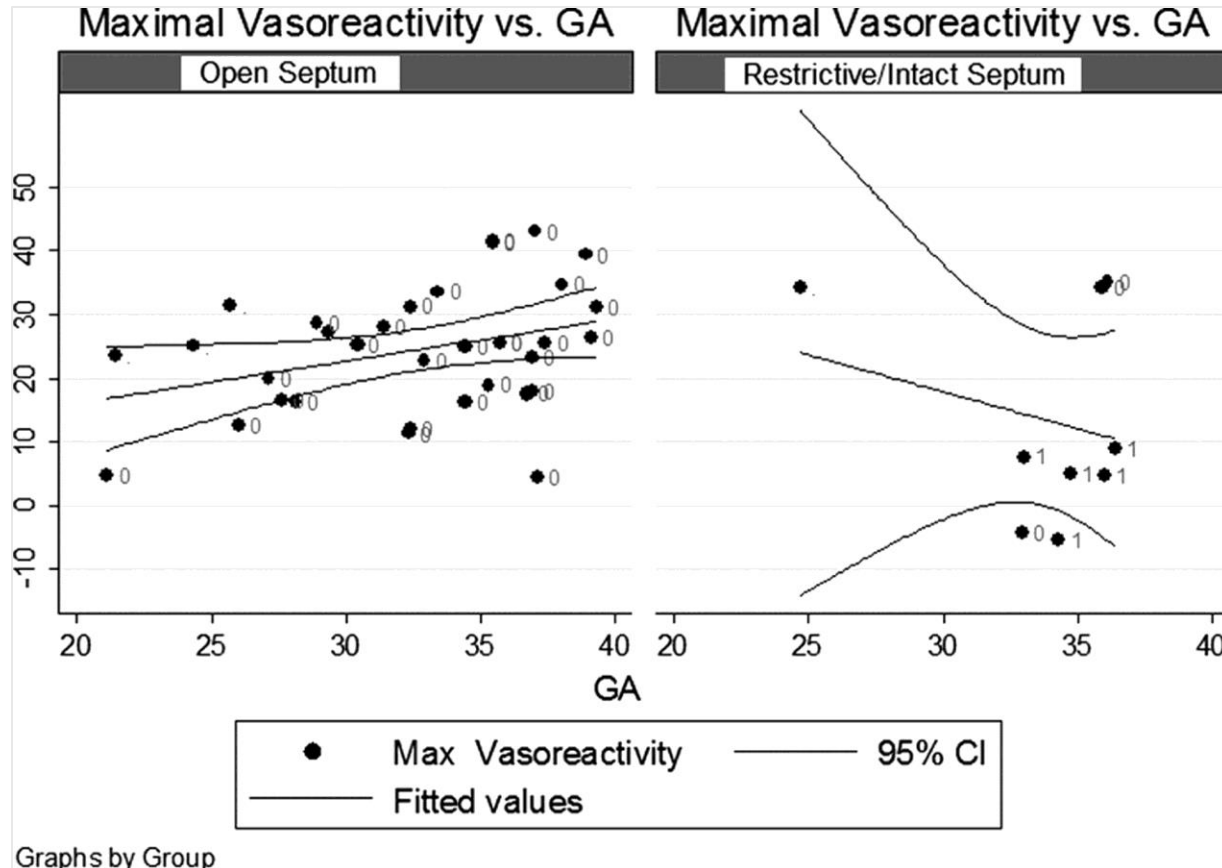


- ~5–6% of HLHS; historical mortality 48–69% with restrictive or intact septum
- Pulmonary vein forward/reverse VTI ratio (PV f/r): $<5 \rightarrow \text{LOC } 3$; $<3 \rightarrow \text{LOC } 4$
- Foramen ovale <3 mm; thick atrial septum
- MHO: no fall in PA PI; PV f/r <7 on MHO (CFCC)
- Fetal MRI “nutmeg lung” = lymphangiectasia from long-standing LA hypertension



Donofrio MT, et al. Am J Cardiol. 2013 (Figure 3 reproduced). Michelfelder E, et al. Circulation. 2005. Divanović A, et al. J Thorac Cardiovasc Surg. 2011. Mardy C, et al. Fetal Diagn Ther. 2021. Saul D, et al. Pediatr Radiol. 2016.

The evidence: vasoreactivity and prediction



Open septum: reactivity rises toward term. Restrictive/intact: essentially none.

Szwast A, et al. Circ Cardiovasc Imaging. 2010 (figure). Donofrio MT, et al. Am J Cardiol. 2013. Moray A, et al. J Am Heart Assoc. 2024.

100%

Moray sensitivity (specificity 95%)

80%

Donofrio sensitivity for PV f/r <3
(specificity 100%)

In practice

- All 4 of Moray's cardiac-OR HLHS fetuses tested showed little or no MHO response
- Donofrio: 3 fetuses with PV f/r <3 were kept at LOC 3; 2 still needed intervention → PV f/r <3 is now always LOC 4
- Decompressing veins and asymmetric veins cause the misses

Fetal therapy

Fetal aortic valvuloplasty

EMERGING

- For critical aortic stenosis evolving to HLHS
- Goal: preserve a biventricular circulation
- Select centers; technical success and safety have improved over 20 years
- Referral window is mid-gestation so please refer early

Fetal atrial septoplasty / stent

INVESTIGATIONAL

- For restrictive or intact septum; select centers
- Although fetal atrial stenting may allow for an easier perinatal transition, long term impact on reversing maladaptive lung development is controversial

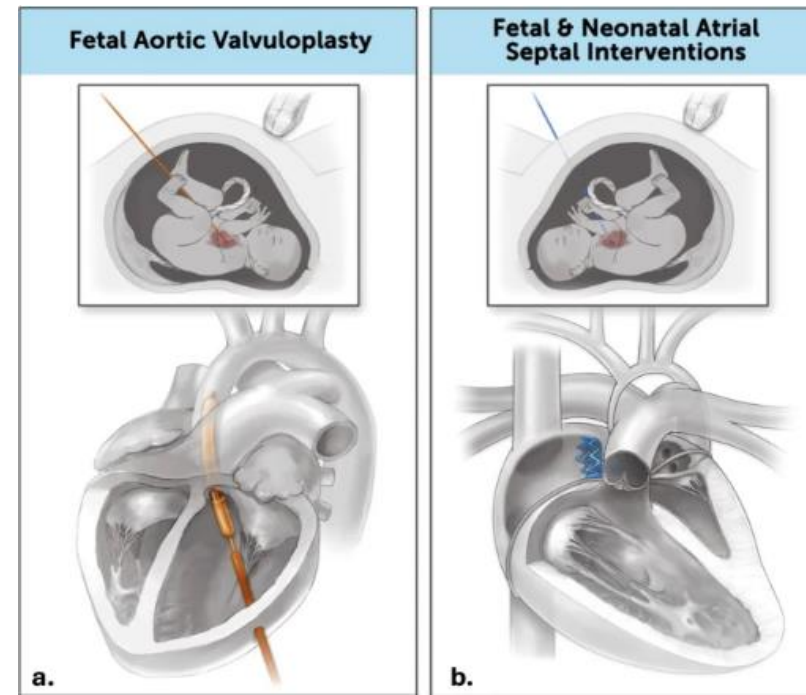
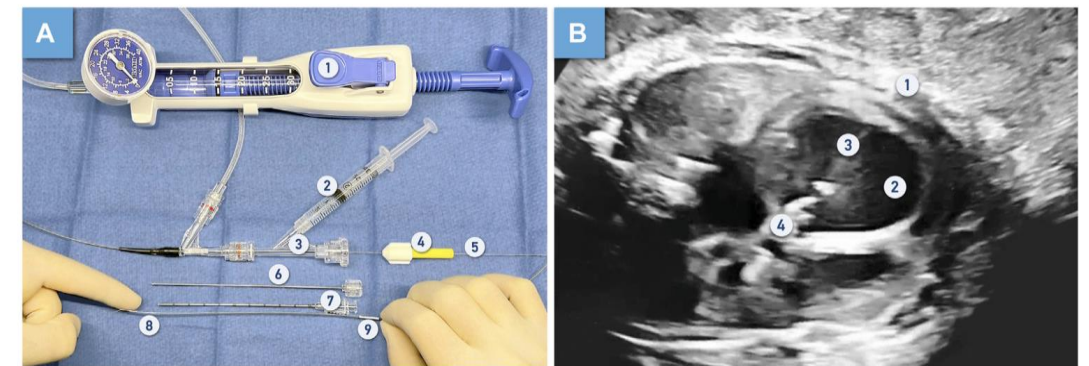


FIGURE 1 Fetal Aortic Valvuloplasty



(A) Interventionalist setup for fetal aortic valvuloplasty: 1. Inflation device, 2. preloaded resuscitation medication, 3. Y-adapter, 4. handle (tightened) for wire manipulation, 5. 0.014-inch support wire, 6. stylet, 7. cannula, 8. balloon, 9. marker (delineates exact exit point of balloon from cannula). (B) Fetal echocardiographic image of a fetal aortic valvuloplasty: 1. Fetal chest wall, 2. left ventricular cavity, 3. cannula, 4. inflated balloon across aortic valve.

