

FETAL CARDIOLOGY · MATERNAL-FETAL MEDICINE

Fetal Heart Views

Patterns that Should Trigger Referral

Samantha Holmes, MD

Department of Pediatrics, Division of Cardiology
University of Colorado
Children's Hospital Colorado | Colorado Fetal Care Center



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Disclosures



No disclosures.

I have no relevant financial relationships or conflicts of interest to disclose.



TODAY

Learning Objectives

1

When to refer

Identify fetal cardiac findings that warrant referral to fetal cardiology.

2

What it looks like

Recognize key abnormal patterns on the four-chamber, outflow tract, and three-vessel trachea views, and abnormal fetal rhythms.

3

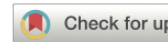
What we can do

Describe the fetal therapies and delivery planning that a prenatal diagnosis makes possible.



Indications for Fetal Echo

Guidelines and Recommendations for Performance of the Fetal Echocardiogram: An Update from the American Society of Echocardiography



Anita J. Moon-Grady, MD (Co-Chair), Mary T. Donofrio, MD (Co-Chair), Sarah Gelehrter, MD, Lisa Hornberger, MD, Joe Kreeger, ACS, RCCS, RDCS, Wesley Lee, MD, Erik Michelfelder, MD, Shaine A. Morris, MD, Shabnam Peyvandi, MD, Nelangi M. Pinto, MD, Jay Pruetz, MD, Neeta Sethi, MD, John Simpson, MD, Shubhika Srivastava, MD, and Zhiyun Tian, MD, *San Francisco and Los Angeles, California; Washington, District of Columbia; Ann Arbor, Michigan; Edmonton, Alberta, Canada; Atlanta, Georgia; Houston, Texas; Salt Lake City, Utah; Durham, North Carolina; London, United Kingdom; Wilmington, Delaware; and Philadelphia, Pennsylvania*

Always refer

Any suspected structural abnormality, abnormal heart rate or rhythm, or hydrops on screening ultrasound.

Moon-Grady AJ, et al. *J Am Soc Echocardiogr.* 2023;36(7):679-723.



WHEN TO REFER

Potential Indications for Fetal Echo

Table 1 Potential indications for fetal echocardiography

	ASE 2023 recommendation	AIUM 2020 ⁴	AHA 2014 ^{2*}
Maternal factors (absolute risk)[†]			
Pre-gestational diabetes (3%-5%)	Is indicated	Is indicated	I (indicated)
Gestational diabetes diagnosed after second trimester (<1%)	Not indicated	Not indicated	III (no benefit)
Phenylketonuria (12%-14%)	Is indicated	Is indicated	I (indicated)
Autoimmune disease: SSA/SSB positive (1%-5%) [‡]	Is indicated	Is indicated	IIa (probably indicated)
In vitro fertilization (1.1%-3.3%)	May be considered [§]	Is indicated	IIa (Probably indicated)
Maternal infection: rubella (3%-4%)	Is indicated	Is indicated	I (indicated)
Family history of CHD: first-degree relative (3%-20%) [¶]	Is indicated	Is indicated	I (indicated)
Family history of CHD: second-degree or more distant relative (<2%)	Not indicated	May be indicated	IIb (may be indicated)
Obesity (BMI > 30 kg/m ²) (1-2%)	Not indicated	Not indicated	—
Retinoids (8%-20%)	Is indicated	Is indicated	I (indicated)
ACE inhibitors (3%)	May be considered [§]	May be indicated	IIa (probably indicated)
Paroxetine (3%)	May be considered [§]	May be indicated	IIb (may be indicated)
Other selective serotonin reuptake inhibitors (1%-2%) ^{6,7}	Not indicated	Not indicated	III (no benefit)
Anticonvulsants (1%-2%)	Not indicated	May be indicated	IIb (may be indicated)
Lithium (1%-2%)	Not indicated	May be indicated	IIb (may be indicated)
Warfarin (<1%) ⁸	Not indicated	Not indicated	III (no benefit)
Fetal factors identified during screening (absolute risk)			
Fetal hydrops (15%-20%) ⁹	Is indicated	Is indicated	I (indicated)
Extracardiac anomaly (20%-45%) ^{10,11}	Is indicated	Is indicated	I (indicated)
Chromosomal abnormalities (10%-90%)	Is indicated	Is indicated	I (indicated)
Monochorionic twinning (2%-10%)	Is indicated	Is indicated	I (indicated)
Nuchal translucency 3.0-3.4 mm (~3%)	May be considered [§]	May be indicated	IIa (probably indicated)
Nuchal translucency ≥3.5 mm (6%-60%)	Is indicated	Is indicated	I (indicated)
Single umbilical artery in isolation (1.2%-1.8%) ¹²	Not indicated	Not indicated	IIb (may be indicated)

Moon-Grady AJ, et al. J Am Soc Echocardiogr. 2023.



When to Refer to Fetal Cardiology

1

Risk of fetal morbidity or demise

Findings raise the risk of fetal morbidity or demise, and in-utero intervention may be possible.

EXAMPLES

- Sustained tachycardia or bradycardia
- Hydrops
- Ebstein anomaly with circular shunt
- Obstructive cardiac mass

2

Complex CHD

Complex congenital heart disease (CHD) that may require neonatal ICU management or surgical intervention.

EXAMPLES

- Tetralogy of Fallot, truncus arteriosus
- AVSD
- Suspected coarctation
- Duct-dependent lesions needing PGE1

3

Critical CHD

Critical CHD that will require emergency intervention immediately at birth.

EXAMPLES

- TGA with restrictive atrial septum
- HLHS with restrictive or intact atrial septum
- Obstructed TAPVR
- Complete heart block with low ventricular rate



The Screening Sweep: What Each View Can Catch

1

Abdominal situs

- Stomach & heart on the left
- Aorta / IVC relationship
- → Heterotaxy, situs inversus

2

Four-chamber

- HLHS, HRHS / tricuspid atresia
- AVSD, large VSD
- Ebstein anomaly
- Masses, cardiomegaly, effusion

3

Outflow tracts (LVOT & RVOT)

- Great arteries must cross → TGA (parallel vessels)
- Tetralogy of Fallot
- Truncus arteriosus, DORV
- VSD
- Aortic or pulmonary stenosis / atresia

4

3-vessel / 3VT

- PA vs aorta size
- Coarctation, arch anomalies
- Extra vessel: L-SVC

A normal four-chamber view does not exclude CHD. TGA, tetralogy, truncus, coarctation and TAPVR can all look normal at the four-chamber level — sweep through the outflows. Add M-mode/Doppler whenever the rate or rhythm is abnormal.



FOUR-CHAMBER VIEW

Four-Chamber View

LOOK FOR

- Chamber symmetry and size
- Atrioventricular valve offsetting
- Cardiac axis and position
- Ventricular contractility



FOUR-CHAMBER VIEW

What do you see?



FOUR-CHAMBER VIEW

Hypoplastic Left Heart Syndrome

WHAT YOU'LL SEE

Small, non-apex-forming LV; diminutive or absent mitral/aortic valves; tiny ascending aorta.

WHY IT MATTERS

Systemic flow is duct-dependent — prenatal diagnosis allows PGE1 at birth and delivery at a surgical center.

RED FLAG

A restrictive or intact atrial septum makes this critical CHD needing emergency intervention at birth.