Callahan R, et al. Technical success and serious adverse events for fetal aortic valvuloplasty in a large 20-year cohort. JACC Adv. 2024;3:100835 (image). Lara DA, et al. UOG. 2016. Edwards LA, et al. Fetal Diagn Ther. 2019. Sperotto F et al. Innovations in transcatheter interventions for hypoplastic left heart syndrome and borderline left heart structures from fetal to neonatal life: current evidence and systematic review of recent advances (2016-2025). Eur J Pediatr. 2026 Aug 21;185(9):686

Perinatal management

	Plan
LOC	4 (PV f/r <3, intact septum, non-reactive MHO)
Where	Delivery with cath lab immediately available
Timing	Planned, ~38–39 wk unless compromise
Mode	Planned cesarean in both published models
Delivery room	PGE; immediate transfer to cath lab for septostomy or stent
Intact septum	Consider Delivery-to-ECMO; ECMO on standby

80% vs 0% 82%

survival with vs without cardiac OR delivery (4/5 vs 0/2; Moray) HLHS survival (Donofrio) vs historical 48–69%

For MFM

- At least 2 scans after 35 weeks
- Offer genetic testing before an aggressive plan
- Labor plan: call cardiology the moment she presents
- The 2 babies not delivered as planned both died

Donofrio MT, et al. Am J Cardiol. 2013;111:737–747. Moray A, et al. J Am Heart Assoc. 2024;13:e031184.

Fetal assessment: why coarctation is hard to call

Why prediction is hard

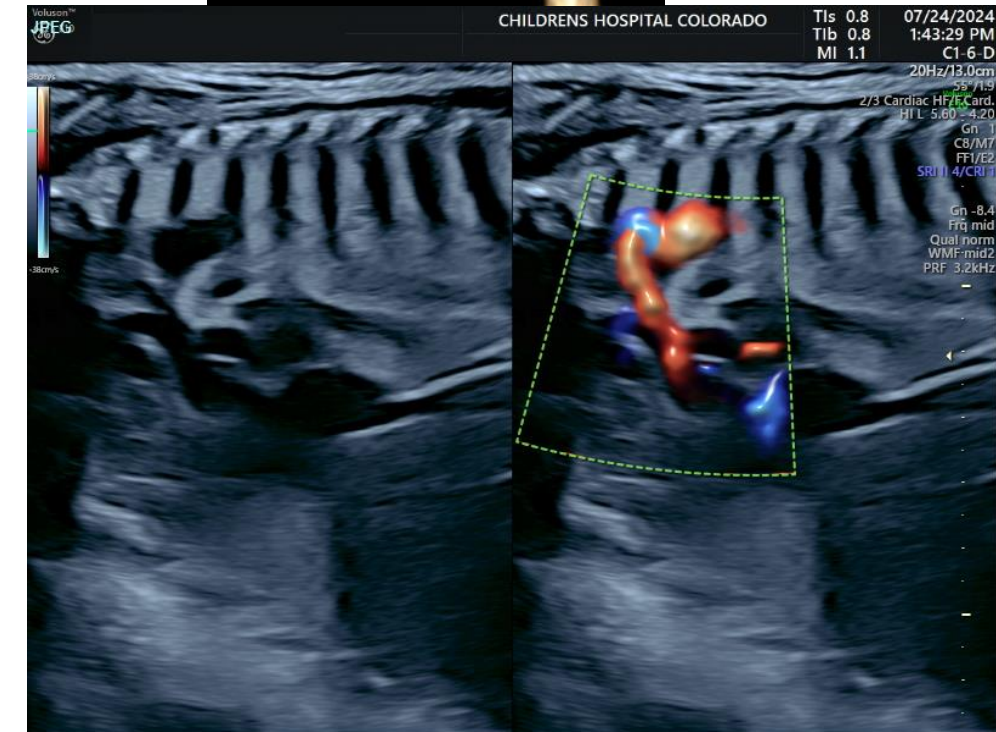
- The open ductus masks the degree of arch obstruction
- Altered fetal flow causes ventricular, valve, or great-artery disproportion — without postnatal coarctation
- Arch size can change through gestation and again after ductal closure

Markers (multicenter)

- **Posterior shelf**
- **Hypoplastic transverse arch**
- Tricuspid/mitral disproportion
- Isthmus/ductal diameter <0.7
- Isthmus z-score <-2
- PA/ascending Ao >1.4

Mimics and adjuncts

- Atrial septal aneurysm can obstruct mitral inflow: borderline arch or retrograde isthmus flow without coarctation
- Maternal hyperoxygenation: better arch flow is reassuring; persistent retrograde flow raises concern



Familiari A, et al. Risk factors for coarctation of the aorta on prenatal ultrasound: a systemic review and meta-analysis. *Circulation*. 2017;135(8):772-85.

Channing A, et al. *Ultrasound Obstet Gynecol*. 2015.

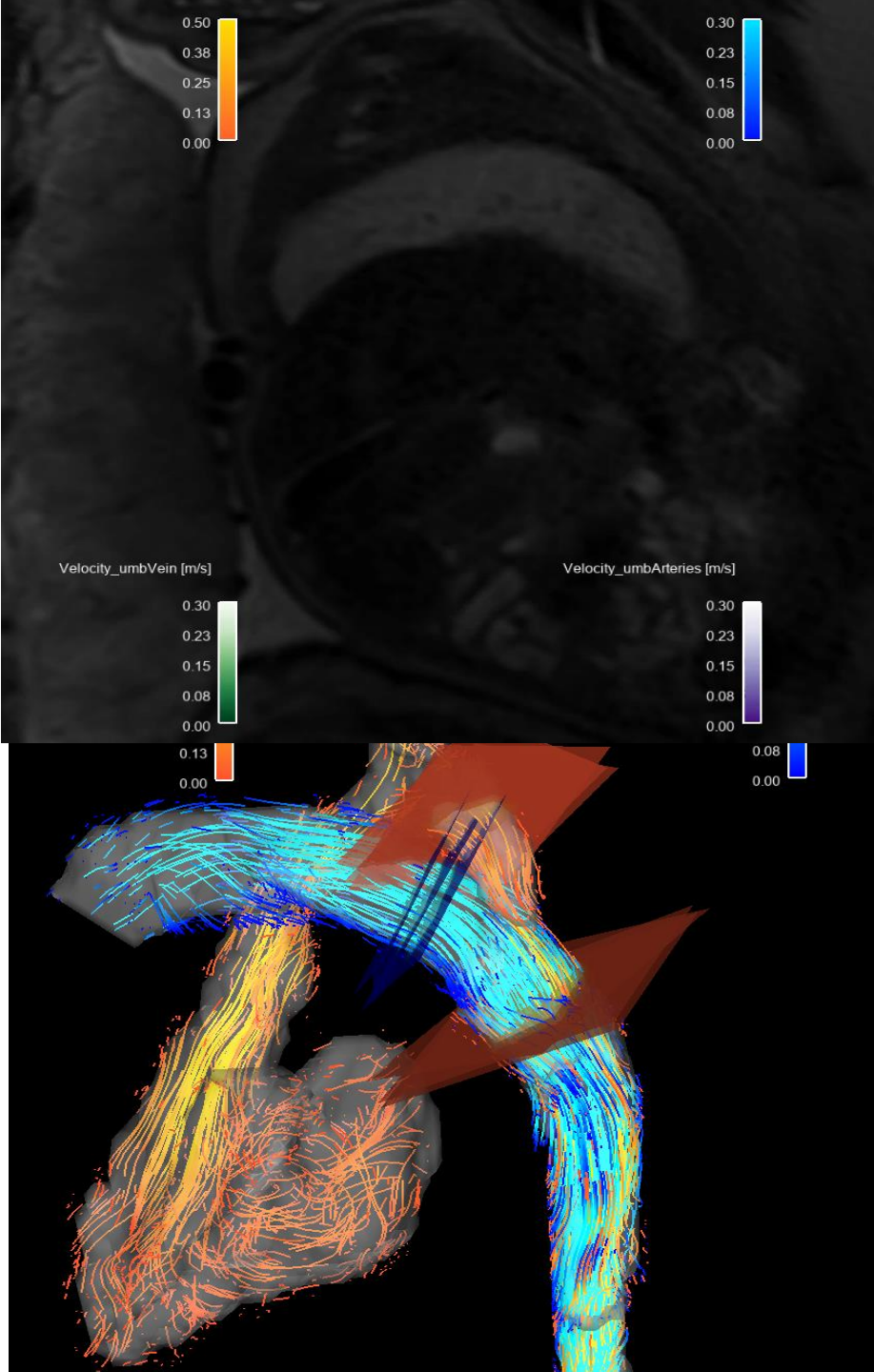
Fetal cardiac MRI: 4D flow

What our fetal CMRI team adds

- Doppler-ultrasound-gated 4D-flow cardiac MRI (CU / Children's Colorado: Friesen, Browne, Barker, and colleagues)
- 3D arch anatomy at multiple sites plus quantitative fetal blood-flow measurement
- Isthmus morphology combined with indexed aortic and arch flow may better discriminate true coarctation
- Good measurement precision and repeatability in the third trimester

Risk tier	Role of fetal CMRI
Borderline, low suspicion	Not routinely needed
Indeterminate	Consider if it would change counseling, delivery site, or management
Probable coarctation	Anatomic and flow detail before delivery planning
High-risk	Characterize the arch before delivery (plan already at CFCC/CHCO)

Englund EK, Fujiwara T, Smith SA, Meyers ML, Friesen RM, Browne LP, Barker AJ. Reliability of 4D Flow MRI for Investigation of Fetal Cardiovascular Hemodynamics in the Third Trimester. Radiol Cardiothorac Imaging. 2024 Dec;6(6):



Perinatal management

	Plan (Dependent on severity)
LOC	2, ductal-dependent until proven otherwise.
Where	Where serial postnatal echo is available
Timing / mode	~39 weeks; obstetric indications
PGE	Start at birth vs observe with serial echo
Discharge	Not until the ductus has closed and the arch is reassessed

For MFM

- Counsel that prenatal CoA is a probability, not a diagnosis
- Plan delivery where the baby can be watched through ductal closure

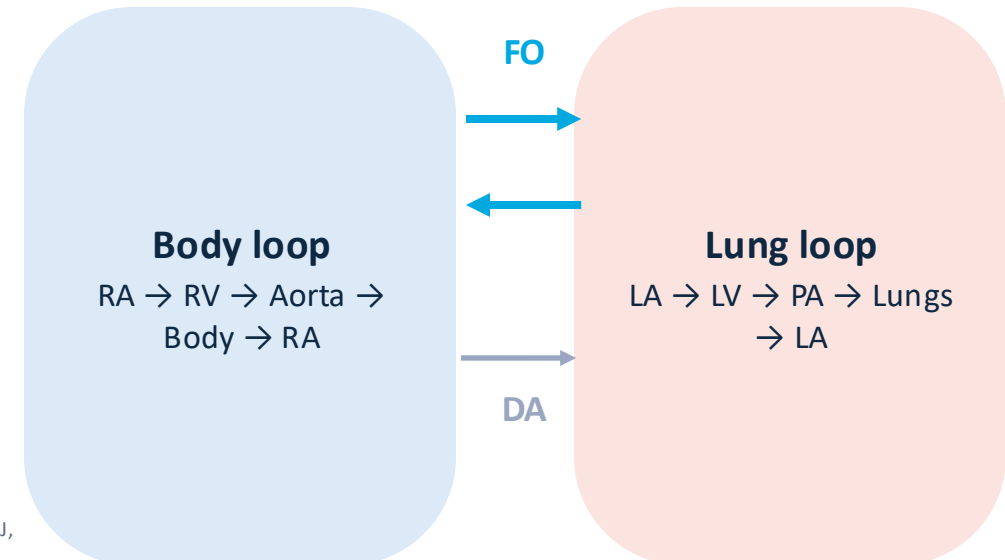
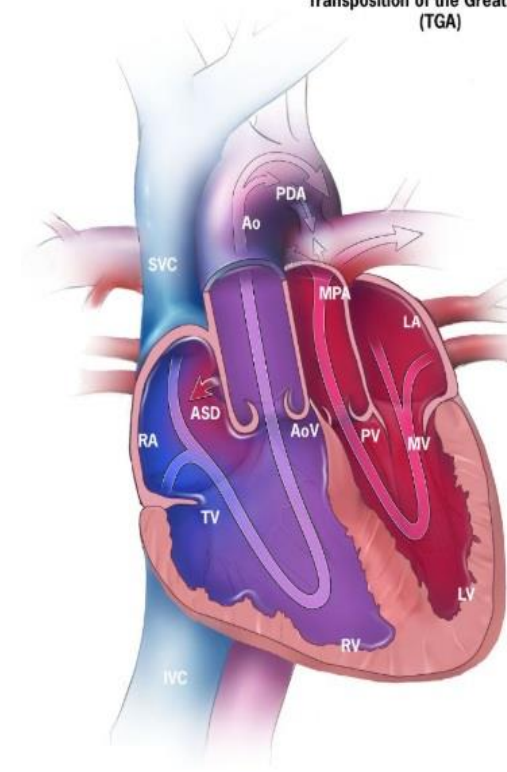
Why it's missed, and why it matters

- The four-chamber view is usually normal
- Detection requires outflow tract and 3-vessel-trachea views: parallel great arteries, no criss-cross
- In Alberta, prenatal detection of TGA rose after outflow views were added to obstetric screening guidelines

Two separate circuits with limited areas of communication

Blue blood circles the body; red blood circles the lungs. The FO and ductus are the only interchanges. Before birth the placenta feeds both loops; after the cord is clamped, the FO is the main interchange — if it's narrow, traffic stops.

d-Transposition of the Great Arteries (d-TGA).” *Centers for Disease Control and Prevention*, 15 May 2024, <https://www.cdc.gov/heart-defects/about/d-tga.html>. Ravi P, et al. *Ultrasound Obstet Gynecol*. 2018;51:659–664. Moon-Grady AJ, et al. *J Am Soc Echocardiogr*. 2023;36:679–723.



Mixing between parallel circulations depends on the interchanges.

Fetal assessment: the sonographer checklist

Atrial septum

- Fixed / flat: septum primum angle $<30^\circ$
- Redundant: bows $>50\%$ across the LA
- Hypermobility vs fixed motion

Foramen ovale size

- Flow orifice ≤ 3 mm
- FO : total septal length <0.5

Ductus arteriosus

- Small or constricted
- Flow direction, including diastolic reversal
- Velocities (acceleration = constriction)

Pulmonary veins

- Peak velocity >41 cm/s associated with Rashkind within 24 h (single study)



Punn R, Silverman NH. JASE. 2011 [confirm definitions source]. Vigneswaran TV, et al. Am J Cardiol. 2017. Słodki M, et al. UOG. 2018;51:531–536. Maeno YV, et al. Circulation. 1999. Donofrio MT, et al. Am J Cardiol. 2013 (Figure 4).

What restriction looks like before and after birth

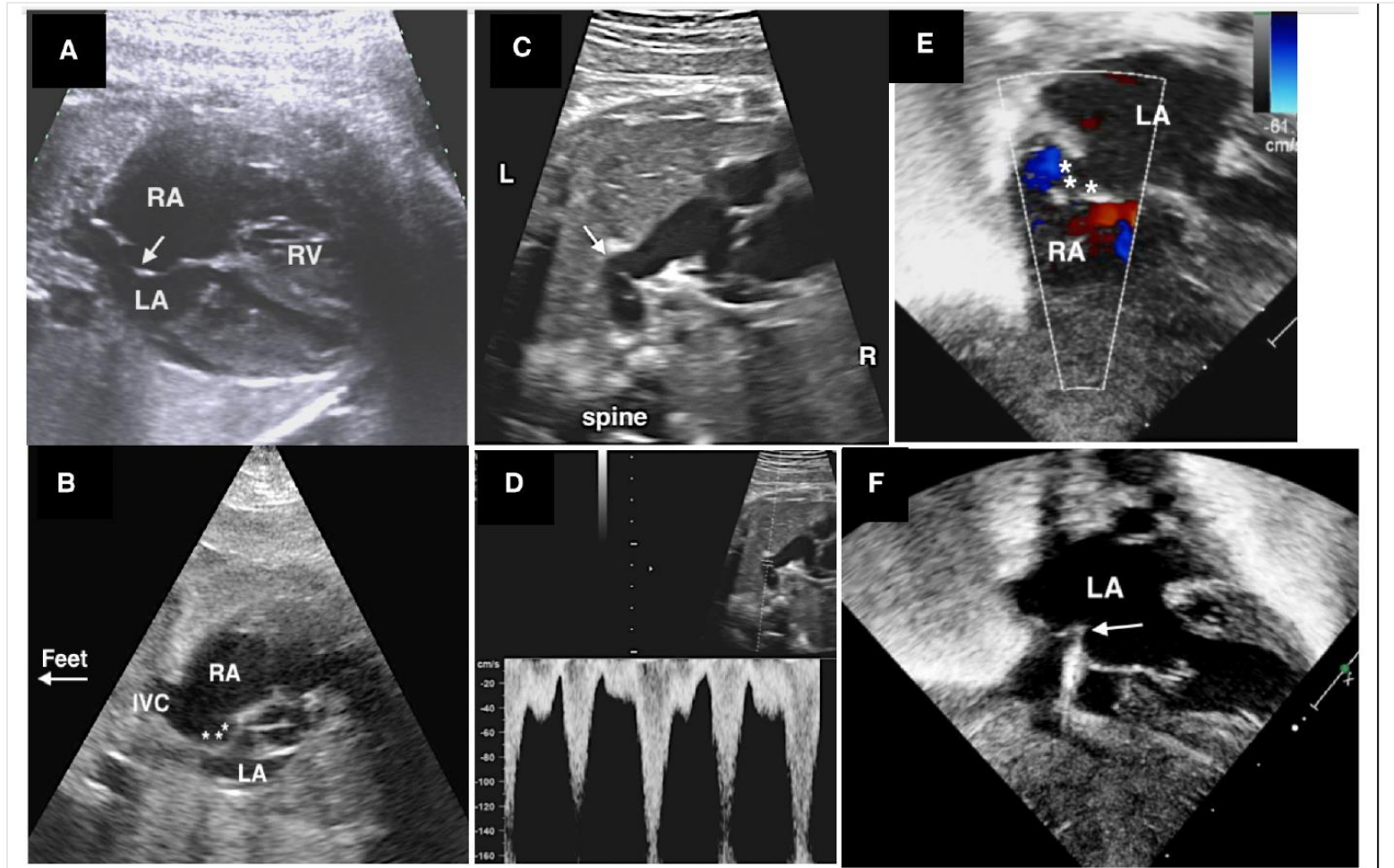


Figure 3 reproduced from Moray A, et al. J Am Heart Assoc. 2024;13:e031184 (CC BY-NC-ND 4.0).