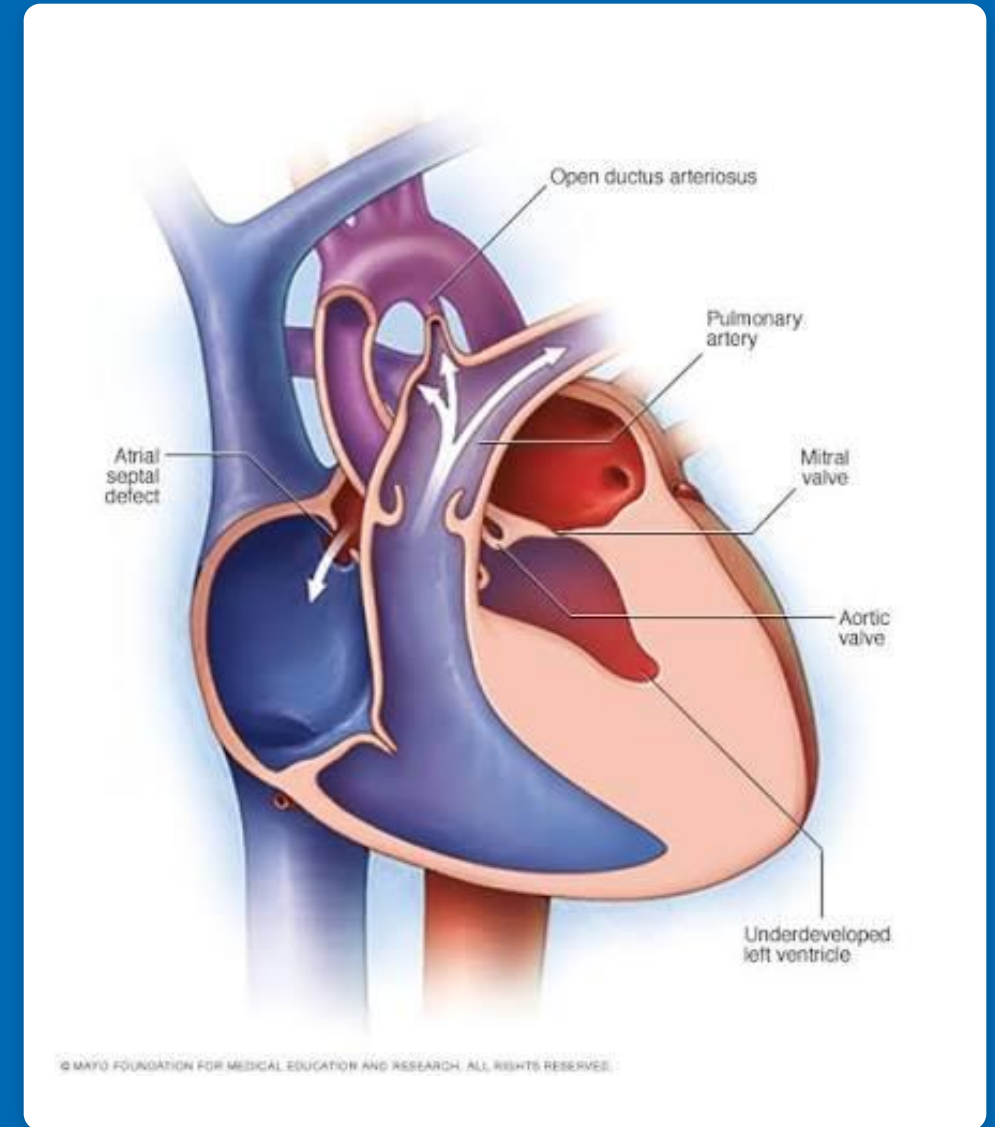
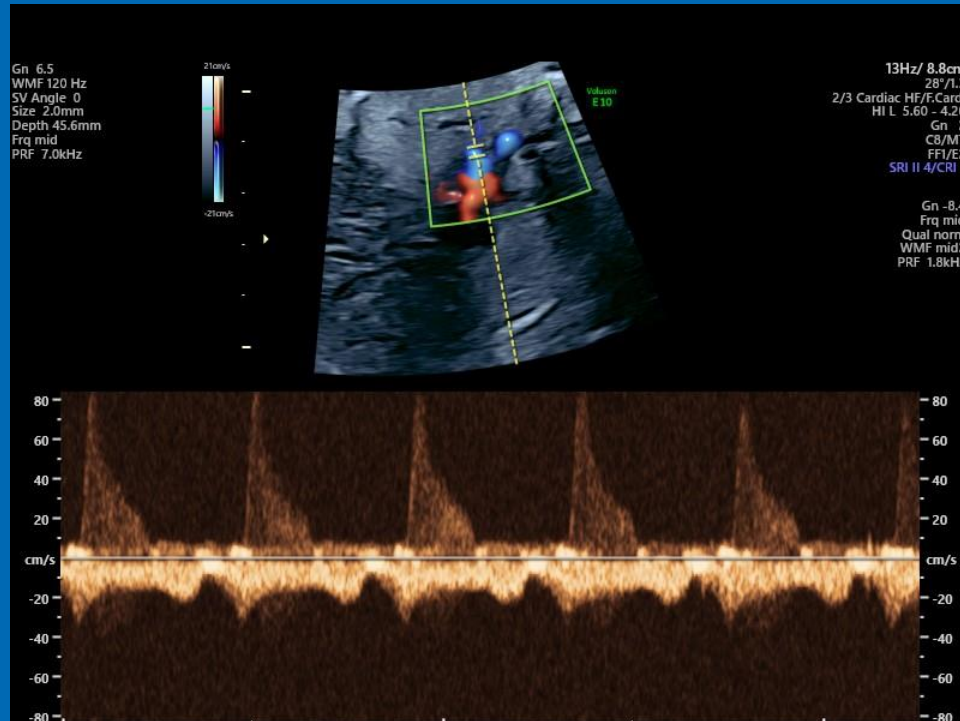


Illustration: Mayo Clinic

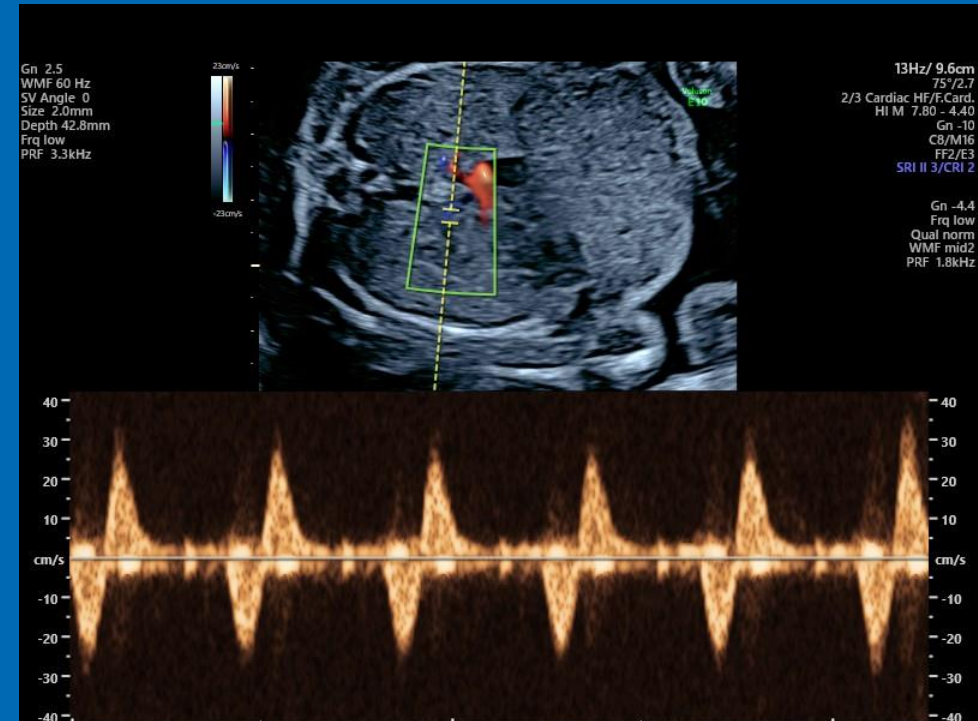


FOUR-CHAMBER VIEW

HLHS: Pulmonary Vein Doppler



Normal: predominantly forward flow; small reversal with atrial contraction



Abnormal: prominent to-and-fro reversal, suggesting a restrictive atrial septum

Why it matters: a restrictive atrial septum in HLHS means emergent atrial septal intervention at birth — it changes the delivery plan.



FOUR-CHAMBER VIEW

What do you see?



FOUR-CHAMBER VIEW

Tricuspid Atresia

WHAT YOU'LL SEE

Small right ventricle; diminutive or absent tricuspid valve (and often pulmonary valve).

KNOW THE GREAT ARTERIES

Great artery relationship determines whether pulmonary or systemic flow is at risk.

WHY IT MATTERS

If pulmonary (or systemic) flow is inadequate, the baby is duct-dependent — PGE1 at birth and delivery at a surgical center.

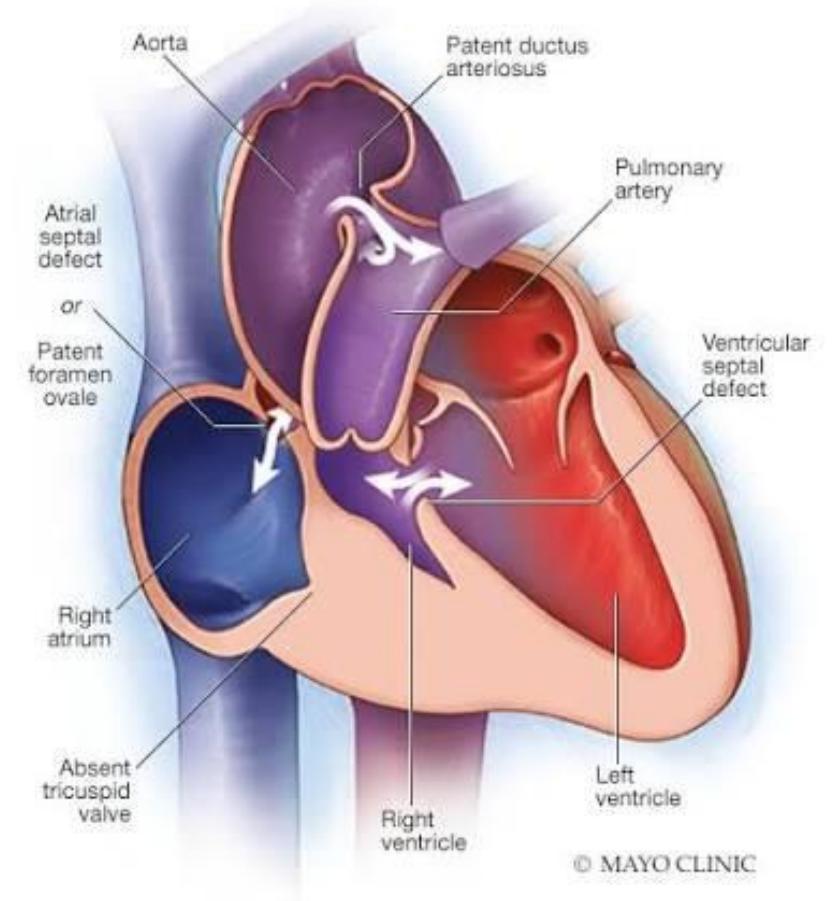
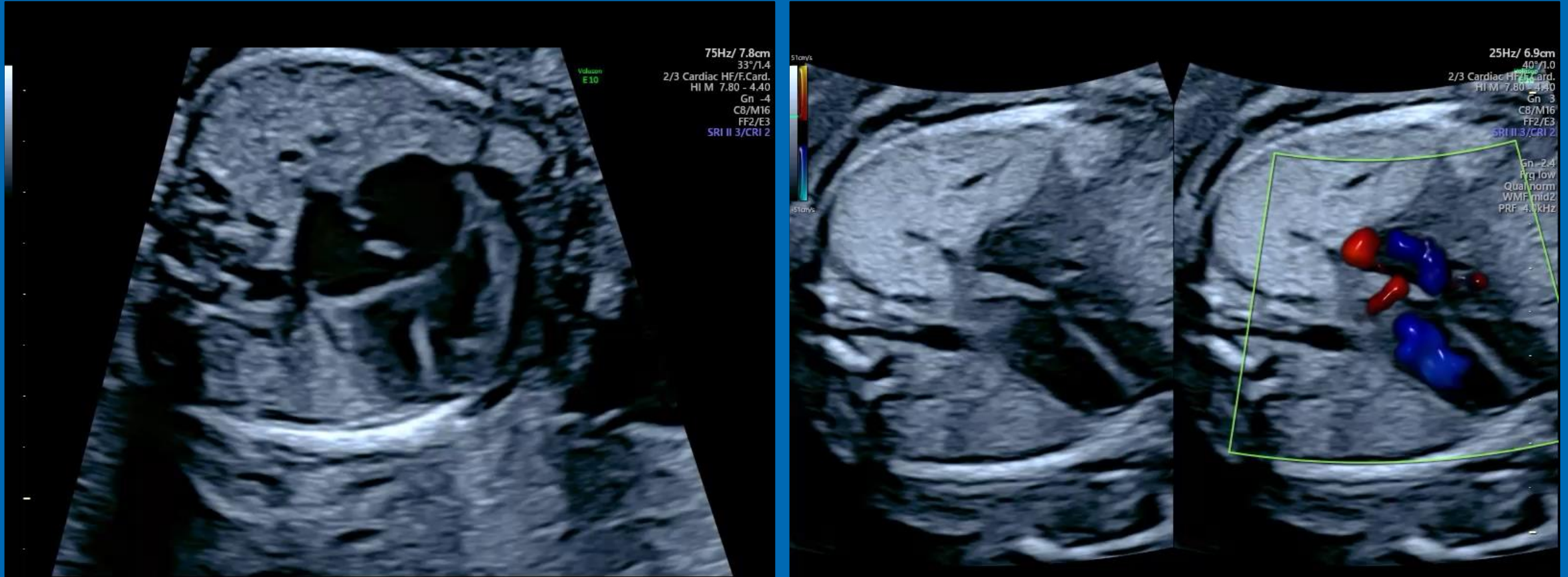


Illustration: Mayo Clinic



Septal Defects



Most VSDs are benign — but size, location, and associated lesions guide follow-up. AVSD: offer genetic testing (trisomy 21).



FOUR-CHAMBER VIEW

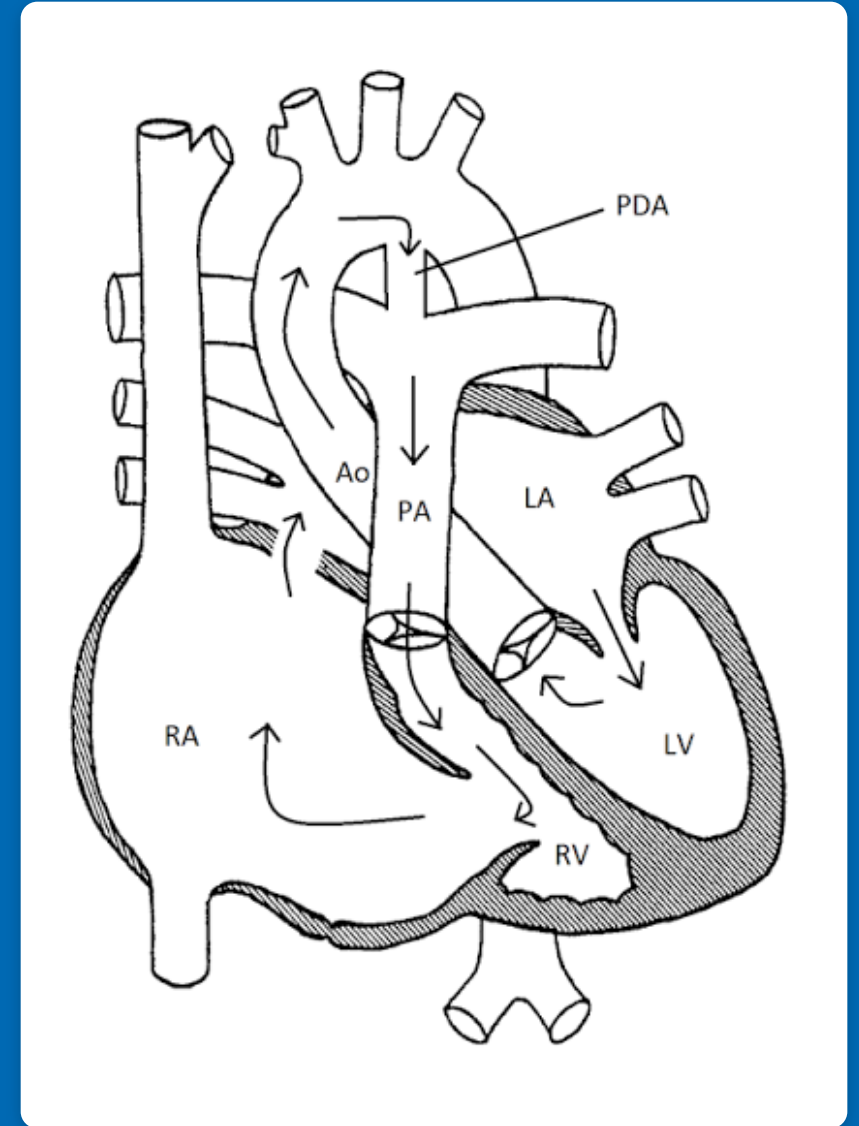
What do you see?



Ebstein Anomaly: The Circular Shunt

- 1 Aorta
- 2 Ductus arteriosus (reversed flow)
- 3 Main pulmonary artery
- 4 Pulmonary regurgitation → RV
- 5 Tricuspid regurgitation → RA
- 6 Foramen ovale → LA → LV → aorta

Result: blood loops through the heart instead of reaching the body and placenta — low systemic output, hydrops, and risk of fetal demise.