This case

- Tethered septum with an aneurysmal flap; no atrial communication seen
- Discretely narrowed ductus with high velocities
- After birth: the septostomy catheter could only tent the septum
- Severe cyanosis and acidosis → arterial switch at <1 hour

How often is an urgent balloon septostomy needed?

Study	n	Urgent BAS / intervention
Vaujois 2017	45	27% (desaturation group)
Vigneswaran 2017	40	30% emergency BAS
Tuo 2017	40	50% urgent BAS at birth
Słodki 2018	51	56% Rashkind within 24 h
Moray 2024	61	30% at ≤2 h; 48% by 24 h

1 in 3

babies who needed intervention within 2 h had been labeled low-risk (6 of 18; Moray)

5 of 5

high-risk TGA who labored at the OB hospital needed immediate BAS; 1 arrested on arrival (ECMO)

Vaujois L, et al. Cardiol Young. 2017. Vigneswaran TV, et al. Am J Cardiol. 2017. Tuo G, et al. Fetal Diagn Ther. 2017. Słodki M, et al. UOG. 2018. Moray A, et al. JAHA. 2024 (counts from Figure 2 and text).

Same sign, opposite conclusions

Fetal sign	Supports prediction	Does not / excluded
Fixed / flat / tethered septum primum	Vigneswaran, Słodki, Moray, Donofrio	— (consensus)
Small FO (≤ 3 mm; FO:TSL < 0.5)	Vigneswaran, Vaujois (2.9 vs 4.0 mm), Moray	— (consensus)
Small or constricted ductus	Maeno, Moray, Donofrio	—
Hypermobility / redundant septum	Tuo ($p=0.002$); Donofrio (now LOC 4)	Vigneswaran (not predictive); Moray (excluded: subjective)
Retrograde diastolic ductal flow	Vaujois (92% vs 23%); Donofrio (abnormal DA $\rightarrow$ LOC 4)	Moray (excluded: nonspecific)
PV peak velocity > 41 cm/s	Słodki (specificity / PPV 100%)	Single study, not replicated

Fetal Heart Society multicenter study (n=105): preliminary analysis found no baseline feature that predicted neonatal hypoxia; the MHO arm is pending.

Maeno 1999; Tuo 2017; Vigneswaran 2017; Vaujois 2017; Słodki 2018; Donofrio 2013; Moray 2024 (see references).

Perinatal management

	Plan
Fetal therapy	None
LOC 3	Hypermobility or tethered septum, or septum primum angle $<30^\circ$
LOC 4	Bowing $>50\%$ into LA, intact septum, or restrictive FO with abnormal ductus
Where	Where BAS is available within minutes
When	Planned daytime delivery, 38–39 wk
Delivery room	PGE; BAS (bedside or cath lab)
If still hypoxemic	Think PPHN: iNO; ECMO ready

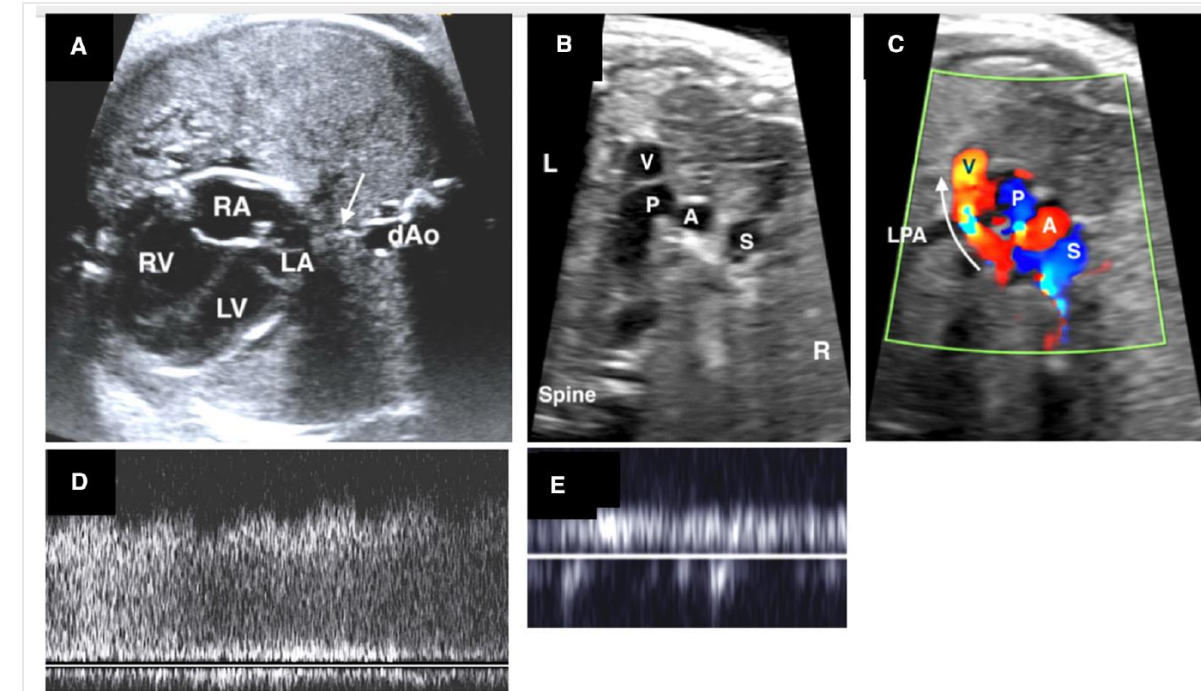
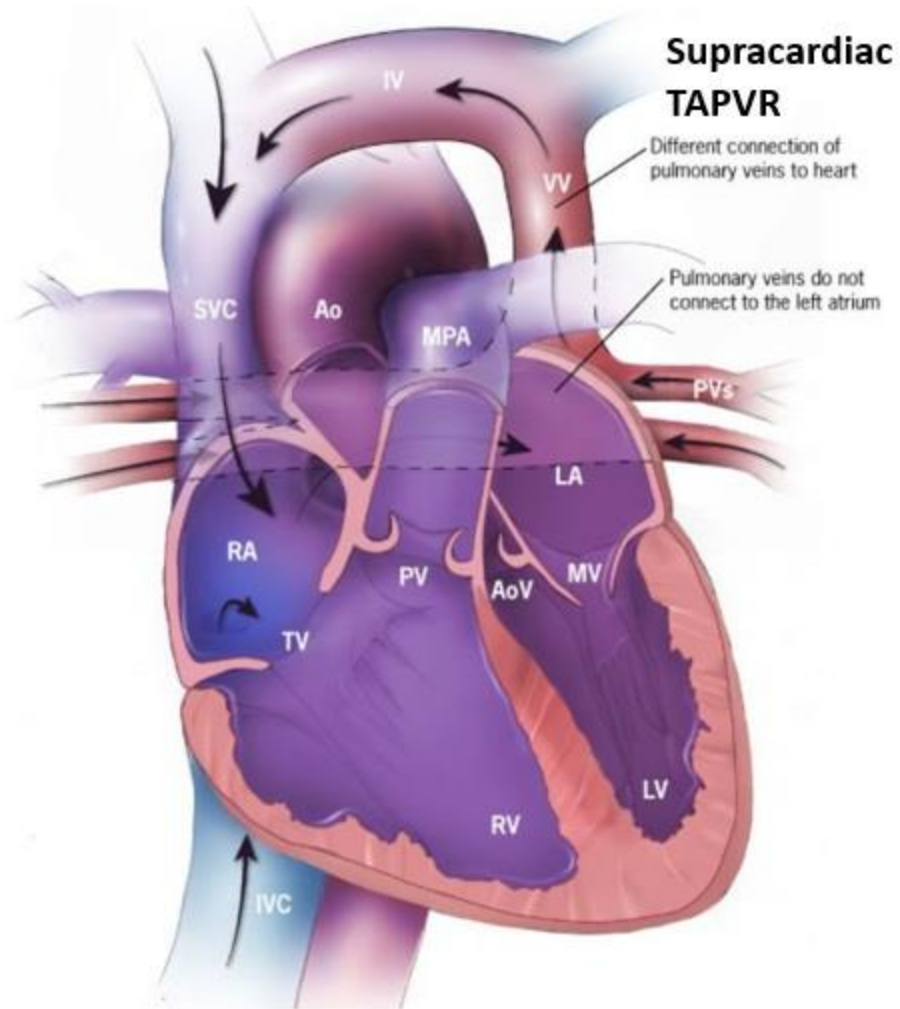
Why daytime and planned?

- A brief “honeymoon” before profound hypoxemia lets BAS happen calmly
- Persistent pulmonary hypertension occurs in up to 5% (Maeno)
- Arterial switch at a median of 5 days; 14/15 high-risk survived (Moray)

Bottom line

We don't predict BAS well — so plan every d-TGA/IVS as though it will need one in the first hour.