Breaking the Circular Shunt: Maternal NSAIDs



Recognize

- Severe TR with cardiomegaly
- Pulmonary regurgitation
- Retrograde flow in the ductus arteriosus
- Hydrops or abnormal UA/DV Doppler



Treat

- Maternal ibuprofen or indomethacin
- Goal: constrict the ductus arteriosus
- Stops retrograde ductal flow and breaks the loop



Monitor

- Serial fetal echo: ductal flow, PR/TR, ventricular function
- Maternal tolerance
- Deliver at a cardiac surgical center



Watch the rhythm

- Accessory pathways (WPW) are common in Ebstein anomaly
- Enlarged RA adds risk of SVT and atrial flutter
- Tachycardia further strains a fragile circulation

82%

achieved ductal constriction with NSAIDs

6% vs 50%

fetal demise with vs. without constriction

50%

of hydropic fetuses had resolution of hydrops

Peña FL, et al. J Neonatal Perinatal Med. 2024;17(1):63-69 (review of published cases). Walsh EP. JACC Clin Electrophysiol. 2018;4(10):1271-1288.



OUTFLOW TRACTS

Is this four-chamber view normal?



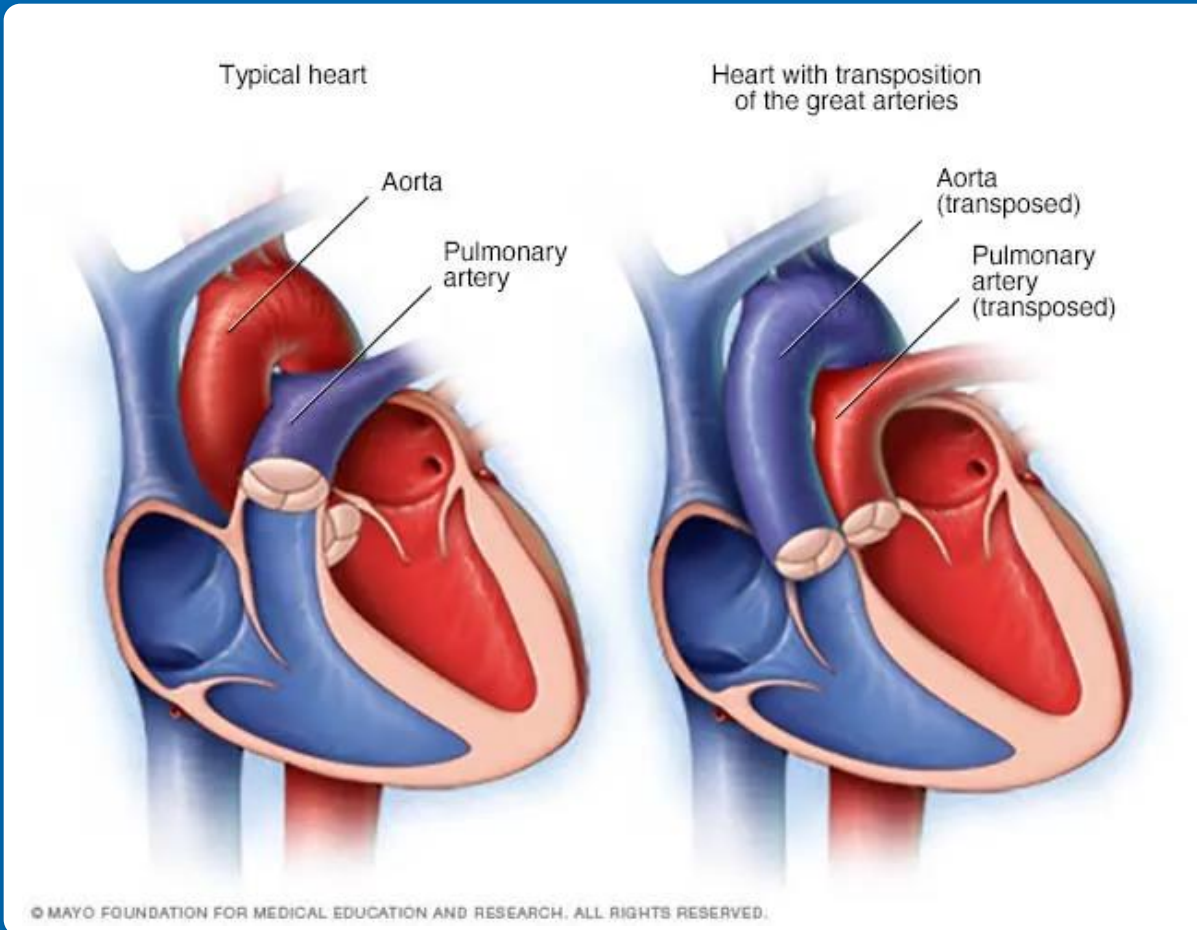
OUTFLOW TRACTS

Now sweep up to the outflow tracts. What do you see?



OUTFLOW TRACTS

Transposition of the Great Arteries



Illustrations: Mayo Clinic

WHAT YOU'LL SEE

Great arteries run parallel instead of crossing; the four-chamber view often looks normal.

WHY IT MATTERS

Circulations don't mix without a shunt. A restrictive atrial septum means emergency septostomy at birth; planned delivery avoids severe hypoxemia.

CAN WE PREDICT IT?

Not reliably. No fetal echo measure has been shown to consistently predict a restrictive atrial septum or the need for urgent septostomy — so every TGA should deliver where septostomy is immediately available.

Gottschalk I, et al. Arch Gynecol Obstet. 2024;309:1353-1367.



How Many Outflow Tracts Do You See?



One Outflow Tract: Truncus vs. Tetralogy with Pulmonary Atresia

Truncus arteriosus

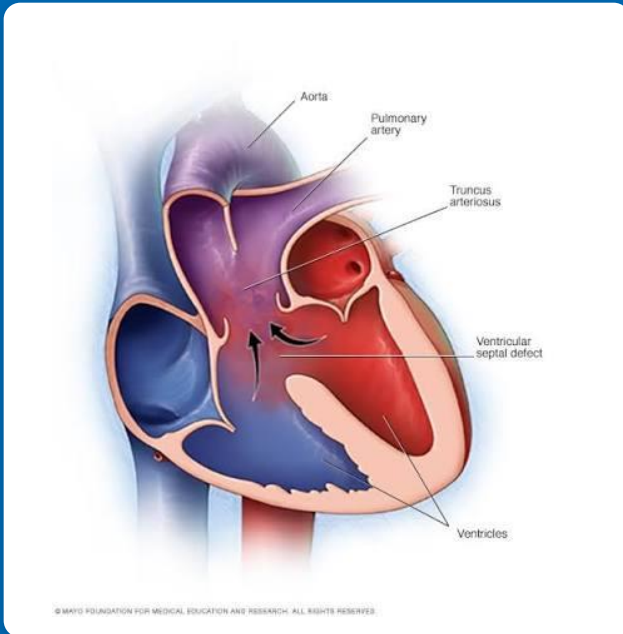


Illustration: Mayo Clinic

- A single great vessel arises over a VSD
- Pulmonary arteries arise from the truncus
- Usually not duct-dependent

Tetralogy with pulmonary atresia

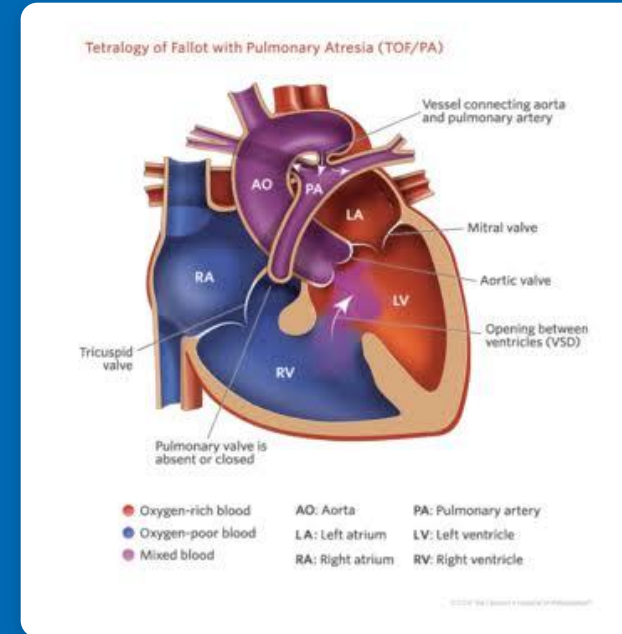
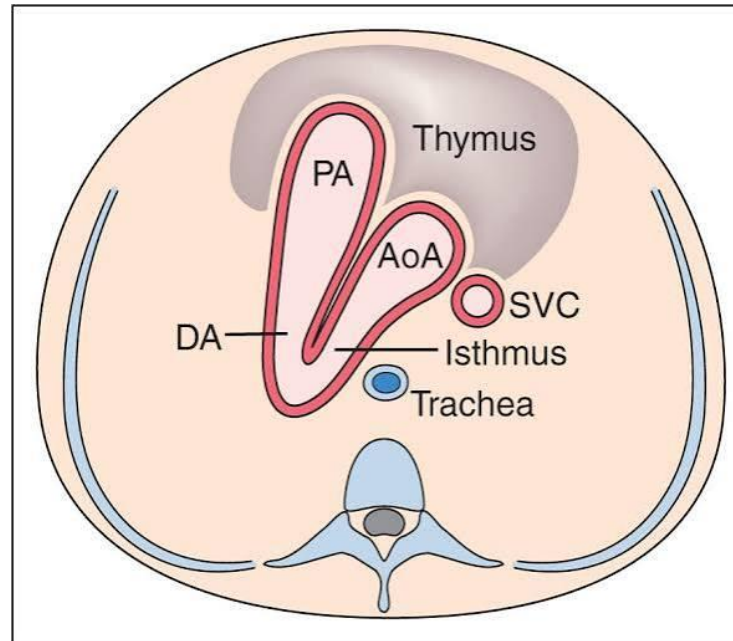


Illustration: Children's Hospital of Philadelphia

- Aorta overrides a VSD
- No antegrade flow from RV to PA
- Pulmonary flow is duct-dependent (or via collaterals)

Both: linked to 22q11.2 deletion — offer genetic counseling and testing; both need intervention or surgery early in life.

Three-Vessel Trachea View



NORMAL

- PA slightly larger than the aorta
- Both arches left of the trachea, forming a “V”
- SVC to the right

REFER IF

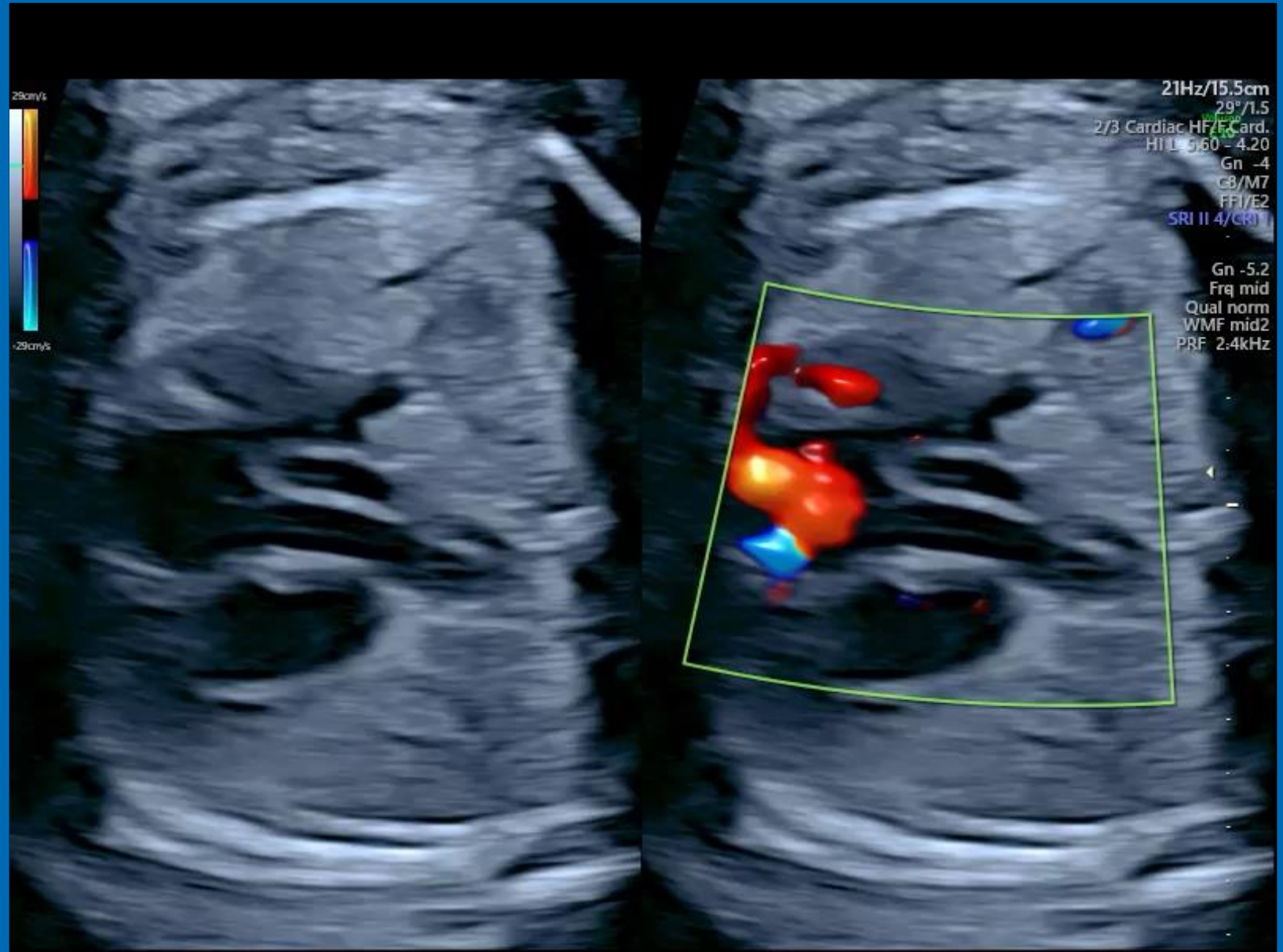
- Size discrepancy (small aorta or small PA)
- Arch on the wrong side of the trachea
- An extra vessel

THREE-VESSEL VIEW

Aortic Stenosis

LOOK FOR

- Thickened, doming aortic valve with turbulent, high-velocity flow
- Small ascending aorta relative to the PA
- Retrograde flow in the transverse arch = LV can no longer support systemic output

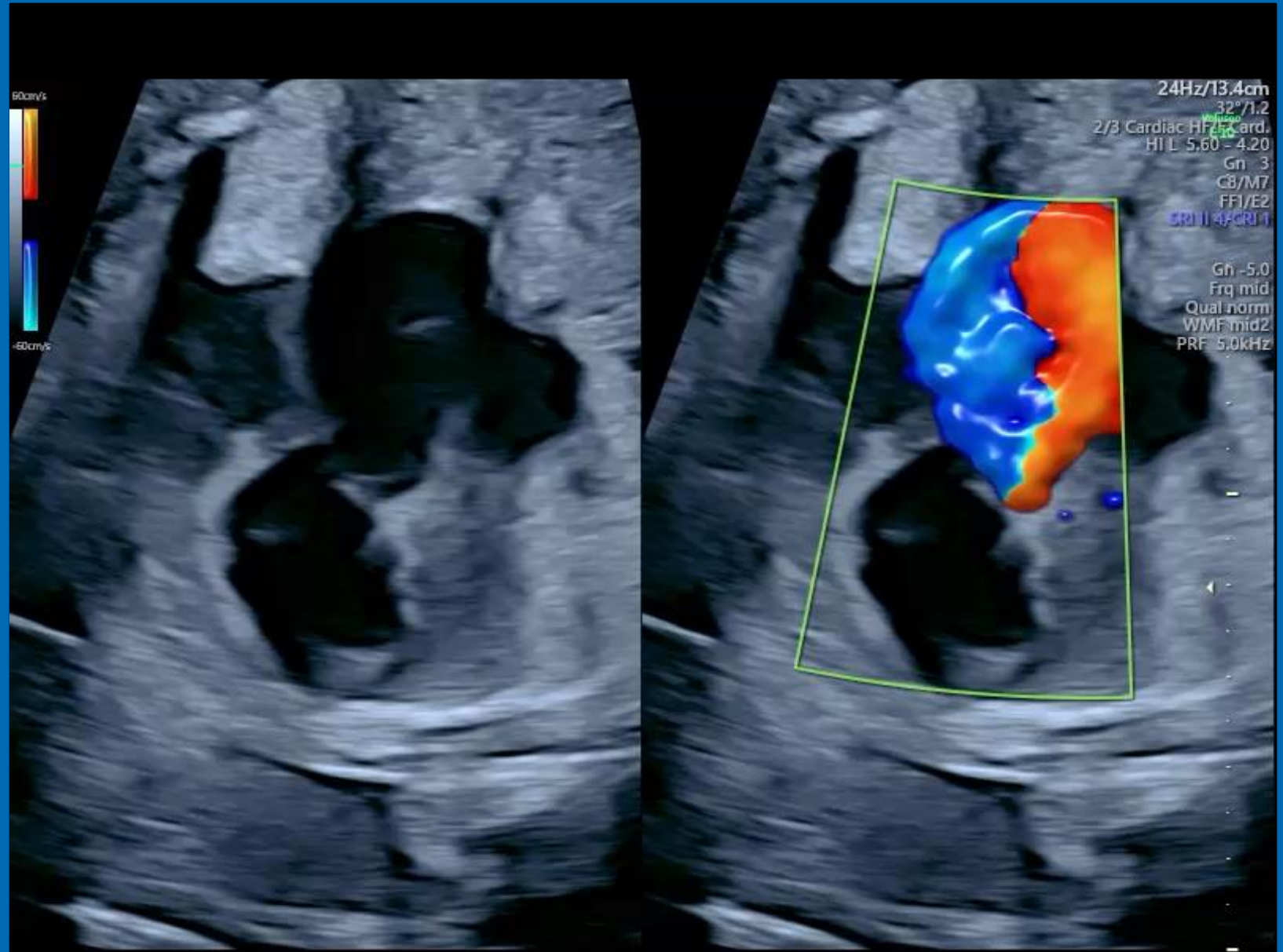


FOUR-CHAMBER VIEW

Aortic Stenosis with Severe MR

LOOK FOR

- Severe mitral regurgitation from a pressure-loaded LV
- Dilated left atrium
- LV dysfunction; echobright endocardium (EFE)
- Risk of hydrops — refer promptly