Fetal assessment



Supracardiac TAPVD: vertical vein obstructed over the LPA (mean gradient 5 mmHg in utero)

- RV > LV; increased LA–descending aorta distance
- Vertical vein in the 3VT view; color aliasing where it crosses the LPA
- Low-velocity, non-pulsatile pulmonary vein flow
- Echogenic lungs (pulmonary edema); nutmeg lung on MRI
- MHO: vertical vein gradient 2 → 12 mmHg predicted obstruction (case)

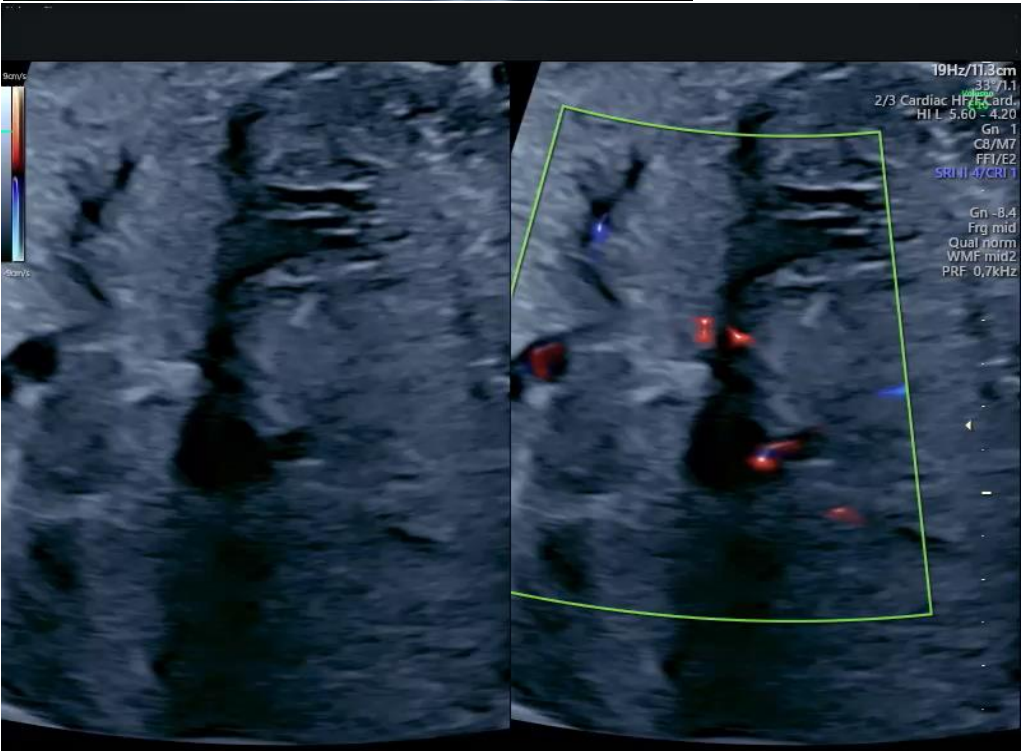
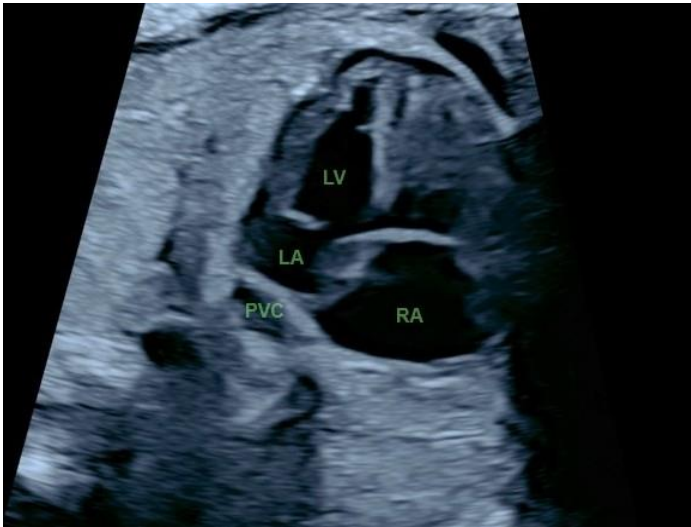
Perinatal management

	Plan
Fetal therapy	None
LOC	4: accelerated flow in the connecting vein
Delivery	With surgery and ECMO immediately available
First hour	Early imaging; emergent repair or ECMO
Caution	PGE won't relieve venous obstruction; iNO can worsen pulmonary edema

CHCO: Supracardiac obstructed TAPVD

- C-Section at 39 weeks
- Repair at 1 hour
- Transitioned to ECMO
- Mild pulmonary vein confluence gradient
- Residual pulmonary hypertension, on sildenafil

Donofrio MT, et al. Am J Cardiol. 2013 (Table 2). Moray A, et al. J Am Heart Assoc. 2024;13:e031184.



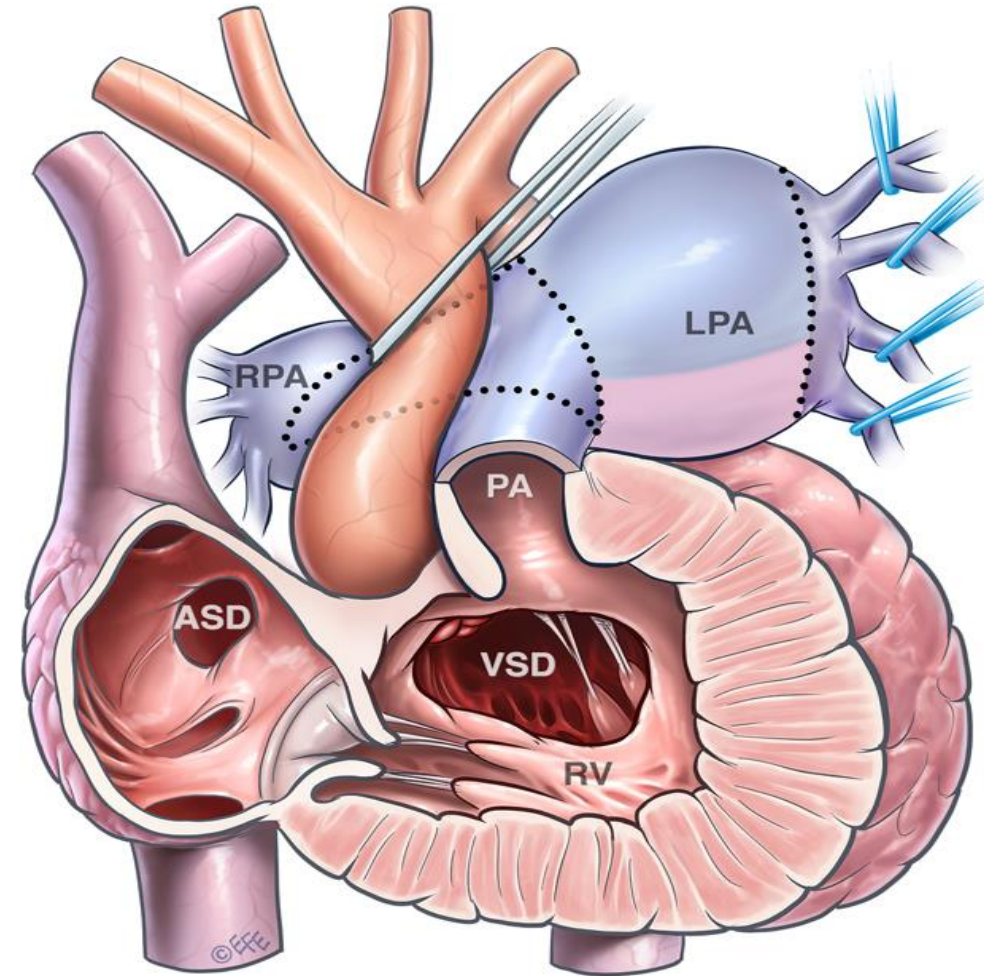
Assessment and perinatal management

Fetal assessment

- Aneurysmal branch PAs compress the bronchi
- Hyperexpanded or echogenic lung, cardiac shift, polyhydramnios
- Fetal MRI to assess the airway
- Genetics: in Moray, the only genetic diagnoses were in TOF/APV (22q11 x2, trisomy 21 x1)

Delivery room

- Intubation expected; prone positioning helps
- Specialized ventilation; ECMO ready
- Moray: 4/4 survived (intubated, CPAP, or none)
- Donofrio: planned ECMO for air trapping; surgery day 2



Chelliah A, et al. J Am Heart Assoc. 2021;10:e019713. Donofrio MT, et al. Am J Cardiol. 2013. Moray A, et al. J Am Heart Assoc. 2024;13:e031184. Konstantinov IE, Rodrigues C, Konstantinov SI, Fricke TA. Absent pulmonary valve syndrome: Valvular reconstruction with autologous pulmonary arterial wall. JTCVS Tech. 2025 May 30;32:105-107.