Critical Aortic Stenosis: Fetal Aortic Valvuloplasty

Signs of evolving HLHS

- Retrograde flow in the transverse arch
- Left-to-right flow across the foramen ovale
- Monophasic mitral inflow
- LV dysfunction $\pm$ EFE

The procedure

- Ultrasound-guided needle through the maternal abdomen into the fetal LV
- Balloon dilation of the aortic valve
- Mid-gestation

The goal

- Relieve LV outflow obstruction
- Preserve LV growth and function
- Keep a biventricular circulation possible

100

fetuses attempted (Boston)

77%

technically successful

38

achieved a biventricular circulation

11%

fetal loss after the procedure

Freud LR, et al. Circulation. 2014;130(8):638-645. Mäkilä K, et al. Circulation. 2006;113(11):1401-1405. Refer early — the window for intervention is narrow.

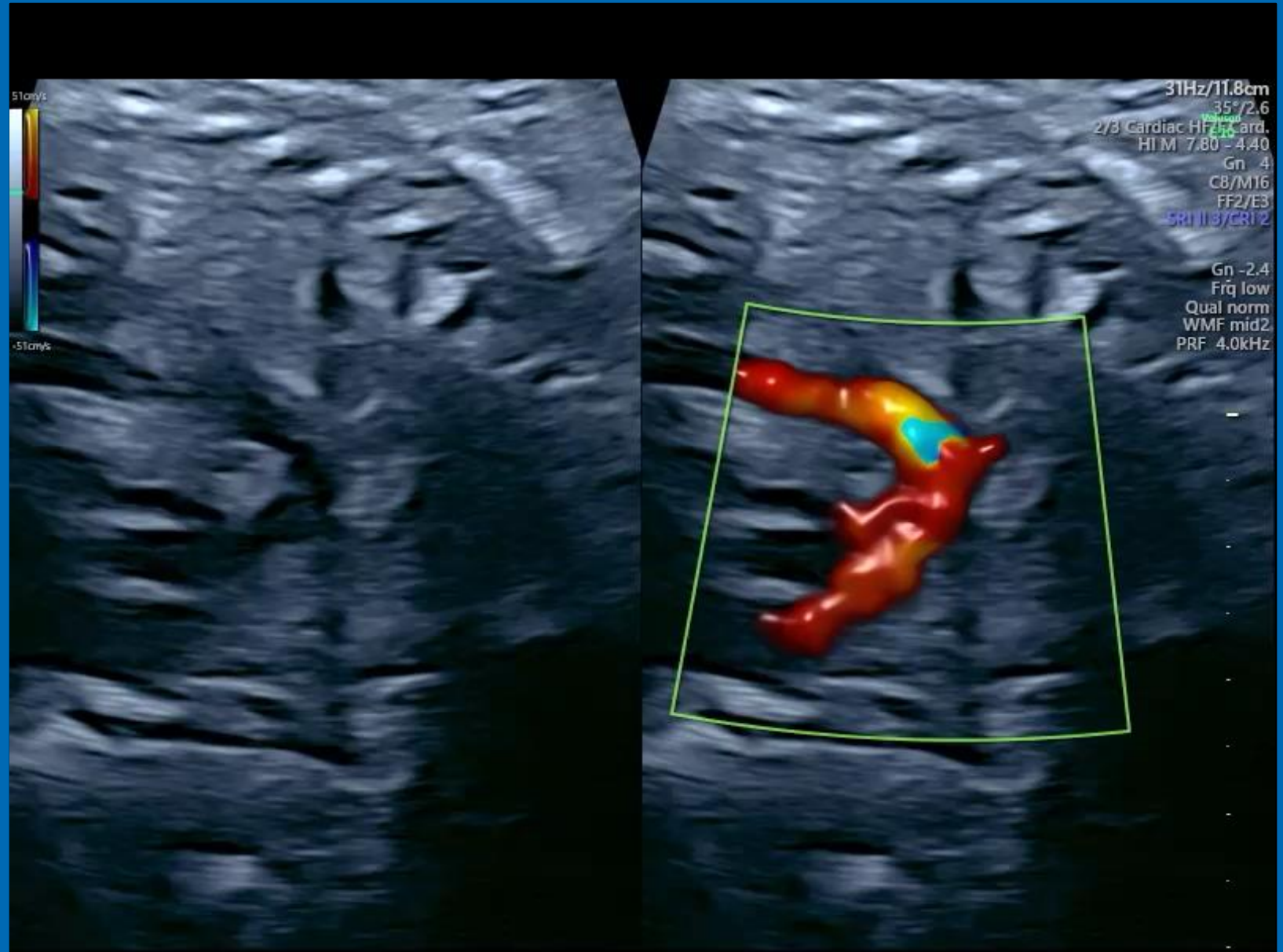


AORTIC ARCH

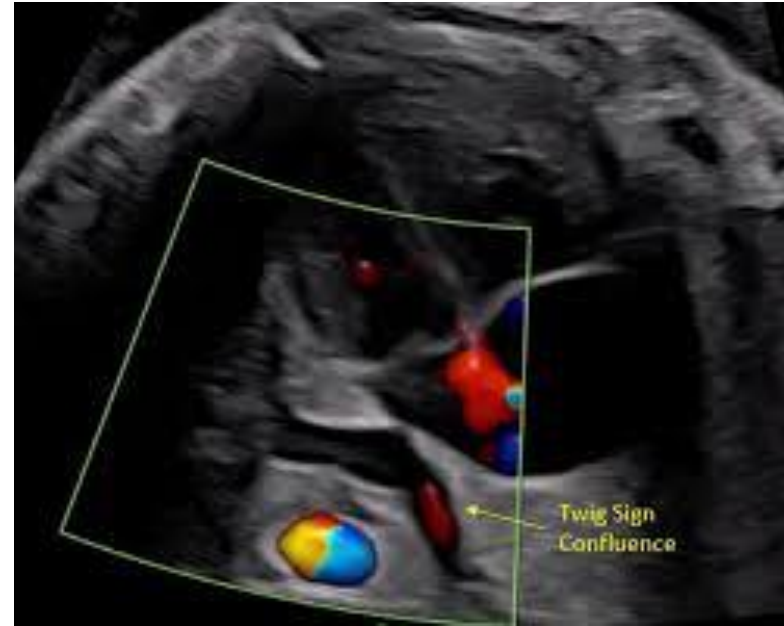
Coarctation of the Aorta

WHY IT'S HARD

- The ductus is large in fetal life and inserts where coarctation occurs
- Clues: small arch/isthmus; RV > LV; PA > aorta
- Suspected → postnatal “coarctation watch” and PGE1 readiness



Pulmonary Veins



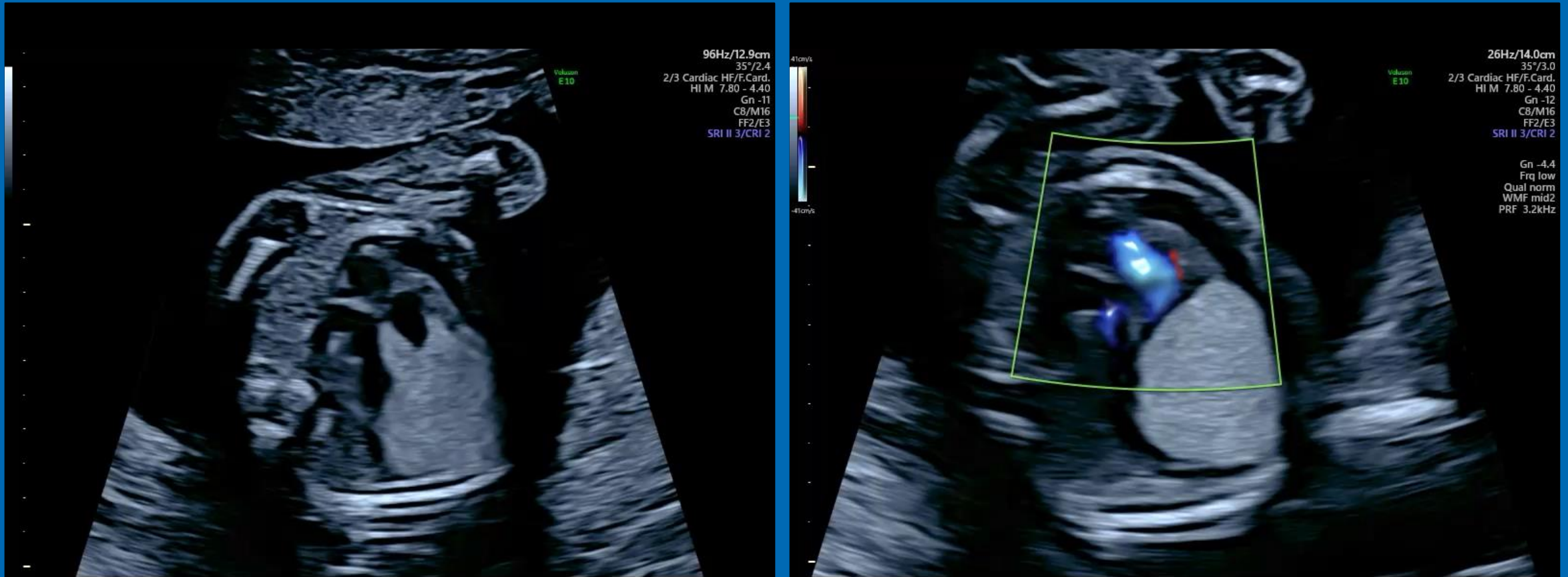
PITFALL

Tiny fetal veins can look like they enter the LA on 2D and color when they don't — always confirm with pulsed Doppler (at least one vein on each side).

WHY IT MATTERS

Obstructed TAPVR is a surgical emergency at birth — prenatal suspicion (though difficult) allows delivery at a surgical center.

Cardiac Masses



Large echogenic mass — what is it, and is it obstructing inflow or outflow?



Cardiac Rhabdomyoma: Sirolimus and Surveillance

Natural history

- Most common fetal cardiac tumor
- Strongly associated with tuberous sclerosis complex (TSC), especially multiple tumors
- Tend to grow until ~30–32 weeks, then regress — often after birth
- Refer to genetics; consider fetal brain MRI for TSC

Watch for

- Inflow or outflow tract obstruction
- Arrhythmia — SVT (including WPW) and ventricular arrhythmias
- Ventricular dysfunction or pericardial effusion
- Hydrops
- Serial fetal echo as the tumor grows

Transplacental sirolimus

- mTOR inhibitor — targets the pathway disrupted in TSC
- Considered for obstruction, arrhythmia, dysfunction, or impending hydrops
- Reported tumor regression within weeks
- Maternal trough levels and labs; watch for mucositis, transaminitis, hyperlipidemia, infection



RHYTHM

Rhythm Evaluation

Is this heart rate normal?