What predicts outcome after fetal diagnosis

19 centers, 10 years, 100 fetuses

64%

overall survival with
intent to treat
(n=78)

9%

died in utero

27%

died after birth; none
after 13 months

46%

of 80 tested had a
chromosomal
abnormality (22q11.2:
35%)

Fetal echo red flags

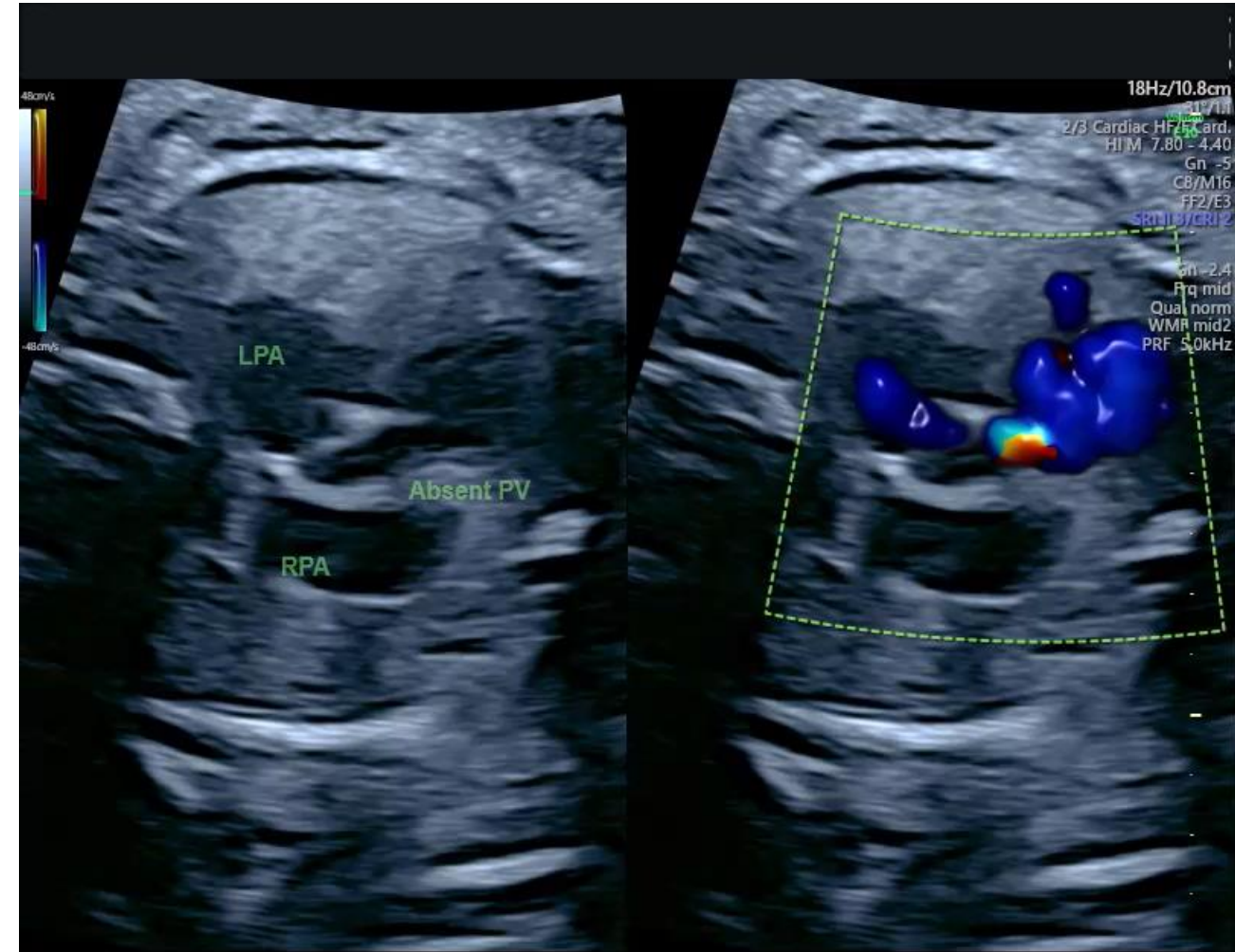
- **RV dysfunction:** overall mortality (OR 7.9)
- **LV dysfunction:** fetal demise (OR 7.4)
- **Mediastinal shift:** present in all 7 fetal deaths
- Also: RV/LV dilation, TR, Doppler abnormalities, earlier delivery

What didn't predict

Pulmonary artery size
— the most dramatic
finding on the scan

For MFM

Offer genetics; get fetal MRI
of the airway; LV dysfunction
may change delivery timing
and place



Fetal assessment

45%

perinatal mortality (multicenter)

33%

neonatal mortality

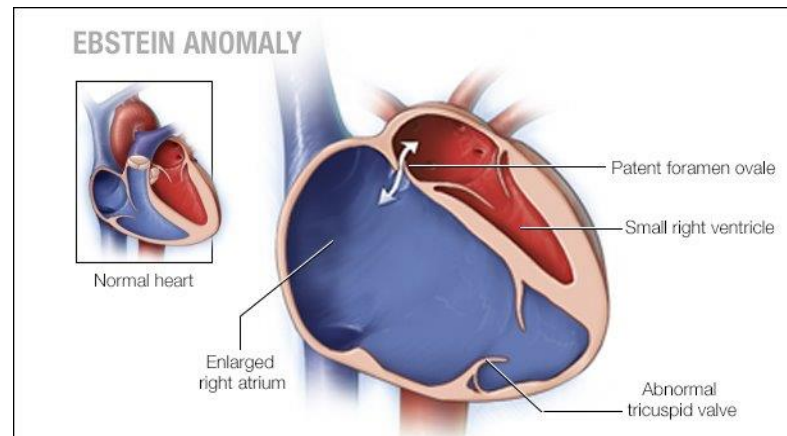
Predictors of death at diagnosis (Freud)

- Diagnosis <32 weeks
- Cardiothoracic ratio; TV annulus z-score
- No forward flow across the pulmonary valve
- Pulmonary regurgitation; retrograde ductal flow
- Pericardial effusion

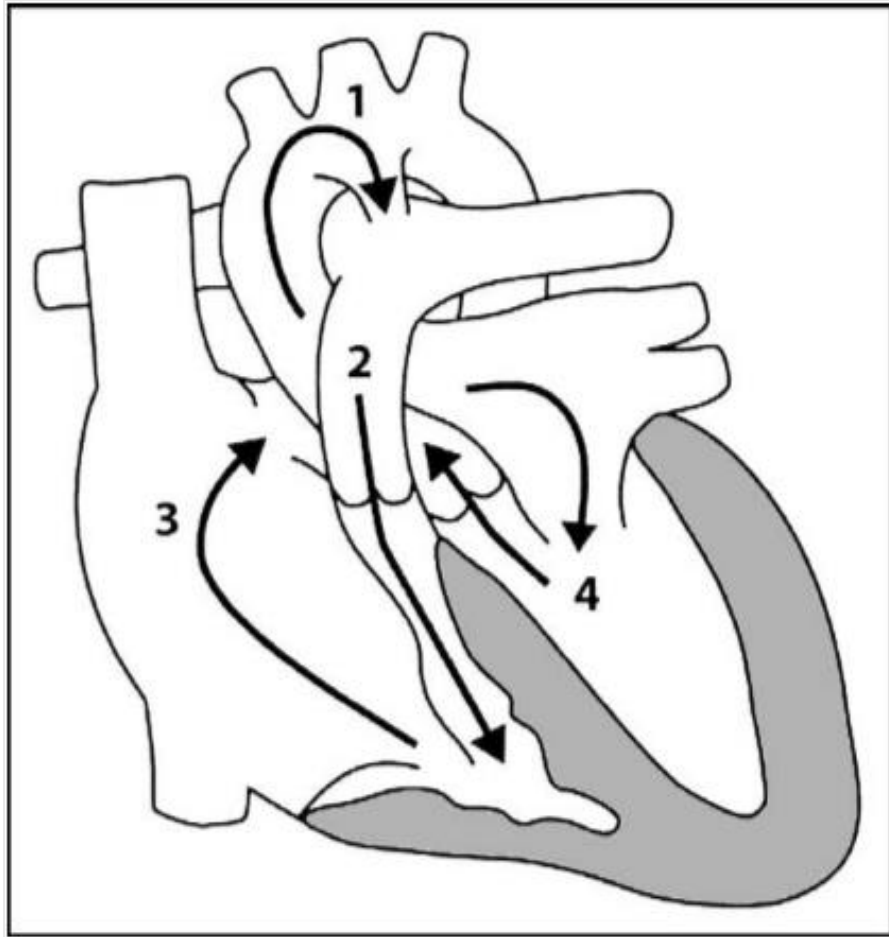
Progression markers (diagnosed <24 wk)

- No antegrade PV flow; pulmonary regurgitation
- CT area ratio >0.48; LV dysfunction; TV annulus z >5.6
- Sonographer: every visit — CT ratio, TR jet velocity (low = bad), RVOT color, ductal direction, effusions, rhythm

Freud LR, et al. Circulation. 2015;132:481–489. Can J Cardiol. 2016;32:1500–1506 [confirm authors]. Am J Cardiol. 2017;119:106–111 [confirm authors].



Circular shunt



- Needs both tricuspid and pulmonary regurgitation
- Aorta → ductus (reversed) → PA → PR into RV → TR into RA → FO → LA → LV → aorta
- Blood circles the heart instead of reaching the body and placenta
- Umbilical artery and MCA Dopplers become abnormal (a systemic “steal”)

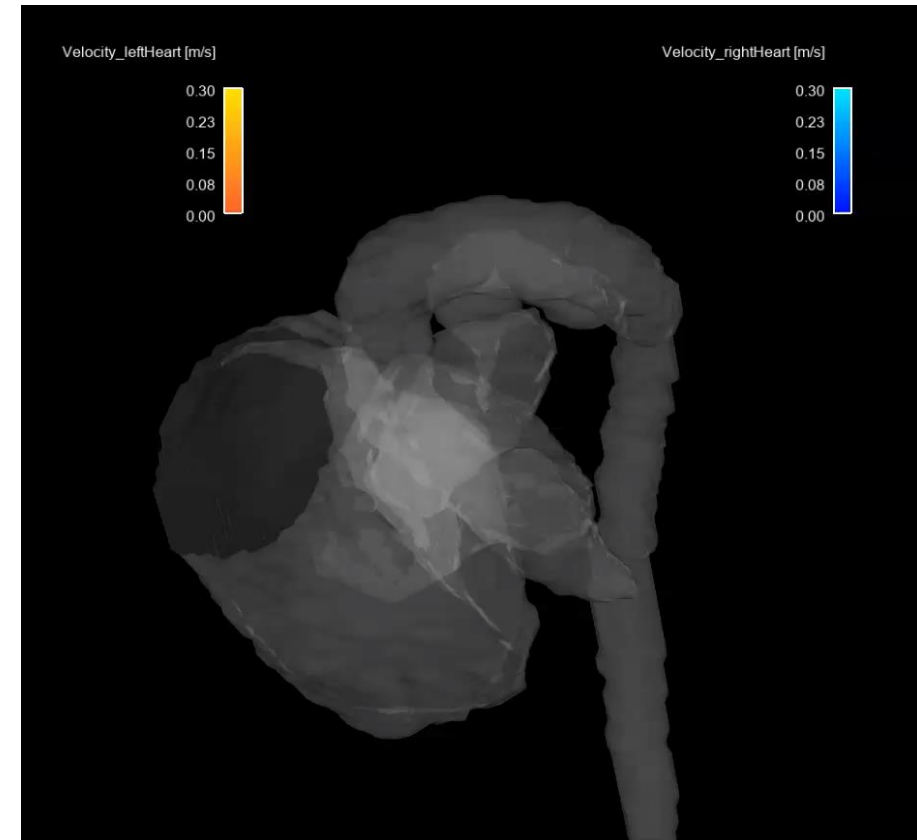


Diagram: Arunamata A, et al. Ann Pediatr Cardiol. 2017. Freud LR, et al. Circulation. 2015.

Fetal therapy

Transplacental NSAIDs

INVESTIGATIONAL

- Constrict the ductus to break a circular shunt; CFCC's preferred approach
- MFM co-management: amniotic fluid, fetal renal effects

Chronic maternal hyperoxygenation

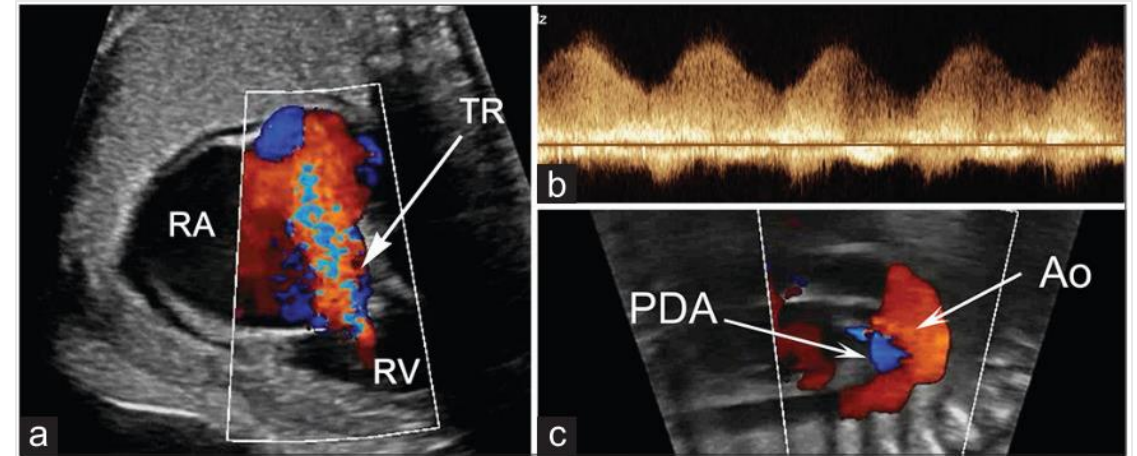
CASE REPORT ONLY

- Case report: ductal restriction after 30 weeks — feasible, not effective enough

Treat arrhythmias; counsel

- Accessory pathways → SVT; discuss comfort-care pathways for high scores

Arunamata A, et al. Chronic antepartum maternal hyperoxygenation in severe fetal Ebstein anomaly with circular shunt. Ann Pediatr Cardiol. 2017 (image).



Perinatal management

	Plan
LOC 3	Dilated heart with decreased function or significant arrhythmias
LOC 4	Hydrops → consider early delivery
Delivery room	Intubation expected (lung hypoplasia); O ₂ and iNO to lower PVR (being cognizant of the presence of PDA)
PGE	Physiology-dependent: helps functional PA atresia, worsens circular shunt

Rescue ECMO standby; ductal ligation, Starnes, later Cone

For MFM

Outcome is driven by the heart and lungs. Ebstein's is a spectrum and the severity dictates the time to response in the neonatal period. Deliver at 39 wk unless hydrops.

Donofrio MT, et al. Am J Cardiol. 2013 (Table 2). Moray A, et al. J Am Heart Assoc. 2024;13:e031184.

Picking up where Dr. Holmes left off: delivery by rhythm

Rhythm status	LOC	Timing / mode	Delivery room
SVT / flutter controlled on therapy	1–2	Term; mode by obstetric indications	Standard team; ECG after birth
Persistent tachycardia at ≥37 wk	2–3	Deliver rather than keep treating	Cardioversion ready
Uncontrolled, decreased function	3	Planned; specialist present	Adenosine, synchronized cardioversion, drugs pre-drawn
Hydrops	4	Consider early delivery	Cardioversion or drugs in the DR; ventilation
Bradycardia <3rd %ile, 2:1 block, torsades (suspect LQTS)	3–4	Planned	Magnesium, lidocaine, β-blocker, pacing ready
Complete heart block	3–4	See CHB chart	See CHB chart

Why plan at all? Labor catecholamines can worsen tachycardia, and a weak ventricle can fail when systemic resistance jumps at cord clamping. Mortality: 0% without hydrops vs 17% with hydrops (Jaeggi).

Donofrio MT, et al. Am J Cardiol. 2013 (Table 2). Jaeggi ET, et al. Circulation. 2011;124:1747–1754. [Align the 36 vs 37 wk cutoff with Dr. Holmes.]

The first 72 hours after birth

Breakthrough as drug levels fall

Controlled in utero can recur on days 1–3:
monitored bed, not a normal-newborn disposition

ECG in sinus rhythm

Look for pre-excitation (WPW); changes drug
choice and counseling

Flutter vs SVT diverge

Flutter: cardiovert (synchronized or
transesophageal pacing), often no recurrence. SVT:
maintenance therapy through infancy

Transplacental drug exposure

Amiodarone → neonatal thyroid tests;
digoxin/flecainide/sotalol → ECG intervals ± levels;
breastfeeding compatibility varies

Suspected LQTS

Postnatal QTc; avoid QT-prolonging drugs; parental
ECGs and genetics (MFM can start this)

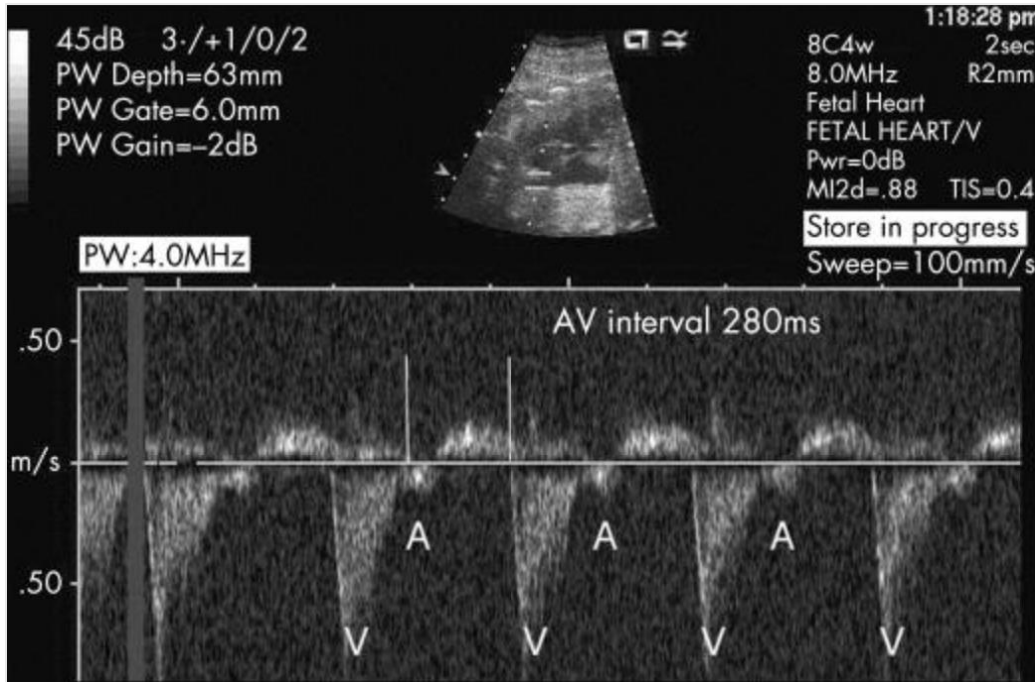
Hydropic neonate

Ventilation, drain effusions, inotropes; rhythm
control alone won't reverse hydrops quickly

Cases: Moray — SVT with hydrops at 34 wk, cardiac OR delivery, early antiarrhythmics, controlled. Donofrio — torsades needing magnesium, lidocaine, and esmolol in the DR, then pacemaker/ICD.