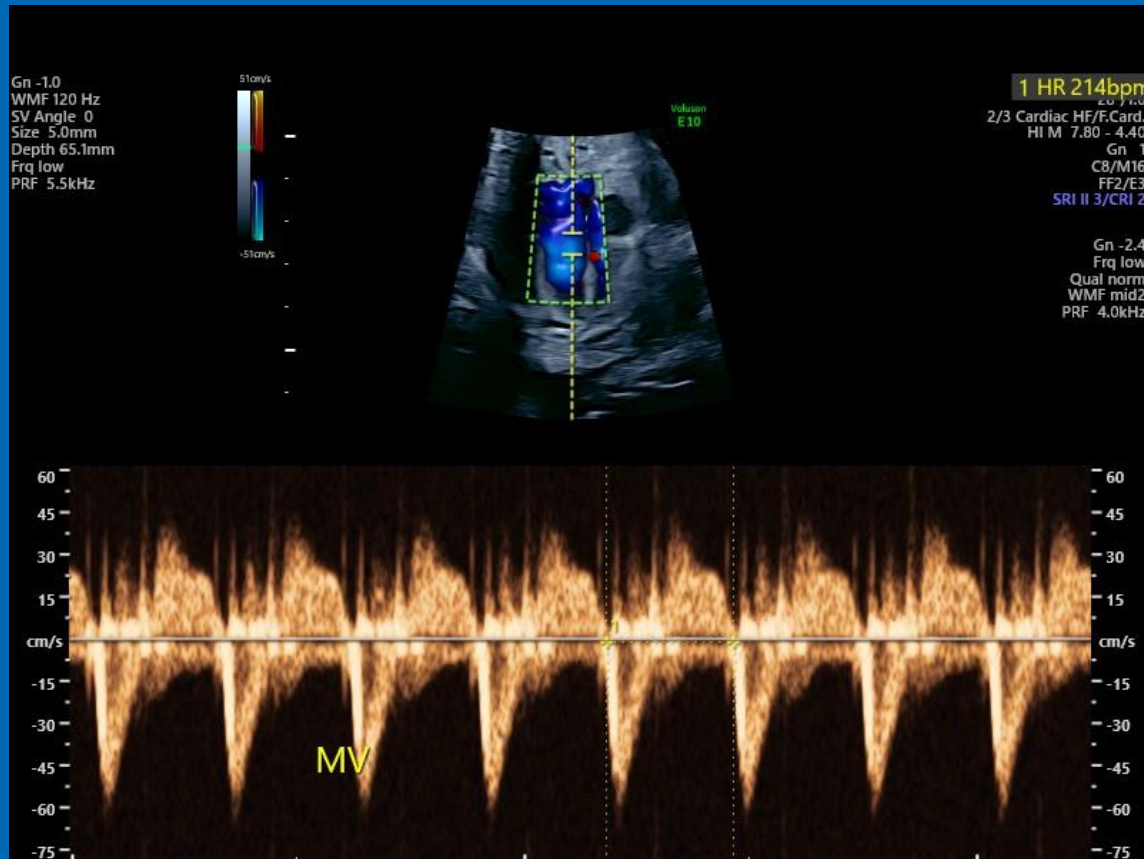
NORMAL FETAL HEART RATE

- ≈110–160 bpm, regular

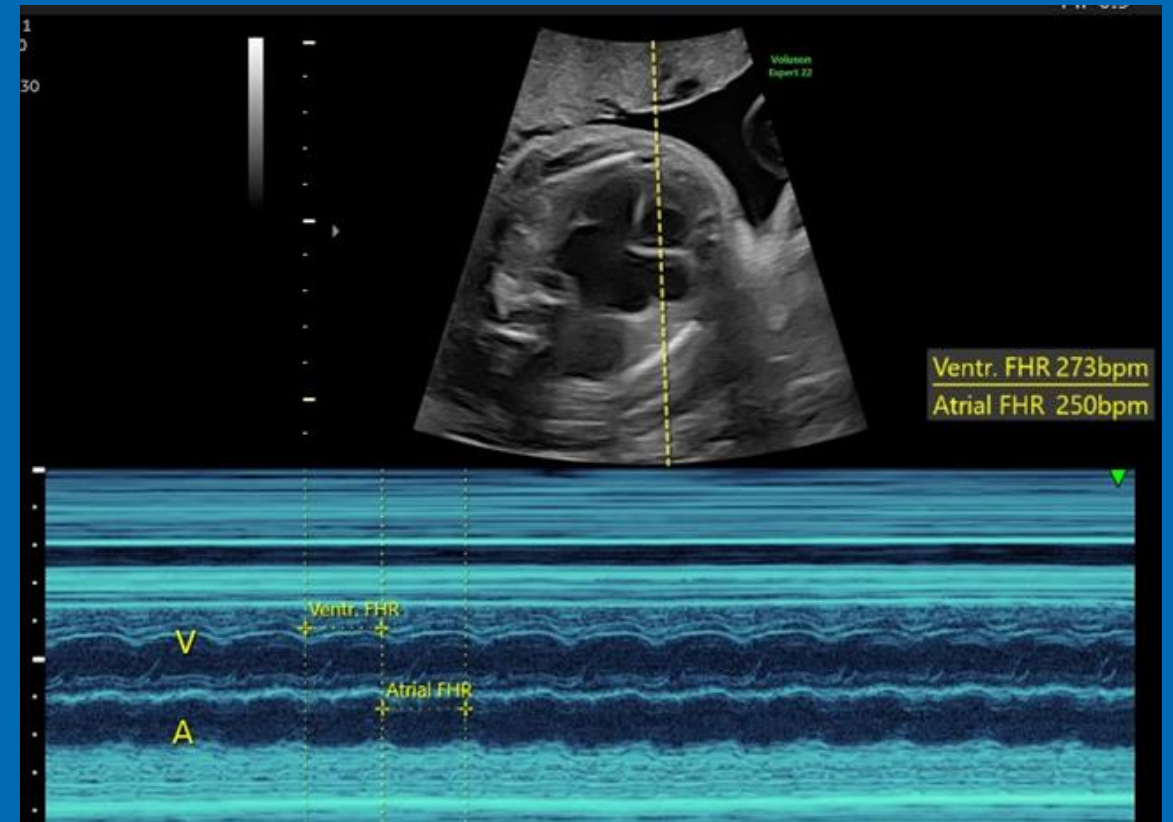


RHYTHM

Defining Tachycardia without an ECG



Inflow / outflow Doppler: 1:1 AV relationship at ≈ 214 bpm



M-mode: atrial and ventricular rates — 1:1, consistent with SVT



Treating Fetal Tachyarrhythmia

Who to treat

- Sustained or frequent tachycardia
- Any ventricular dysfunction, AV valve regurgitation, or hydrops
- Brief, intermittent runs with normal function: close surveillance

Goal: control the rhythm in utero and aim for a full-term delivery.



Flecainide

First- or second-line for SVT. Good placental transfer, including with hydrops. Monitor maternal ECG.



Digoxin

First- or second-line for SVT. Poor placental transfer once hydropic. Follow maternal levels; monitor maternal ECG.



Sotalol

Often used for atrial flutter. Monitor maternal ECG.



Amiodarone

Refractory tachycardia, often in combination. Monitor maternal ECG and thyroid; risk of fetal hypothyroidism.

Maternal safety: baseline ECG and electrolytes; inpatient initiation with telemetry and serial ECGs.



RHYTHM

Rhythm Evaluation

And this one?



Heart Block: Surveillance and Treatment

Anti-SSA/SSB (Ro/La) pregnancies

- Refer to fetal cardiology even with a normal heart rate
- Higher risk of heart block — highest if a prior child was affected
- Close surveillance (AV interval) until about 27 weeks
- Any new bradycardia or irregular rhythm: same-day evaluation

Dexamethasone

CONTROVERSIAL

Some use for evolving 1st/2nd-degree block. Does not reverse 3rd-degree block.

IVIG

LIMITED EVIDENCE / ENCOURAGING

For myocarditis or endocardial fibroelastosis (EFE).

β-agonists

LIMITED EVIDENCE

Terbutaline or salbutamol to raise the rate (usually when <55 bpm).

Hydroxychloroquine

EMERGING

Recurrence prevention in anti-Ro+ mothers with a prior affected child. PATCH: 400 mg/day by 10 wk → 7.4% (4/54) vs 18% historical.

PATCH: Izmirly P, et al. J Am Coll Cardiol. 2020;76(3):292-302.



Planning for Fetal Arrhythmias

Rhythm status	Fetal management	Delivery plan
SVT / flutter controlled on therapy	Continue therapy; serial fetal echo for rhythm and function	Aim for term; mode by obstetric indications
Persistent tachycardia at term	Weigh delivery against escalating therapy	Deliver rather than keep treating
Uncontrolled tachycardia, decreased function	Escalate or combine agents; inpatient monitoring	Planned delivery at a cardiac center
Hydrops	Aggressive transplacental therapy; close surveillance	Consider early delivery
Bradycardia <3rd %ile, 2:1 block, torsades (suspect LQTS)	Evaluate for long QT (family history, parental ECGs); avoid QT-prolonging drugs	Planned delivery; pacing available
Complete heart block	Serial echo: ventricular rate, function, EFE, hydrops; β -agonist if very slow	Planned delivery; pacing available

Why plan at all? Labor catecholamines can worsen tachycardia, and a weak ventricle can fail when systemic resistance jumps at cord clamping.



What We Can Offer in Utero

Finding	Fetal therapy	Goal
SVT / atrial flutter	Transplacental flecainide, digoxin, sotalol, amiodarone	Restore sinus rhythm or control rate; prevent or reverse hydrops
Anti-Ro-mediated heart block	Dexamethasone, IVIG, β -agonists; hydroxychloroquine for recurrence prevention	Halt progression; treat myocarditis/EFE; support ventricular rate
Ebstein anomaly with circular shunt	Maternal NSAID (ibuprofen, indomethacin)	Constrict the ductus to break the circular shunt
Rhabdomyoma (tuberous sclerosis)	Transplacental sirolimus	Shrink the tumor; relieve or prevent obstruction
Critical aortic stenosis with evolving HLHS	Fetal aortic valvuloplasty	Preserve a biventricular circulation
HLHS with restrictive / intact atrial septum	Fetal atrial septoplasty or stent	Decompress the left atrium before birth

For every diagnosis: delivery planning — location, timing, and delivery-room level of care.



SUMMARY

Pattern Recognition: Quick Reference

Diagnosis	What you'll see	Why it matters prenatally
HLHS	Small, non-apex-forming LV; diminutive or absent mitral/aortic valves; tiny ascending aorta	Systemic flow is duct-dependent — prenatal diagnosis allows PGE1 at birth and delivery at a surgical center; possible balloon atrial septostomy after birth if the atrial septum is restrictive
HRHS (e.g., tricuspid atresia)	Small right ventricle; diminutive or absent tricuspid/pulmonary valves	Pulmonary flow is often duct-dependent — prenatal diagnosis allows PGE1 at birth and delivery at a surgical center
VSD / AVSD	Septal dropout on the 4-chamber view or sweep, confirmed with color Doppler shunt flow	Most VSDs are benign, but size, location, and associated lesions (e.g., AVSD with trisomy 21) guide genetic testing and follow-up
Ebstein anomaly	Cardiomegaly (increased CT ratio); apically displaced tricuspid valve with severe TR; enlarged RA	Risk of circular shunt and fetal demise — maternal NSAIDs (e.g., ibuprofen) can constrict the ductus and break the shunt
TGA	Great vessels run parallel instead of crossing; the 4-chamber view often looks normal	Circulations don't mix without a shunt — planned delivery for possible urgent septostomy and arterial switch avoids severe hypoxemia
Truncus arteriosus	A single great vessel arises over a VSD instead of two separate outflow tracts	Mixing lesion linked to 22q11 deletion — prompts genetic counseling and planning for early repair



SUMMARY

Pattern Recognition: Quick Reference

Diagnosis	What you'll see	Why it matters prenatally
Coarctation of the aorta	Aortic arch small relative to the pulmonary artery/ductus on arch or 3VT views	Can cause neonatal shock as the ductus closes — prenatal suspicion guides ductal monitoring and PGE1 readiness
TAPVR	Pulmonary veins drain to a posterior confluence rather than the LA; subtle on the 4-chamber view	Obstructed TAPVR is a surgical emergency at birth — prenatal suspicion (though difficult) allows delivery at a surgical center
Cardiac masses	Echogenic intracardiac mass(es) that may obstruct inflow/outflow	Monitor all masses for obstruction or arrhythmia — transplacental sirolimus for rhabdomyoma (tuberous sclerosis) with obstruction, arrhythmia, or hydrops
Tachycardia	Sustained rate >180 bpm on M-mode/Doppler (1:1 AV in SVT; 2:1–4:1 in atrial flutter)	Can cause hydrops and fetal distress — transplacental flecainide, digoxin, sotalol, or amiodarone can prevent fetal demise
Bradycardia (e.g., heart block)	Sustained ventricular rate <100 bpm with AV prolongation or dissociation on M-mode	Often maternal anti-SSA/SSB or structural CHD — maternal workup; dexamethasone, IVIG, or β -agonists in select cases; delivery planning for pacing if complete heart block



When to Call Fetal Cardiology

1

Risk of fetal morbidity or demise

In-utero therapy may be possible

2

Complex CHD

May need ICU care or surgery

3

Critical CHD

Emergency intervention at birth

Pearls

- A normal four-chamber view does not exclude CHD — always sweep through the outflows and 3VT view.
- Sustained fetal heart rate >180 or <100 bpm, or hydrops: urgent referral.
- Anti-SSA/SSB positive: refer for surveillance, even with a normal heart rate.
- When in doubt, refer — prenatal diagnosis lets us plan therapy, delivery location, and the team in the room.



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Thank you!



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Here, it's different.™

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