Moray A, et al. J Am Heart Assoc. 2024;13:e031184. Donofrio MT, et al. Am J Cardiol. 2013;111:737–747.

Fetal assessment and therapy



Prolonged mechanical AV interval (280 ms) on SVC-aorta Doppler — heart rate alone misses this.

Red flags: rate <55 bpm · EFE · dysfunction · effusion/hydrops · CV profile score <7

Dexamethasone

CONTROVERSIAL

Some use for evolving 1st/2nd-degree block; does not reverse 3rd-degree
 Recurrence prevention in anti-Ro+ mothers with a prior affected child

IVIG

LIMITED EVIDENCE/ ENCOURAGING

For myocarditis or EFE

β-agonists

LIMITED EVIDENCE

Terbutaline or salbutamol to raise the rate (usually <55 bpm); modest effect

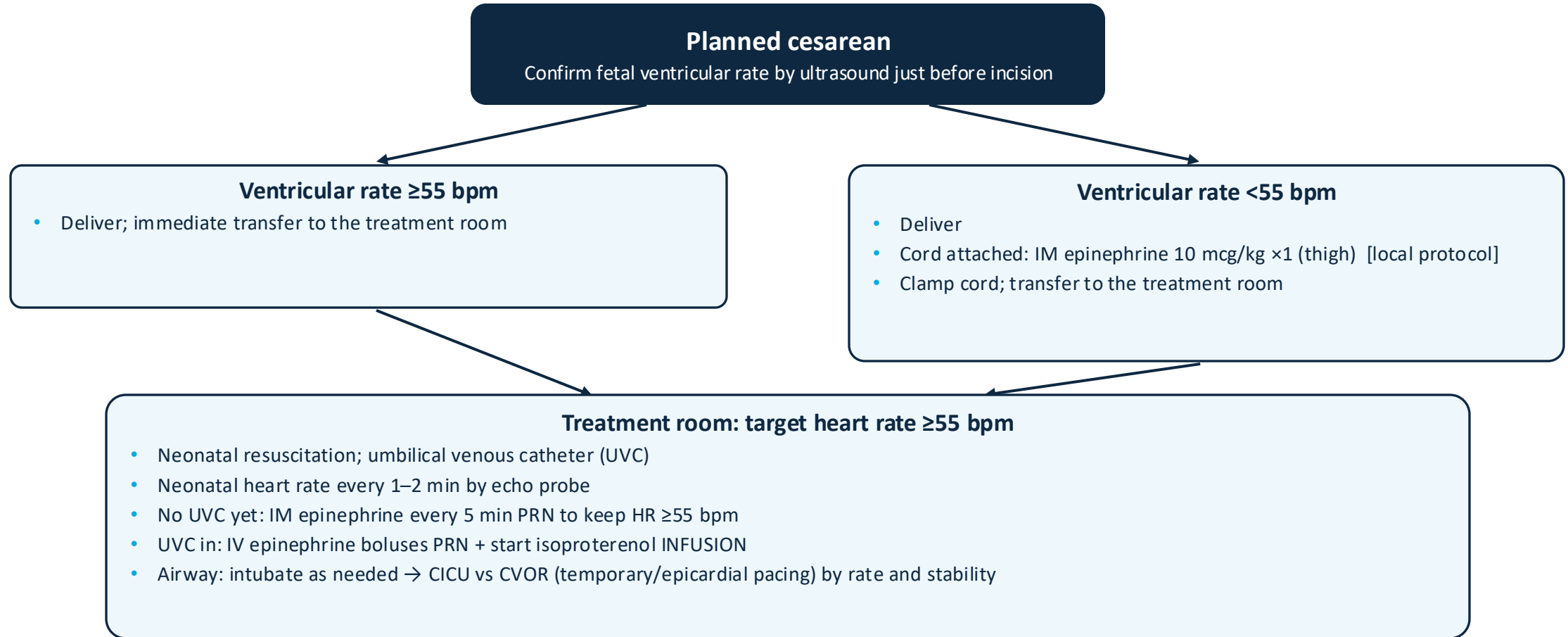
Hydroxychloroquine

EMERGING

Recurrence prevention in anti-Ro+ mothers with a prior affected child

Anti-Ro exposure: 1–5% develop complications (CHB, EFE, cardiomyopathy). Donofrio MT, et al. Am J Cardiol. 2013 (Table 2). Donofrio MT, et al. Circulation. 2014;129:2183–2242.

Delivery room algorithm (example protocol)



Team: MFM/OB, OB anesthesia, EP, NICU, CICU, echo, $\pm$ CT surgery and cardiac anesthesia. **Doses and routes:** verify with NICU pharmacy and EP.

Example institutional protocol — adapt locally. Threshold consistent with Donofrio MT, et al. Am J Cardiol. 2013 (Table 2: ventricular rate < 55 with decreased function).

Perinatal management

	Plan
LOC	3: rate <55 with decreased function, CVP score <7; 4: hydrops
Timing	[CFCC planned gestational age] — consider early delivery with hydrops
Mode	Cesarean: fetal heart rate monitoring can't assess well-being in CHB
Delivery room	Chronotropes vs pacing per algorithm
After birth	Neonatal lupus labs; IVIG/steroids if myocarditis or EFE; pacemaker

Case (Moray)

- Rate in the low 50s, EFE, early hydrops
- Cardiac OR delivery at 37 weeks
- HR 50s with acidosis after birth
- Pacemaker at ~10 h; IVIG and steroids
- Alive and stable

Donofrio MT, et al. Am J Cardiol. 2013 (Table 2). Moray A, et al. J Am Heart Assoc. 2024;13:e031184 (Table).

What you can do

Putting it together for MFM, OB, and sonographers

04

The playbook on one page

Lesion	Fetal red flag	LOC	First-hour plan
HLHS, restrictive septum	PV f/r <3; FO <3 mm; no MHO response	4	Cath lab: septostomy/stent; ECMO ready
d-TGA/IVS	Fixed/small FO; abnormal ductus — but prediction is poor	3–4	PGE; BAS within minutes; iNO/ECMO
Obstructed TAPVR	Low-velocity non-pulsatile PV flow; VV gradient	4	Emergent repair or ECMO
TOF / absent PV	Airway compression; hydrops; CLE on MRI	3–4	Intubate, prone; ECMO ready
Ebstein / TVD	CT ratio, no PV forward flow, PR, circular shunt	3–4	Lower PVR; iNO; ECMO; Starnes
Tachyarrhythmia	Uncontrolled; decreased function; hydrops	3–4	Cardioversion or drugs in the DR
Complete heart block	Rate <55; EFE; dysfunction; hydrops	3–4	Chronotropes, then pacing
Critical AS	LV dysfunction/EFE; restrictive FO; hydrops	3–4	PGE; balloon valvuloplasty ± septostomy
Right-sided obstruction	Reversed ductal flow; sinusoids; restrictive FO	2–4	PGE; cath or surgery
Coarctation / arch	Disproportion; hypoplastic arch	2	PGE vs observe; watch ductal closure

Adapted from Donofrio MT, et al. Am J Cardiol. 2013 (Table 2) and Moray A, et al. J Am Heart Assoc. 2024.

Concrete actions for MFM, OB, and sonographers

1

Refer early, rescan late

Get the outflows; ≥ 2 fetal echos after 35 weeks for LOC 3–4 lesions

2

Timing

Plan 38–39 weeks (both published series) unless hydrops or obstetric indication

3

Mode

Obstetric indications; planned cesarean only where the LOC 4 plan requires it (and for CHB)

4

Genetics

Offer testing before an aggressive LOC 4 plan

5

Labor plan

If a LOC 3–4 patient presents in labor: call cardiology immediately

6

Counsel together

Joint visits, including comfort-care options for the highest-risk lesions

Moray: no major maternal complications after cardiac-OR cesarean (median maternal stay 2 days; 85% primary cesarean). Donofrio 2013; Moray 2024.

Born into a plan

- 1 A fetal diagnosis only helps if the first hour is planned: LOC, team, algorithm, simulation, debrief.
- 2 HLHS with restrictive septum is predictable; d-TGA is not — plan every d-TGA as if it needs a BAS.
- 3 Findings worsen late: the delivery plan is set after 35 weeks, not at 24.
- 4 Maternal hyperoxygenation previews the first breath — acute testing only.
- 5 Planning matters more than the building: a specialist in the room and a rapid path to intervention.

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